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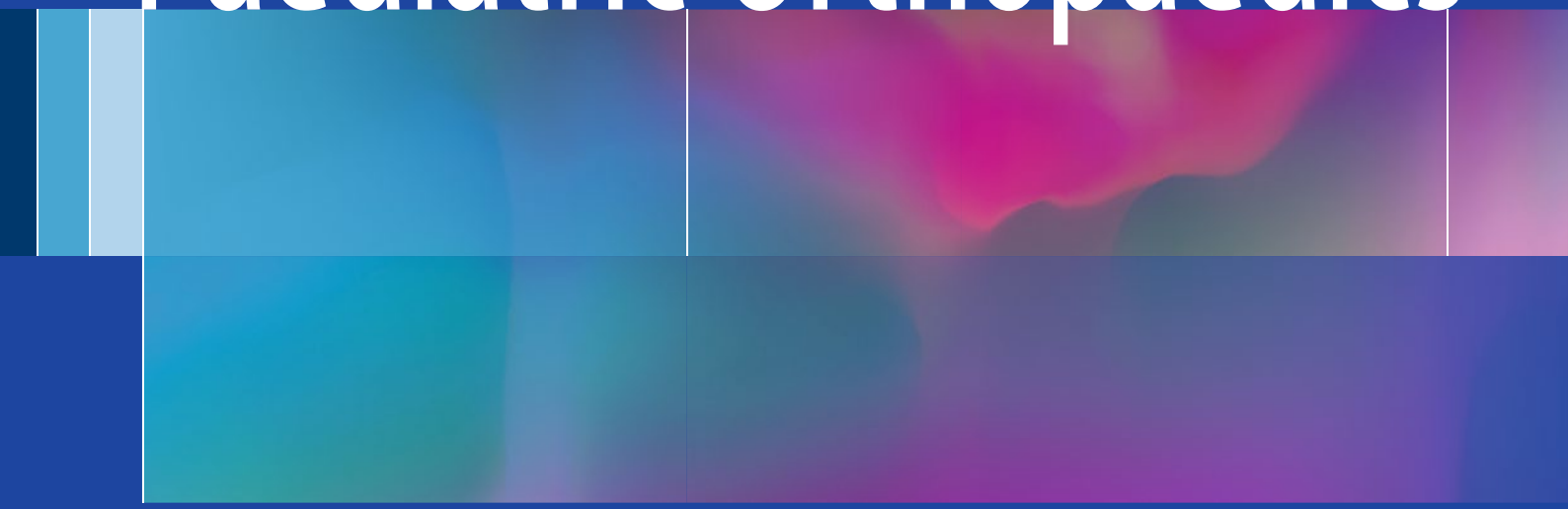
Editors

Paediatric Orthopaedics



An Evidence-Based Approach to Clinical Questions

Springer

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To every child and parent including mine: Daniel, Hannah, Jasmine, Ania, Maria and Jaber.

Sattar Alshryda

For my parents, for their steadfast support.

James S. Huntley

To my parents for their support and encouragement.

Paul Banaszkiewicz

Foreword

Surgeons have always wanted to do the best for their patients. However, determining what is best for the patient is not always clear leading to many treatments options for most orthopaedic conditions. Moreover, because Paediatric Orthopaedics is one of the most varied of the Orthopaedic disciplines due to the many rare conditions with myriad presentations often afflicting the entire child, deciding with the child and parents among the many treatment options is particularly complex.

Treatment options need to provide on average better health outcomes than alternative or no treatment. Each treatment option comes with a range of benefits and risks, dependant on the child, setting, skill and experience of the surgeon. To make these complex decisions surgeons and families need the best evidence. While there are many factors that enter into critical appraisal of research studies and deciding what constitutes best evidence, surgeons recognise that, in general, randomised are better than non-randomised studies, prospective are better than retrospective studies, and controlled are better than uncontrolled studies.

In recent years there has been a focus on Evidence-based Orthopaedics. Evidence-based orthopaedics, with an emphasis on systematic overviews and randomized clinical trials, uses the best evidence to make the decisions with families. However, randomised trials are difficult in Paediatric Orthopaedics for several reasons including the rarity of many conditions therefore requiring multicentre studies that further increases the complexity of any trial. However, the quality of orthopaedic literature continues to improve and this should result in better outcomes for children.

While not every condition is covered and not every area has definitive evidence, this text will provide surgeons with practical advice on the best treatment, or treatment options, for most paediatric orthopaedic conditions. The key to each chapter is found at the end where treatment recommendations are provided with an attached strength of recommendation. Not only will this book be an invaluable and useful text for practising surgeons, it also highlights areas of research for the future. Please enjoy reading this book as much as I have enjoyed being a part.

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List of Abbreviations

Order	Abbreviations	Meaning
A	ABD	Abduction, Pronation abduction type injury
	AFO	Ankle Foot Orthoses
	AK	Above Knee
	AAOS	The American Academy of Orthopaedic Surgeons
	AHA	Assisted Hand Assessment
	AHO	Acute Hematogenous Osteomyelitis
	AIS	Adolescent Idiopathic Scoliosis
	AAI	Atlanto-Axial Instability
	AARF	Atlanto-Axial Rotatory Fixation
	ADM	Abductor Digiti Minimi
	AE	Adverse Event
	AER	Apical Epidermal Ridge
	AMC	Arthrogryposis Multiplex Congenita
	ANC	Absolute Neutrophile Count
	AP	Antero-Posterior
	ARIF	Arthroscopic Reduction and Internal Fixation
	ASIS	Anterior Superior Iliac Spine
	ATFL	Anterior Talo-Fibular Ligament
	ATiFL	Anterior Tibio-Fibular Ligament
	AVN	Avascular Necrosis (see also ON)
	AP	Antero-Posterior
B	BrAIST	Bracing in Adolescent Idiopathic Scoliosis Trial
	BK	Below Knee
C	C	Centigrade
	CDK	Congenital knee dislocation
	CEA	Centre Edge Angle
	CFL	Calcaneo-ofibular Ligament
	CFD	Congenital Femoral Deficiency
	CI	Confidence Interval
	CMC	Carpametacarpal Joint
	CNS	Central Nervous System
	CORA	Centre of Rotation of Angulation
	CP	Cerebral Palsy
	CR	Closed Reduction
	CRPP	Closed Reduction and Percutaneous Pinning
	CRUS	Congenital Radio-Ulnar Synostosis
	CSF	Congenital Short Femur
	CTEV	Congenital Talipes Equinovarus (Club foot)
	CV	Coxa Vara
	CVS	Cardiovascular System

Order	Abbreviations	Meaning
D	DASH	Disabilities of the Arm, Shoulder and Hand
	DFO	Dorsal First metatarsal Osteotomy
	Dexa	Dual-energy X-ray absorptiometry
	DMD	Duchene Muscular Dystrophy
	DVT	Deep Venous Thrombosis
E	ED	Emergency Department
	EWS	Classic Ewing's Sarcoma
	ESFT	Ewing's Sarcoma Family Tumours
	ESIN	Elastic Stable Intramedullary Nailing (see also FIN)
F	FAV	Femoral Anteversion
	FDS	Flexor Digitorum Superficialis
	FIN	Flexible Intramedullary Nailing (see also ESIN)
	FPA	Foot Progression Angle
G	g	Gram
	GA	General Anaesthetic
	GCS	Glasgow Coma Scale
	GHJ	Gleno-Humeral Joint
	GP	General Practitioner
I	IGHL	Inferior Gleno-Humeral Ligament
	IJO	Idiopathic Juvenile Osteoporosis
	II	Image Intensifier
	IMN	Intramedullary Nailing
	Intraop	Intra-operative
	ITB	Ilio-Tibial Band
	IU	International Unit
K	Kg	Kilogram
	KAFO	Knee Ankle Foot Orthoses
L	LoE	Level of Evidence
	LOS	Length of Stay
	LLD	Leg Length Discrepancy
	LRR	Lateral Retinacular Release
M	M	Meter(s)
	MBD	Metabolic Bone Diseases
	MCPJ	Metacarpaphalangeal Joint
	MD	Mean Difference
	MED	Metaepiphyseal Dysplasia
	MGHL	Middle Gleno-Humeral Ligament
	min	Minute(s)
	MOI	Mechanism of Injury
	mPFL	Medial Patella-Femoral Ligament
	MS	Marfan Syndrome
	MSKI	Musculoskeletal Infection
	MTPJ	Metatarsal Phalangeal Joint
	MVC	Motor Vehicle Collision
N	NB	Nerve Block
	NAT	Non-Accidental Trauma
	NF	Neurofibromatosis
	Ng	nanogram
	NSAID	Non Steroidal Anti-inflammatory Drugs

Order	Abbreviations	Meaning
O	OA	Osteoarthritis
	OCF	Occipitocervical Fracture
	ON	Osteonecrosis (see also AVN), Ossific Nucleus
	OR	Odds Ratio; Open Reduction; Operating Room
	ORIF	Open Reduction and Internal Fixation
	OSA	Obstructive Sleep Apnoea
P	P	P-value
	PB	Peroneus Brevis
	PCR	Polymerase Chain Reaction
	PE	Pulmonary Embolism
	PETS	Percutaneous Ephiphysiodesis with Transphyseal Screws
	PF	Patellofemoral/Planter Fasciotomy
	PH	Pavlik Harness
	PIPJ	Proximal Interphalangeal Joint
	PL	Peroneus Longus
	POA	Periacetabular Osteotomy
	PPC	Premature Physeal Closure
	PNET	primitive Neuroectodermal Tumours
	PTFL	Posterior Talo-Fibular Ligament
	PTiFL	Posterior Tibio-Fibular Ligament
	Postop	Postoperative
	Preop	Preoperative
	PSACH	Pseudoachondroplasia
	PTH	Parathyroid Hormone
R	RTA	Road Traffic Accident
	RCT(s)	Randomised Controlled Trial(s)
	RD	Risk Difference
	ROM	Range of Motion
	RR	Risk Ratio
S	SA	Spinal Anaesthetic
	SBC	Simple Bone Cyst
	SBSQ	Spina Bifida Spine Questionnaire
	SCIWORA	Spinal Cord Injury Without Radiological Abnormality
	SD	Standard Deviation
	SED	Spondoepiphyseal Dysplasia
	SH	Salter-Harris
	SRS	Scoliosis Research Society
T	TAL	Tendoachilles Lengthening
	TAT	Transfer of Tibialis Anterior Tendon
	TCC	Talocalcaneal Coalition
	TT	Tourniquet Time
	TL	Thoracolumbar
	TLSO	Thoracolumbosacral Orthosis
	TT-GT	Tibial Tubercle-Trochlear Groove Distance
V	vs.	Versus
	VDRO	Varus Derotation Osteotomy
	VMO	Vastus Medialis Obliqus
U	UCL	Ulnar Collateral Ligament
	UV	Ultraviolet
X	χ^2	Chi square
z	ZPA	Zone of Polarising Activity

Part I

General Guidance

Introduction to Evidence-Based Orthopaedics

1

Sattar Alshryda, James S. Huntley, and Paul Banaszkiewicz

Abstract

Practising surgeons may be unable to keep up with current practice – what seems up to date today can be redundant tomorrow. In paediatric orthopaedics, and the setting of a busy clinical practice, it is challenging to find time to retrieve the best available studies, let alone analyse them, or synthesise the resulting information into a form applicable to one's own practice. We are hardly alone in this regard (eg, Narayanan and Wright [1]), so this book is a collective effort to probe the common questions arising in our speciality – and look critically for answers within the literature. In this work, we are proud to have brought together the thinking of more than fifty leading paediatric orthopaedic surgeons to assemble the evidence underpinning elements of current practice. The approach has been that of the 'evidence-based medicine' pragmatist, written by frontline practitioners.

Keywords

Orthopaedics • Levels of evidence • Grades of recommendation

Practising surgeons may be unable to keep up with current practice – what seems up to date today can be redundant tomorrow. In paediatric orthopaedics, and the setting of a busy clinical practice, it is challenging to find time to retrieve the best available studies, let alone analyse them, or synthesise the resulting information into a form applicable to one's own practice. We are hardly alone in this regard (eg, Narayanan and Wright [1]), so this book is a collective effort to probe the common questions arising in our speciality – and look critically for answers within the literature. In this work, we are proud to have brought together the thinking of more than fifty leading paediatric orthopaedic surgeons to assemble the evidence underpinning elements of current practice. The approach has been that of the 'evidence-based medicine' pragmatist, written by frontline practitioners.

Evidence-based medicine is *'the conscientious, explicit, and judicious use of current best evidence in making decisions about the care of individual patients'* (Sackett et al. [2]). Therefore, it involves the thoughtful integration of the best available research, the clinical circumstances and patients' own values and preferences. A clearly defined relevant question is required, followed serially by (i) identification of the studies/evidence by a thorough search of the literature, (ii) a critical appraisal of available evidence and its applicability to the clinical situation, and (iii) a balanced conclusion to the clinical problem and particular patient. A mnemonic for the process is the 5 'A's (Wright et al. [3]): *Assess* the patient to determine the clinical issues. *Ask* a clear question to be pursued. *Acquire* the evidence. *Appraise* the evidence for its worth/validity. *Apply* the evidence to the particular patient. Broadly, this has been the suggested approach for the text, albeit with generalisation of the patient, and followed by a synthesis of the conclusions according to both level of evidence and grade of recommendation.

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Editing this book has been a challenge on several levels. Authors vary in their understanding, experience of evidence-based medicine and modes of analysis of the literature. As far as possible we have sought to maintain the general format and quality of questions and chapters, whilst recognizing that, as with most multi-author texts, there are bound to be differences in emphasis, analysis and style. We have tried not to allow our own prejudices to influence the authors' 'open hand' to potentially contentious areas of practice.

In some areas, rather than there being a definitive guide to practice, it is clear that there is a lack of evidence; 'confusion' may simply represent the current state of knowledge, not least because: *'development of new treatment choices has far outpaced our capacity for determining what procedures are beneficial…'* (Wenger [4]). Paediatric orthopaedics, as a field of endeavour, has moved forward at a rapid pace over the last two decades, often outstripping our abilities to define best practice *per se*. Defining a dearth of evidence is a natural pre-requisite to addressing it in the future. Conversely where evidence does exist, it is vital that those who care for children keep up to date.

We are immensely grateful to all the authors who have given so generously of their time, effort and expertise in assembling the material for this book.

References

1. Narayanan U, Wright JG. Evidence-based medicine: a prescription to change the culture of pediatric orthopaedics. J Pediatr Orthop. 2002;22:277–8.
2. Sackett DL, Rosenberg WMC, Gray JAM, Haynes RB, Richardson WS. Evidence-based medicine: what it is and what it isn't. BMJ. 1996;312:71–2.
3. Wright JG, Kocher MS, Sanders JO. Evidence-based pediatric orthopaedics: an introduction, part 1. J Pediatr Orthop. 2012;32:S83–90.
4. Wenger DR. Limitations of evidence-based medicine: the role of experience and expert opinion. J Pediatr Orthop. 2012;32:S187–92.

Critical Appraisal of a Published Paper

2

Raymond Pollock and Arkan S. Sayed-Noor

Abstract

Critical appraisal is a systematic process used to identify the strengths and weaknesses of a research article. It enables the validity of research findings to be determined. It is just one step in the process of evidence based medicine – the use of best evidence in making decisions about patient care. The validity of a research study depends on its design and quality. Study designs are classified according to a hierarchical ranking called levels of evidence. To facilitate critical appraisal, checklists that ask questions about the research have been developed enabling the reader to judge its validity. Critical appraisal checklists can be divided into generic and study type specific lists. In this chapter a generic tool is described that is appropriate for the novice reviewer. For the experienced reviewer study specific checklists for each of the main study types are then described. Not only does this chapter provide the tools needed for critical appraisal of published work, it should also be of help when conducting and publishing research by ensuring that the checklists for that study type are taken into account during the design phase.

Keywords

Critical appraisal • Evidence based medicine • Hierarchy of evidence • Checklist • Case report • Case series • Cross-sectional study • Case control • Cohort • Randomized controlled trial • Systematic review • Meta-analysis

Critical appraisal (CA) is a systematic process used to identify the strengths and weaknesses of a research article and thereby determine the study's validity. It is important when reading a published paper to keep a degree of skepticism. This means being open-minded and willing to be convinced but only if authors can adequately back up their claims. The critical reader is not put off by the limitations of a study but will expect authors to interpret their results in a way that takes account of the limitations.

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The validity of a research study depends on its design and quality. Study designs are classified according to a hierarchical ranking called levels of evidence. The classification varies depending on whether the study type is therapeutic, prognostic, diagnostic or economic/decision analysis.

For therapeutic studies randomized controlled trials (RCTs) and meta-analyses of RCTs are at the top of the hierarchy and can be considered best evidence. Descriptive studies (qualitative studies, case reports, case series, cross sectional studies) are the lowest level of evidence. Observational studies (case-control and cohort) are in the middle (Fig. 2.1). These study types will be described in detail later.

Low-level evidence is more likely to be subject to bias. Bias is a systematic error that can make the results invalid. There are many kinds of bias but important ones in orthopedics are selection bias, response bias, recall bias and bias due

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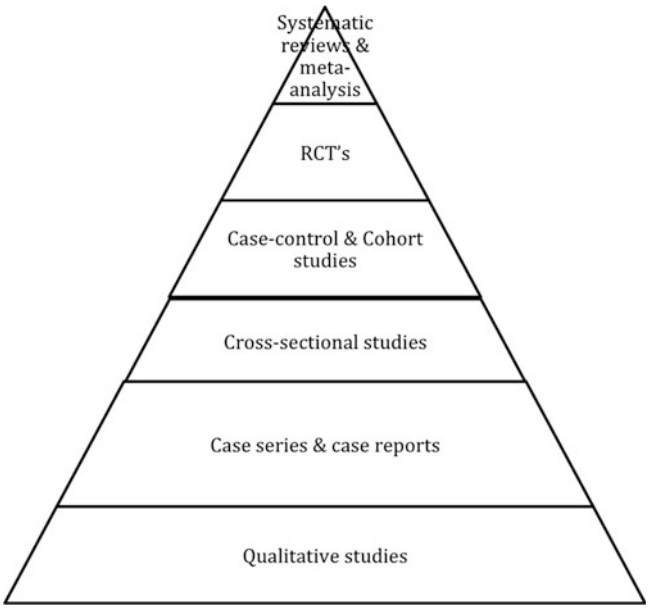

Fig. 2.1 Levels of evidence hierarchy for therapeutic studies

to confounding. These will be explained later on. This systematic error only results in bias when the inaccuracy affects comparison groups unequally. In theory a well conducted RCT should be free of bias because the process of randomisation is used to assign patients to treatment groups and this should result in the groups being balanced in all factors.

As the level of evidence hierarchy is descended, bias is increasingly likely and you need to be aware of this when you critically appraise articles.

Selection bias, sometimes referred to as sampling bias is error due to the improper process of selecting a study population i.e. the way subjects were identified, selected and included in a study.

Response bias or loss to follow-up bias can result in differences in the characteristics e.g. socio-demographic characteristics, of patients included in a study and those excluded or between selected comparison groups e.g. case and control. For example, response to follow-up may be dependent on socio-demographic characteristics of patients (sex, age, ethnicity, social class). Those who respond may be different from those who do not, leading to bias in the results. Response bias is common in case series and cross sectional study types. Wherever possible, analysis of the demographics of non-responders should be carried out to determine if they differ significantly from responders. In all study types response rates should be high, at the very least 70 % to minimize this type of bias.

Another type of bias is recall bias. This is particularly common in cross-sectional and case-control studies. Patients may not be able to remember correctly past events. Wherever possible any information obtained from patients should be verified from other sources such as patient records.

Confounding bias occurs when part of an observed relationship between 2 variables or factors involved in a disease is due to the action of a third, which is the actual factor responsible. Confounding arises because many aspects of behavior and health are related. Frequent confounders are gender, age, socio-economic status and co-morbidity. In RCTs randomisation ensures that potential confounding factors, known or unknown are evenly distributed among the study groups. This is why this study type is highly regarded.

The Process of Critical Appraisal

Critical appraisal to determine the validity of research findings is an established method used in evidence-based medicine (EBM). It is just one step in the process of EBM – the use of best evidence in making decisions about patient care. To facilitate CA, checklists that ask questions about the research have been developed enabling the reader to judge its validity.

Critical appraisal checklists can be divided into generic and study type specific lists. For the novice reviewer a generic tool is appropriate until more experience is gained. When you are confident and able to identify the study type, you will be able to progress to using the study specific checklists described later in this chapter. The checklists in this chapter are generally from our own experience.

The Anatomy of a Scientific Manuscript

Manuscripts in orthopedic journals have a standard format as follows but with minor variations depending on the particular journal:

- Abstract (structured or unstructured with Medical Subject Heading (MeSH) keywords – keywords are sometimes at the end of the article before the references instead). MeSH keywords are used to describe precisely the content of journal articles
- Sponsorship/competing interests (usually on the title page).
- Introduction.
- Methods (or materials and patients, materials and methods, patients and methods).
- Results.
- Discussion.
- Conclusion (sometimes absent).
- References.

Appraisal for the New Reviewer

For those new to CA it may be best to start with a more general appraisal until confidence is gained. Read through the whole paper quickly first. Does it seem clearly written and easy to understand or does it appear that it has been rushed?

You will probably find papers describing RCTs and meta-analyses the most structured because journals usually have guidelines as to how they should be formatted.

Next, you should be aware of the quality of the journal in which the research is published. This is partly measured by its impact factor (IF). Because it is based upon the number of citations of its papers it is not a fixed value but can vary from year to year.

For orthopedic journals an IF of 2.8 is regarded as high (e.g. JBJS (Br), recently renamed as The Bone & Joint Journal). For general medical journals it is much higher – the BMJ is currently about 16.

Basically a journal is considered to be of good quality if is peer reviewed – that is each paper is reviewed by at least one expert in the subject area prior to acceptance of publication in the journal. In the higher impact journals it is often reviewed independently by 3 experts including a statistician if the work involved statistical tests.

To be accepted, a paper usually has to be approved by all reviewers although the editor does have the final decision, for example if one of the reviewers is doubtful.

Look also at the author names as given on the title page. You may be familiar with certain authors from your attendance at conferences etc. Related to the authors, is the institution – is it a center of excellence in orthopedics? This will give you more confidence in the validity of the research.

Conflict of interest is particularly important to look out for. This is usually at the bottom of the title page along with authors' affiliations. The most common conflict of interest is that the authors have a financial affiliation with a company that manufactures the products used in the research. For example many orthopedic surgeons are actively involved in design of new implants for which they get remuneration or gifts (e.g. holidays) for their involvement. This is an important part of the evolution of new devices for patient benefit. But it can lead to conscious or unconscious behavior that undermines the integrity and validity of research that involves such appliances.

When there is conflict of interest it is important that it has been recognized and dealt with. For example it might be stated that sponsors had no input into the protocol or conduct of the study. The reader must then decide whether any conflicts are important and might have influenced the validity of the study findings.

After reading through the whole paper, look in detail at each section as follows:

Introduction

What were the aims of the study? Look for this in the introduction or discussion (where it is often reiterated). It may be stated as a formal hypothesis (the null hypothesis). For example "there is no difference in outcome between patients in the two treatments" that the study aims to reject. More usually it is stated as a general research question such as "the purpose of this study was to determine if treatment A is more effective than treatment B".

Papers that do not have a clearly focused research question may be data dredging i.e. performing multiple statistical tests on the resulting data to see if anything of significance surfaces. This is bad science.

Methods

How were the patients selected for the study? Remember selection/sampling bias here. What were the exclusion criteria?

Are the details of statistical analysis described and appropriate? If so, what types of tests were used e.g. t-test, Pearson's, and were they the most appropriate for the data types? For continuous data were efforts made to check the data for normality and if they were then a parametric test should have been used. If the data were non-normal, then the median rather than the mean should be quoted and non-parametric tests used. Statistical significance should be stated and is almost always given as $P < 0.05$ with confidence intervals (CI) at 95 %.

Was a sample size calculation made? This will not be relevant for case reports and series but applies for cross sectional and other studies higher in the hierarchy of evidence. If the sample size is too small for the effect size difference expected, then the study is unlikely to show statistical significance. A sample size calculation estimate should include the size of the minimum difference between the groups that is considered clinically significant (the effect size). For example, a 3-point decrease on the 0 to 10 visual analogue scale (VAS) for pain, would be regarded as clinically significant.

In case series, where a number of patients are reviewed and comparisons are made between subgroups of patients, it is possible to perform a post-hoc (after the analysis) power analysis. This may show that the results of no difference in the groups for a particular outcome may be due to insufficient sample size rather than the difference does not actually exist.

Other things to look out for in the methods are details of the surgical or other interventions used. Are they adequately described? What about the measures used to assess outcome? For example, if questionnaires have been used are they established ones or did the authors use their own questionnaires specifically designed for the study that may have not been validated?

Methods of measurements should be described in detail e.g. was a goniometer used for measuring straight leg raise or the less reliable visual estimation.

Be particularly critical of cross sectional survey type studies that the questions used are valid and reliable. How

was the questionnaire developed? Was it piloted for reliability and validity? For example does the questionnaire have content validity i.e. are the questions asked relevant and how did the results compare with similar validated questionnaires (criterion validity)? Did the same questionnaire give similar results when repeated soon after on the same patients (reliability)? Where standard questionnaires have been used (e.g. SF-36) then this should not be a problem assuming they have been used before in that particular patient group: questionnaires designed for adults may not be valid and reliable when used with children. Were answers to questions involving recall verified using other data sources such as patient records? This raises confidence in the results.

Results

Are the demographics of patients described in detail (age, sex, pathology etc) and a breakdown for the groups where relevant e.g. study and control groups. Remember confounding factors.

What was the response rate? It is recommended to be at the very least 70 %, otherwise there could be sampling bias. Were the demographics of the non-responders, where known, given and were they similar to the responders. Otherwise this means the responders may be atypical and the results will be biased.

Are any deviations from the protocol described e.g. unexpected events, patient drop out?

Are confidence intervals (CI) given for values? These are more informative than just P values as they indicate the possible range of values in the general population. Just a note here: if a P value is not significant then the CI should include zero (i.e. no difference) so just check these – It may be that the statistics are not up to scratch. Always look at the statistical tables and figures carefully (e.g. graphs – see below on interpreting tables and graphs) and see if there are unusual values that don't quite (metaphorically) add up (e.g. CI and P values – see above). Concerning P values, the smaller the value the less likely the result is due to chance, e.g. a $P < 0.01$ rather than $P = 0.048$ which is only bordering on significance. Not all papers quote the exact P value but use the expression $P < 0.05$.

Look for data dredging involving post-hoc analysis where tests are done on the data to look for interesting results – only tests should be performed that were stated in the original hypothesis e.g. to look for age or sex differences. Otherwise some of the significant results may be due to chance. This is because $P = 0.05$ signifies that chance could create the result 1 time in 20.

Do the tables, figures and graphs match up with any description in the main text? Do the values add up within the tables, figures and graphs?

Discussion

Have the authors discussed how their findings fit in with what is already known about the subject? Do the results fit in with previous findings and if not is there an explanation by the authors. Are you aware of similar studies that have been omitted and are contradictory to their findings? Do the findings appear plausible from a medical viewpoint.

Look for any overstatement of the findings i.e. over-extrapolation of the results which may only be the authors opinion.

Have they discussed the strengths and weaknesses of their findings?

Interpreting Figures, Tables and Graphs

Tables and graphs are time consuming and difficult to produce. Even with the help of word processor templates it is easy for errors to creep in. But they often improve the clarity of a paper. Tables often contain a lot of information and may be difficult to decipher. Look for:

- Self-explanatory title with units of measurements.
- Labelling of rows and columns.
- Are the rows/columns ordered e.g. by age.
- Numbers rounded to 2 significant figures e.g. 72.8 not 72.799. This will give you some indication of the standard of statistical input to the paper.

For figures make sure the appropriate type of figure has been used, that is graphs, histograms, bar charts, scatter plots or box plots. The axes should be labeled with the units.

With graphs watch out for scales that don't start at zero – this may deceptively emphasize an effect. Histograms are for continuous grouped data e.g. age groups. They show the symmetry of the data and give some indication of the normality. This symmetry is related to the use of an appropriate statistical test. Parametric tests should be used for normal (symmetrical) data and non-parametric for non-normal (skewed) data.

Scatter plots and graphs show how 2 variables relate to each other. Bar charts are for discrete data e.g. blood groups, whereas graphs are for continuous data e.g. blood pressure or age. When data is grouped e.g. age in 5-year intervals, information is lost within the groups and this may hide important information that was in the original raw data. Scatter plots are for showing the relationship between 2 variables and often a correlation coefficient is given for the strength of the relationship. With scatter plots look out for outliers (extreme values e.g. age 99 instead of 9) that may have distorted summary values (e.g. means) listed in tables. Such outliers may be erroneous values that should have been screened out during data cleaning or an explanation given for their inclusion.

Advanced Critical Appraisal

Although the foregoing generic appraisal guidelines are relevant for the less experienced reader, detailed study specific checklists are needed for a more robust critical appraisal.

The main epidemiological study types are:

- Qualitative research.
- Case reports.
- Case series.
- Cross sectional.
- Case control.
- Cohort.
- RCT.
- Systematic review/meta-analysis.

Advanced critical appraisal necessitates identifying the study type used in the paper. This may be given in the title or in the introduction or methods section. Often apart from RCTs and systematic review/meta-analyses it may not be explicitly mentioned and you will need to decide for yourself. This will only come from experience.

If the paper has keywords and these are MeSH terms, then the type of study should be stated so it is best to look their first. But not all study types have a MeSH term e.g. case series is not given a term although case report is.

Most of the checklists detailed below are the ones we use from our own experience of carrying out research and publication. Before submission we ensure that we have covered the relevant check points mentioned for each study type.

The Study Types

Qualitative Research

This is rarely found in the orthopedic literature.

It provides information on qualities that are difficult to measure for example patient experience, emotions, social interactions, attitudes, and behaviour. Qualitative studies have their own study types such as descriptive, phenomenology and ethnography. Qualitative studies are often combined with quantitative methods.

Qualitative studies are prone to bias and for this reason are at the bottom of the hierarchy. A description of these and their detailed appraisal is beyond the scope of this chapter.

Case Reports

This type of study is common in orthopedics. It is a type of qualitative research and in the hierarchy of evidence it is at a low level. This study type is easy to identify as often they are in a separate section of a journal. They are regarded as having low validity but have been important in alerting clinicians to unusual events such as adverse reactions to treatment or conditions not seen before.

They are communicated in a narrative fashion e.g. “a 11 year old girl suffered a fall from standing and subsequently developed pain and …” that has unusual or novel outcome. They often do not have the standard format of research papers, usually having a discussion and conclusion after the case report itself.

Case reports can lead to generation of new hypotheses that can be tested using a study higher in the hierarchy of evidence for example by an RCT. They also have a strong educational component providing unusual things to watch out for in your own patients. This is enhanced by the fact that many case reports also include a literature review of the subject.

There are journals (such as Case Connector of the JBJS (Am), www.caseconnector.jbjs.org) entirely devoted to case reports while some other journals do not include them at all e.g. International Orthopaedics. Because there publication adversely affects impact factor, their numbers are restricted. Because of their simplicity case reports usually do not have conflict of interest statements.

The following checklist for the CA of a case report is modified from Chan and Bhandari [1]:

- Does the case report include a literature review usually in the discussion section?
- Does it describe how its findings fit in with what is already known?
- What makes this case different from other cases? How is it unusual?
- Does the case challenge or confirm currently held beliefs?
- Are the results biologically plausible?
- Does the study reveal principles that might be applied to other patients?
- How was diagnosis made? How were treatments chosen?
- How accurate and thorough was the data collection and reporting?

Case Series

This is another common type of study in orthopedics and is an extension of case reports but of multiple patients. Again, it is low level evidence due to lack of any controls. Because of its frequency in the orthopedic literature, it is important to be able to appraise these articles. Many case series reports are quite large with hundreds of cases but they are prone to bias, particularly selection bias, as they are not a random sample of all cases. For example, a female clinician may attract more female patients who feel more comfortable with her.

Case series are easy to conduct and require less time and financial resources than other studies higher in the hierarchy, hence their popularity. In most cases they are the only practical way to determine the effectiveness of surgical procedures because of the impracticality of conducting a RCT, for example comparing a surgical procedure with no (sham) procedure.

Case series can either be prospective in which patients are followed through their treatment while outcomes are recorded or retrospective in which archived records of patient outcomes are correlated with their treatment. They may describe the outcomes of a particular method of treatment or of complications related to treatment for example.

Case series should be reported scientifically based on a protocol with a hypothesis. They are of value by acting to generate hypothesis that can be tested using other studies higher in the hierarchy of evidence.

This study type can be identified by not having controls and either follows consecutive patients through their treatment prospectively or looks back at their treatment retrospectively using patient records. There is no MeSH term for case series.

Checklist for Case Series

Introduction

Is there a clear study aim or research question (this may be stated as a null hypothesis)?

Methods

Is the study prospective or retrospective? The former is less liable to recall bias.

Are the inclusion/exclusion criteria there?

What was the time interval for recruitment? This should be over a short time as possible to minimize the effect of changes in care. For example pre and post-operative care may change over time even though the surgical procedure is the same.

There should be consecutive patient enrollment to avoid selection bias.

Full details of the intervention(s) used.

Details of outcome measure(s) used – were they validated ones?

Details of statistical methods and were they appropriate for that data type?

Results

Demographics analysis such as age and sex included often in a table?

Look for a high follow-up rate. This avoids selection bias.

Discussion

Are the strengths and weaknesses of the study listed? How do the authors interpret and discuss their results in relation to those of related studies. Are the conclusions justified considering the inherent limitations of this type of study?

Cross-Sectional Studies

These are commonly called surveys and measure the prevalence of a problem at one particular period in time, which may be a single day or the time period it takes to collect the data. Surveys take a sample of the population and the results should be representative of the wider population providing the survey has been carried out correctly. The way the sample is obtained is crucial to the validity of the results. Look for the MeSH term "cross-sectional".

Checklist for Cross-Sectional Studies

Introduction

Is the reason why the survey is being carried out clearly stated?

Are there any hypotheses that the survey will answer? There should be clear questions about what the survey hopes to answer. This will avoid data dredging during the analysis phase.

Methods

Is the sampling frame (i.e. population from which sample is taken) and sample described? This is important to minimize selection/sampling bias.

How was the sample selected? Were patients selected from a register and if so, how complete was it? Ideally the register should contain the majority of patients who have undergone that procedure or with that disease. For example if the trust/hospital records for that region were used, it is important that efforts were made to check on the completeness and validity of the data.

Was a sample size calculation included? This is important and although for surveys it is a relatively crude calculation – it should be included. It is usually calculated on the basis of an expected minimum response rate (e.g. 70 %) and power to detect a yes/no response to questions of 50 %. This ensures that negative results to questions will not be due to an underpowered study.

Was the method used to obtain the sample from the larger population (e.g. a register) described – was some form of random selection used (e.g. using a random number table/generator). This means a probability sample has been used. If a convenience sample has been used then it is unlikely the results can be relied upon. For example interviewing people who only attend a private clinic will recruit atypical patients.

Surveys are usually carried out using questionnaires which may be self-administered or by an interviewer. Is the questionnaire a validated one? If not was the development of

the questionnaire described and how was it tested for reliability and validity. Was it piloted on a small sample first? If physical measurements were made e.g. muscle strength, was the technique standardized among investigators?

Results

What was the response rate? It should be at least 70 %. Lower than this suggest the results cannot be relied on.

Demographics of both responders and non-responders should have been compared to determine if selection bias has occurred e.g. non-responders may be of lower socioeconomic status. If so, this should be acknowledged in the discussion on how this may have affected the results. Was a sensitivity analysis performed in order to determine the effect on the results?

Surveys are prone to data dredging; performing numerous statistical tests to see if anything interesting drops out. Look out for this.

Confidence intervals in addition to P values should be included for all statistical tests performed. The CI gives an idea of the range in the general population so is more informative than a P value.

Discussion and Conclusion

Are the weaknesses of the study discussed e.g. possible selection bias? How do the results compare with previous studies? If similar, this instills confidence in the study.

Case Control Study

A case control study is a study that starts from the outcome (disease) and looks backwards to see what may have caused this. Patients with a disease (the cases) are compared to those without the condition (the controls) but are as similar as possible to the cases (e.g. age, sex, i.e. are matched) in order to determine what (exposure) may have caused the disease. It attempts to elucidate a potential cause from observing an effect (the disease).

Case control studies are appropriate when the disease has a long latency period because the study looks backwards making it unnecessary to wait for the disease to develop. It is also suitable for rare diseases because the investigator selects patients on the basis of their disease status rather than having to follow a large number of people and wait for the disease to develop as in cohort studies. It is also suitable for investigating outbreaks e.g. infections as it enables a quick answer to be found. It cannot be used for looking at the possible causes of multiple diseases – for this a cohort study should be used.

As with other study types, case control studies are susceptible to bias, the main ones of which are selection and recall bias.

Selection bias can arise if the diagnostic criteria used to identify the cases are not precisely defined. Selection bias can also arise if the corresponding controls are not representative and dissimilar to the cases. These must be selected from a similar population as the cases. For example hospital cases should have hospital controls (i.e. patients with other illness) and be similar in age, sex, socioeconomic status and other possible confounding variables.

In this type of study patients often have to recall past events from memory such as medication use or dietary habits or the data can be obtained from records. Both these sources of information may be inaccurate or incomplete leading to recall bias.

To identify this type of study look for the MeSH term "case-control study" in the keywords if it is not explicitly stated in the title or introduction. Be aware though that case-control study is used loosely in orthopedics and what is stated as a case-control study is actually only comparing a case series with a few unmatched controls. It is really only a case series study.

Checklist for Case Control Studies

Introduction

Again as with all epidemiological studies a hypothesis or aim of the study should be stated here.

Was a case control study appropriate? Or maybe a RCT would have been better?

Methods

There should be a sample size calculation. This should be based on the minimum odds ratio (OR) to be detected. For example an OR of 1.5 means the cases are 1.5 times more likely to develop the disease than controls.

Selection of Cases

The diagnostic criteria for selecting the cases should be precisely defined, for example the stage or grading of the illness.

Were the majority of the cases who were contacted included in the study? Otherwise bias may arise because patient's who do not consent to inclusion tend to differ from those who do. So 90 % inclusion should be the minimum.

Selection of Controls

Were the controls selected from a similar population as the cases e.g. if hospital cases then the controls should be hospital patients with unrelated disease.

Were they similar in e.g. age, sex and time of hospitalisation …etc. (matched) to the cases except for not having the outcome? This will be given in the tables in the results section so look these over to determine this.

How was the exposure identified, from records or interviews/questionnaires? Interviewers/data collectors should be

blinded to patient status (i.e. case or control). This eliminates interviewer bias in which interviews or record searches may be influenced by knowledge of whether a patient is case or control.

Results

Are the demographics of cases and controls included? Also demographics of those cases and controls that refused to participate, where known, should be given in order to determine if selection bias had occurred. Data dredging is indicated if a large number of possible causative agents have been tested for significance and the same problem of chance significance occurs as it does with cross sectional studies.

The results should be expressed as odds ratio (OR). This compares the odds of the disease/outcome occurring in the cases and controls. A CI for the OR should be included. The tested association is not significant if the CI includes 1.0.

Cohort Studies

In epidemiology, this is a group of people with a common characteristic e.g. exposure, followed up over time to determine what illness happens to them because of this.

There are 2 ways of conducting a cohort study. Prospectively, in which study participants e.g. patients who have had metal implants, are followed up over time and observed as to whether the outcome e.g. cancer, occurs. This rate can be compared to the known rate in a similar general population (the control group). In retrospective cohort studies exposure and outcome has already occurred. Therefore, from medical records, patients with metal implants would be identified and if they developed cancer, again determined from records e.g. the cancer registry and whether this incidence was higher than in a similar population (e.g. matched for age, sex etc). Often a prospective study is carried out after a retrospective study has provided evidence of a link between exposure and outcome.

The main bias that cohort studies are susceptible to is loss to follow-up and this can severely affect the validity of the results. In any cohort study whether retrospective or prospective, it is necessary to exhaustively trace all patients e.g. from records or by following all members of the cohort from point of exposure to development of the outcome disease. This needs to be done for all members otherwise the results could be invalid. This can be difficult as cohort members may migrate, die or decide not to continue participation in the study.

To identify this type of study, look in the keywords for the MeSH term “cohort study”. Again beware that cohort study is used loosely in the orthopedic literature to mean a case series with follow-up either prospectively or retrospectively.

Checklist for Cohort Studies

Introduction

There should be a clear hypothesis either as the null hypothesis or as a statement of the aim of the study e.g. in patients who had a prosthesis made of metal (the cohort) is there an increased risk of cancer (the outcome) compared to the general population (the control group).

Was a cohort study appropriate or would a case control have been better? Case control studies provide much greater power than cohort studies to detect differences and thus need fewer participants. Would a RCT have been better if this was ethically possible, for example comparing non-metal vs. metal implant?

Methods

A sample size calculation should be given so that the study has sufficient power to detect the outcome of interest. The sample size depends on the incidence in the non-exposed population and the minimum relative risk (RR) regarded as important.

Were the data sources (e.g. registers) complete and accurate so that they can be relied on? Was data cleaning and checks for the correct diagnostic coding done? When using death certificates the recorded cause of death may hide the outcome of interest.

Were the appropriate data sources for assessing the outcome used? For cancer or death this will be less of a problem but for other outcomes such as juvenile arthritis a variety of data sources will have to be used e.g. GP records, hospital admissions.

Was the comparison (control) group of non-exposed individuals appropriate? Often the control group can be the general population providing the demographics are similar.

Was the follow-up time long enough for the outcome to develop? For cancer this will be many years. For treatment side effects this may only be months.

Results

The major bias in cohort studies will be due to loss to follow-up. Were the majority of participants followed up with every effort made to contact all patients using multiple sources? A follow up of less than 70 % is liable to have seriously affected the validity of the results. Where there was loss to follow-up, was a sensitivity analysis performed? For example, assuming those lost to follow-up all developed the outcome – the effect on the results can be determined.

Discussion and Conclusion

Is there a discussion of the limitations of the study, particularly the comparison group used and how they may differ from the cohort?

Randomized Controlled Trials

These are high in the hierarchy of the evidence pyramid and if done properly will provide results that are free of all bias. RCTs are essentially experiments in which patients are allocated to a treatment by the investigator. There are a variety of trial designs but the most common in therapeutic interventions is the parallel 2-arm trial in which patients are allocated to one of 2 treatments or to a placebo. The essence of RCTs is random assignment of patients to the intervention. This has the effect of balancing out all known and unknown confounders. However just because a study is randomized does not mean it is unbiased. If they lack methodological rigor, a variety of types of bias such as selection or loss to follow-up can creep in.

The CONSORT statement was published in 1996 with an update in 2010 [2] in order to improve the reporting of RCTs and prevent bias during trial conduct. It includes a checklist of points that should be reported so that it is possible to determine to what extent the results are valid. Although CONSORT was designed to improve the reporting of trials and was mainly developed for assessment of pharmacological treatments, it can be used as a CA tool for all types of RCTs. The checklist presented below is based on CONSORT together with that of the Cochrane bone, joint and muscle trauma group [3].

In general the quality of reporting of orthopedic trials is poor [4] so it is important to be critical when appraising them. Conducting surgical orthopedic trials presents special problems such as with blinding of treatments and surgeon learning curve when comparing a novel treatment to the traditional method for example. This is particularly true for multi-center trials.

Although randomisation should eliminate bias due to selection (allocation) and confounding, it is possible during the trial for other types of bias to creep in. In orthopedic trials, the possible sources of bias are due to lack of blinding, loss to follow-up, learning curve and surgeon expertise bias, bias due to patient crossover (when patients have a preference for one treatment over another e.g. surgery vs. conservative therapy) or patients not included in a study due to their severe illness or old age. In addition surgical orthopedic interventions are complex and usually consist of several components such as pre-treatment, anesthesia and rehabilitation such as physiotherapy. These may not be standardized between the groups and/or the participating centers in the trial thereby introducing further bias. These points need special attention when an orthopedic trial is critically appraised in addition to those that generally apply such as the details of randomisation.

Because of the logistical and practical difficulties of conducting orthopedic surgical trials, they are less commonly reported than in other fields such as drug treatments.

RCTs are easily identified from the title and MeSH keywords but it is important to be aware that not all reported trials have used randomization but are "quasi-randomized" or "stratified". This uses, for example, date of birth or medical record number, to allocate patients to the intervention and as such are prone to selection bias and therefore confounding.

Checklist for RCTs

Introduction

A clear hypothesis should be stated preferably as a null hypothesis.

Methods

The following should be provided:

Type of trial such as parallel with 2 arms and equal allocation to each group.

Trial setting, location and time period.

A detailed description of inclusion/exclusion criteria.

A sample size calculation including power of the study to detect a difference in the primary outcome. This is very important. Otherwise the study may not have had sufficient power to detect a difference in outcome where one actually exists.

Description of primary and secondary outcomes and details of how they were measured e.g. with questionnaires.

Details of the minimum clinically significant change in outcome measures. This is required for the sample size calculation. Were these outcome measures validated and clinically relevant for this study or would alternative ones have been more appropriate?

How was randomisation achieved? This is important and should always be included. For example, use of a random number table with the numbers concealed in consecutive envelopes or a telephone randomisation service. Use of any form of quasi-randomisation or stratification puts the validity of the whole study in doubt.

Were participants blinded to their assignment status? This is not always possible in orthopedic trials.

Were the treatment providers blinded? The surgeon, nursing staff, the outcome assessors and data analyst ideally should all be blinded as to allocation of patients. In surgical trials it almost impossible for the surgeon to be blinded, but it is possible for the nurses, outcome assessor and data analyst to be.

Was there a description of how patients in the groups were cared for before (e.g. pre-operative education) and after the intervention (e.g. rehabilitation regimes) and were these identical for the trial arms?

Was follow up of sufficient duration to measure the outcome?

Were the intervention procedure(s) described and details of surgical expertise for the procedure(s) given?

There should be full details of statistical analysis e.g. checking for normality of the data and the tests that were used.

Results

Look for a chart showing the flow of participants through the trial. This should include numbers allocated to each intervention and any loss to follow-up with reasons and numbers analyzed. Check all patients have been accounted for in the flow chart.

Were baseline characteristics of the groups given and are they comparable? Although randomisation should result in the groups being very similar at the start of the study, this is not guaranteed so it is best to check they were. Possible confounders are age, sex, acute/chronic condition.

Were results analyzed by intention to treat (ITT)? Patients may change treatments during the trial or withdraw part way through. Even though this may seem illogical, they should be included in the analyses under the original arm they were randomized to. Failure to follow this rule can invalidate the randomization process and thereby introduce bias.

Were there any side effects that were different between the groups? This could affect the outcome of one group and negate any beneficial effects.

Systematic Reviews and Meta-Analysis of RCTs

These are at the top of the levels of evidence hierarchy and therefore should have least bias.

As the name suggests, a systematic review is simply a review of the literature, which may be observational studies or trials, that has been conducted in a systematic way by following an organized protocol usually that of the Cochrane collaboration [5].

Meta–analysis goes one step further and uses statistical methods to combine the results of selected studies that meet strict inclusion criteria in order to give an overall result that is an "average" of the individual results of the included studies. The general aim of meta-analysis is to increase the overall power by using results from what may be underpowered studies.

Not all systematic reviews include a meta-analysis, for example if the reviewed studies are of insufficient quality or few in number, but a meta-analysis cannot be done without an initial systemic review. Meta-analysis has become increasingly common in the orthopedic literature, as the quest for EBM has intensified. In addition, reviews have become an important way of keeping up to date with research output in orthopedics without having to read every new article published.

In orthopedics, meta-analysis plays an important part. They are conducted where there is doubt about alternative procedures for example minimally invasive vs. standard methods in surgery. Individual research papers may provide an inconclusive answer often due to their small sample sizes.

Systematic review and meta-analysis can be performed for all study types not just RCTs. In orthopedics they are increasingly being done on observational studies such as case series. Most commonly though they combine the outcomes of RCTs and this is what we will concentrate on. These are generally conducted according to the guidelines in the Cochrane handbook for systematic reviews of Interventions [5] and reported according to the PRISMA statement (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) [6].

The most important thing in a meta-analysis is that apples are not combined with oranges, in other words, the trials combined are of high quality, have a similar hypothesis and the results reported are generally similar i.e. the studies are homogenous (see later). The aim of PRISMA is to ensure that only similar high quality studies are combined so that the conclusion will be valid.

Publication bias can be an important source of bias in meta-analyses. Unpublished studies (e.g. internal reports from companies, dissertations) may report different findings from published ones so it is important to exhaustively search the literature. Often RCTs that show no effect are less likely to be published, again leading to publication bias. There are ways of detecting this bias either using a graphical method (funnel plot) or with statistical tests (see later).

Once the research question has been formulated, the procedure of performing a meta-analysis essentially involves:

- Carry out an exhaustive literature research.
- Select those studies that meet the study selection criteria.
- Critically appraise the methodological quality of included studies.
- Extract the outcome data from included studies.
- Determine publication bias using a funnel plot if there are sufficient studies.
- Perform the meta-analysis for example using Review Manager (RevMan) software. A simplified description of this can be found at: http://www.mtm.uoi.gr/PracticalRevMan.pdf
- Present the data using forest plots.

These steps form the basis for the critical appraisal of a meta-analysis.

Meta-analyses are easily identified as they are always stated in the title.

Checklist for Systematic Reviews and Meta-Analysis of RCTs

Introduction

This should include why the analysis is being done especially if there has been a previously published one with inconclusive results. Often as more trials on a particular subject accumulate then there is justification for performing a

new meta-analysis to see if earlier conclusions have changed. It should be stated in the methods section that a search was made for previous similar meta-analyses on the subject.

Methods

How was the analysis conducted and reported, e.g. using Cochrane and PRISMA guidelines?

Are there details of the literature search? This should include the search strategy, bibliographic databases searched (e.g. Medline, Embase) and the MeSH terms used. Were efforts made to find unpublished and ongoing studies e.g. by searching the current controlled trials register? Also look in relevant orthopedic journals for articles "published ahead of print" – these are available on the journal website and may have not yet have been indexed in the bibliographic databases. Were efforts made to search the grey literature i.e. unpublished reports, of limited (in-house) distribution and that are not included in bibliographic databases e.g. dissertations, literature from health departments? Also abstracts presented at conferences may give a clue to work that is in progress and that may be published soon. The important thing is that an exhaustive search was made to locate all relevant material. If there is an acknowledgement of librarian help this should instill confidence in the search.

Were the study inclusion criteria given? This is needed for selecting relevant articles from the literature search for further detailed appraisal and suitability for inclusion. This should include types of patients e.g. adolescents undergoing treatment for idiopathic scoliosis, types of intervention e.g. bracing vs. surgery and types of outcome measures e.g. duration of surgery, questionnaires used. This step results in a large number of abstracts that will need to be filtered for possible inclusion in the analysis.

Once the abstracts have been reviewed for inclusion, full text articles of those papers for possible final inclusion will need to be assessed for methodological quality. Were their details of how this was done? What basis was a study deemed to be fit to be included in the analysis e.g. was each paper given a numerical score? This can be done with a formal checklist as described in the critical appraisal of RCTs e.g. that of the Cochrane bone joint and muscle trauma group [3] mentioned earlier.

There should be details of the statistical analysis and the software used e.g. RevMan (see above). Sometimes data e.g. standard deviations, that are needed for the meta-analysis are missing from a paper. In this case did the authors attempt to contact the authors or calculate it from other data (e.g. the range) given in the results?

Results

A description of the number of studies identified, the number excluded and the number of full text articles selected should be described. Ideally there should be a flow chart of the whole process from record identification to the number of studies included.

Details of all included studies together with the data on the selected outcome measures for each study should be presented in a table.

Was a funnel plot or a statistical test included to determine if publication bias was present? This is a graph that plots the size of the treatment effect against the standard error (SE). This is normally plotted for the most frequently cited outcome measure (e.g. duration of surgery) when there are multiple outcomes measures. In the absence of publication bias, this plot should be funnel shaped with the neck of the funnel at the top of the graph and the included trials approximately symmetrical about the zero point of the x-axis. This plot is only relevant when there are about 10 or more studies.

The results of a meta-analysis are expressed graphically as a forest plot with an accompanying table describing the statistics of the outcome in detail for the treatment groups. There should be a forest plot for each outcome. For a meta-analysis with a large number of outcomes not all forest plots will be included due to editorial space limitations. The plot (Fig. 2.2 shows an example) essentially shows the value of the treatment effect for each study as a square, the size of which depends on the statistical weighting given to the study

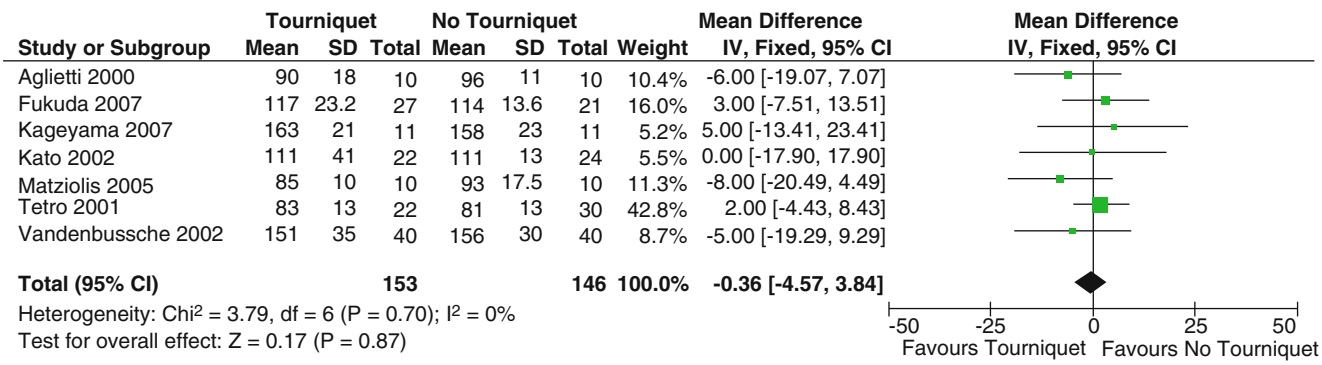

Fig. 2.2 An example forest plot of a meta-analysis of randomized controlled trials for total knee arthroplasty with and without a tourniquet and outcome of duration of surgery [7]. *Weighted mean difference: −0.36: 95 % CI: 4.6 to 3.8; n = 299; P = 0.87*

together with its CI as a horizontal line. A diamond summarizes the treatment overall for all the studies and whether it favors one treatment over the other. Its lateral points are its CI. The vertical line corresponds to no effect.

The treatment effect should be expressed as mean differences (MD) or odds ratio (OR) for continuous data e.g. duration of surgery and risk difference (RD) for dichotomous data e.g. number of complications. Look for the position of the diamond shape on the plot and check its position as to whether it favors one treatment or the other and that this correlates with its value and CI.

Are the included studies homogenous? Statistical heterogeneity arises from differences in socioeconomic factors such as age or gender, disease factors such as severity and duration and completeness of follow-up between the different studies. This can be assessed using the result of the chi-square test (also called Cochrane Q test) that will be given in the forest plot's accompanying table. A p value of <0.1 (rather than the usual 0.05) is considered suggestive of heterogeneity. Whenever heterogeneity is present, the analysis for that outcome may be invalid.

Because of the complex statistical methods involved in meta-analysis, it is important that all data and the forest plots are carefully checked over before the results are accepted as valid. This also applies to those studies included in the analysis – it may be that the authors have omitted a study that you are aware off.

A meta-analysis is a major project and it is easy for mistakes to creep in, so it is important that the conclusions are interpreted with caution.

Discussion

Did the authors mention any shortcomings in their analysis that may affect the validity of their conclusions? There may language bias if the literature search included English language studies only. Were there any deficiencies in the detailed aspects of included studies (e.g. randomization, blinding, variations in operative technique)? How did the findings compare with any previous meta-analyses on the subject? If they disagree, has an explanation been given? It may be that more recent studies are methodologically more rigorous than those in a previous meta-analysis.

What are the strengths? Was the aim of the analyses achieved e.g. for or against a procedure? Did the authors give a recommendation from their results so that an informed decision as to whether to change practice can be made?

This chapter has hopefully provided the tools that you need for critical appraisal of published work. It should also be of help when you conduct and publish your own research by ensuring that the checklist questions for your study type are taken into account during the design phase.

References

1. Chan K, Bhandari M. Three-minute critical appraisal of a case series article. Indian J Orthop. 2011;45(2):103–4.
2. Schulz KF, Altman DG, Moher D, for the CONSORT Group. CONSORT 2010 Statement: updated guidelines for reporting parallel group randomised trials. Ann Int Med. 2010:152.
3. Cochrane bone, joint and muscle trauma group. http://bjmtg.cochrane.org. Accessed 8 Mar 2013.
4. Chaudhry H, Mundi R, Singh I, Einhorn TA, Bhandari M. How good is the orthopaedic literature? Indian J Orthop. 2008;42(2):144–9.
5. Higgins JPT, Green S. Cochrane handbook for systematic reviews of interventions. The Cochrane Library. Chichester: John Wiley & Sons Ltd; 2008.
6. Moher D, Liberati A, Tetzlaff J, Altman DG, The PRISMA Group. Preferred reporting items for systematic reviews and meta-analyses: the PRISMA statement. PLoS Med. 2009;6(7).
7. Alcelik I, Pollock R, Sukeik M, Bettany-Saltikov J, Armstrong PM, Fismer P. A comparison of outcomes with and without a tourniquet in total knee arthroplasty: a systematic review and meta-analysis of randomised controlled trials. J Arthroplasty. 2012;27:331–40.

Part II

The Hip

Clinical Surveillance, Selective or Universal Ultrasound Screening in Developmental Dysplasia of the Hip

3

Jonathan Wright and Deborah M. Eastwood

Abstract

Early detection of the child with developmental dysplasia of the hip (DDH) allows treatment to be commenced promptly, with potentially fewer requirements for surgical intervention and a better final outcome. Clinical examination has been the mainstay of early detection but the introduction of ultrasound examination provided the possibility for greater sensitivity. The ideal process for detection is still a subject of debate, with surveillance practices varying both between and within countries. Most screening programmes attempt to find a balance between missing a dislocated hip and overtreatment of ultrasonographic findings. This chapter considers the evidence related to screening programmes for DDH and looks at the efficacy of each method of detection, the risks of a screening programme and the question of whether late presentation of developmental dysplasia is a preventable occurrence.

Keywords

Developmental dysplasia of the hip • Congenital dislocation of the hip • Screening • Surveillance • Clinical examination • Selective or universal ultrasound

Introduction

Developmental dysplasia of the hip (DDH) represents a spectrum of disorders in which the normal relationship between the femoral head and the acetabulum is disturbed, leading to abnormal development of the joint. This can range from mild dysplasia to complete dislocation of the hip. Persistent subluxation and dysplasia is associated with an increased risk of early osteoarthritis [1, 2].

Neonatal hip instability is common and it is assumed that many such cases are associated with hip dysplasia. Clinical and ultrasound assessments of the infant hip have differing abilities to detect dysplasia and/or instability and the sensitivity of the examination changes according to its timing. Most neonatal instability is physiological with spontaneous resolution by 6–8 weeks of age [3].

The reported incidence of DDH varies, not only through well-documented geographic and racial backgrounds [4–6], but also through the means of detection. Use of ultrasound facilitates detection of immaturity related "dysplasia" which, similar to clinical neonatal instability, may resolve without treatment and lead to normal or near normal hip development [7]. Within a northern European population the incidence of hip dysplasia has been estimated as between 120/1000 live births [8–11]. The incidence of a disease within a population is essential information when developing a screening test, as for a rare disease, even with high levels of specificity, a number of false positive results will be expected.

The basic principle of treatment for DDH is that placement and maintenance of the femoral head within the acetabulum restores the normal conforming forces, which will encourage normal joint development. As the remodelling

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potential of the acetabulum is greatest in the early months of life [12, 13], timing of any intervention is important. The aim of early detection is to commence simple treatment such as the Pavlik harness [14] at an appropriately early time, avoiding the increased risks of surgical intervention with later diagnosis [15].

Clinical Examination

It is a legal requirement in the UK that all newborns are examined according to the NIPE protocol [16]. Clinical examination is based on the Ortolani [17] and Barlow manoeuvres [3] and aims to detect instability by asking the following questions.

1. Is the hip dislocated at rest?
   (a) If so, is it reducible?
   (b) If it reduces, it is Ortolani positive, if it does not do so, it is Ortolani negative.

If it is not dislocated then:

2. Is the hip dislocatable/subluxatable at rest?
   (a) If so, it is Barlow positive

If it is not dislocatable then:

3. Is the hip clinically normal?
   (a) If so, are there sufficient risk factors in the history that mandate further assessment of the hip despite the negative clinical findings?

Both the Barlow and Ortolani tests become less sensitive in the older infant: it becomes more difficult to detect instability (in an otherwise neurologically normal child) and the dislocated hip is now often irreducible (Ortolani negative). Over the age of 2–3 months, examination includes assessment of leg length discrepancy, asymmetry of thigh creases (unreliable [18]) and most importantly limitation and/or asymmetry of abduction in flexion [19, 20].

Ultrasound Examination

A static demonstration of hip joint anatomy uses the 'standard plane' described by Graf [21] whilst a dynamic assessment will demonstrate joint instability [22, 23]: a combination of both methods is often helpful [23]. Both techniques may identify hip abnormalities that would not have been detected on clinical examination alone. The management of minor degrees of instability and/or anatomical dysplasia remains controversial [24]. Dynamic ultrasound assessment could be considered an extension of the clinical examination for instability.

Table 3.1 Value of the current DDH screening programmes

Has the presence of a screening programme improved outcomes for patients with DDH?
How effective is clinical examination in the early detection of DDH and who should be performing the clinical examination?
What is the effect of a universal ultrasound surveillance programme on the detection of DDH?
Is selective ultrasound surveillance as effective as universal at detection of DDH?
Who is at highest risk of DDH and who is at risk of being missed?
Can late presentation of DDH be prevented?

Screening or Surveillance?

The WHO criteria for a screening programme were described by Wilson & Jungner [25] in 1968 but the current screening systems for the early detection of DDH only fulfil some of these criteria. The condition is "important" with an accepted treatment and suitable facilities available for that treatment. There is an early stage in which DDH can be picked up, with the test (both clinical and ultrasound) being acceptable to the population. However, there is no universal agreement as to what is the best screening test. The natural history is not fully understood, and thus there is no universal agreement on who should receive treatment and who should not. Finally, the cost of the screening programme in comparison to the full costs of delayed detection (including medicolegal costs) has not been established for any individual country.

The severity of DDH can change over time with come cases improving and others deteriorating and the serial examinations required to assess this would be better described as "a surveillance process", rather than a true screening test. In each case the examination findings must be related to the clinical history.

There are many questions to be asked in relation to the value of the current DDH screening programmes and the following discussion aims to address the points outlined in Table 3.1.

Has the Presence of a Screening Programme Improved Outcomes for Patients with DDH?

There is no evidence that the current or historical screening programmes for DDH have improved the functional outcomes of adult patients with DDH partly because there has never been a clear gold standard definition of what constitutes a case of DDH.

Rates of surgery have been used as a surrogate endpoint for the effectiveness of a screening programme with the assumption that early diagnosis and treatment of DDH will

allow stabilisation through conservative measures, reducing the requirement for surgical management at a later date. However, as the indications for surgical intervention are not universally agreed, results may be affected by changing thresholds for surgery over a period of time, or in different geographic regions.

Similarly, the incidence of "late presenting" DDH in a screened population has been used as an endpoint, where late presentation represents a failure of screening: again, there is significant variability in the definition of 'late' and in how long the follow up is. Observational studies, assume a "captive population" or at least one in which children moving into the geographic area are of similar number and have the same rates of late presentation to those leaving the area.

Finally, rates of splintage/treatment have been used to judge the success of surveillance programmes based on the assumption that an increased splintage rate should be reflected in a subsequent decrease in the rates of late presenting DDH. Historically, some programmes saw an increased treatment rate with little reduction in late presentation [26, 27], raising concerns about a high false positive rate and over treatment, with a potential risk of resultant AVN. More recent trials have gone on to demonstrate the opposite [28, 29], perhaps reflecting improved implementation of screening programmes.

How Effective Is Clinical Examination Alone in the Early Detection of DDH and Who Should Be Performing the Clinical Examination?

There are no studies directly comparing an unscreened cohort with that of a cohort undergoing a clinical screening programme alone. Table 3.2 lists a number of observational studies that have reported the rates of late presenting DDH in different patient populations. These studies suggest that the effectiveness of a clinical screening programme is improved when the person performing the examination has experience. Hadlow [30] demonstrated reductions in the rates of late presentation of DDH when a Consultant Orthopaedic surgeon, rather than a junior paediatrician performed clinical examination. These rates remained low when experienced paediatricians performed the examinations in subsequent years. This work was supported by similar studies from Krikler [31] and Tegnander et al. [32]. Interestingly, Moore hypothesised that the act of examination itself may cause instability through forceful or repeated attempts to dislocate an otherwise stable hip [33]. There was no evidence in this paper (and none since) to support this hypothesis.

What Is the Effect of a Universal Ultrasound Surveillance Programme on the Detection of DDH?

The use of ultrasound as part of surveillance for DDH is well established, with certain centres electing to use universal surveillance of newborns. A number of observational studies have compared the introduction of universal surveillance to that of historic controls in order to estimate the effect on rates of late presentation and splintage [27, 28, 37–40]. These demonstrate a variable increase in splintage rates, with improvement in the rates of late presenting DDH, with one study stating that no cases have presented late since the introduction of universal screening [39]. There are presently only two controlled trials (level II evidence) comparing the techniques directly.

Two arms of the 1994 Rosendahl et al. [41] controlled trial compared clinical examination alone to clinical examination and universal ultrasound in neonates. The study reported increased rates of splintage in the universal ultrasound group without a statistically significant reduction in the rates of late diagnosed DDH (Table 3.3). **Even with the large numbers used in this study, due to the low rates of late presentation, later power calculations suggested that the original study was underpowered to demonstrate significance in late diagnosis [42].

In 2002, Elbourne [29] compared the use of ultrasound vs serial clinical examination in patients with clinical instability

Table 3.2 Summary of late presentation rates for clinical screening programmes

Authors	Study type	Type of screening	Patients, *n*	Results
MacKenzie and Wilson [34]	Observational	Clinical by physician	53,033	Late presentation rate: 1.1/1000
Dunn et al. [35]	Observational	Clinical by junior paediatrician	103,431	Late presentation rate: 0.88/1000
Macnicol [36]	Observational	Clinical by junior paediatrician	117,256	Late presentation rate: 0.5/1000
Hadlow [30]	Observational	Clinical by experienced orthopaedic surgeon	20,657	0.2/1000
Krikler and Dwyer [31]	Cohort study	Clinical by junior paediatrician *or* experienced physiotherapist	68,861	0.35/1000 0.1/1000
Tegnander et al. [32]	Cohort study	Clinical by experienced paediatrician *or* inexperienced clinician	363,508	2.6/1000 5.3/1000

Table 3.3 Summary of studies comparing clinical examination alone with clinical examination supplemented by universal ultrasound screening

Author	Study type	Clinical examination only				Universal ultrasound				Conclusions
		n	Splinting rate, %	Late diagnosis	Surgical procedures	*n*	Splinting rate, %	Late diagnosis	Surgical procedures	
Elbourne et al. [22]	RCT of children with clinically abnormal hip exam	315	48	n/a	7.9 %	314	37	n/a	6.7 %	Lower rates of splintage in US group. No difference in rates of surgical procedures
Rosendahl et al. [41]	RCT of unselected newborns	3924	1.8	2.6/1000	1.2/1000 (5 h ips)	3618	3.4	1.4/1000	0.3/1000 (1 hip)	Higher splintage rate in US group. Lower rates of late diagnosis (NS)
Roovers et al. [40]	Cohort study of unselected newborns	5170	2.7	8/1000	3/1000	2066	4.7	6/1000	1/1000	Higher splintage rate. Lower rates of late diagnosis/surgery

as part of a multicentre randomised controlled trial. The study demonstrated lower rates of splintage in hips, which were initially clinically abnormal, when ultrasound was used as part of the assessment. There was no difference seen in later surgery rates, suggesting that the ultrasound group was not undertreated.

Is Selective Ultrasound Surveillance as Effective as Universal at Detection of DDH?

Ultrasound assessment programmes can either be universal or selective with the latter aiming to identify those patients at highest risk of DDH. Risk factors for DDH have been well described, with a recent meta-analysis demonstrating a significant increase in relative risk for female infants, breech presentation, first born and those with a positive family history [43]. Historically, a foot deformity has often been considered to have an association with DDH, but the meta-analysis in 2012 showed this did not reach statistical significance [44]. Selective screening programmes reserve ultrasound examination for those infants who have an abnormal clinical examination and/or one or more risk factors. The most frequently used risk factors are a positive family history, breech positioning and foot deformities [41, 45–49]: female gender is not considered a risk factor for screening programmes.

Two large controlled trials (Table 3.4) have compared selective ultrasound screening with universal ultrasound [41, 48]. Neither demonstrated statistically significant differences in the rates of late diagnosis between the two patient groups. In Holen's study [48], 5 patients were diagnosed late in the selective group (all without risk factors) and one patient from the universal group (where the protocol was not adhered to and no ultrasound actually took place). The authors of both trials [41, 48] emphasise that their ultrasound surveillance programmes supplemented a clinical programme where the examination was performed by an experienced physician, suggesting that their results are dependent on a combination of clinical and ultrasound assessments. Inexperienced clinical examination may lead to a loss of patients from the selective ultrasound screening group [31, 32] and a higher late diagnosis rate.

Who Is at Highest Risk of DDH and Who Is at Risk of Being Missed?

Three observational studies have considered the characteristics of late presenting DDH/dysplasia, which require intervention; all studies were carried out in areas with selective

Table 3.4 A comparison of selective and universal ultrasound programmes in patients who had already undergone an expert clinical examination

Author	Study type	Clinical examination + selective ultrasound				Clinical examination + Universal ultrasound				Conclusions
		n	Splinting rate, %	Late diagnosis	Surgical procedures	*n*	Splinting rate, %	Late diagnosis	Surgical procedures	
Holen et al. [48]	RCT of unselected newborns	7689	0.86	0.65/1000 (5 pts)	0.13/1000	7640	0.96	0.13/1000 (1 pt)	0.26/1000	No significant difference between groups
Rosendahl et al. [41]	RCT of unselected newborns	4388	2	2.1/1000	0.7/1000	3618	3.4	1.4/1000	0.3/1000	No significant difference between groups

ultrasound screening [47, 49, 50]. Sanghrajka et al. [50] analysed 65 consecutive open reductions, demonstrating that 88 % of late presentations did not have any risk factors to trigger ultrasound screening. The remaining 12 % had risk factors, which should have triggered a scan but the screening programme failed to identify them. The study infers that in order to detect these cases early, universal good quality ultrasound screening was required. Similarly, Azzopardi et al. [51] suggested that the selection criteria used in a screening programme affected the risk factors for late presenting DDH: those presenting late had an absence of risk factors.

Laborie et al. [52] reported the long term follow up of a group of patients involved in the Rosendahl et al. [41] controlled trial: they looked for residual dysplasia (due to under diagnosis) and avascular necrosis (as a sequela of the increased treatment rates associated with ultrasound screening). In these skeletally mature individuals, no significant dysplasia was identified and there was no statistical difference in the radiographic parameters used to define hip dysplasia between the cohorts screened by different methods. The patients reviewed represented only 17 % of the original cohort and thus with a condition of low incidence, the risk of a Type II error is high.

Sink et al. [53] reviewed 68 adults with symptomatic hip dysplasia, requiring operative intervention. In this group, 85 % did not have risk factors that would have triggered a scan in infancy, had they been born at the time of the study. The suggestion of the authors is that dysplasia in adulthood may be predicted by an abnormal ultrasound in infancy, although it is possible that dysplasia in adulthood may represent a disease process outside that which presents in infancy and can be detected by a screening programme.

Can Late Presentation of DDH Be Prevented?

Two papers have suggested that late presenting DDH can be prevented with universal ultrasound screening of captive/local populations. In their study over 5 years, Marks et al. [39] found no cases of late presentation of DDH following the introduction of universal ultrasound screening. Similarly, the only case of late diagnosed DDH in the universal arm of the controlled trial by Holen et al. [48] was due to a failure of the study protocol and no scan took place. Observational studies in countries where national universal ultrasound programmes are used [28, 54] have failed to completely eliminate late presentations, perhaps due to factors related to migration, and incomplete compliance with the protocol. These studies highlight the difficulties of 'rolling out' a good regional programme to a national level.

Table 3.5 Recommendations

Statement	Grade of evidence
DDH surveillance programmes in infancy improve functional outcomes in adulthood	I
For the clinical component of a surveillance programme to be effective, examination must be performed by practitioners experienced in the technique	B
Where a programme of high quality clinical examination is present, the trend is that fewer cases of DDH are missed as more ultrasound examinations are performed This does not reach statistical significance	C
In areas of selective ultrasound surveillance, patients without risk factors are at risk of late presentation Girls are at risk although gender is not considered a risk factor	C
Neither clinical nor ultrasonographic surveillance has been able to prevent late presenting DDH across a population	C

Conclusions

Several recent systematic reviews [46, 47, 55] have attempted to define the value of the differing surveillance programmes for DDH: each has been limited in their conclusions. There is a lack of high-level evidence that demonstrates an improvement in clinical outcomes and alternative, often heterogenous, endpoints such as late presentation have been used to monitor efficacy. The conclusions drawn must be made on the best evidence available, with the acceptance that sensible first principles must be applied (Table 3.5).

It is reassuring to see that the NIPE programme includes an emphasis on teaching and quality assurance to ensure that the clinical programmes are robust in terms of governance. Similar standards should also be applied to ultrasound programmes.

References

1. Murphy SB, Ganz R, Muller ME. The prognosis in untreated dysplasia of the hip. A study of radiographic factors that predict the outcome. J Bone Joint Surg Am. 1995;77–7:985–9.
2. Terjesen T. Residual hip dysplasia as a risk factor for osteoarthritis in 45 years follow-up of late-detected hip dislocation. J Child Orthop. 2011;5–6:425–31.
3. Barlow TG. Early diagnosis and treatment of congenital dislocation of the hip. J Bone Joint Surg Br. 1962;44–B–2:292–301.
4. Coleman SS. Congenital dysplasia of the hip in the Navajo infant. Clin Orthop Relat Res. 1968;56:179–93.
5. Edelstein J. Congenital dislocation of the hip in the Bantu. J Bone Joint Surg Br. 1966;48–7:257.
6. Walker JM. Congenital hip disease in a Cree-Ojibwa population: a retrospective study. Can Med Assoc J. 1977;116–5:501–4.
7. Tegnander A, Holen KJ, Terjesen T. The natural history of hip abnormalities detected by ultrasound in clinically normal newborns: a 6–8 year radiographic follow-up study of 93 children. Acta Orthop Scand. 1999;70–4:335–7.
8. Bialik V, Bialik GM, Blazer S, Sujov P, Wiener F, Berant M. Developmental dysplasia of the hip: a new approach to incidence. Pediatrics. 1999;103–1:93–9.
9. Kokavec M, Bialik V. Developmental dysplasia of the hip. Prevention and real incidence. Bratisl Lek Listy. 2007;108–6:251–4.
10. Loder RT, Skopelja EN. The epidemiology and demographics of hip dysplasia. ISRN Orthop. 2011;2011:238607.
11. Wynne-Davies R. The epidemiology of congenital dislocation of the hip. Dev Med Child Neurol. 1972;14–4:515–7.
12. Harris NH. Acetabular growth potential in congenital dislocation of the hip and some factors upon which it may depend. Clin Orthop Relat Res. 1976;119:99–106.
13. Lindstrom JR, Ponseti IV, Wenger DR. Acetabular development after reduction in congenital dislocation of the hip. J Bone Joint Surg Am. 1979;61–1:112–8.
14. Ramsey PL, Lasser S, MacEwen GD. Congenital dislocation of the hip. Use of the Pavlik harness in the child during the first six months of life. J Bone Joint Surg Am. 1976;58–7:1000–4.
15. Price KR, Dove R, Hunter JB. Current screening recommendations for developmental dysplasia of the hip may lead to an increase in open reduction. Bone Joint J. 2013;95–b–6:846–50.
16. NHS newborn and infant physical examination (NIPE) screening programme.
17. Ortolani M. Congenital hip dysplasia in the light of early and very early diagnosis. Clin Orthop Relat Res. 1976;119:6–10.
18. Ando M, Gotoh E. Significance of inguinal folds for diagnosis of congenital dislocation of the hip in infants aged three to four months. J Pediatr Orthop. 1990;10–3:331–4.
19. Roposch A, Liu LQ, Hefti F, Clarke NM, Wedge JH. Standardized diagnostic criteria for developmental dysplasia of the hip in early infancy. Clin Orthop Relat Res. 2011;469–12:3451–61.
20. Jari S, Paton RW, Srinivasan MS. Unilateral limitation of abduction of the hip. A valuable clinical sign for DDH? J Bone Joint Surg Br. 2002;84–1:104–7.
21. Graf R. Classification of hip joint dysplasia by means of sonography. Arch Orthop Trauma Surg. 1984;102–4:248–55.
22. Harcke HT, Clarke NM, Lee MS, Borns PF, MacEwen GD. Examination of the infant hip with real-time ultrasonography. J Ultrasound Med. 1984;3–3:131–7.
23. Clarke NM, Harcke HT, McHugh P, Lee MS, Borns PF, MacEwen GD. Real-time ultrasound in the diagnosis of congenital dislocation and dysplasia of the hip. J Bone Joint Surg Br. 1985;67–3:406–12.
24. Wood MK, Conboy V, Benson MK. Does early treatment by abduction splintage improve the development of dysplastic but stable neonatal hips? J Pediatr Orthop. 2000;20–3:302–5.
25. Wilson JM, Jungner YG. Principles and practice of mass screening for disease. Bol Oficina Sanit Panam. 1968;65–4:281–393.
26. Godward S, Dezateux C. Surgery for congenital dislocation of the hip in the UK as a measure of outcome of screening. MRC Working Party on Congenital Dislocation of the Hip. Medical Research Council. Lancet. 1998;351–9110:1149–52.
27. Clarke NM, Clegg J, Al-Chalabi AN. Ultrasound screening of hips at risk for CDH. Failure to reduce the incidence of late cases. J Bone Joint Surg Br. 1989;71–1:9–12.
28. Thallinger C, Pospischill R, Ganger R, Radler C, Krall C, Grill F. Long-term results of a nationwide general ultrasound screening system for developmental disorders of the hip: the Austrian hip screening program. J Child Orthop. 2014;8–1:3–10.
29. Elbourne D, Dezateux C, Arthur R, Clarke NM, Gray A, King A, Quinn A, Gardner F, Russell G. Group UKCHT. Ultrasonography in the diagnosis and management of developmental hip dysplasia (UK Hip Trial): clinical and economic results of a multicentre randomised controlled trial. Lancet. 2002;360–9350:2009–17.
30. Hadlow V. Neonatal screening for congenital dislocation of the hip. A prospective 21-year survey. J Bone Joint Surg Br. 1988;70–5:740–3.

31. Krikler SJ, Dwyer NS. Comparison of results of two approaches to hip screening in infants. J Bone Joint Surg Br. 1992;74–5:701–3.
32. Tegnander A, Terjesen T, Bredland T, Holen K. Incidence of late-diagnosed hip dysplasia after different screening methods in newborns. J Pediatr Orthop B. 1994;3:86–8.
33. Moore FH. Examining infants' hips–can it do harm? J Bone Joint Surg Br. 1989;71–1:4–5.
34. MacKenzie IG, Wilson JG. Problems encountered in the early diagnosis and management of congenital dislocation of the hip. J Bone Joint Surg Br. 1981;63–B–1:38–42.
35. Dunn PM, Evans RE, Thearle MJ, Griffiths HE, Witherow PJ. Congenital dislocation of the hip: early and late diagnosis and management compared. Arch Dis Child. 1985;60–5:407–14.
36. Macnicol MF. Results of a 25-year screening programme for neonatal hip instability. J Bone Joint Surg Br. 1990;72–6:1057–60.
37. Clegg J, Bache CE, Raut VV. Financial justification for routine ultrasound screening of the neonatal hip. J Bone Joint Surg Br. 1999;81–5:852–7.
38. Grill F, Muller D. Results of hip ultrasonographic screening in Austria. Orthopade. 1997;26–1:25–32.
39. Marks DS, Clegg J, al-Chalabi AN. Routine ultrasound screening for neonatal hip instability. Can it abolish late-presenting congenital dislocation of the hip? J Bone Joint Surg Br. 1994;76–4:534–8.
40. Roovers EA, Boere-Boonekamp MM, Castelein RM, Zielhuis GA, Kerkhoff TH. Effectiveness of ultrasound screening for developmental dysplasia of the hip. Arch Dis Child Fetal Neonatal Ed. 2005;90–1:F25–30.
41. Rosendahl K, Markestad T, Lie RT. Ultrasound screening for developmental dysplasia of the hip in the neonate: the effect on treatment rate and prevalence of late cases. Pediatrics. 1994;94–1:47–52.
42. Patel H, Canadian Task Force on Preventive Health C. Preventive health care, 2001 update: screening and management of developmental dysplasia of the hip in newborns. CMAJ. 2001;164–12:1669–77.
43. Ortiz-Neira CL, Paolucci EO, Donnon T. A meta-analysis of common risk factors associated with the diagnosis of developmental dysplasia of the hip in newborns. Eur J Radiol. 2012;81–3:e344–51.
44. de Hundt M, Vlemmix F, Bais JM, Hutton EK, de Groot CJ, Mol BW, Kok M. Risk factors for developmental dysplasia of the hip: a meta-analysis. Eur J Obstet Gynecol Reprod Biol. 2012;165–1:8–17.
45. Clarke NM, Reading IC, Corbin C, Taylor CC, Bochmann T. Twenty years experience of selective secondary ultrasound screening for congenital dislocation of the hip. Arch Dis Child. 2012;97–5:423–9.
46. Shipman SA, Helfand M, Moyer VA, Yawn BP. Screening for developmental dysplasia of the hip: a systematic literature review for the US Preventive Services Task Force. Pediatrics. 2006;117–3:e557–76.
47. Woolacott NF, Puhan MA, Steurer J, Kleijnen J. Ultrasonography in screening for developmental dysplasia of the hip in newborns: systematic review. BMJ. 2005;330–7505:1413.
48. Holen KJ, Tegnander A, Bredland T, Johansen OJ, Saether OD, Eik-Nes SH, Terjesen T. Universal or selective screening of the neonatal hip using ultrasound? A prospective, randomised trial of 15,529 newborn infants. J Bone Joint Surg Br. 2002;84–6:886–90.
49. Talbot CL, Paton RW. Screening of selected risk factors in developmental dysplasia of the hip: an observational study. Arch Dis Child. 2013;98–9:692–6.
50. Sanghrajka AP, Murnaghan CF, Shekkeris A, Eastwood DM. Open reduction for developmental dysplasia of the hip: failures of screening or failures of treatment? Ann R Coll Surg Engl. 2013;95–2:113–7.
51. Azzopardi T, Van Essen P, Cundy PJ, Tucker G, Chan A. Late diagnosis of developmental dysplasia of the hip: an analysis of risk factors. J Pediatr Orthop B. 2011;20–1:1–7.
52. Laborie LB, Engesaeter IO, Lehmann TG, Eastwood DM, Engesaeter LB, Rosendahl K. Screening strategies for hip dysplasia: long-term outcome of a randomized controlled trial. Pediatrics. 2013;132–3:492–501.
53. Sink EL, Ricciardi BF, Torre KD, Price CT. Selective ultrasound screening is inadequate to identify patients who present with symptomatic adult acetabular dysplasia. J Child Orthop. 2014;8–6:451–5.
54. von Kries R, Ihme N, Altenhofen L, Niethard FU, Krauspe R, Ruckinger S. General ultrasound screening reduces the rate of first operative procedures for developmental dysplasia of the hip: a case-control study. J Pediatr. 2012;160–2:271–5.
55. Shorter D, Hong T, Osborn DA. Screening programmes for developmental dysplasia of the hip in newborn infants. Cochrane Database Syst Rev. 2011;9:Cd004595.

Evidence-Based Management of Developmental Dysplasia of the Hip

4

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Abstract

This chapter aims to summarise the best available evidence for decision making in treating children with developmental dysplasia of the hip. While the treatment of DDH is not without risk, it is universally accepted that the clinician should do what is necessary to achieve and maintain a gentle, concentric and stable reduction. Several challenges became apparent during the preparation of this chapter. Many of the studies are retrospective case series with suboptimal methodology, small numbers and inconsistent treatment methods. Radiological appearance (using the Severin classification and scores to identify the pattern of growth disturbance) are consistently used as surrogates for poor outcome. There are several aspects in the treatment of DDH where uncertainties remain. We hope that this chapter will help guide the surgeon through these controversies.

Keywords

Developmental dysplasia of the hip • DDH • Congenital dislocation of the hip • CDH • Infant hip • Dislocated hip • Baby hip • Pavlik harness • Von Rosen splint • Hip spica • Closed reduction • Open reduction • Avascular necrosis • Growth disturbance

Background

Developmental dysplasia of the hip (DDH) is a spectrum of disorders in the developing hip that encompass [1, 2]:

1. Hip abnormalities found on ultrasound and radiograph with no clinical abnormalities. If untreated this may present later as a frank dislocation.
2. Hip instability, such that the femoral head can be dislocated partially or fully from the acetabulum by an examiner but relocates spontaneously.
3. Dislocated but reducible hip.
4. Dislocated hip that cannot be reduced.

Accordingly, the incidence varies from 8/100 birth (as ultrasound abnormality), 3/100 birth (abnormal clinical findings) to 1.4/1000 (as frank dislocation). Clinical examination of the newborn can detect hip instability but not acetabular dysplasia. The development of ultrasound has advanced our understanding of the normal development of the hip and allows monitoring of the dislocated, unstable and dysplastic hip. Static ultrasound parameters of the neonatal hip have been classified by Graf [3] (Table 4.1 and Fig. 4.1). Screening for dynamic instability is considered to be an important part of the assessment of the infant hip [4–7].

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Table 4.1 Graf sonographic grading for DDH (see Fig. 4.1)

Type	Alpha angle (α)			Beta angle (β)		Descriptions
I	> 60°			<55°	Ia	Normal hip (at any age). This grade is further divided into (Ia; β < 55°) and (Ib; β > 55°). The significance of this subdivision is not yet established. Patient does not need follow-up.
				> 55°	Ib	
II	50–59°	IIa		< 77°		If the child is <3 months. This may be physiological and does not need treatment; however, Follow up is required.
		IIb		< 77°		> 3 months, delayed ossification.
	43–49°	IIc	Stable	< 77°		Critical zone, labrum not everted. This is further divided into stable and unstable by provocation test.
			Unstable			
D	43–49°			> 77°		This is the first stage where the hip becomes decentred (subluxed).
III	<43°	IIIa				Dislocated femoral head with the cartilaginous acetabular roof is pushed **upwards**. This is further divided into IIIa and IIIb depending on the echogenicity of the hyaline cartilage of the acetabular roof (usually compared to the femoral head) which reflects the degenerative changes.
		IIIb				
IV	<43°					Dislocated femoral head with the cartilaginous acetabular roof is pushed **downwards**

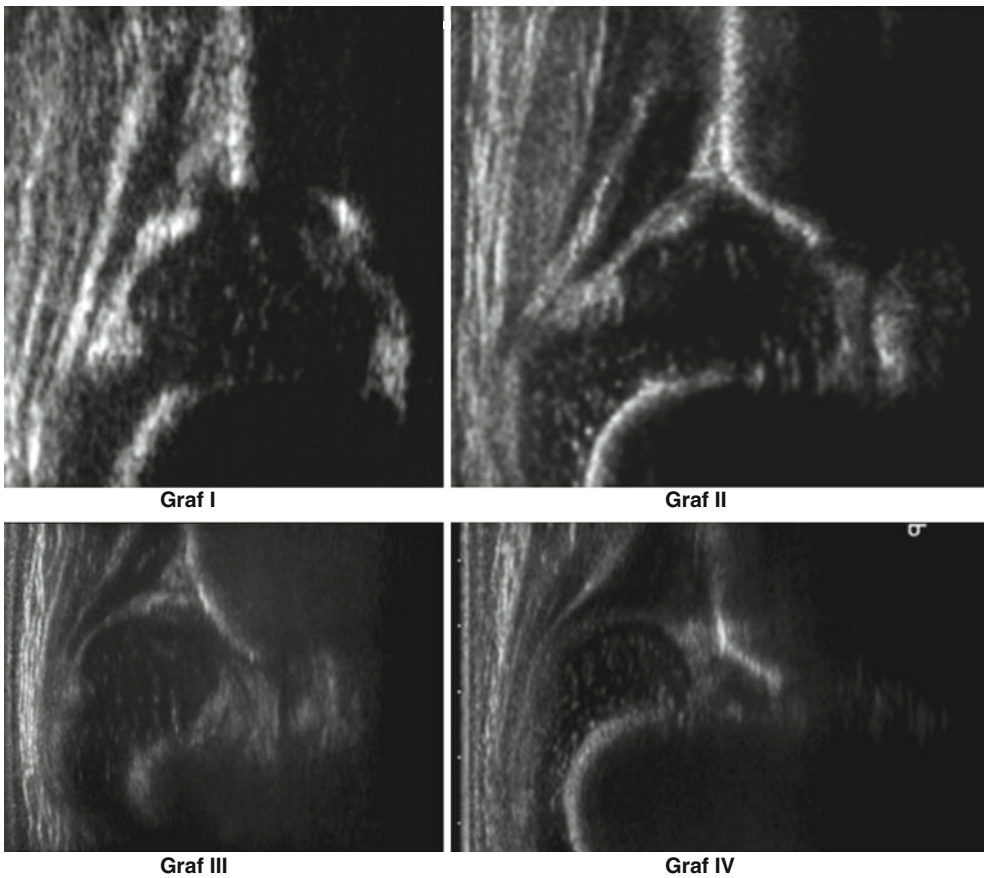

Fig. 4.1 Hips ultrasound; Graf hip types

The principles of treating DDH can be summarised in 4 steps:

1. Achieve a concentric reduction without excessive force.
2. Maintain the concentric reduction for an optimum time.
3. Promote the normal growth and development of the hip.
4. Minimise complications.

Although surgeons around the world agree on the above principle, they adapted different approaches to achieve the above principles. In this chapter, we have explored the evidence behind these approaches concentrating on the common questions that clinicians face in the management of children with DDH.

Table 4.2 Severin classification

Group	Description
I	Well developed hip joint
II	Moderate deformity of the femoral head, neck or acetabulum in otherwise well developed joint
III	Dysplasia, but not subluxation
IV	Subluxation
V	Femoral head is articulating with secondary acetabulum
VI	Re-dislocation

The hip continues to develop throughout childhood and complications of treatment in the young child may not become evident for many years. Large prospective studies with long-term outcomes are scarce and the majority has used surrogates such as avascular necrosis (AVN), failure of treatment, re-dislocations or further surgery etc. AVN is a major cause of long-term disability and is directly related to the treatment – it does not occur in untreated DDH. There are several classifications of AVN including Kalamachi and MacEwen [8], Bucholz and Ogden [9] and Salter's classification [10].

The Severin classification (Table 4.2) describes the radiographic appearance of the hip 5 years after treatment for dislocations. It has been used by many authors as a surrogate for clinical outcome [11]. Several studies have questioned the reliability [12, 13] and showed unacceptably low levels of inter-observer and intra-observer reliability and agreement.

DDH Management in the Infant (0–6 months of Age)

Is Universal Ultrasound Screening Program to Detect Hip Dysplasia Necessary?

This question was addressed in Chap.3.

When Should Treatment Be Commenced for the Dislocatable Hip?

While it is accepted that treatment should not be delayed with the dislocated but reducible hip but should the same apply to the neonate with a dislocatable hip or a stable but dysplastic hip? When should 'watchful waiting' be applied? Treatment with abduction splintage is not entirely benign with a 2–3 % rate of avascular necrosis (AVN) reported in children treated at less than 2 months of age, compared with 1 % after 6 months [14, 15]. Furthermore, AVN has also been shown to occur in the contralateral hip [16, 17].

It has been established that many unstable hips at birth will stabilise without the need for abduction splintage [18]. The fixed flexion contracture of the neonatal hip spontaneously resolves and rapid resolution of acetabular dysplasia also occurs in the majority of cases [19]. Therefore, at what age should treatment be initiated to avoid over-treatment in the clinically unstable hip?

Gardiner and Dunn [7] randomised 79 neonates with unstable (dislocatable) hips to receive either immediate splinting or ultrasound surveillance. At 2 weeks follow-up, 60 % of hips in the surveillance group had no further sonographic signs of hip instability. The remainder with persistent instability or acetabular dysplasia underwent abduction splinting at this stage. At 6 and 12 months follow-up, there was no difference in clinical or radiological appearance in the two initial groups. They recommended that a 2-week delay, prior to initiating treatment for the dislocatable hip is advisable to minimize over treatment. However, due to the small sample size, it would have required a major adverse outcome to be statistically significant.

Elbourne et al. [20] undertook a multi-centre randomised trial to determine whether ultrasonographic surveillance could reduce the number of children undergoing abduction splintage for hip instability, without resulting in a significant increase in the incidence of late treatment. Following a clinical diagnosis of hip instability, infants were randomised to receive treatment based on clinical examination alone (n = 315) or treatment decision based on a hip ultrasound at 2 weeks of age or older (n = 314). In the ultrasound group, those hips that were significantly displaced or unstable were splinted, while minor displacement or instability was monitored. If abnormality persisted until 8 weeks of age, splintage was initiated. In the clinical examination group, abduction splintage was prescribed based on clinical suspicion. Splintage was undertaken earlier in the clinical examination group (81 % by 2 weeks of age vs. 63 % in the ultrasound group) with more hips requiring treatment with clinical examination alone (50 vs. 40 % respectively). Radiographic parameters at 2 years of age and the need for surgical intervention were similar in both groups. The study concluded that ultrasound examination reduced the rate of abduction splintage by a third without increasing the need for late treatment.

While the study does not directly address the age at which treatment should be initiated in the unstable hip, it suggests that the decision on abduction splintage should be based on the ultrasound appearance, not just clinical examination. Furthermore, immediate abduction splintage (<2 weeks of age) was not deemed to be necessary.

These studies indicate that the decision on whether to treat the unstable hip should include ultrasound examination. Treatment prior to 2 weeks of age does not appear to be necessary as many hips will spontaneously stabilise. Grade of recommendation: B.

Should an Ultrasound-Confirmed Dysplastic but Stable Hip Be Treated?

Ultrasound examination allows us to quantify the degree of acetabular dysplasia, but should all neonates with stable dysplastic hips be treated? Neonates that have evidence of

acetabular dysplasia at birth can demonstrate rapid remodeling over the first few weeks of life, without the need for abduction splinting [15]. There are four relevant studies:

Wood et al. [5] reported on a study with 44 infants aged 2–6 weeks with dysplastic stable hips (<40 % coverage on ultrasound examination). Random allocation was used to allocate patients to receive splintage or surveillance. At 3 months the acetabular coverage measured by ultrasound improved in both groups but the greatest improvement was found in those infants placed in an abduction splint (36.7–54.3 % in the splinted group and 32.8–48.6 % without splint, $p < 0.03$). However, at 3 months the acetabular index on plain x-ray was similar (24.79 vs. 24.28). There was a 69 % follow-up at 24 months – mean acetabular index 21.6° (splinted) and 23.5 (no splint). They concluded that in the 2–6 week old child with dysplastic but stable hips, abduction splinting confers no benefit. However, the sample size in the study was small.

Sucato et al. [21] investigated the predictive value of an abnormal ultrasound in an infant under 1 month of age. They performed a retrospective review of 112 infants (192 hips) less than one month of age with a normal hip examination but abnormal hip ultrasound (Graf IIa-III). Pavlik harness treatment was selected at the discretion of the treating physician allowing review of two groups: those treated (43 hips) and untreated (149 hips). The two groups had similar demographics and Graf classification, but the treated group demonstrated less femoral head coverage on stress maneuvers. At final follow-up (15.9 months), in the treated group, no hip had evidence of dysplasia while 2 hips (1.3 %) were considered dysplastic in the untreated group. These 2 hips were initially classified as Graf IIc and it was reported that other more dysplastic hips (Graf IId/III) in the untreated group had normal acetabular indices at final review. They concluded that Pavlik harness management did not appear to influence the incidence of acetabular dysplasia in the stable hip.

Rosendahl et al. [22] conducted a blinded RCT involving 128 neonates (less than 2 weeks of age) with mild hip dysplasia (alpha angle 43–49°). Patients were allocated to receive 6 weeks of abduction splinting or ultrasound surveillance alone. There was no loss to follow-up. 29 children (47 %) in the 'active-surveillance' group received abduction splinting at 6 weeks of age due to persistent dysplasia. At 12 months of age, the mean acetabular inclination was 24.2° in both groups. The authors concluded that in the dysplastic stable hip, an active surveillance policy reduces the need for treatment by 50 % compared with immediate abduction splintage.

In a prospective study of 8638 hips (4319 neonates assessed in the immediate postnatal period) from Israel [23], 8030 hips (93 %) were normal by clinical examination and ultrasound findings. These babies were discharged when they did not have risk factors. Babies with clinically stable hips but with either risk factors or with a Graf type IIa or IIc sonographic appearance were re-examined clinically and sonographically at 6 weeks of age. Those neonates with unstable hips or a stable hip with Graf type D appearance (or worse) were re-examined at 2 weeks of age. If the sonographic appearance showed no improvement of the unstable hips at 2 weeks, treatment with the Pavlik harness was commenced.

Table 4.3 Summary of Bialik's study findings

Type	Stable	Unstable	Total	Needed treatment (%)
Type IIa	255	6	261	7 (2.6 %)
Type IIc	93	19	112	12 (10.7 %)
Type D	24	28	52	12 (23 %)
Type III	4	13	17	3 (17.6 %)
Type IV	1	14	15	12 (80 %)

There were 479 hips (5.53 %) with abnormal sonographic findings placing them in Graf class IIa or worse; of these 81 hips (0.9 %) were unstable on clinical examination.

At the end of the established waiting periods, 90 % of the abnormal hips had become normal without treatment. Less than 3 % of the Graf IIa hips failed to normalise without treatment, whereas 17 % of Graf III hips and 80 % of Graf IV hips failed to normalise (Table 4.3).

These studies suggest that immediate abduction splintage in the dysplastic hip could result in some hips being unnecessarily treated. In the infant with a stable but dysplastic hip, the decision to treat may be delayed to 6 weeks of age without risk of unsatisfactory outcome. Grade of recommendation: B.

How Should a Dysplastic or Dislocated Hip Be Treated in Children Who Are Less Than 6 Months of Age?

The Pavlik harness is the most widely used method of treatment in this age group. The reported success rate ranged from 59 % to 97 % (Table 4.4). Likely reasons for such variation are the threshold for treatment, the length of treatment and definition of failure by reporting authors. As noted previously, 93 % of Graf IIa hips resolve spontaneously without any treatment. If these hips are included in a treatment protocol the reported success will be high.

A variety of other splints and braces have also been used to treat DDH. Comparable success rates have been reported in treating DDH with the Von Rosen splint (Table 4.5). Comparative studies between the above braces are contradictory and most are suboptimal.

These studies suggest that the dislocated or dysplastic hip can be treated with a Pavlik harness or a Von Rosen splint with a high success rate and low incidence of AVN. Grade of recommendation: B.

Table 4.4 Summary of Pavlik Harness treatment

Study		Hips	Success rate (%)	AVN rate (%)	LOE
Pavlik [24]		1912	84	0	IV
Wada [25]		2481	80	14.3	IV
Walton [26]		123	90	2.4	IV
Cashman [27]		546	97	1	IV
Grill [28]		3611	92	2.4	IV
Johnson [29]		91	90	0	IV
Filipe [30]		74	NR	5.4	IV
Santos [31]		159	93.7	16	IV
Murnaghan [32]		1218	94	9	IV
Nakamura [33]		130	81.6	12.3	IV
van der Sluijs [34]		62	60	16	IV
Walton [26]		123	95.2	2.3	IV
Westacott [35] [a]	Groups A	80	88	9	III
	Group B	48	71	4	
Wilkinson [36]		43	76.7	0	III

[a]See the text in question 5 for details

Table 4.5 Summary of Von Rosen splint treatment

Study	Hips	Success rate (%)	AVN rate (%)	LOE
Finlay [37]	56	93	1.7	IV
Fredensborg [38]	111	98	0.9	IV
Heikkila [39]	180	98.3	0.6	IV
Mitchell [40]	100	95	2	IV
Wilkinson [36]	26	100	0	III
Lauge-Pedersen [41]	247	97.5	NR	III

Does Weaning the Pavlik Harness Treatment Provide an Advantage Over Immediate Discontinuation?

Pavlik harness treatment is usually discontinued when ultrasound examination confirms a normal hip morphology and the clinical examination is negative. However, some authors prefer to wean the infant from the harness with a period of part-time wear that occurs over several weeks or months.

Westacott et al. [35] compared the outcome of Pavlik harness treatment between two centres. Eighty children in Centre A underwent staged weaning of the Pavlik harness once three consecutive weekly ultrasounds demonstrated a Graf Grade I hip. Forty-eight children were treated in Centre B where Pavlik harness treatment discontinued immediately with no weaning period. No statistically significant difference was found in the success rate (88 % vs. 71 %) although there was a non-significant trend towards higher intervention when the harness was immediately stopped. While there was also no significant difference in AVN rate, there was a trend towards a lower AVN rate with the immediate cessation (9 % vs. 4 %).

This single study suggests that there is no obvious advantage of weaning the Pavlik harness treatment over the immediate cessation and there may be an associated higher AVN rate. Grade of recommendation: C.

How Should Femoral Nerve Palsy During Pavlik Harness Treatment Be Managed?

Femoral nerve palsy is an uncommon complication of Pavlik harness treatment. In a study [32] of 1218 patients treated by Pavlik harness, 30 cases of femoral nerve palsy were identified (incidence of 2.5 %). 87 % presented within one week of application of the harness. Femoral nerve palsy was more likely in older, larger patients in whom the developmental dysplasia of the hip was of higher severity. Nineteen patients were treated with temporary suspension of harness treatment and subsequent reapplication when femoral nerve function returned, 6 were treated with adjustment of harness to reduce hip flexion, 5 were managed with complete abandonment of the harness, with 4 requiring subsequent closed or open reduction of the hip. There was no correlation between the method of management of the femoral nerve palsy and the success of treatment. Of those 19 patients who did have reinstitution of harness therapy, only 3 developed a recurrence of the palsy. Pavlik harness treatment was abandoned in all patients who demonstrated recurrence of femoral nerve palsy. All patients had eventual complete return of full quadriceps function, with no clinically evident long-term motor or sensory deficit. Patients whose femoral nerve palsy resolved within 3 days had a 70 % chance of having successful treatment with harness, whereas those who had not recovered by 10 days had a 70 % chance of having treatment failure. Notably, the success rate associated with treatment with a Pavlik harness was 94 % in the control group and 47 % in the palsy group.

The published evidence to guide on the best treatment for femoral nerve palsy associated with Pavlik harness is limited. The above study suggested that it is reasonable to temporarily stop the Pavlik harness treatment until the nerve recovers or alternatively reduce the amount of hip flexion. The presence of femoral nerve palsy is associated with a higher failure rate. Grade of recommendation: C.

What Is the Next Step in the Hip That Fails to Reduce and Stabilise in a Pavlik Harness?

Several factors have been associated with failure of Pavlik harness management: breech presentation, bilateral dislocation, age at application, incorrect application, poor parental compliance and an initially irreducible hip [42–46].

The clinically irreducible hip can be successfully treated in a Pavlik harness, but close monitoring is essential. The harness can gradually reduce the abduction contracture and with ongoing active motion and flexion, the femoral head can relocate. It is the most commonly used device in the early management of DDH [27, 47, 48]. However, prolonged positioning of the dislocated hip in adduction and flexion can potentiate femoral head and posterolateral acetabular dysplasia causing greater difficulty in successfully obtaining stability with future closed or open reduction [49].

If the hip is reducible but fails to stabilise in a Pavlik harness, many specialists will elect to proceed with closed reduction ± adductor tenotomy and spica cast. However, there are increasing concerns on the effect of general anaesthesia on the developing brain [50, 51] and a viable alternative to a procedure under anaesthesia is appealing. Furthermore, closed reduction has been associated with a highly variable rate of AVN (see next section). Semi-rigid abduction bracing is an alternative and several authors have reported their experience in using the orthosis in those hips that fail to stabilise in a Pavlik harness. In particular the Ilfield orthosis is gaining in popularity. The Ilfield orthosis holds the hip in less flexion than in a Pavlik harness and in 50° abduction. It reduces the amount of hip motion compared to the Pavlik harness. Sankar et al. [47] postulate that the semi-rigid device may be particularly effective in those hips that are reducible and lie inferiorly or are excessively lax. Parental compliance due to ease of re-application may also be a factor.

Sankar et al. [47] reviewed two retrospective cohorts of patients who had failed Pavlik harness management at the same institution. Nineteen infants who had undergone Ilfield bracing following persistent hip instability following Pavlik harness management were compared to a consecutive retrospective cohort of 16 infants who had a closed reduction and spica cast application. The groups were comparable prior to the secondary intervention. The hips stabilized in 82 %vs 91 % of cases respectively and radiographic appearance was similar at one year. Notably, hips that were dislocated and irreducible were excluded from the study. Three hips in the closed reduction cohort had evidence of AVN at 12 months follow-up.

Hedequist et al. [52] reviewed their experience of using an abduction orthosis after failed Pavlik management in 14 infants. At the time of brace application, 12 of the hips were dislocated but reducible and 2 hips were unstable. There were no irreducible hips. 12/14 hips successfully stabilized in the abduction orthosis and 2 hips failed, requiring closed reduction. One of these developed radiographic evidence of grade 1 AVN at 3-year follow-up. The mean time for the hip stabilisation was 24 days (14–63) with duration in the splint of 46 days (18–91).

Swaroop et al. [53] performed a retrospective review of their experience in managing the dislocated but reducible hip. In the cohort 41/44 hips were successfully stabilised in a Pavlik harness and of the remaining 3 hips who failed Pavlik harness, 2 (67 %) were successfully treated in an abduction orthosis.

Ibrahim et al. [42] performed a retrospective review of 7 patients at a single institution who had failed Pavlik harness management and were then treated in an Ilfield abduction brace. In contrast to the experience of the previous authors, all hips failed to stabilize with the abduction splint and thus closed or open reduction was then required. Of note, three of the patients had a dislocated irreducible hip at initial presentation (unchanged post-Pavlik management) and one patient could not tolerate the brace and it was discontinued after three days. The remaining hips were unstable on commencement of the brace.

The experience of these authors suggest that while an abduction orthosis will not successfully stabilize the irreducible dislocated hip, it can be considered for the dislocated but reducible or unstable hip that fails Pavlik harness management. The evidence thus far are level 3 and 4 studies. Prospective trials are needed to further clarify the indications. Grade of recommendation: B/C.

Failure of Early Treatment or Late Presentation Between 6–18 Months of Age

When Should a Closed Reduction Be Considered?

Closed reduction of the dislocated hip with adductor +/– psoas tenotomy and spica cast is an accepted technique in the management of DDH. It can be performed as the initial procedure in the child over 6 months of age, or following failure post Pavlik harness/abduction brace management. However, it is not a benign procedure with wide variation in AVN rates reported from 4 % to 60 % [54–60]. Case selection, surgical technique, pre-operative traction and the presence of the ossific nucleus are thought to be the contributive factors. Excessive hip abduction in the post-operative spica is likely to be a cause of AVN, due to vascular occlusion and diminished blood supply to the femoral epiphysis – care should be taken to keep the degree of hip abduction within the 'safe-zone', as described by Ramsey [61].

Senaran et al. [62] hypothesised that reducing dislocated hips which fail Pavlik harness treatment within 3 months of age will result in a lower incidence of AVN. To support their hypothesis, they reviewed 21 consecutive cases (35 hips) that failed Pavlik harness treatment and underwent closed reduction before the age of three months. Successful closed reduction

was achieved in 33 (94 %) of 35 hips, and open reduction required in 2 (6 %) of 35 hips. At latest follow-up, one (3 %) of 35 hips had AVN – it should be noted that follow-up duration was just 36 months. At the time of reporting, 1 (3 %) of the 35 hips has required an additional procedure (Pemberton osteotomy) for residual dysplasia. They concluded that the study supports their hypothesis that an early closed reduction following Pavlik harness management minimizes the rate of AVN.

Novais et al. [63] recently published a systematic review and meta-analysis (level III) which provided some answers to our question. They included 66 studies in the systematic review and 24 in the meta-analysis. Data on 481 hips treated by closed reduction and 584 hips treated by open reduction were available to evaluate the association between AVN and age. The association between AVN and operative approach was assessed using data on 364 hips treated by medial open reduction and 220 hips treated by anterior open reduction. Novais reported that the overall, adjusted incidence of AVN (≥ Grade II) was 8.0 % (95 % CI, 2.8 %–20.6 %) among patients who underwent closed reduction at or before 12 months of age and 8.4 % (95 % CI, 3.0 %–21.5 %) among those who had closed reduction after 12 months. The difference between the two age groups was not significant (OR, 1.1; 95 % CI, 0.4–3.2; p = 0.9).

It is of note that this meta-analysis was based on the results of observational studies and potential confounding variables such as failure of previous treatment, associated procedures including adductor tenotomy, length of immobilisation, and degree of abduction in a spica cast were not accounted for.

The authors conducted a systematic review of the literature to investigate the incidence and predictors of AVN and the radiographic outcome in children who had a closed reduction under 2 years of age (unpublished) [64]. The study included 7 papers that had a 5-year minimum follow-up. The review included 539 hips across the studies. At a mean follow-up of 7.6 years there was a 10 % rate of AVN.

Although these studies indicate that a closed reduction is not a benign procedure with 8–10 % rate of AVN, it remains an effective treatment for the hip that fails to stabilise in a Pavlik harness. While careful positioning of the hip in a spica is important to minimize AVN, age at reduction is not conclusively associated with AVN.

Is Preliminary Period of Traction Necessary Before Closed Reduction of a Dislocated Hip?

Proponents of preliminary traction claim that traction reduces the risk of AVN and the need for open reduction. To reduce the cost and inconvenience of hospital admission, portable home traction devices may be used. Opponents cite the increased cost for this additional step in the management and argue that the evidence for reducing the risk of AVN has not been substantiated in recent studies. Several comparative studies (level III) have been identified and their findings have been summarised in Table 4.6. Crude pooling of the AVN rate is slightly lower with traction. Some retrospective case series tried to address the value of the preliminary traction in treating DDH but their findings were inconclusive [56, 70].

Table 4.6 AVN rate and traction

Study	Traction	Hips	AVN
Segal et al. [65]	Yes	48	16
	No	6	1
Langenskiold et al. [66]	Yes	176	65
	No	86	33
Sibinski et al. [67]	Yes	66	20
	No	31	20
Brougham et al. [68]	Yes	42	19
	No	168	80
Kutlu [69]	Yes	89	4
	No	52	0
Crude pooled	Yes	421	124 (28 %)
	No	343	134(37 %)

The published evidence for the value of traction prior to closed reduction is inconclusive. The potential benefit should be weighed against the cost and inconvenience of the traction. Grade of recommendation: C.

Should Treatment Be Delayed Until the Ossific Nucleus Is Visible?

In his above mentioned study [65], Segal also investigated the effect of several factors that might influence the rate of AVN including the presence of the ossific nucleus (ON). The study included 49 children (57 dislocated hips) who were <12 months old. Eighteen hips developed AVN. There was no significant difference in the occurrence of AVN with respect to variables such as preliminary traction, closed versus open reduction, Pavlik harness use, and age at the time of operative intervention. However, the presence of the ossific nucleus before reduction, detected either by radiographs ($p < 0.001$) or sonography ($p = 0.033$) was statistically significant in predicting AVN; one (4 %) of 25 hips with an ossific nucleus developed AVN, whereas 17 (53 %) of 32 hips without an ossific nucleus before reduction developed AVN.

Another study [71] of 48 patients who underwent successful closed reduction showed similar findings. At 2 years follow up, AVN was noted post-reduction in 17 hips (35 %): 4 of 23 hips that had a visible ossific nucleus prior to reduction (17 %), compared with 13 of 25 hips reduced before the ossific nucleus was visible (52 %).

Clarke et al. [72] conducted a prospective study of 50 hips that presented late or had failed conservative treatment. In 28 hips treatment was intentionally delayed until either ossific nucleus became visible or the child reached 13 months of age. In 22 hips the ossific nucleus was present at clinical presentation and treatment proceeded without delay. Six hips reached the age of 13 months without an ossific nucleus appearing and so underwent to treatment. The significant AVN rate (more than grade 1) was 7 % for closed reduction and 14 % for open reduction. The authors concluded that the presence of the ossific nucleus is an important factor in the prevention of AVN and delaying the surgical intervention until it either appears, or the child reaches 13 months of age, is justified. However, there was a higher AVN rate (14 % vs. 4.5 %, not statistically significant) and higher open reduction rate (61 % vs. 23 %) in the delayed group vs. immediate treatment group, indicating that the benefit of delaying treatment is far from clear.

Several other studies did not support the notion that the visibility of the ossific nucleus has a protective effect against the development of the AVN. Findings from these studies have been summarised in Table 4.7.

Our literature search has identified an ongoing phase III randomised trial to assess the effect of timing of surgical intervention on the occurrence of AVN in children with DDH. The target number is 636 children and 3 % have been recruited. The anticipated completion date is 30/6/2024 [78].

The current evidence does not support delaying DDH treatment until the ossific nucleus becomes visible. Grade of recommendation B/C.

Table 4.7 Association of the presence of ossific and AVN

Study	ON	Hips	AVN
Segal et al. [65]	Yes	25	1
	No	32	17
Carney et al. [71]	Yes	23	4
	No	25	13
Konigsberg et al. [73]	Yes	13	4
	No	27	7
Sllamniku et al. [74]	Yes	84	12
	No	150	4
Roposch et al. [75]	Yes	63	20
	No	42	17
Luhmann et al. [76]	Yes	90	1
	No	63	4
Cooke [77]	Yes	24	2
	No	24	2
Crude pooling	Yes	322	44 (13.6 %)
	No	363	64 (17.6 %)

When Should a Medial Open Reduction Be Considered?

Open reduction of the dislocated hip through a medial approach was initially described by Ludloff [79]. He described using the interval between adductor brevis and pectineus to allow direct access to the structures that prevent reduction – the anteromedial capsule, psoas tendon and transverse acetabular ligament. Ferguson [80], followed by Weinstein and Ponseti [81] described alternative surgical approaches – between adductor brevis and gracilis and between pectineus and the neurovascular bundle respectively. The approach is cosmetically acceptable and avoids disturbing the abductor muscles and iliac apophysis. Capsulorraphy cannot be performed, thus it is predominantly used in children prior to walking age (<18 months).

The reported incidence of AVN following medial open reduction varies widely from 0–67 % [82, 83, 84]. It is postulated that this could be due to injury of the medial femoral circumflex artery as it passes over the anteromedial capsule of the hip. Type 2 AVN (similar in the Bucholz and Ogden [9] and Kalamchi and MacEwen [8] classifications) appears to be the most common. It represents a growth disturbance due to a tether to the lateral physis of the femoral head causing a late-onset progressive coxa valga and caput valgus.

A recent systematic review investigated the incidence of AVN following the medial open reduction [85]. Studies included had >5 years follow-up to minimize the underdiagnosis of late-onset growth disturbance and ten or more cases in the series [73, 86–98]. Type 1 AVN (temporary, irregular ossification with minimal clinical significance in the adult hip) was not included in the analysis. The 14 papers that met the inclusion criteria included 734 hips. Detailed, individual information was available on 221 hips allowing further analysis. With a mean follow-up of 10.9 years (2–28 years), the rate of AVN was 20 %.

The analysis of those hips on which specific data was available, type 2 AVN predominated and the rates increased with duration of follow-up (up to 24 % at skeletal maturity). Considering Severin grades 3–6 as an unsatisfactory outcome at skeletal maturity, 55 % of hips with AVN were unsatisfactory compared to 20 % without AVN. Postoperative immobilization, revision surgery and age <12 months at the time of the initial surgery were associated with increased rates of AVN.

This review has limitations – all identified studies were case series and there was variability in the data recorded. There are significant challenges to performing a long-term randomized controlled trial in the operative management of DDH and, for the majority; most evidence is level 3 or 4. In conclusion, MOR is an effective technique of reducing the dislocated hip following failure of Pavlik harness management, or in late diagnosis of DDH <18 months of age. However, the incidence of late onset growth disturbance in >20 % of patients is of concern.

When Should Anterior Open Reduction Be Considered?

The anterior approach to perform open reduction of the hip has withstood the test of time [2] and it allows excellent access to the hip, the obstacles to reduction and permits capsulorraphy. It is usually used as a primary treatment for the dislocated hip after 18 months [99–103] or revision surgery for failed closed or medial open reduction of the hip [104]. The reported AVN rates ranged from 6–23 % and re-dislocation rate ranged 1–11 % (Table 4.7).

Closed Reduction Treatment in Comparison to Open Reduction?

Although closed reduction can be a technically easy procedure, it is not benign and requires experience and attention to the detail. Cooper et al. [105] reported a wide variation in failure rate (0–25 %), AVN rates (0–14 %) and re-dislocation rate (2–60 %) [54–60, 105, 106–109]. The above variations do not come as surprise given the spectrum of the condition, the different surgeon experience, treatment rationales, thresholds for surgical intervention, differences in the definitions of AVN and the timing of follow-up evaluation.

Novais et al. [63] reported a higher incidence of AVN in open reduction in comparison to closed reduction. For open reduction the incidence of AVN (≥Grade II) was 18.3 % (95 % CI, 11.7 %–27.4 %) among patients who had open reduction at or before 12 months of age and 20.0 % (95 % CI, 13.1 %–29.4 %) among those who had index open reduction after 12 months of age.

Interestingly, they found no difference in the risk of AVN between the medial and the anterior approaches (OR, 1.1; 95 % CI, 0.5–2.2; p = 0.9). After controlling for age at reduction, the incidence of AVN (≥ Grade II) after open reduction was 18.7 % (95 % CI, 11.0 %–30.0 %) for patients treated using a medial operative approach and 19.6 % (95 % CI, 12.4–29.5 %) for those who had an anterior approach. The role of pelvic osteotomies in treating dislocated hips.

The role of pelvic osteotomies in treating dislocated hips

Several pelvic osteotomies have been described to treat DDH depending on several factors such as age of the child (skeletal maturity), size and orientation of the acetabulum and congruency. In the context of early treatment for DDH, the most frequently used pelvic osteotomies are the Salter Innominate osteotomy [110–114], Pemberton osteotomy [115–118] and Dega osteotomy [119, 120] (Fig. 4.2).

The Salter osteotomy is a complete transiliac reorientation osteotomy that hinges on the symphysis pubis and provides anterolateral coverage at the expense of posterior coverage. It is advocated as part of the surgical treatment of DDH in children over the age of 18 months – after this age adequate acetabular remodeling may not occur [110]. Thomas et al. [111] reviewed the hip survivorship (hip arthroplasty defined as the end point) at 30, 40 and 45 years after reduction with survival rates of 99 %, 86 % and 54 % respectively. Barrett et al. [114] and Haidar et al. [121] reported good outcomes following a combined open reduction and Salter osteotomy while Bohm et al. [112] reported a higher probability of a better long-term result when the open reduction was performed separately prior to the Salter osteotomy.

The Pemberton and Dega osteotomies are incomplete transiliac osteotomies. They are volume-reducing osteotomies that are held open by either iliac crest graft or bone taken from a femoral shortening. The intrinsic elastic recoil obviates the need for fixation. Advocates highlight improved superior cover due to the proximity of the osteotomy to the origin of the acetabular dysplasia [116, 118].

The Pemberton osteotomy extends from 10 to 15 mm above the ASIS, curves posteriorly and ends at the ilioischial limb of the triradiate cartilage, midway between the sciatic notch and the posterior acetabular rim [117] . It hinges on the triradiate cartilage. The Dega osteotomy breaches a variable amount of the medial cortex depending on the degree of lateral coverage required and hinges on the intact posteromedial cortex [122].

Follow-up studies of the Pemberton and Dega osteotomies report generally good outcomes. Acetabular indices show marked improvement for both osteotomies and variable AVN rates [118, 122–124].

In a review article, Cooper [105] showed that the published AVN rates in Salter osteotomy ranged 3.8–44 %, 2.7–51 % in Pemberton osteotomy and 8–25 % in Dega osteotomy. However, attributing AVN to the pelvic osteotomy is difficult when confounding causes such as previous treatment (e.g. Pavlik harness, closed reduction) open reduction technique and degree of abduction post-operatively have been clearly shown to be causative factors.

Comparison studies also show good outcomes for each pelvic osteotomy and offer little to choose one over another [125]. One study did note a difference: López-Carreño [126] reported on 93 children (99 hips) with developmental dysplasia of the hip, retrospectively comparing the Salter osteotomy and the Dega. They noted a greater correction in acetabular indices with the Dega osteotomy as well as gait and joint mobility.

In conclusion, there is little evidence to recommend one osteotomy over another. The available studies are retrospective case series and show generally good outcomes with each osteotomy.

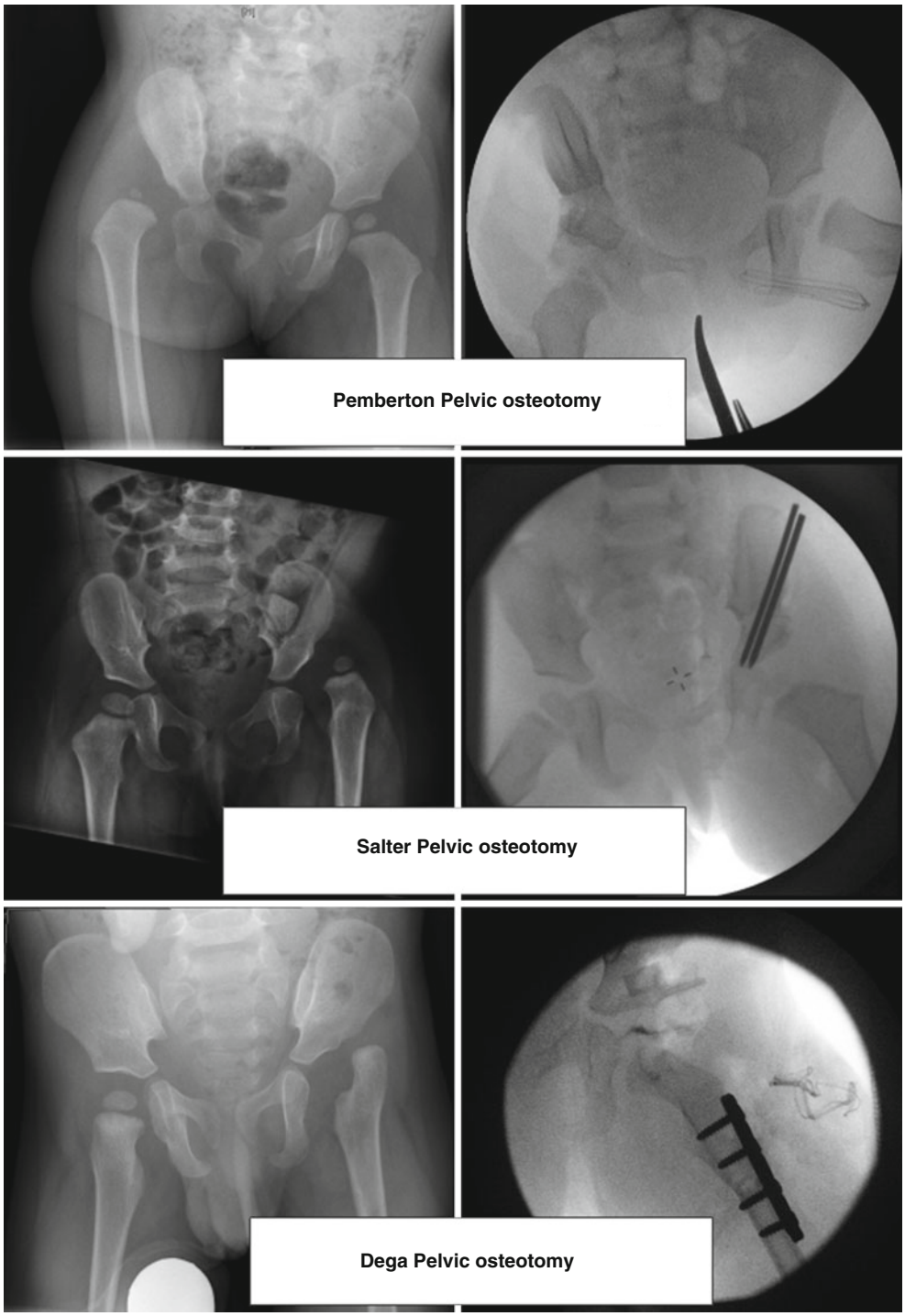

Fig. 4.2 Pelvic osteotomies in treating dislocated hips

The Role of Femoral Osteotomies in Treating Dislocated Hips

Several studies have reported the use of the femoral osteotomy in conjunction with open reduction for DDH – either as a shortening, derotation or varus osteotomy or as a combination. The indication is to reduce pressure on the reduced femoral head and improve stability [2]. Schoenecker [127] analysed the complications and the radiographic and functional outcomes of treatment in 39 hips comparing preoperative traction to femoral shortening. All of the patients were three years or older and they had received no previous treatment. From 1961 to 1975, 17 children (26 hips) were treated with preoperative skeletal traction prior to any operative procedures (group 1). From 1976 to 1980, 8 children (13 hips) were treated with a femoral shortening osteotomy at the time

of open reduction (group 2). AVN was documented in 14 hips in Group 1 and none in Group 2. Re-dislocation occurred in 8 hips in Group 1 and in 1 hip in Group 2. Similar outcomes were reported by Westin [128, 129]. Two groups (traction vs. femoral shortening) were again compared over two time frames. The incidence of AVN was 57 % vs. 0 % respectively and a poor Severin outcome was noted in 43 % vs. 0 % of cases respectively. Galpin [130] reported on 25 patients (33 hips) who were two years or older with DDH. None of the patients had had previous treatment. Femoral shortening was used instead of traction and the results were good or excellent in 82 % of hips with an AVN rate of 9 %.

While there are no prospective studies available for analysis, the current evidence is in favour of intra-operative femoral shortening over pre-operative traction as a means of improving stability and minimizing the incidence of AVN in the older child.

What Is the Upper Age Limit for Reducing a Dislocated Hip?

Although it is well known that the outcome of reducing a dislocated hip becomes poorer with older age, the cut off age has not been agreed on. Several cases series and comparative studies demonstrated good results in children below the age of 8 [129–135, 136].

Yagmurlu [131] reported on 21 children (27 hips) who had one stage open reduction, femoral shortening and Salter or triple pelvic osteotomy at older age. They were grouped into one older and one younger than 8 years of age with the were divided into two groups: those older than 8 years of age and those younger. The younger group fared better according to according to the Severin and Mc Kay classifications. Ning et al. [137] reported on a retrospective case series of 652 children who underwent hip reduction in China; all were above the age of 18 months and underwent a single stage open reduction, pelvic and femoral osteotomies. Children were divided into 3 groups: young (1.5–2.5 years), middle (2.5–8 years) and old group (>8 years). The worst clinical and radiographic outcomes were in the old groups ($P < 0.001$). Table 4.8 summarises their findings.

El-Tayeby [138] reported on his experience in treating hip dislocation in 16 patient who were above the age of 8 (range 8–18 years). Fifteen hips (79 %) were clinically excellent to good, while four hips (21 %) were fair to poor according to the McKay modified criteria. Sixteen hips (84 %) were excellent to good and 3 hips (16 %) were fair to poor according to Severin classification.

The above findings should be contrasted with the natural history of a dislocated hip. Pain develops in approximately half of the patients with untreated DDH and in some individuals; fully dislocated hips may never become painful. The outcome is usually worse for a subluxated hip or unilateral dislocation [2].

In summary, there is a reasonable evidence to support reducing congenital dislocation of a hip in children up to the age of 8 (grade B/C). Older children have a poorer outcome and may be worse than the natural history.

Table 4.8 Age and outcomes of hip reduction

Characteristics	Items	Young (1.5–2.5 years)	Middle (2.5–8 years)	Old (> 8 years)
Baseline	Patients (hips)	183(206)	391(576)	78(82)
	Age (y)	1.9 ± 0.23	4.6 ± 0.65	9.6 ± 1.2
	Follow-up (y)	6.4 ± 2.81	6.12 ± 2.23	6.15 ± 2.45
McKay Clinical classification n (%)	Excellent	112 (54.4)	362 (62.8)	6 (7.3)
	Good	62 (30.1)	137(23.9)	8 (9.8)
	Fair	28 (13.6)	56(9.7)	43 (52.4)
	Poor	4 (1.9)	21(3.6)	25 (30.5)
Severin classification n (%)	Ia	94(45.6)	196(34.5)	8(9.7)
	Ib	36(17.5)	158(27.8)	13(15.9)
	II	42(20.4)	177(30.7)	8(9.7)
	III	25(12.1)	31(5.4)	13(15.9
	IV	9(4.4)	8(1.4)	6(7.3)
	V	0	0 6(1.2)	21(25.6)
	VII	0	0	13(15.9)
Kalamchi and MacEwen AVN classification n (%)	Absent	393(68.3)	12(14.7)	494(57.2)
	I	82(14.2)	5(6.1)	133(15.4)
	II	74(12.8)	24(29.3)	136(15.7)
	III	15(2.6)	26(31.7)	66(7.6)
	IV	12(2.1)	15(18.2)	35(4.1)

Table 4.9 Recommendations

Statement	Grade of recommendation
Hip ultrasound is important to guide DDH treatment. Treatment prior to 2 weeks of age does not appear to be necessary as many hips will spontaneously stabilise.	B
Dislocated or dysplastic hips can be treated with a brace with a high success rate and low AVN rate. Pavlik harness and the von Rosen splint are the most widely used.	B
Is the von Rosen splint better than the Pavlik harness?	I
There is no benefit of weaning the Pavlik harness treatment over the immediate cessation. There may be a higher AVN rate.	C
When femoral nerve palsy developed in a child who is being treated with Pavlik harness, it is reasonable to temporarily stop the treatment until the nerve recover or reduce the amount of hip flexion. The femoral nerve palsy was associated with a high failure rate.	C
If Pavlik harness fails to keep hip reduce, an abduction orthosis will not successfully stabilize the irreducible dislocated hip but it can be considered for the dislocated but reducible or unstable hip	B/C
Closed reduction should not be delayed until the ossific nucleus becomes visible.	B/C
The value of traction is inconclusive and the potential benefit should be weighed against the cost and inconvenience of the traction	C
Surgical treatment of a dislocated hip should be considered in children younger than 8 years old	B/C

Authors' Preferred Method of Management

In concluding this chapter, it is apparent that many controversies remain. However, the underlying principle of managing a child with DDH is universally accepted: the clinician must do what is necessary to achieve and maintain a gentle, concentric and stable reduction. In this final section, we have summarized our approach in managing the child with DDH (Table 4.9). We do appreciate that in some areas surgeons familiar with the management of DDH will take an alternative approach.

We advocate that treatment should not be delayed in the neonate with a dislocated but reducible hip. However, the neonate with a dislocatable hip should be re-examined at 2 weeks of age as many hips will stabilize during this period. If the hip remains unstable, we commence treatment with a Pavlik harness. When a neonate has been referred with ultrasonographic evidence of dysplasia, we delay treatment until 6 weeks of age and commence a Pavlik harness for the IIb (and worse) hip. The harness is retained for 12 weeks until the hip is ultrasonographically normal. We do not wean the harness at the end of treatment. For the dislocated reducible hip that fails Pavlik harness we switch to a semi-rigid orthosis.

Should the child present after six months of age, or the hip fail to stabilize with the Pavlik harness (with or without a supplementary semi-rigid orthosis), we routinely proceed with a closed reduction. We perform an open adductor longus tenotomy and directly feel the femoral head reducing into the acetabulum via the Ludloff approach. The capsule is not opened. If the hip is unstable in less than 60° of flexion, we perform a psoas tenotomy at the lesser trochanter via the same approach. The child is immobilized in a hip spica for 12 weeks in 100° of hip flexion and 50° (or less) hip abduction, depending on the safe-zone of the hip.

We do not delay treatment if the ossific nucleus is not visible and proceed when the child can be safely placed under general anaesthesia. Achieving a gentle, concentric and stable reduction remains the most important principle and we believe that delaying closed reduction may increase the pressure on the vulnerable femoral head and increase the need for open reduction.

In the event of the hip failing to stabilize with a closed reduction, we do not proceed with a medial open reduction. The long-term studies demonstrate an unacceptably high level of type 2 AVN. We prefer to wait until the child is 9–12 months of age and perform an open reduction and capsulorrhaphy via an anterior approach. We then immobilise the child in a hip spica for 6–8 weeks in 20° of abduction, flexion and internal rotation.

We do not use pre-operative traction but are judicious with the use of femoral shortening to avoid excess pressure on the femoral head. In a child 18 months and older, we perform a Salter osteotomy in addition to the open reduction and capsulorrhaphy for two reasons – we recognise that the remodeling capability of the acetabulum is often insufficient at this age and the osteotomy improves the antero-lateral cover to improve hip stability. In hips with more severe acetabular dysplasia with an acetabular index of >40° and a lateral trough via which the femoral head can be felt to slide out after open reduction, we perform an acetabuloplasty in preference to a Salter osteotomy due to the ability to achieve a greater magnitude of correction.

References

1. Alshryda S, Jones S, Banaszkiewicz P. Postgraduate paediatric orthopaedics: the candidate's guide to the FRCS (Tr and Orth) examination. Cambridge: Cambridge University Press; 2014.
2. Herring JA, editor. Tachdjians, pediatric orthopaedics, vol. 1. Philadelphia: Saunders Elsevier; 2013.
3. Graf R. Fundamentals of sonographic diagnosis of infant hip dysplasia. J Pediatr Orthop. 1984;4(6):735–40.
4. Clarke NM et al. Real-time ultrasound in the diagnosis of congenital dislocation and dysplasia of the hip. J Bone Joint Surg (Br). 1985;67(3):406–12.
5. Wood MK, Conboy V, Benson MK. Does early treatment by abduction splintage improve the development of dysplastic but stable neonatal hips? J Pediatr Orthop. 2000;20(3):302–5.
6. Engesaeter LB et al. Ultrasound and congenital dislocation of the hip. The importance of dynamic assessment. J Bone Joint Surg (Br). 1990;72(2):197–201.
7. Gardiner HM, Dunn PM. Controlled trial of immediate splinting versus ultrasonographic surveillance in congenitally dislocatable hips. Lancet. 1990;336(8730):1553–6.
8. Kalamchi A, MacEwen GD. Avascular necrosis following treatment of congenital dislocation of the hip. J Bone Joint Surg Am. 1980;62(6):876–88.
9. Bucholz, R. O., JA, *Patterns of ischemic necrosis of the proximal femur: nonoperatively managed congenital hip disease*. The hip: proceedings of the sixth scientific meeting of the hip society, 1978: p. 43–63.
10. Salter RB, Kostuik J, Dallas S. Avascular necrosis of the femoral head as a complication of treatment for congenital dislocation of the hip in young children: a clinical and experimental investigation. Can J Surg. 1969;12(1):44–61.
11. Severin E. Congenital dislocation of the hip; development of the joint after closed reduction. J Bone Joint Surg Am. 1950;32-A(3):507–18.
12. Ward WT, V. M, Grudziak JS, Tumer Y, Cook PC, Fitch RD. Severin classification system for evaluation of the results of operative treatment of congenital dislocation of the hip. A study of intraobserver and interobserver reliability. J Bone Joint Surg Am. 1997;79(5):656–63.
13. Ali AM et al. Reliability of the Severin classification in the assessment of developmental dysplasia of the hip. J Pediatr Orthop B. 2001;10(4):293–7.
14. Lehmann HP et al. Developmental dysplasia of the hip practice guideline: technical report. Committee on Quality Improvement, and Subcommittee on Developmental Dysplasia of the Hip. Pediatrics. 2000;105(4):E57.
15. Burger BJ et al. Neonatal screening and staggered early treatment for congenital dislocation or dysplasia of the hip. Lancet. 1990;336(8730):1549–53.
16. Bradley J, Wetherill M, Benson MK. Splintage for congenital dislocation of the hip. Is it safe and reliable? J Bone Joint Surg (Br). 1987;69(2):257–63.
17. Langkamer VG, Clarke NM, Witherow P. Complications of splintage in congenital dislocation of the hip. Arch Dis Child. 1991; 66(11):1322–5.
18. Castelein RM et al. Natural history of ultrasound hip abnormalities in clinically normal newborns. J Pediatr Orthop. 1992;12(4):423–7.
19. Gardiner HM, Duncan AW. Radiological assessment of the effects of splinting on early hip development: results from a randomised controlled trial of abduction splinting vs sonographic surveillance. Pediatr Radiol. 1992;22(3):159–62.
20. Elbourne D et al. Ultrasonography in the diagnosis and management of developmental hip dysplasia (UK Hip Trial): clinical and economic results of a multicentre randomised controlled trial. Lancet. 2002;360(9350):2009–17.
21. Sucato DJ et al. Outcome of ultrasonographic hip abnormalities in clinically stable hips. J Pediatr Orthop. 1999;19(6):754–9.
22. Rosendahl K et al. Immediate treatment versus sonographic surveillance for mild hip dysplasia in newborns. Pediatrics. 2010;125(1):e9–16.
23. Bialik V, Bialik GM, Wiener F. Prevention of overtreatment of neonatal hip dysplasia by the use of ultrasonography. J Pediatr Orthop B. 1998;7(1):39–42.
24. Pavlik A. The functional method of treatment using a harness with stirrups as the primary method of conservative therapy for infants with congenital dislocation of the hip. Clin Orthop Relat Res. 1957;1992(281):4–10.
25. Wada I et al. The Pavlik harness in the treatment of developmentally dislocated hips: results of Japanese multicenter studies in 1994 and 2008. J Orthop Sci. 2013;18(5):749–53.
26. Walton MJ et al. The success of management with the Pavlik harness for developmental dysplasia of the hip using a United Kingdom screening programme and ultrasound-guided supervision. J Bone Joint Surg (Br). 2010;92(7):1013–6.
27. Cashman JP et al. The natural history of developmental dysplasia of the hip after early supervised treatment in the Pavlik harness. A prospective, longitudinal follow-up. J Bone Joint Surg (Br). 2002;84(3):418–25.
28. Grill F et al. The Pavlik harness in the treatment of congenital dislocating hip: report on a multicenter study of the European paediatric orthopaedic society. J Pediatr Orthop. 1988;8(1):1–8.
29. Johnson AH et al. Treatment of congenital hip dislocation and dysplasia with the Pavlik harness. Clin Orthop Relat Res. 1981;155:25–9.
30. Filipe G, Carlioz H. Use of the Pavlik harness in treating congenital dislocation of the hip. J Pediatr Orthop. 1982;2(4):357–62.
31. Mafalda Santos M, Filipe G. Treatment of congenital hip dislocation using Pavlik's harness. Long term results. Rev Chir Orthop Reparatrice Appar Mot. 1997;83(1):41–50.
32. Murnaghan ML et al. Femoral nerve palsy in Pavlik harness treatment for developmental dysplasia of the hip. J Bone Joint Surg Am. 2011;93(5):493–9.
33. Nakamura J et al. Treatment for developmental dysplasia of the hip using the Pavlik harness: long-term results. J Bone Joint Surg (Br). 2007;89(2):230–5.
34. van der Sluijs JA et al. Prolonged treatment with the Pavlik harness in infants with developmental dysplasia of the hip. J Bone Joint Surg (Br). 2009;91(8):1090–3.
35. Westacott DJ et al. Staged weaning versus immediate cessation of Pavlik harness treatment for developmental dysplasia of the hip. J Pediatr Orthop B. 2013;23(2):103–6.
36. Wilkinson AG, Sherlock DA, Murray GD. The efficacy of the Pavlik harness, the Craig splint and the von Rosen splint in the management of neonatal dysplasia of the hip. A comparative study. J Bone Joint Surg (Br). 2002;84(5):716–9.
37. Finlay HV, Maudsley RH, Busfield PI. Dislocatable hip and dislocated hip in the newborn infant. Br Med J. 1967;4(5576):377–81.
38. Fredensborg N. The results of early treatment of typical congenital dislocation of the hip in Malmo. J Bone Joint Surg (Br). 1976;58(3):272–8.
39. Heikkila E. Comparison of the Frejka pillow and the von Rosen splint in treatment of congenital dislocation of the hip. J Pediatr Orthop. 1988;8(1):20–1.
40. Mitchell GP. Problems in the early diagnosis and management of congenital dislocation of the hip. J Bone Joint Surg (Br). 1972;54(1):4–12.

41. Lauge-Pedersen H, Gustafsson J, Hagglund G. 6 Weeks with the von Rosen splint is sufficient for treatment of neonatal hip instability. Acta Orthop. 2006;77(2):257–61.
42. Ibrahim DA, Skaggs DL, Choi PD. Abduction bracing after Pavlik harness failure: an effective alternative to closed reduction and spica casting? J Pediatr Orthop. 2013;33(5):536–9.
43. Mubarak S et al. Pitfalls in the use of the Pavlik harness for treatment of congenital dysplasia, subluxation, and dislocation of the hip. J Bone Joint Surg Am. 1981;63(8):1239–48.
44. Harris IE, Dickens R, Menelaus MB. Use of the Pavlik harness for hip displacements. When to abandon treatment. Clin Orthop Relat Res. 1992;281:29–33.
45. Viere RG et al. Use of the Pavlik harness in congenital dislocation of the hip. An analysis of failures of treatment. J Bone Joint Surg Am. 1990;72(2):238–44.
46. Atalar H et al. Indicators of successful use of the Pavlik harness in infants with developmental dysplasia of the hip. Int Orthop. 2007;31(2):145–50.
47. Sankar WN, Nduaguba A, Flynn JM. Ilfeld abduction orthosis is an effective second-line treatment after failure of Pavlik harness for infants with developmental dysplasia of the hip. J Bone Joint Surg Am. 2015;97(4):292–7.
48. Guille JT, Pizzutillo PD, MacEwen GD. Development dysplasia of the hip from birth to six months. J Am Acad Orthop Surg. 2000;8(4):232–42.
49. Jones GT, Schoenecker PL, Dias LS. Developmental hip dysplasia potentiated by inappropriate use of the Pavlik harness. J Pediatr Orthop. 1992;12(6):722–6.
50. Ing C et al. Long-term differences in language and cognitive function after childhood exposure to anesthesia. Pediatrics. 2012;130(3):e476–85.
51. Hays SR, Deshpande JK. Newly postulated neurodevelopmental risks of pediatric anesthesia: theories that could rock our world. J Urol. 2013;189(4):1222–8.
52. Hedequist D, Kasser J, Emans J. Use of an abduction brace for developmental dysplasia of the hip after failure of Pavlik harness use. J Pediatr Orthop. 2003;23(2):175–7.
53. Swaroop VT, Mubarak SJ. Difficult-to-treat Ortolani-positive hip: improved success with new treatment protocol. J Pediatr Orthop. 2009;29(3):224–30.
54. DeRosa GP, Feller N. Treatment of congenital dislocation of the hip. Management before walking age. Clin Orthop Relat Res. 1987;225:77–85.
55. Joseph K, MacEwen GD, Boos ML. Home traction in the management of congenital dislocation of the hip. Clin Orthop Relat Res. 1982;165:83–90.
56. Kahle WK et al. The value of preliminary traction in the treatment of congenital dislocation of the hip. J Bone Joint Surg Am. 1990;72(7):1043–7.
57. Quinn RH, Renshaw TS, DeLuca PA. Preliminary traction in the treatment of developmental dislocation of the hip. J Pediatr Orthop. 1994;14(5):636–42.
58. Race C, Herring JA. Congenital dislocation of the hip: an evaluation of closed reduction. J Pediatr Orthop. 1983;3(2):166–72.
59. Zionts LE, MacEwen GD. Treatment of congenital dislocation of the hip in children between the ages of one and three years. J Bone Joint Surg Am. 1986;68(6):829–46.
60. Malvitz TA, Weinstein SL. Closed reduction for congenital dysplasia of the hip. Functional and radiographic results after an average of thirty years. J Bone Joint Surg Am. 1994;76(12):1777–92.
61. Ramsey PL, Lasser S, MacEwen GD. Congenital dislocation of the hip. Use of the Pavlik harness in the child during the first six months of life. J Bone Joint Surg Am. 1976;58(7):1000–4.
62. Senaran H, Bowen JR, Harcke HT. Avascular necrosis rate in early reduction after failed Pavlik harness treatment of developmental dysplasia of the hip. J Pediatr Orthop. 2007;27(2):192–7.
63. Novais, E.N., et al., Is age or surgical approach associated with osteonecrosis in patients with developmental sysplasia of the hip? A Meta-analysis. Clin Orthop Relat Res. 2015.
64. Perry, B.J., Bradley, C.S., Wedge, L.H., Murnaghan, M.L., Kelley, S.P., Avascular necrosis following closed reduction for the treatment of developmental dysplasia of the hip: a systematic review. Currently unpublished.
65. Segal LS et al. Avascular necrosis after treatment of DDH: the protective influence of the ossific nucleus. J Pediatr Orthop. 1999;19(2):177–84.
66. Langenskiold A, Paavilainen T. The effect of prereduction traction on the results of closed reduction of developmental dislocation of the hip. J Pediatr Orthop. 2000;20(4):471–4.
67. Sibinski M, Murnaghan C, Synder M. The value of preliminary overhead traction in the closed management of DDH. Int Orthop. 2006;30(4):268–71.
68. Brougham DI et al. Avascular necrosis following closed reduction of congenital dislocation of the hip. Review of influencing factors and long-term follow-up. J Bone Joint Surg (Br). 1990;72(4):557–62.
69. Kutlu A et al. Preliminary traction as a single determinant of avascular necrosis in developmental dislocation of the hip. J Pediatr Orthop. 2000;20(5):579–84.
70. Daoud A, Saighi-Bououina A. Congenital dislocation of the hip in the older child. The effectiveness of overhead traction. J Bone Joint Surg Am. 1996;78(1):30–40.
71. Carney BT, Clark D, Minter CL. Is the absence of the ossific nucleus prognostic for avascular necrosis after closed reduction of developmental dysplasia of the hip? J Surg Orthop Adv. 2004;13(1):24–9.
72. Clarke NM, Jowett AJ, Parker L. The surgical treatment of established congenital dislocation of the hip: results of surgery after planned delayed intervention following the appearance of the capital femoral ossific nucleus. J Pediatr Orthop. 2005;25(4):434–9.
73. Konigsberg DE et al. Results of medial open reduction of the hip in infants with developmental dislocation of the hip. J Pediatr Orthop. 2003;23(1):1–9.
74. Sllamniku S et al. Correlation between avascular necrosis and the presence of the ossific nucleus when treating developmental dysplasia of the hip. J Child Orthop. 2014;7(6):501–5.
75. Roposch A et al. The presence of an ossific nucleus does not protect against osteonecrosis after treatment of developmental dysplasia of the hip. Clin Orthop Relat Res. 2011;469(10):2838–45.
76. Luhmann SJ et al. The prognostic importance of the ossific nucleus in the treatment of congenital dysplasia of the hip. J Bone Joint Surg Am. 1998;80(12):1719–27.
77. Cooke SJ et al. Ossification of the femoral head at closed reduction for developmental dysplasia of the hip and its influence on the long-term outcome. J Pediatr Orthop B. 2010;19(1):22–6.
78. Clarke NMP. Hip 'Op. Timing of surgical intervention for developmental dysplasia of the hip. [cited 2015]; Available from: http://public.ukcrn.org.uk/Search/StudyDetail.aspx?StudyID=16882
79. Ludloff K. Zur blutigen Einrenkung der angeborenen Hüftluxation. Z Orthop Chir, 1908: p. 272–276 (in German).
80. Ferguson Jr AB. Primary open reduction of congenital dislocation of the hip using a median adductor approach. J Bone Joint Surg Am. 1973;55(4):671–89.
81. Weinstein SL, Ponseti IV. Congenital dislocation of the hip. J Bone Joint Surg Am. 1979;61(1):119–24.
82. Kalamchi A, Schmidt TL, MacEwen GD. Congenital dislocation of the hip. Open reduction by the medial approach. Clin Orthop Relat Res. 1982;169:127–32.
83. O'Hara JN, Bernard AA, Dwyer NS. Early results of medial approach open reduction in congenital dislocation of the hip: use before walking age. J Pediatr Orthop. 1988;8(3):288–94.

84. Mergen E et al. Medial approach open reduction for congenital dislocation of the hip using the Ferguson procedure. A review of 31 hips. Arch Orthop Trauma Surg. 1991;110(3):169–72.
85. Gardner RO et al. The incidence of avascular necrosis and the radiographic outcome following medial open reduction in children with developmental dysplasia of the hip: a systematic review. Bone Joint J. 2014;96-B(2):279–86.
86. Bache CE et al. Ligamentum teres tenodesis in medial approach open reduction for developmental dislocation of the hip. J Pediatr Orthop. 2008;28(6):607–13.
87. Bicimoglu A et al. Posteromedial limited surgery in developmental dysplasia of the hip. Clin Orthop Relat Res. 2008;466(4): 847–55.
88. Castillo R, Sherman FC. Medial adductor open reduction for congenital dislocation of the hip. J Pediatr Orthop. 1990;10(3): 335–40.
89. Erturk C et al. Medial open reduction of developmental dysplasia of the hip using the Weinstein-Ponseti approach. Saudi Med J. 2011;32(9):901–6.
90. Kiely N et al. The ferguson medial approach for open reduction of developmental dysplasia of the hip. A clinical and radiological review of 49 hips. J Bone Joint Surg (Br). 2004;86(3):430–3.
91. Koizumi W et al. Ludloff's medial approach for open reduction of congenital dislocation of the hip. A 20-year follow-up. J Bone Joint Surg (Br). 1996;78(6):924–9.
92. Mankey MG, Arntz GT, Staheli LT. Open reduction through a medial approach for congenital dislocation of the hip. A critical review of the Ludloff approach in sixty-six hips. J Bone Joint Surg Am. 1993;75(9):1334–45.
93. Matsushita T et al. Open reduction for congenital dislocation of the hip: comparison of the long-term results of the wide exposure method and Ludloff's method. J Orthop Sci. 1999;4(5): 333–41.
94. Morcuende JA et al. Long-term outcome after open reduction through an anteromedial approach for congenital dislocation of the hip. J Bone Joint Surg Am. 1997;79(6):810–7.
95. Okano K et al. Long-term outcome of Ludloff's medial approach for open reduction of developmental dislocation of the hip in relation to the age at operation. Int Orthop. 2009;33(5):1391–6.
96. Sosna A, Rejholec M. Ludloff's open reduction of the hip: long-term results. J Pediatr Orthop. 1992;12(5):603–6.
97. Tumer Y, Ward WT, Grudziak J. Medial open reduction in the treatment of developmental dislocation of the hip. J Pediatr Orthop. 1997;17(2):176–80.
98. Ucar DH et al. Open reduction through a medial approach in developmental dislocation of the hip: a follow-up study to skeletal maturity. J Pediatr Orthop. 2004;24(5):493–500.
99. Doudoulakis JK, Cavadias A. Open reduction of CDH before one year of age. 69 hips followed for 13 (10–19) years. Acta Orthop Scand. 1993;64(2):188–92.
100. Szepesi K et al. Preliminary results of early open reduction by an anterior approach for congenital dislocation of the hip. J Pediatr Orthop B. 1995;4(2):171–8.
101. Varner KE et al. Surgical treatment of developmental hip dislocation in children aged 1 to 3 years: a mean 18-year, 9-month follow-up study. Orthopedics. 2010;33(3).
102. Cordier W et al. Long-term results after open reduction of developmental hip dislocation by an anterior approach lateral and medial of the iliopsoas muscle. J Pediatr Orthop B. 2005;14(2): 79–87.
103. Dhar S et al. Early open reduction for congenital dislocation of the hip. J Bone Joint Surg (Br). 1990;72(2):175–80.
104. McCluskey WP et al. Treatment of failed open reduction for congenital dislocation of the hip. J Pediatr Orthop. 1989;9(6):633–9.
105. Cooper AP, Doddabasappa SN, Mulpuri K. Evidence-based management of developmental dysplasia of the hip. Orthop Clin North Am. 2014;45(3):341–54.
106. Aksoy MC et al. Treatment of developmental dysplasia of the hip before walking: results of closed reduction and immobilization in hip spica cast. Turk J Pediatr. 2002;44(2):122–7.
107. Ishii Y, Ponseti IV. Long-term results of closed reduction of complete congenital dislocation of the hip in children under one year of age. Clin Orthop Relat Res. 1978;137:167–74.
108. Murray T et al. Closed reduction for treatment of developmental dysplasia of the hip in children. Am J Orthop (Belle Mead NJ). 2007;36(2):82–4.
109. Terjesen T, Halvorsen V. Long-term results after closed reduction of latedetected hip dislocation: 60 patients followed up to skeletal maturity. Acta Orthop. 2007;78(2):236–46.
110. Salter RB, Dubos JP. The first fifteen year's personal experience with innominate osteotomy in the treatment of congenital dislocation and subluxation of the hip. Clin Orthop Relat Res. 1974;98:72–103.
111. Thomas SR, Wedge JH, Salter RB. Outcome at forty-five years after open reduction and innominate osteotomy for late-presenting developmental dislocation of the hip. J Bone Joint Surg Am. 2007;89(11):2341–50.
112. Bohm P, Brzuske A. Salter innominate osteotomy for the treatment of developmental dysplasia of the hip in children: results of seventy-three consecutive osteotomies after twenty-six to thirty-five years of follow-up. J Bone Joint Surg Am. 2002;84-A(2): 178–86.
113. Bohm P, Klinger HM, Kusswetter W. The Salter innominate osteotomy for the treatment of developmental dysplasia of the hip in young adults. Arch Orthop Trauma Surg. 1999;119(3–4):186–9.
114. Barrett WP, Staheli LT, Chew DE. The effectiveness of the Salter innominate osteotomy in the treatment of congenital dislocation of the hip. J Bone Joint Surg Am. 1986;68(1):79–87.
115. Berkeley ME et al. Surgical therapy for congenital dislocation of the hip in patients who are twelve to thirty-six months old. J Bone Joint Surg Am. 1984;66(3):412–20.
116. Pemberton PA. Pericapsular osteotomy of the ilium for the treatment of congenitally dislocated hips. Clin Orthop Relat Res. 1974;98:41–54.
117. Pemberton PA. Pericapsular osteotomy of the ilium for treatment of congenital subluxation and dislocation of the hip. J Bone Joint Surg. 1965;47-A(1):65–86.
118. Faciszewski T, Kiefer GN, Coleman SS. Pemberton osteotomy for residual acetabular dysplasia in children who have congenital dislocation of the hip. J Bone Joint Surg Am. 1993;75(5):643–9.
119. Karlen JW et al. The Dega osteotomy: a versatile osteotomy in the treatment of developmental and neuromuscular hip pathology. J Pediatr Orthop. 2009;29(7):676–82.
120. Aksoy C et al. Evaluation of acetabular development after Dega acetabuloplasty in developmental dysplasia of the hip. J Pediatr Orthop B. 2013;22(2):91–5.
121. Haidar RK et al. Simultaneous open reduction and Salter innominate osteotomy for developmental dysplasia of the hip. J Bone Joint Surg (Br). 1996;78(3):471–6.
122. Grudziak JS, Ward WT. Dega osteotomy for the treatment of congenital dysplasia of the hip. J Bone Joint Surg Am. 2001;83-A(6): 845–54.
123. Szepesi K et al. Pemberton's pericapsular osteotomy for the treatment of acetabular dysplasia. J Pediatr Orthop B. 1996;5(4): 252–8.
124. Aydin A et al. The results of Pemberton's pericapsular osteotomy in patients with developmental hip dysplasia. Acta Orthop Traumatol Turc. 2012;46(1):35–41.

125. El-Sayed M et al. The effect of Dega acetabuloplasty and Salter innominate osteotomy on acetabular remodeling monitored by the acetabular index in walking DDH patients between 2 and 6 years of age: short – to middle-term follow-up. J Child Orthop. 2013;6(6):471–7.
126. Lopez-Carreno E, Carillo H, Gutierrez M. Dega versus Salter osteotomy for the treatment of developmental dysplasia of the hip. J Pediatr Orthop B. 2008;17(5):213–21.
127. Schoenecker PL, Strecker WB. Congenital dislocation of the hip in children. Comparison of the effects of femoral shortening and of skeletal traction in treatment. J Bone Joint Surg Am. 1984;66(1):21–7.
128. Westin GW, Ilfeld FW, Provost J. Total avascular necrosis of the capital femoral epiphysis in congenital dislocated hips. Clin Orthop Relat Res. 1976;119:93–8.
129. Westin GW et al. Skeletal traction vs. femoral shortening in treatment of older children with congenital hip dislocation. Isr J Med Sci. 1980;16(4):318–22.
130. Galpin RD et al. One-stage treatment of congenital dislocation of the hip in older children, including femoral shortening. J Bone Joint Surg Am. 1989;71(5):734–41.
131. Yagmurlu MF et al. Clinical and radiological outcomes are correlated with the age of the child in single-stage surgical treatment of developmental dysplasia of the hip. Acta Orthop Belg. 2013;79(2):159–65.
132. Umer M et al. Outcome of triple procedure in older children with developmental dysplasia of hip (DDH). J Pak Med Assoc. 2007;57(12):591–5.
133. Inan M et al. Successful treatment of high congenital dislocated hips in older children by open reduction, pelvic and femoral osteotomy with external fixator stabilization (average 8.2 years of age). J Pediatr Orthop B. 2005;14(6):405–9.
134. Klisic P, Jankovi L, Basara V. Long-term results of combined operative reduction of the hip in older children. J Pediatr Orthop. 1988;8(5):532–4.
135. Klisic P, Jankovic L. Combined procedure of open reduction and shortening of the femur in treatment of congenital dislocation of the hips in older children. Clin Orthop Relat Res. 1976;&NA(119):60–9.
136. Vallamshetla VR, Mughal E, O'Hara JN. Congenital dislocation of the hip. A re-appraisal of the upper age limit for treatment. J Bone Joint Surg (Br). 2006;88(8):1076–81.
137. Ning B et al. Analyses of outcomes of one-stage operation for treatment of late-diagnosed developmental dislocation of the hip: 864 hips followed for 3.2–8.9 years. BMC Musculoskelet Disord. 2014;15:401.
138. El-Tayeby HM. One-stage hip reconstruction in late neglected developmental dysplasia of the hip presenting in children above 8 years of age. J Child Orthop. 2009;3(1):11–20.

What Is the Best Treatment for Perthes' Disease?

5

Daniel Perry and David Bodansky

Abstract

There are few more enigmatic conditions than Perthes' disease. The aetiology, mechanisms and optimal treatments all remain the source of controversy. The wide variety of available treatments provides the clearest demonstration of the uncertainty amongst clinicians treating the disease. Treatment regimens vary considerably from years of not weight bearing, to months of abduction cast treatment, to femoral and pelvic surgical procedures, to observation alone without any specific intervention. The orthopaedic community has little understanding of how treatment influences the natural history of this disease. This review considers the best available evidence, to formulate an evidence-based approach.

Keywords

Perthes' disease • Legg-Calvé-Perthes disease • LCPD • Osteonecrosis • Avascular Necrosis • Hip • Arthritis

Introduction

Perthes' disease of the hip was described independently by Arthur T Legg (USA), Jacques Calvé (France) and Georg Perthes (Germany) in 1910. It is an idiopathic avascular necrosis of the proximal femoral epiphysis, which results in flattening of the femoral head. The child presents with a pain and/or a limp. Perthes' disease has a cumulative incidence of approximately 1 in 1200 individuals with a male preponderance (5 male: 1 female) [1, 2].

A wide range of treatments have been used, from observation alone, to restrictions on weight bearing, to physiotherapy, to a variety of braces to constrain the femoral head within the acetabulum, to surgery [3–6]. Surgical interventions most commonly include a femoral varus osteotomy, an innominate osteotomy or both. There is no treatment consensus and a lack of controlled or randomised studies. Most evidence is retrospective with few prospective studies. Here, we review the options for treatment, focusing primarily on studies with more robust methodology, i.e. levels of evidence I or II.

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The Radiographic Stage, Outcome and Grades of Disease

A clear awareness of the radiologic descriptors used is fundamental to understand the treatment of Perthes' disease. The terms that describe the stage, grade and outcome should not be confused. Each radiographic term should be considered when considering the evidence and optimal treatment regimen.

Radiographic Stage

Perthes' disease progresses through several radiologic stages in the disease cycle described by Waldenström: 'initial', 'sclerosis', 'fragmentation', 'reossification' and 'healed' [7]. The stage of disease is important because the head of the femur is plastic and deformable in the early stages of disease, and is not so once healing has occurred. Perhaps

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unsurprisingly there is evidence to suggest that intervening early in the disease (i.e. prior to deformation of the femoral head), has better outcomes than intervening late [8]. These stages have recently been further refined by a collaborative international group into subcategories of each stage, which are a useful research tool though should not detract from the day-to-day utility of Waldenström's descriptions [9].

Radiographic Outcomes

The radiographic outcomes generally refer to the hip shape at skeletal maturity. Classically, hip shape is defined using the Stulburg classification, a qualitative measure of hip shape that broadly divides hips into those that are 'spherical', 'aspherical' and 'flat' [10, 11]. A simple algorithm to classify hips according to Stulberg is illustrated in Fig. 5.1, however there remains debate about the reproducibility of Stulberg outcomes as inter-rater reliability is generally poor [10, 12].

More recently, hip shape has been defined using a quantitative measure of sphericity, termed the sphericity deviation score. This has been shown to closely correlate with the Stulberg outcome [13]. Quantitative measures of outcome, if reproducible, have particular advantages over qualitative measures in terms of greater efficiency in powering future studies of outcomes.

Radiographic Grade

The grade of disease is an attempt to predict long-term outcome, based on the radiographic appearance of the hip at a particular point in the cycle of disease. The grade of the disease is perhaps where greatest controversy exists. There are classifications that are used to help determine the prognosis: Catterall, Salter-Thomson, and Herring.

The Catterall classification is an assessment of the extent of the radiological involvement of the femoral head on the lateral radiograph [14]. Greater involvement of the head indicates a greater severity of disease, and a worse prognosis. Catterall suggested that involvement of >50 % of the head has poorer outcomes, compared to those with <50 % head involvement. However, the classification may only be applied at the point of maximal collapse, which is usually some months into fragmentation.

The Salter-Thompson classification recognises that approximately 30 % of hips will have a subchondral fracture early in disease [15]. If evident, a subchondral fracture reliably delineates the amount of collapse anticipated, which therefore maps onto Catterall's descriptors, i.e. a subchondral fracture >50 % indicates a poor prognosis.

The Herring lateral pillar classifications assesses the integrity of the lateral portion of the epiphysis (Fig. 5.2). Herring et al. broadly described that hips with no lateral column collapse have good outcomes ('Herring A Hips'),

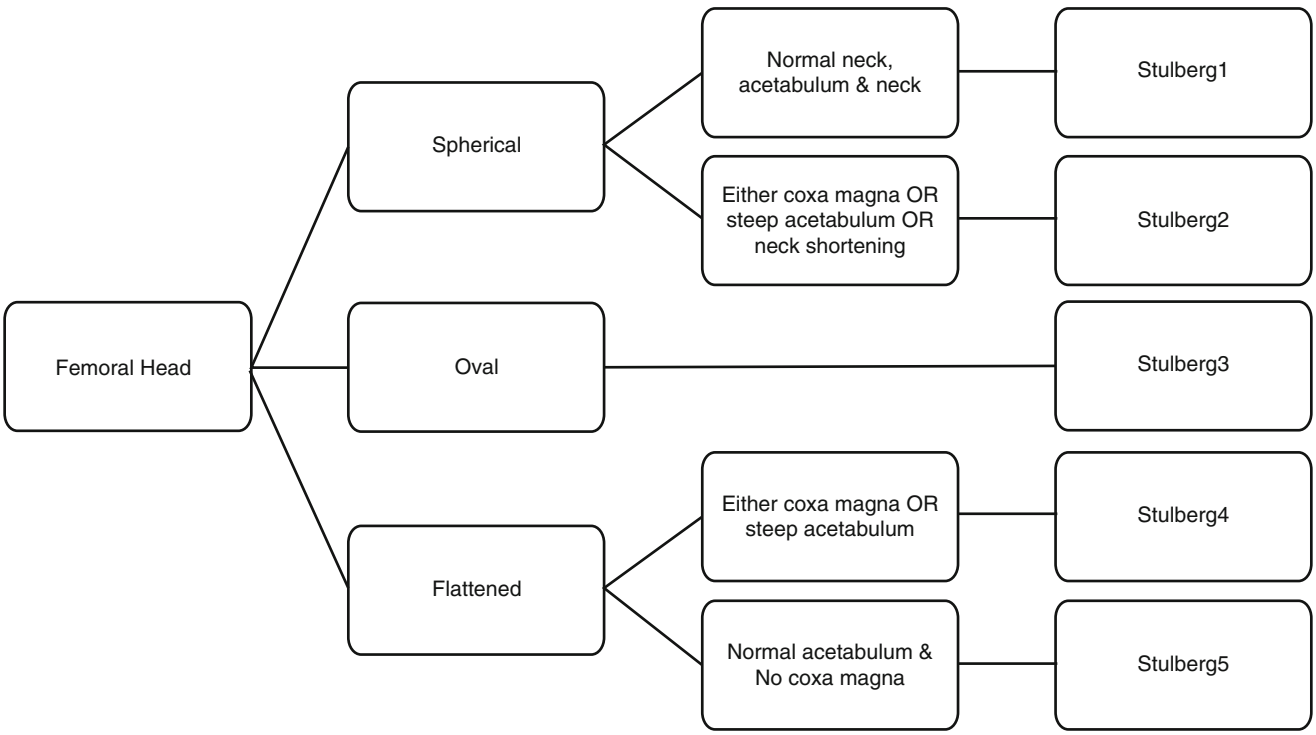

Fig. 5.1 Algorithm to determine the Stulberg outcomes in Perthes' disease (Adapted from [12])

and those with greater than 50 % of lateral column collapse have poor outcomes (Herring 'C' Hips). Those with some collapse though less than 50 % (Herring 'B' Hips), may be amenable to intervention. However, like the Catterall classification, this may only be applied at the point of maximal collapse, which is some months into fragmentation [16].

Radiographic grades of disease may therefore be useful to guide treatment; however, they are most useful at the end of the radiographic stage of collapse. There is also concern over the reproducibility of these descriptors [17, 18].

Early Predictors of Prognosis

In addition to the radiographic appearances of the hip there are other factors that must be considered when determining the eventual prognosis.

There is general consensus that younger children have better outcomes than older children. An arbitrary cut-off of 6 years old is often used, suggesting that children under 6 years have a good prognosis, and those over 6 years have a poor prognosis. This is clearly an oversimplification with many reports of young children with poor outcomes [19–22].

The sex of the child also appears to have a bearing on the prognosis. Boys generally have better outcomes than girls, which is thought to be a consequence of girls having greater skeletal development at any given chronologic age [14, 17].

The range of motion of the hip is also widely believed to have a bearing on prognosis, though it has seldom been investigated. A recent paper investigating outcomes amongst those under 6-years old, suggested range of motion to be more important than radiographic grade in predicting outcome [19].

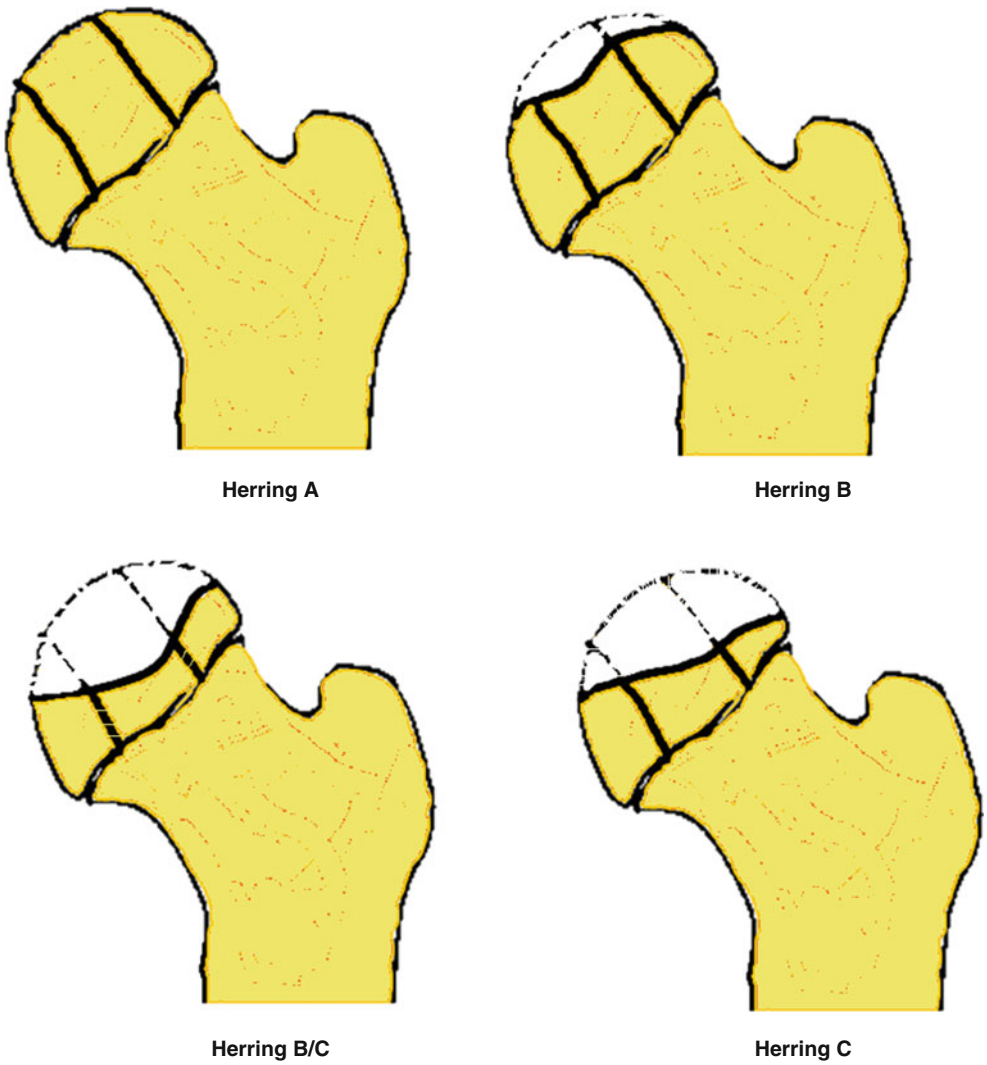

Fig. 5.2 Herring classification

Considerations When Forming the Treatment Plan

When considering the treatment of the patient one must therefore consider their age, sex, range of motion and the radiographic stage of disease. The radiographic grade is important; the optimal time to intervene may before significant deformity occurs, i.e. prior to the fragmentation stage because significant deformity has already occurred at this stage. It can therefore be argued that classifying a hip according to Catterall and Herring therefore occurs too late to guide treatment, although the presence of a subchondral fracture described by Salter-Thompson may have some useful bearing on the decision.

Critical Appraisal of the Evidence for the Management of Perthes' Disease

We searched for studies that have reached level I or II evidence, i.e. prospectively collected cases with pre-defined inclusion and exclusion criteria and predefined outcomes, with or without an intervention.

There is no level I evidence in the management of Perthes' disease, although there is an ongoing randomised controlled trial in Australia. This study is addressing whether the novel intervention of systematic bisphosphonate administration may prevent femoral head collapse [23].

There are two important level two studies addressing the question "What is the best treatment for Perthes' disease?"

In 2004, Herring et al. reported findings from a prospective, multicentre study in North America and New Zealand. Thirty-nine paediatric surgeons enrolled 438 patients between 6 and 12 years of age across five treatment arms [17]. Hips that had reached the reossification stage at presentation were excluded. Stratification of disease severity was undertaken using the lateral pillar classification, and outcomes were defined according to Stulberg. 345 hips in 337 patients were followed to skeletal maturity. Each surgeon agreed to employ one of five treatment approaches for patients under their care (either no specific treatment, range of motion exercises, brace, Salter pelvic osteotomy or varus femoral osteotomy) (Figs. 5.3 and 5.4).

The majority (218, 63 %) of cases were Herring type B and 87 % were younger than eight at onset. The study found no superiority between the three non-surgical treatment options: range of motion exercises, Atlanta braces and no treatment. Further, no radiological difference was seen between femoral osteotomy and innominate osteotomy. Overall, patients receiving surgery had better outcomes than those treated non-operatively ($p = 0.02$), in particular those patients with Herring B and those older than eight at diagnosis. Herring C patients with severe disease (60, 17 %) generally had poor outcomes regardless of treatment. A new subgroup of the Herring classification was used to describe hips that were exactly 50 % collapse, or with a number of features deemed worse than a typical 'Herring B' hip – "the B/C border hip". A summary of the recommendations from Herring is available in Table 5.1.

This study is useful, however prone to selection bias as no attempt was made to randomise treatments. Although there were assurances that surgeons would employ a single treatment protocol, some patients crossed-over to other treatment regimens demonstrating that surgeons did not employ a single treatment supporting the possibility of selection bias. Eighty-percent of the hips operated on during this study received surgery prior to fragmentation, therefore prior to the maximum extent of lateral pillar collapse becoming apparent. Consequently it is not possible to advocate waiting for the lateral pillar collapse to become apparent based on this study. Perhaps the biggest criticism of this study is that there were no a-priori hypotheses relating to the grade of disease, which is particularly important to those hips that were newly classified to strengthen the statistical conclusions – the 'B/C border hips' [24]. The lateral pillar classification was chosen over Catterall and Salter-Thompson because it had a stronger correlation with outcome, and was therefore selected because of its statistical significance. However criticism can be made when using a dataset to derive a classification and then using this classification to make sense of the findings in the same dataset. This is therefore a hypothesis generating approach, rather than a hypothesis testing method. The overarching purpose of the study was to demonstrate the benefit of surgery, yet the results were far from conclusive.

Wiig and colleagues undertook a national Norwegian prospective study between 1996–2000 [25]. In this instance, 368 unilateral cases were assigned to treatment according to the surgeon's selection; to physiotherapy, bracing or a femoral varus osteotomy. 358 were available for outcome assessment at 5 years. Radiographs taken at one and 5 years of the 345 patients achieving the 5 year follow up were reviewed separately considering the lateral pillar grade (excluding the B/C border subset), a modified two group Catterall grade (necrosis comprising less or more than 50 % of the femoral head) and Stulberg outcomes.

This study demonstrated that a simplified two-group Catterall grade was reproducible, and was the strongest predictor of Stulberg outcomes. Age was the next strongest predictor of outcome, followed by the lateral pillar classification score [25].

There were 152 patients older than six with more than 50 % femoral head necrosis. Of these, 55 (36 %) received physiotherapy, 26 (17 %) a brace and 71 (47 %) surgery.

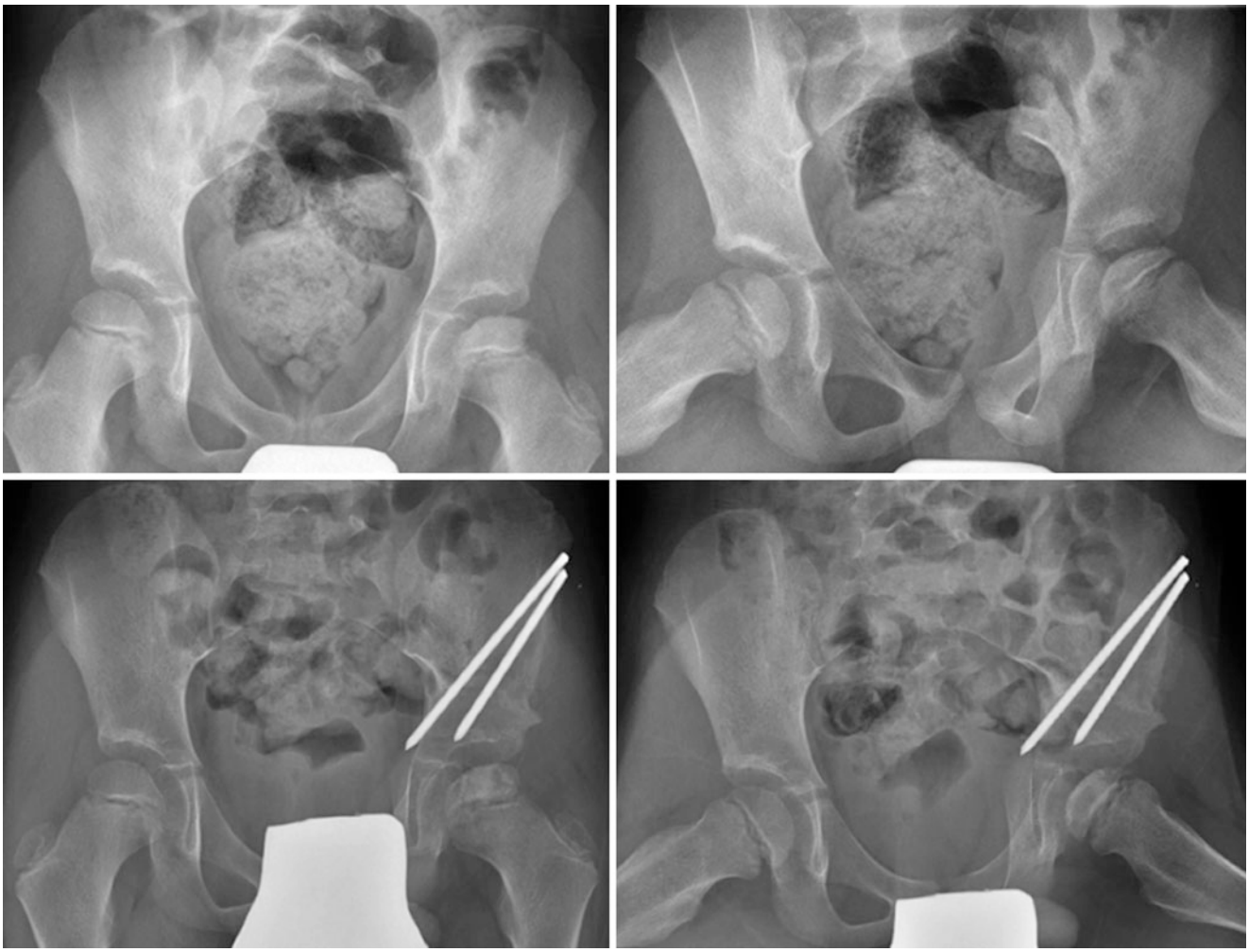

Fig. 5.3 Containment surgery using Salter's pelvic osteotomy

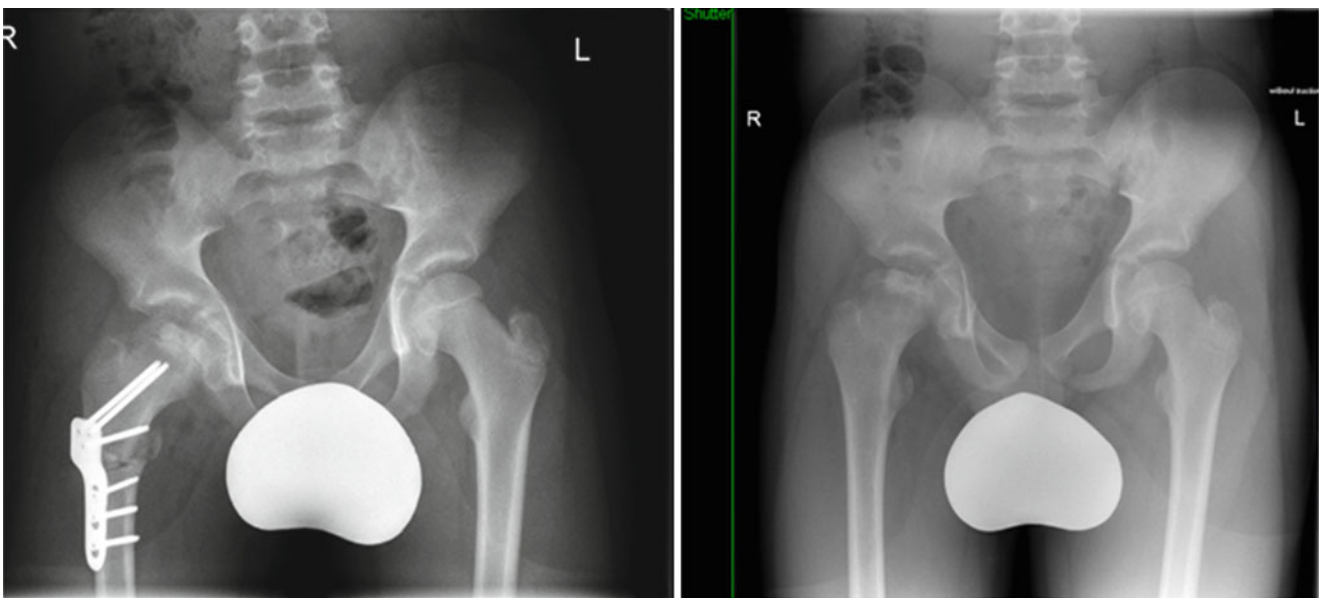

Fig. 5.4 Containment surgery using femoral osteotomy

There was no difference in the Stulberg outcomes between patients receiving physiotherapy or a brace (p = 0.36). However, those patients receiving surgery had better outcomes than either physiotherapy (p = 0.001) or a brace (p = 0.001). For patients younger than six, there was no outcome superiority between intervention and no intervention (p = 0.73). This study did not identify a difference in outcome by gender, which differed to previous studies.

This study therefore demonstrated an overall benefit of surgery amongst older children (>6 years old). More severe disease appeared to have the greatest benefit of intervention. As per the study of Herring et al., a prospective cohort of this nature is not immune to bias, yet a priori hypotheses did appear more readily apparent. It was therefore of interest that the utility of the lateral pillar classification was less than demonstrated by Herring, instead offering strong support the Catterall classification.

Ancillary Evidence

Evidence from studies of lower levels of evidence up to 2012 were summarised well by meta-analysis, seeking to address whether operative or non-operative treatment is the optimal intervention for Perthes' disease. This meta-analysis included 1232 patients (1266 hips) across 23 studies [26]. Here, 783 hips were treated non-operatively and 483 received surgery. This study supported the suggestion that children under six at diagnosis were as likely to have a good radiographic outcome regardless of treatment approach. It also suggested that after controlling for age, sex and disease severity, those treated with operative interventions were about twice as likely to have a good outcome (OR 2.0 (95 % CI 1.3–3.0), although there did not appear any clear difference between the choice of operative intervention chosen.

Meta-analyses of lower level evidence are clearly fraught with bias, with each of the included studies introducing additional challenges. Nevertheless, the findings are supportive of the level II evidence and therefore are of value. A summary of recommendations and their evidence is shown in Table 5.2.

Conclusion

There is a paucity of high-level evidence for the treatment of Perthes' disease, with only two level II studies that have begun to adequately addressed the question of 'operative or non-operative treatment'. The results appear to suggest that younger children do well irrespective of the intervention chosen, although the age of six has only arbitrarily been chosen as the 'cut-off'. It is unclear if hip stiffness has any prognostic significance. A number of prognostic grading systems have been used in the disease, although there is no certainty as to which is most reliable. Indeed there is suggestion that surgery may be most useful very early in disease, which is prior to any radiographic grading system becoming apparent. A general treatment approach may therefore be to offer surgery to all older children early in disease, irrespective of the radiographic appearance. There is no evidence to suggest of superiority of one surgical intervention over others. Research is urgently required to develop an early prognostic grading system for Perthes' disease. However, more crucially, a pragmatic trial is needed to investigate whether operative or non-operative interventions offer the optimal outcome in early Perthes' disease.

Table 5.1 Suggested treatment options dependent on severity of disease and age of onset

Age of onset	Lateral pillar A	Lateral pillar B	Lateral pillar B/C	Lateral pillar C
Below 8 years	Good outcome	No superiority between treatment options	No superiority between treatment options	Poor outcome independent of treatment option
Above 8 years	Good outcome	Osteotomy	Osteotomy	Poor outcome independent of treatment option

Table 5.2 Recommendations

Statement	Grade of recommendation
Patients with significant collapse (such as lateral pillar type C disease) will have a poor outcome, regardless of treatment	B
The optimal outcomes are achieved if surgery is undertaken prior to maximal fragmentation	B
Patients older than eight at diagnosis should receive a surgical intervention	C
There is no superiority between femoral osteotomy and innominate osteotomy	B
There is no superiority between different types of braces and no treatment at all	C
Younger children will have a good outcome regardless of treatment	B

References

1. Perry DC1, Bruce CE, Pope D, Dangerfield P, Platt MJ, Hall AJ. Legg-Calvé-Perthes disease in the UK: geographic and temporal trends in incidence reflecting differences in degree of deprivation in childhood. Arthritis Rheum. 2012;64:1673–1679.
2. Perry DC, Machin DM, Pope D, Bruce CE, Dangerfield P, Platt MJ, Hall AJ. Racial and geographic factors in the incidence of Legg-Calvé-Perthes' Disease: a systematic review. Am J Epidemiol. 2012;175(3):159–66.
3. Harrison H, Turner M, Nicholson F. Coxa plana: results of a new form of splinting. J Bone Joint Surg Am. 1969;51:1057–69.
4. Bobechko WP. The Toronto brace for Legg-Perthes disease. Clin Orthop Relat Res. 1974;102:115–7.
5. Purvis J, Dimon 3rd JH, Meehan PL, Lovell WW. Preliminary experience with the Scottish Rite Hospital abduction orthosis for Legg-Perthes disease. Clin Orthop Relat Res. 1980;150:49–53.
6. Martinez AG, Weinstein SL, Dietz FR. The weight-bearing abduction brace for the treatment of Legg-Perthes disease. J Bone Joint Surg Am. 1992;74:12–21.
7. Waldenström H. The Definite Form of the Coxa Plana. Acta Radiol. 1922;1:384.
8. Joseph B. Management of Perthes' disease. Indian J Orthop. 2015;49:10–6.
9. Hyman JE, Trupia EP, Wright ML, Matsumoto H1, Jo CH, Mulpuri K. Interobserver and intraobserver reliability of the modified Waldenström classification system for staging of Legg-Calvé-Perthes disease. J Bone Joint Surg Am. 2015;97:643–650.
10. Wiig O, Terjesen T, Svenningsen S. Inter-observer reliability of the Stulberg classification in the assessment of Perthes disease. J Child Orthop. 2007;1:101–5.
11. Stulberg S, Cooperman D, Wallensten R. The natural history of Legg-Calvé-Perthes disease. J Bone Joint Surg Am. 1981;63:1095–108.
12. Neyt J, Weinstein SL, Spratt KF, Dolan L, Morcuende J, Dietz FR, et al. Stulberg classification system for evaluation of Legg-Calvé-Perthes disease: intra-rater and inter-rater reliability. J Bone Joint Surg Am. 1999;81:1209–16.
13. Shah H, Siddesh ND, Pai H, Tercier S, Joseph B. Quantitative measures for evaluating the radiographic outcome of Legg-Calvé-Perthes disease. J Bone Joint Surg Am. 2013;95:354–61.
14. Catterall A. The natural history of Perthes' disease. J Bone Joint Surg Br. 1971;53:37–53.
15. Salter RB, Thompson GH. Legg-Calvé-Perthes disease. The prognostic significance of the subchondral fracture and a two-group classification of the femoral head involvement. J Bone Joint Surg Am. 1984;66(4):479–89.
16. Herring JA, Kim HT, Brown R. The lateral pillar classification of Legg-Calvé-Perthes disease. J Pediatr Orthop. 1992;12:143–50.
17. Herring JA, Kim HT, Browne R. Legg-Calvé-Perthes disease. Part II: prospective multicenter study of the effect of treatment on outcome. J Bone Joint Surg Am. 2004;86:2121–34.
18. Mahadeva D, Chong M, Langton DJ, Turner AM. Reliability and reproducibility of classification systems for Legg-Calvé-Perthes disease: a systematic review of the literature. Acta Orthop Belg. 2010;76(1):48–57.
19. Nakamura J, Kamegaya M, Saisu T, Kakizaki J, Hagiwara S, Ohtori S. Outcome of patients with Legg-Calvé-Perthes onset before 6 years of age. J Pediatr Orthop. 2015;35(2):144–50.
20. Rosenfeld S, Chao J, Herring J. Legg-Calve-Perthes disease: a review of cases with onset before six years of age. J Bone Joint Surg Am. 2007;89(12):2712–22.
21. Canavese F, Dimeglio A. Perthes disease: prognosis in children under six years of age. J Bone Joint Surg Br. 2008;90(7):940–5.
22. Schoenecker P, Stone J, Capelli A. Legg-Perthes disease in children under 6 years old. Orthop Rev. 1993;22(2):201–8.
23. Cowell C. Australian New Zealand Clinical Trials Registry: avascular Necrosis due to Perthes disease. 2009; cited 2015; Available at: https://www.anzctr.org.au/Trial/Registration/TrialReview.aspx?id=335166
24. Herring J, Kim HT, Browne R. Legg-Calvé-Perthes disease. Part I: classification of radiographs with use of the modified lateral pillar and Stulberg classification. J Bone Joint Surg Am. 2004;86(10):2103–20.
25. Wiig O, Terjesen T, Svenningsen S. Prognostic factors and outcome of treatment in Perthes' disease: a prospective study of 368 patients with five-year follow-up. J Bone Joint Surg Br. 2008;90-B(10):1364–71.
26. Nguyen N, Klein G, Dogbey G, McCourt JB, Mehlman CT. Operative versus nonoperative treatments for Legg-Calvé-Perthes disease: a meta-analysis. J Pediatr Orthop. 2012;32:697–705.

Evidence-Based Treatment for Slipped Upper Femoral Epiphysis

6

Sattar Alshryda, Kai Tsang, and Gavin De Kiewiet

Abstract

Slipped upper femoral epiphysis (SUFE), though not common, is an important paediatric disorder. It has a reported incidence of 1–10 per 100,000. Some aspects of management of SUFE are controversial and evolving with advancing surgical skills and expertise. The infrequency of cases, the various classifications in use, the various surgical treatments, and lack of robust evidence for outcomes, has resulted in the lack of clear, evidence-based recommendations for treatment. The following review examined the current evidence for treating SUFE and concluded that pinning in situ is the best treatment for mild and moderate stable slip (grade B). Surgical dislocation may give better results than pinning in situ for severe stable slip (grade C). Urgent gentle reduction, capsulotomy and fixation is the best current treatment for unstable slip (grade C). Routine prophylactic pinning of the contralateral asymptomatic side is not recommended (grade C)

Keywords

SUFE • SCFE • Slipped • Stable slip • Unstable slip • Loders classification • AVN • Osteonecrosis • FHO • Slipped upper femoral epiphysis

Background

Slipped upper femoral epiphysis (SUFE) is one of the most important paediatric and adolescent hip disorder. Incidence is 1–10:100,000. Patients usually presented with painful hip and or knee with affected leg is short and externally rotated (Fig. 6.1). The plain x-ray is usually diagnostic (Fig. 6.2). The cause is poorly understood, it is believed that increased shear forces and/or weak growth plate (the physis) in adolescence predispose to slips.

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Although rare, endocrine disorders must be considered in every patient with SUFE. Loder [1] identified two types of SUFE; idiopathic type and atypical type where there is an underlying endocrine disorders or other aetiology. He studied the demographics of 433 patients with 612 SUFEs (285 idiopathic, 148 atypical) and found that weight and age were predictors for atypical SUFE and he recommended the *age-weight test:* the test was defined as negative when age younger than 16 years and weight ≥50th percentile and positive when beyond these boundaries. The probability of a child with a negative test result having an idiopathic SUFE was 93 %, and the probability of a child with a positive test result having an atypical SCFE was 52 %.

Slipped upper femoral epiphysis was traditionally classified as (1) pre-slip: patient has symptoms with no anatomical displacement of the femoral head, (2) acute: there is an abrupt displacement through the proximal physis with symptoms and signs developing over a short period of time (<3 weeks), (3) Chronic: present with pain in the groin, thigh, and knee of more than 3 weeks, often ranging from months to years and (4) acute on chronic: initially, patient

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has chronic symptoms, but develops acute symptoms as well following a sudden increase in the degree of slip [2, 3].

However, in a classic paper by Loder [4, 5] a new, clinically more relevant classification was introduced. SUFE was classified based on the patient weight-bearing status into stable when patient is able to ambulate and bear their weight and unstable when patient is unable to ambulate with or without crutches. In his series of 55 SUFEs, Loder showed that avascular necrosis (AVN) developed in 47 % of unstable slips but none of stable hips. However, unintentional reduction of the slip occurred in 26 unstable slips (out of 30) and in only 2 of the stable slips (out of 25). [4]. Several other papers confirmed Loder's findings [6, 7–9].

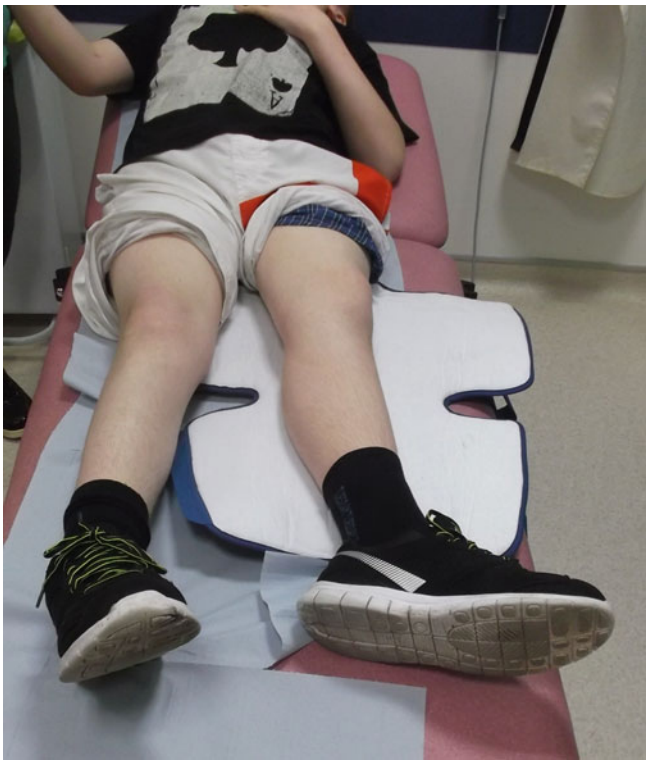

Fig. 6.1 A child with SUFE. A clinical photograph of a child with SUFE, notice the short and externally rotated left leg (mimic fracture neck of femur). Patient was investigated and treated for knee pain

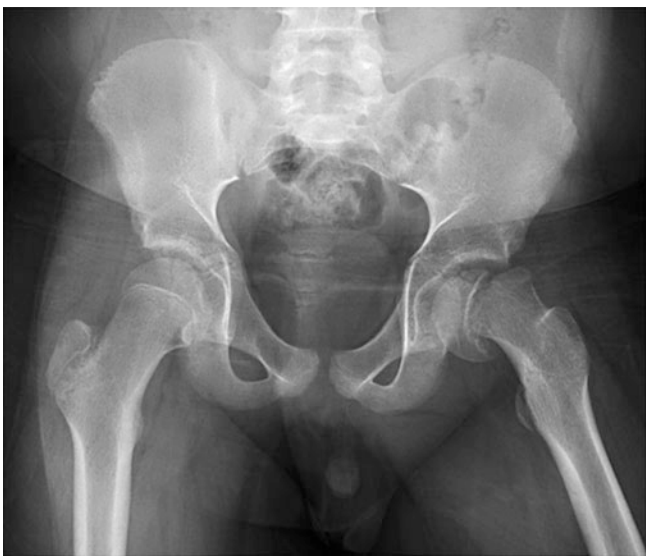

Fig. 6.2 A pelvis x-ray with left slipped upper femoral epiphysis. The x-ray shows a severe slipped upper epiphysis

Grading the severity of the slip is usually based on the radiographic findings. The Southwick angle is the most commonly used [10]. The angle is measured on the lateral view of the both hips by drawing a line perpendicular to a line connecting the posterior and anterior tips of the epiphysis at the physis. The angle between the perpendicular line and the femoral shaft line is called the lateral epiphyseal shaft angle. The Southwick angle is the difference between the lateral epiphyseal shaft angle of the slipped and the non slipped sides (Fig. 6.3). In patients with bilateral involvement, 12° is subtracted from each of the measured lateral epiphyseal angles. Mild slip (grade I) has an angle difference of less than 30°, moderate slip (grade II) has an angle difference of between 30 and 50 degrees and severe slip has a difference of over 50 degrees.

Treatment aim is to prevent progression of the slip without complications. Reduction of the slip to near anatomical position is desirable but this is tempered by the higher risk of AVN and chondrolysis (CL) which are surrogates for bad outcomes. The choice of treatment depends on the type of slip, its severity, and surgical expertise.

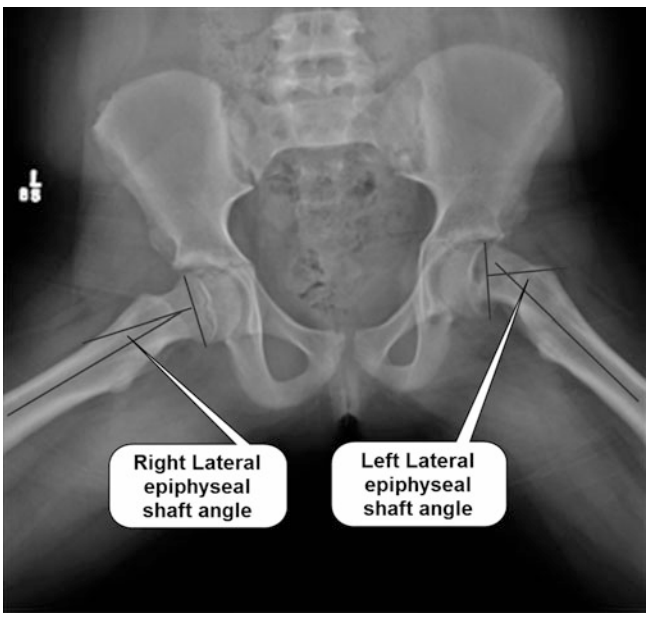

Fig. 6.3 SUFE radiological grading. The Southwick angle is the difference between the lateral epiphyseal shaft angle of the slipped and the non slipped sides. Mild slip (grade I) has an angle difference of less than 30°, moderate slip (grade II) has an angle difference of between 30° and 50° and severe slip has a difference of over 50°

What Is the Best Treatment for a Stable Slip?

There is a consensus that the best treatment for mild and most moderate stable slip is pinning-in-situ (PIS) using a single cannulated screw (SS). This has been supported by a comprehensive review paper by Loder [11]. If the slip is severe, pinning can be technically difficult. Gentle reduction is often unsuccessful in a stable slip and forceful reduction is contraindicated as this increase the risk of AVN. The options are either PIS with re-alignment procedure later if remodeling is suboptimum or primary corrective osteotomy.

Realignment procedures can be performed at one of three levels: subcapital, femoral neck and intertrochanteric region. The ability to correct a deformity is greatest with subcapital osteotomy (where the CORA is), least with an intertrochanteric osteotomy. The risk of AVN is the highest with subcapital osteotomy and the lowest with intertrochanteric osteotomy.

We performed an extensive literature search for the best available evidence to support various treatments of stable slips. We could not find level I or II evidence. There were 16 comparative studies and several case series with a follow-up more than a year. With a few exceptions all these studies were unmatched; mild and moderate slips were treated with pinning whereas severe slips were treated with reduction (either close or open reduction) and stabilisation undermining the comparison between pinning in situ and reduction.

Tables 6.1 and 6.2 show that pinning using a single screw has the lowest rates of AVN and chondrolysis (CL) and even a better patient's satisfaction when compared with traditional corrective osteotomies namely Dunn's and Fish osteotomies. One point needs further emphasis that patient who had corrective osteotomies were more likely to have severe slips and their outcomes are less favourable anyway.

In the last two decades, the femoro-acetabular impingement (FAI) has become widely recognized as an orthopaedic condition that requires treatments to prevent future osteoarthritis (OA) and premature artificial hip replacement. Ganz (Ganz et al. [54, 55] has been a pioneer in spreading the understanding of the condition and its treatment. He described a new technique of surgical dislocation of the hip involving tro-

Table 6.1 Pooled summary of studies of stable slips treatments

Intervention	Hips	AVN (%)	CL (%)	Satisfactory patients result[a]
Hip spica	101	8 (7.9 %)	21 (20.8 %)	NR
Epiphysiodesis	485	14(2.9 %)	8 (1.6 %)	67 (67 %) excellent 6 (6 %) good 10 (10 %) fair 7 (7 %) poor 7 (7 %) failure
Pinning using single screw	525	8(1.5 %)	12 (2.3 %)	113 (47 %) excellent 86 (36 %) good 19 (8 %) fair 10 (4 %) poor 11 (5 %) failure
Pinning using multiple pins	273	6(2.2 %)	11(4 %)	76 (67 %) excellent 19 (17 %) good 0 (0 %) fair 16 (14 %) poor 3 (3 %) failure
Physeal osteotomy	545	63(11.6 %)	51 (9.4 %)	131 (28 %) excellent 210 (45 %) good 46 (10 %) fair 72 (16 %) poor 3 (6 %) failure
Ganz surgical dislocation	81	3(3.7 %)	2 (2.5 %)	52 (87 %) excellent 2 (3 %) good 0 (0 %) fair 5 (8 %) poor 1 (2 %) failure
Base of neck osteotomy	92	2(2.1 %)	6 (6.5 %)	22 (60 %) excellent 11 (30 %) good 2 (5 %) fair 2 (5 %) poor
Inter-trochanteric osteotomy	336	5 (1.5)	16 (4.8 %)	121 (44 %) excellent 105 (38 %) good 35 (13 %) fair 15 (5 %) poor

[a]Satisfactory patients result based on closely related rating such as Heyman and Herndon classification, Harris hip score or Iowa hip scores.

Table 6.2 studies of various interventions in stable slips

Study	Rx	Patients	Hips	AVN	CL	FAI	OA	Patient satisfaction	Others	Notes
Hip Spica										
Betz [12]	Spica	32	37	0	5	NR	NR	NR		0 acute, 8 acute on chronic and 29 chronic 25 mild, 7 moderate and 5 severe All stable slips
Carney (Carney et al. [13])	Spica		47	8	6			IHS for chronic slips 81 and 71for acute slips		4 acute and 43 chronic. Spica with closed reduction (16 hips) resulted in a mean IHS of 65 points, 6 AVN and 2 CL. Spica cast without reduction (26) resulted in a mean IHS of 83 points, 2 AVN and 4 CL.
Meier (Meier et al. [14])	Spica	13	17	NR	10	NR	9	NR	3 pressure sores 3 Further slipping	
Total		NA	101	8	21					
Epiphysiodesis										
Adamczyk (Adamczyk et al. [15])	Epi	225 (+43)	278 (+45)	4 (+3)	0 (+1)	NR	NR	NR	17 further slipping (+6) 4 deep infection 12 re-operation	45 acute, 0 acute on chronic and 278 chronic Outcomes of acute slips are bracketed
Rao (Rao, et al. [16])	Epi	43	46	3	2	NR	NR	NR	3 infections 7 cases of transient anterolateral thigh hypesthesia 44 hetertopic ossification.	18 unstable (excluded) and 46 stable slips. The average operating time and blood loss per hip were 122 ± ?4 min and 426 ± 238 ml, respectively.
Schmidt (Schmidt et al. [17])	Epi	33	40	1	1	NR	NR	HHS 35 excellent 1 good 2 fair 2 poor	1 femoral neck fracture 1 subt-rochanteric hip fracture 2 coxa vara	31 mild, 9 moderate, 0 severe. 6 unstable and 34 were stable. The average time 1 h 57 min and blood loss averaged 360 ml.
Szypryt (Szypryt et al. [18])	Epi	25	30	2	3	NR	7	MSC 12 excellent 5 good 8fair 4 poor	3 wound infection	1 acute, 13acute-on-chronic, 16 chronic. 0 mild, 12 moderate, 18 severe
Zahrawi (Zahrawi et al. [19])	Epi		28	0	0	NR	NR	HHC 20 excellent 0 good 0 fair 1 poor 7 failure	4 wound infection 2 graft failure 1 further slipping 6 needed further surgery	Severity (mean slip angle 30) LOS 21 Duration of surgery 150 min Blood loss 500 ml

Total		NA	467	13	7			67 (67 %) excellent 6 (6 %) good 10 (10 %) fair 7 (7 %) poor 7 (7 %) failure		
Pinning Using Screws										
Alshryda (Alshryda et al. [7])	Pinning(PIS) (SS)	36	36	1	1	NR	NR	NR	1 loss of fixation	Unstable and uncertain hips were excluded
Aronson [20]	Pinning(PIS) (SS)	34	43	1	0	NR	1	HHC 27 excellent 12 good 2 fair 2 poor	2 loss of fixation 1 Sub-trochanteric fracture 2 failed screw removals	6 acute and 37chronic. 27 mild, 8 moderate and 8 severe
Aronson [21]	Pinning(PIS) (SS)	44	58	1	0	NR	1	HHC 33 excellent 21 good 2 fair 2 poor	2 loss of fixation 1 Sub trochanteric fracture No further slipping.	8 acute, 0 acute-on-chronic, and 50 chronic. 38 mild, 10 moderate and 10 severe.
Blanco (Blanco et al. [22])	Pinning (PIS) (SS)	80	43	0	0	NR	NR	NR	2 Metalware problems 1 reoperation	1 acute , 6 acute on chronic, 36 chronic 23 mild, 12 moderate , 8 severe 1 CRIF
Carlioz [88]	Pinning (PIS) (SS)	34	38	0	2	NR	NR	31 good 10 fair 2 bad 3 Failure	1 Sub-trochanteric fracture	6 patients underwent reduction (1 AVN excluded). Authors did not use “Excellent” in outcomes
Gonzalez-Moran (Gonzalez-Moran et al. [23])	Pinning (PIS) (SS)	25	31	1	0	NR	NR	NR	1 wound infection 3 metalware problems	All received two weeks of skin longitudinal traction then pinning in situ without manipulation 22 case had a single screw and 9 had 2 screws 11 acute, 6 acute on chronic and 14 chronic 1 preslip, 17 mild, 11 moderate and 2 severe
Herman (Herman et al. [24])	Pinning(PIS) (SS)	11	11	0	1	NR	NR	HHS (95 points) 11 excellent 0 good 0fair 0 poor	No further slipping.	4 acute, 11 acute-on-chronic, and 6 chronic.
Kenny [25]	Pinning(PIS) (SS)	40	53	0	1	NR	1	HHC (31)58 % excellent (19)36 % good (2)4 % fair (1)2 % a poor	1 Sub-trochanteric fracture No further slipping.	3 acute, 8 acute-on-chronic and 35 chronic. 80 % mild, 12 % moderate and 2 % severe

(continued)

Table 6.2 (continued)

Study	Rx	Patients	Hips	AVN	CL	FAI	OA	Patient satisfaction	Others	Notes
Koval [26]	Pinning(PIS) (SS)	49	67	2	7	NR	2	NR	1 Growing off fixation 1 Stress fracture of the femoral neck	12 acute, 1 acute-on-chronic, 67 chronic. 55 mild, 19 moderate and 6 severe. 3 CRIF (1AVN)
Lim (Lim et al. [27])	Pinning (PIS) (SS)	13	13	1	0			Aadalen criteria 8excellent 2good 2fair 0 poor 1failure		All underwent preoperative traction All acute or acute on chronic Severity: mean 30° (range 0°-60°).
Novais (Novais et al. [28])	Pinning (PIS) (SS)	15	15	1				HHC 3 excellent 1good 1fair 3 poor 7failure	2 metalware problems 1 further slipping	All patients had stable severe slip revealed better deformity correction with the modified Dunn procedure compared with in situ pinning
Souder (Souder et al. [29])	Pinning (PIS) (SS)		64	0	0	NR	NR	NR	3 metalware problems 1 infection 1 further slipping	Ganz surgical dislocation 7 Unstable cause 3 AVN excluded
Ward (Ward et al. [30])	Pinning(PIS) (SS)	42	53	0	0	NR	NR	NR	Neither chondrolysis nor avascular necrosis developed. 1 HO 2 Metalware problems	2 acute, 3 acute-on-chronic and 48 chronic. 19 mild, 25 moderate and 9 severe 5 CRIF
Total (%)		NA	525	8	12	NA	0.02 %	113 (47 %) excellent 86 (36 %) good 19 (8 %) fair 10 (4 %) poor 11 (5 %) failure		
Pinning Using Multiple Pins										
Aronson [20]	Pinning (MPF)	39	54	2	3	NR	18	HHC 27 excellent 13 good 0 fair 13 poor 1Failure	13 patients had pin protruded through the back of the neck	4 acute and 50 chronic. 34 mild, 14 moderate and 6 severe
Blanco (Blanco et al. [22])	Pinning (MPF)		25	1	0	NR	NR	NR	8 Metalware problems 1 Growing off 4 reoperation	1 Preslip, 4 acute, 6 acute on chronic, 12 chronic 11 mild , 9 moderate, 4 severe. 7 CRIF

Carney (Carney et al. [13])	Pinning (MPF)		37	3	1	NR	NR	IHS for chronic slips 86 and 93 for acute slips		3 acute and 34 chronic. Reduction and pinning resulted in a mean ISH of 75 points, 2 AVN, 1 CL. pinning in situ resulted in a mean IHS of 85 points, 1 AVN, 0 CL.
Dreghorn (Dreghorn et al. [31])	PIS (MPF)		66	0	2	NR	0	NR	1 Growing off fixation	51 mild, 14 moderate and 1 severe
Gonzalez-Moran (Gonzalez-Moran et al. [23])	Pinning (MPF)	28	31	0	3	NR	NR	NR	4 wound infection 9 metalware problems	1 acute, 4 acute on chronic and 26 chronic 0 preslip, 15 mild, 12 moderate and 4 severe
Zahrawi (Zahrawi et al. [19])	PIS (MPF)		60	0	2	NR	NR	HHC 49 excellent 6 good 0 fair 3 poor 2 failure	2 metalware problems 1 further slipping 3 wound infection 2 needed further surgery	Severity (mean slip angle 22) Chronicity and stability NR LOS 17 Duration of surgery 90 min Blood loss 250 ml
Total			273	6	11			76 (67 %) excellent 19 (17 %) good 0 (0 %) fair 16 (14 %) poor 3 (3 %) failure		
Physeal Osteotomy										
Alshryda (Alshryda et al. [7])	PO	7	7	2	1	NR	NR	NR	Hip dislocation	15 unstable hips were excluded (5 AVN)
Barros [32]	PO	23	23	3	2	NR	NR	MSC 9 excellent 9 good 1 fair 4poor	1 metalware problem 0 infection	0 acute, 3 acute-on-chronic, 20 chronic. 0 mild, 0 moderate, 23 severe
Broughton (Broughton et al. [33])	PO	115	115	14	14	1	17	Overall 67 good 9fair 19poor		0 acute, 38 acute-on-chronic, 77 chronic. 0 mild, 15 moderate, 100 severe Patients satisfaction (G/F/B) in the acute-on-chronic (27/5/6); in the chronic with open growth plate (59/3/8) in the chronic slip with closed growth plate (1/1/5) .
Carlioz [88]	PO	26	27	0	3	NR	NR	20 good 3 fair 4 bad 3 Failure	Septic arthritis	

(continued)

Table 6.2 (continued)

Study	Rx	Patients	Hips	AVN	CL	FAI	OA	Patient satisfaction	Others	Notes
Carney (Carney et al. [13])	PO		14	3	6	NR	NR	IHS for chronic slips 76 and 50 for acute slips		26 moderate or severe slips
DeRosa(DeRosa et al. [34])	PO	23	27	4	8	NR	NR	MSC 0 excellent 19 good 4fair 4poor	2 loss of fixation	1 CRIF before PO went into AVN 0 mild, 0 moderate, 27severe
Dreghorn (Dreghorn et al. [31])	PO		3	1	0	NR	0	NR	1 wound infection	0 mild, 5 moderate and 6 severe
Diab (Diab et al. [35])	PO	11	11	2	0	1	NR	NR		
Dunn [36]	PO	69	73	9	3	NR	2	55 good 6 fair 12 poor		Several hips were manipulated under GA somewhere else (CRIF) 0 acute, 33 acute-on-chronic, 40 chronic.
Fish [37]	PO	61	66	3	1	NR	6	55 excellent 6 good 2fair 3poor		0 acute, 16 acute-on-chronic, 50 chronic. Chronic slips (0 mild, 23 moderate, 27 severe)
Fron (Fron et al. [38])	PO	46	50	6	3	NR	NR	34 excellent 10 good 2fair 4 poor	2 hematomas 2 infections 3 pseudarthroses of the greater trochanter 1 HO	0 acute, 17 acute-on-chronic, 30 chronic. 0 mild, 0 moderate, 50 severe
Jerre [39]	PO	22	22	5	1	NR	6	HHC 5 excellent 4 good 1 fair 8 poor	4 THR 1 Hip arthrodesis	1 acute, 1 acute-on-chronic, 20 chronic. 10 mild, 6 moderate, 0 severe, 6 none
Nishiyama [40]	PO	15	18	1	1			13 excellent 1good 1 fair 0poor		0 acute, 0 acute-on-chronic, 18 chronic. 0 mild, 0 moderate, 18 severe
Szypryt (Szypryt et al. [18])	PO	23	23	4	0	NR	5	MSC 15 excellent 2 good 1fair 4 poor	2 wound infection Metalware problems 10	1 acute, 16 acute-on-chronic, 6 chronic. 0 mild, 0 moderate, 23 severe
Velasco (Velasco et al. [41])	PO	65	66	6	8			22 good 16 moderate (fair) 10 poor		8 acute, 29 acute-on-chronic, 29 chronic. All moderate or severe (although table II showed that angles <30° in 5 hips) Full set data in 48 hips

Total			545	63	51			131 (28 %) excellent 210 (45 %) good 46 (10 %) fair 72 (16 %) poor 3 (6 %) failure		
Ganz Surgical Dislocation										
Madan (Madan et al. [42])	PO/G	11	11	0	1	NR	NR	HHS (90.3) NAHS(91.0)		17 unstable hips were excluded (4 AVN) 0 acute, 0 acute-on-chronic, 11 chronic. 3 had previous operations
Masse (Masse et al. [43])	PO/G	18	18	0	0		0	HHS (98.2) 18 excellent 0 good 0 fair 0 poor 0 failure	1 metalware problem	2 unstable hips excluded (no AVN) 2mild, 4 moderate, 12 severe
Novais (Novais et al. [28])	PO/G	15	15	1	NR	NR	NR	HHC 7 excellent 2 good 0 fair 5 poor 1 failure	2 metalware problems	
Souder (Souder et al. [29])	PO/G	NR	10	2	1	NR	NR	NR	1 metalware problems	From a total of 17 hips, 7 were unstable, 2 of these unstable hips went into AVN
Ziebarth (Ziebarth et al. [44])	PO/G	27	27	0	0	1		HHS (96.5) 27 excellent 0 good 0 fair 0 poor 0 failure		25 patients from series A and 2 from series B. 5 unstable/uncertain hips excluded 0mild, 15 moderate, 12 severe
Total			81	3	2			HHS (95) 52 (87 %) excellent 2 (3 %) good 0 (0 %) fair 5 (8 %) poor 1 (2 %) failure		

(continued)

Table 6.2 (continued)

Study	Rx	Patients	Hips	AVN	CL	FAI	OA	Patient satisfaction	Others	Notes
Base of Neck Osteotomy										
Abraham (Abraham et al. [45])	BNO	32	36	0	5	NR	NR	MSC 22 excellent 11 good 2 fair 2poor	5 metalware problems	0 mild, 14 moderate, 22 severe
Kramer (Kramer et al. [46])	BNO	55	56	2	1	NR	NR	NR	5 infections (1required arthrodesis) 2 metalware problems 1 needed distal transfer of the greater trochanter to increase abductor power.	Authors attributed the AVN to falls that patients had. All moderate and severe
Total		87	92	2	6	NA	NA	MSC 22 (60 %) excellent 11 (30 %) good 2 (5 %) fair 2 (5 %) poor		
Intertrochanteric Osteotomy										
Ireland [47]	ITO	32	35	0	NR	NR	4	28good 5fair 2poor		All severe
Kartenbender (Kartenbender et al. [48])	ITO	35	39	2	NR	NR	3	MSC 10 excellent 17 good 7fair 4poor		All severe
Parsch (Parsch et al. [49])	ITO	130	130	1	3	NR	NR	IHS Group A + B (96 hips) 67 Excellent 24 Good 3fair 4 poor Groups C HIS 93.9	8 femoral fractures	0 mild, 111 moderate, 19 severe Authors defined the series into 3 cohorts: A (47 hips; 40 moderate), B (49 hips; 40 moderate) and C (34 hips; 31 moderate). These combined together in this paper
Rao (Rao et al. [50])	ITO	27	29	0	1			MSC 23 excellent 0 good 3fair 1 poor	2 wound infection 1delayed union 1 hip subluxation However, the patient was asymptomatic	0 mild, 10 moderate, 19 severe

Salvati (Salvati et al. [51])	ITO	21	24	1	6	NR	NR	NR	1 infection 1 loss of fixation 1 delayed union 1bone cyst	All severe slips
Schai (Schai et al. [52])	ITO	51	51	1	NR	NR	7	Jerre Criteria 31 good 15 fair 5poor		All moderate (slip angle >30° < 60°)
Southwick [53]	ITO	26	28	0	6	NR	NR	21 excellent 5 good 2 fair 0poor	1 infection 3 further operations	All moderate and severe
		290	336	5	16			121 (44 %) excellent 105 (38 %) good 35 (13 %) fair 15 (5 %) poor		

AVN avascualr necrosis, *BNO* base of neck osteotomy, *CL* chondrolysis, *CRIF* closed reduction and internal fixation, *NR* not reported or suboptimum reporting to provide useful information, *HHS* harris hip score or modified Harris hip score; Excellent 90 to 100 points; good 80 to 89 points; fair 70 to 79 points; and poor <70 points, *HHC* heyman and herndon classification, *ITO* inter-trochanteric or peri-trochnateric osteotomy, *PO* physeal osteotomy, *IH* iowa hip-rating system. Excellent 90 to 100 points; good 80 to 89 points; fair 70 to 79 points; and poor <70 points, *MSC* modified Southwick criteria, *NAHS* non Arthritic Hip scores

chanteric flip osteotomy and anterior capsulotomy preserving the blood supply to the femoral head. Although the technique has similarities to the Dunn's osteotomy [36], hence it is also called the modified Dunn osteotomy, it poses less risk to femoral head blood supply. Six studies (81 hips) assessed the outcomes of surgical dislocation in stable slip. The crude AVN rate and CL were 3.7 % and 2.5 % respectively. Ninety percent had excellent to good results. The Harris hip score (HHS) [56] was the commonest score used in these studies and the mean was 95 points. These are promising preliminary results; however, most experts in the technique express a long learning curve and good results have not been reproduced in every centre (Alves et al. [57]).

What Is the Best Treatment for Unstable Sufe?

In his classic paper, Loder coined the term of "unstable slip". He recognized two types of slips: unstable one where the patient has such severe pain that walking is not possible even with crutches, regardless of the duration of the symptoms and stable slips where the patient can walk with or without crutches. However, this has been misquoted and misapplied in several studies.

Treatment of unstable slip is essentially the same for stable slips; however, there are two important issues to consider:

1. Being unstable, there is an opportunity for spontaneous or unintentional reduction of the severity of the slip.
2. The risk for AVN is very high (50 %). It is interesting how this high risk of AVN influences surgeons' choices differently; some adopted a minimum intervention (PIS) to prevent this risk from going up while others advocated an aggressive approach (open reduction of the slip) to reduce this high risk of AVN.

An extensive literature search revealed 23 studies that provide useful data on the outcome of unstable slips. The studies are summarised in Table 6.3. The crude AVN rates are shown in Table 6.4. The AVN rates as a surrogate for bad outcomes are comparable among various interventions with the exceptions of open reduction and internal fixation which has the lowest AVN rate of 5 %.

Eight four patients with unstable slips were treated with gentle open reduction and fixation within 24 h of the presentation. Four (5 %) only developed AVN. It is of note that this finding was heavily driven by one study (Parsch et al. [58]) of 64 patient and 3 only developed AVN. However, excluding the data of the study did not change the fact that AVN rate was significantly lower in the open reduction and internal fixation group.

The true definition of slip instability has been debated and not yet been satisfactorily defined or agreed on. Ziebarth (Ziebarth et al. [59]) found that clinical stability of SUFE as defined by Loder does not correlate with intra-operative stability. They retrospectively reviewed 82 patients with SUFE treated by open surgery and introduce the concept of "intra-operative stability" which is either intact or disrupted. They found complete physeal disruption at open surgery in 28 of the 82 hips (34 %). With classification as acute, acute-on-chronic, and chronic, the sensitivity for disrupted physes was 82 % and the specificity was 44 %. With the classification of Loder (stable and unstable) the values were 39 % and 76 %, respectively.

Kallio (Kallio et al. [60, 61]) stated that a stable slip should imply an adherent physis during weight-bearing, active leg movements, or gentle joint manipulation. Physeal instability implies that the displaced epiphysis can move in relation to the metaphysis. In a study of 55 SUFEs, he found that physeal instability is better indicated by joint effusion and inability to bear weight. A slip is very unlikely to be unstable in a child who is able to bear weight and has no sonographic effusion. This uncertainty about the definition of instability should be considered when reading the above results.

How Soon Should We Treat Slipped Upper Femoral Epiphysis?

This question is probably more relevant to unstable slips rather than stable because of the low AVN rate in stable slip. The timing of surgery in unstable slip remains controversial. Given the rarity of the condition, most studies that investigated the timing of surgery and outcome are underpowered to answer such a question. Lowndes et al. [8] in a meta-analysis of 5 studies (130 unstable SUFEs; 56 were treated within 24 h and 74 were treated after 24 h of symptoms onset) found that the odds for developing AVN if treatment occurs within 24 h were half if treatment occurs after 24 h. Although the difference was large, it was not statistically significant (P = 0.44) and may be a chance finding.

Peterson et al. ([62])showed early stabilisation within 24 h was associated with less AVN (3/42 = 7 %) in comparison with those stabilised after 24 h (10/49 = 20 %). Kalogrianitis et al. ([63])showed that AVN developed in 50 % (8/16) of the unstable SUFE in their series. All but one were treated between 24 and 72 h after symptom onset. They recommend immediate stabilization of unstable slips presenting within 24 h. If this is not possible, then delaying the operation until at least a week has elapsed. In contradiction, Loder [5] noted more AVN in patients treated within 48 h (7/8 versus 7/21).

Our findings supported Kalogrianitis's findings; there were 210 patients with unstable slips who had their operation within 24 h. Twenty eight (13 %) developed AVN in comparison to (38/95) 40 % and (5/53) 9 % for those who had their operation between 24 and 72 h and those who had their operation after 72 h respectively.

Table 6.3 Studies of various interventions in unstable slips

Study	Hips	Rx	Time	R	Type of reduction	AVN	CL	Others	Notes
Alshryda (Alshryda et al. [7])	22	7 PIS	> 48 h	Y	Spontaneous	2			Severity (2:2:3)
		15 PO (Fish)	> 48 h	Y	OR	5		2 loss of fixation Hip dislocation	Severity (0:1:14)
Alves (Alves et al. [57])	12	6 CRIF	24.3 h (±7.9 h)	Y	CR	2			Severity (1:3:1)
		6 PO/G	22.2 h (±7.9 h)	Y	OR	4			Severity (2:2:?)
Aronson [75]	15	9 CRIF	<24 h	Y	CR	2			All patients went on traction.
		6 ORIF	<24 h	Y	OR	0			All patients went on traction. Authors described a new controlled open reduction and stabilisation using 2 screws
Biring (Biring et al. [76])	25	PO	NR	Y	OR	3		4 Chondrolysis	
Chen (Chen et al. [77])	30	CRIF	<24 h	Y	CR	4	1	1 slip progression	Severity (13:9:8) + 16 percutaneous, 5 open capsulotomy
Fallath [78]	14	CRIF	28 h (range 3.5–72 h).	Y	2 spontaneous, 9 CR 2 PIS	3			10 single screw, 3 Knowles pin, 1 two screws 7acute and 7 acute-on-chronic All AVN in the CR group; fixed with a single screw and duration of symptoms (72, 19 and 36 h)
Gordon (Gordon et al. [79])	16	12 CRIF	7 < 24 h 3 < 72 h	Y	CR	2			Severity (2:4:6) 1 AVN within 24 h and another 168 h
		4 ORIF	<24 h		4 OR	0			Severity (0:4:0) All patients had capsulotomy and 2 screws
Kalogrianitis (Kalogrianitis et al. [63])	16	14 PIS 2PO	5 < 24 h 7 > 24 < 72 h 3 > 8 days	Y	Spontaneous	8			5 < 24 h (1 AVN) 7 > 24 < 72 h (7 AVN) 3 > 8 days (no AVN) No AVN in PO Severity (2:6:8) 10 acute and 6 acute-on-chronic 6 AVN are type III and 2 type II
Kennedy (Kennedy et al. [80])	27	18 pinning	Traction ranged from <24 h to 6 days	Y	1 Spontaneous 14 CR 3PIS	2			19 two screws, 7 single screw, 1 AVN in a mild and the second is in a severe one. Both had traction of <24 h
		9 ORIF		Y	9 OR	2			1 AVN had a failed PIS which was treated with OR and osteotomy. It was moderate slip. The second AVN was severe

(continued)

Table 6.3 (continued)

Study	Hips	Rx	Time	R	Type of reduction	AVN	CL	Others	Notes
Lim (Lim et al. [27])	24	13 PIS	Traction for 6 days 11	N	PIS	1			All acute or acute on chronic 2 of the 14 hips were stable but not clear which group Severity (16:7:1)
		11 CRIF		Y	CR	1			
Loder (Loder et al. [5])	30	26 CRIF	4 ± 3.8 d		4 PIS 26 CR	14			17 acute or 13 acute on chronic Severity (2:9:19) AVN group from presentation to operation was (2 ± 1.8 d) and for non AVN groups (6 + 3.8d) (P = 0.0004). AVN (4/5 hips) that were operated within 24 h and (10/25) were operated after 24 h. AVN (7/8 hips) that were operated within 48 h and (7/22) were operated after 48 h.
Madan (Madan et al. [42])	17	17 PO/G	Traction 11 days	Y	OR	4			9 acute, 8 acute-on-chronic All severe
Palocaren (Palocaren et al. [81])	27	PIS	16 < 24 h 3 < 48 h 3 < 72 5 > 120 h		Spontaneous PIS	6			16 < 24 h (4 AVN) 3 < 48 h (1 AVN) 3 < 72 (1 AVN) 5 > 120 h
Parsch (Parsch et al. [58])	64	64 ORIF	49 < 24 h 15 > 224 h	Y	OR	3		4 metalware problems	Severity (20:24:20) AVN1 (moderate slip, within 24 h), AVN2 (severe slip, > 24 h) AVN3 (severe slip, > 24 h)
Peterson (Peterson et al. [62])	91	91 CR	42 < 24 h 12 > 24 < 48 h 7 > 48 < 72 h 30 > 72 h		41 CRIF 4 spica 31 Epi + spica 15 Epi + IF	13			42 < 24 h (3 AVN) 12 > 24 < 48 h (3 AVN) 7 > 48 < 72 h (4 AVN) 30 > 72 h (3 AVN) 41 CRIF (4 AVN) 4 spica (3 AVN) 31 Epi + spica (2 AVN) 15 Epi + IF (4 AVN) Severity (5:67:19)
Phillips (Phillips et al. [82])	14	12 CRIF 2 PO	<24		12 CRIF 2 PO	0			Severity (0:3:11)
Rao (Rao et al. [16])	18	Epi	4 > 2w traction		OR	1		1 Chondrolysis	Severity (6:7:5) AVN in moderate
Rhoad (Rhoad et al. [83])	10	Pinning	NR	R	8 Spontaneous 2 PIS	5		1 Chondrolysis (and AVN in same patinets) in PIS group	2 PIS (1 AVN)

Sankar (Sankar et al. [84])	70	16 PIS	40 < 24 h 12 < 24 > 48 h 18 > 48	N		3			40 < 24 h (10 AVN) 12 < 24 > 48 h (2 AVN) 18 > 48 (2 AVN)
		38 CRIF		Y	Spontaneous	10			
		8 PO		Y	OR	1			
		8 PO/G		Y	OR	0			Excluded from the final count as some may be a duplicate Sankar's study (Sankar, Vanderhave et al. 2013)
Sankar (Sankar et al. [85])	27	PO/G	20 < 24 h 4 < 48 h 3 > 72 h 35.9 h (Range, 6 to 184 h).	R	27 OR	7		4 metalware problems	20 < 24 h (4 AVN) 4 < 48 h (2AVN) 3 > 72 h (1AVN)
Seller (Seller et al. [86])	29	MPF	<48 h	Y	CR	2		1 Chondrolysis 8 outgrow fixation 4 subtrochanteric osteotomy	
Souder (Souder et al. [29])	14	7 PIS	NR	N		3			
		7 PO/G	NR	Y	OR	2			
Tokmakova (Tokmakova et al. [87])	36	Pinning	NR		21 reduced. 15 unclear	21			Severity (8:20:8) 1 AVN in mild slip 14 AVN in moderate slips 6 AVN in severe slip All AVN patients had reduction 8 in acute slips and 13 in chronic slip
Ziebarth (Ziebarth et al. [44])	12	PG/O	NR	Y	OR	0		0 Chondrolysis	Severity (3:3:4:2)

Table 6.4 Pooled summary of studies of unstable slips treatments

Interventions	Hips	AVN (%)
Epiphysiodesis	64	7 (11 %)
Pinning in situ	115	38(33 %)
Closed reduction and pinning	269	71(26 %)
Open reduction and internal fixation	84	4 (5 %)
Physeal osteotomies (Dunn's or Fish)	59	10 (17 %)
Ganz surgical dislocation	70	13(18 %)
Total	661	143 (22 %)

Should We Treat the Contralateral Non Slipped, Asymptomatic Side?

This is also controversial. One of the main the reason for this controversy is the uncertainty about the incidence of the contralateral slip. The quoted risk of contralateral slip varies from 18 % to 60 %. Jerre (Jerre et al. [39]) reviewed 100 patients treated for SUFE to evaluate the incidence of bilateral slipping of the epiphysis at an average follow-up time of 32 years. Fifty nine patients (59 %) were judged to have had a previous bilateral SCFE; in 42 of these 59 patients (71 %), slipping of the contralateral hip was asymptomatic. In 23 patients (23 %), the diagnosis of bilateral slipping was established at primary admission, in 18 (18 %) later during adolescence, and in 18 (18 %) not until the patients were reexamined as adults and the primary radiographs were reviewed. He concluded that the incidence of bilateral slipping of the epiphysis in patients with SCFE is approximately 60 % in Sweden.

In another long term study of 155 slips by Carney (Carney et al. [13]) the slip was bilateral in 31 patients (25 %). In 14/31patients both hips were symptomatic at presentation. The rest apart from one developed within one year.

Stasikelis et al. [64] performed a retrospective review 50 children who had unilateral SUFE to determine parameters that predict the later development of a contralateral slip. They found that the modified Oxford bone age was strongly correlated with the risk of development of a contralateral slip; contralateral slip developed in 85 % of patients with a score of 16, in 11 % of patients with a score of 21, and in no patient with a score of 22 or more. The modified Oxford bone age is based on appearance and fusion of the iliac apophysis, femoral capital physis, greater and lesser trochanters. Recently, calcaneal scoring (Nicholson et al. [65]) was used to predict an elevated risk of contralateral SUFE. The obvious disadvantage is the need for a calcaneal x-ray.

A recent paper (Phillips et al. [66]) examined the posterior slope angle (PSA) in 132 patients as a predictive for developing a contralateral slip. The mean was 17.2° ± 5.6° in 42 patients who had subsequently developed a contralateral slip, which was significantly higher (P = 0.001) than that of 10.8° ± 4.2° for the 90 patients who had had a unilateral slip. If a posterior sloping angle of 14° were used as an indication for prophylactic fixation, 35 (of 42 = 83.3 %) would have been prevented, and 19 (of 90 = 21.1 %) would have been pinned unnecessarily (Fig. 6.4).

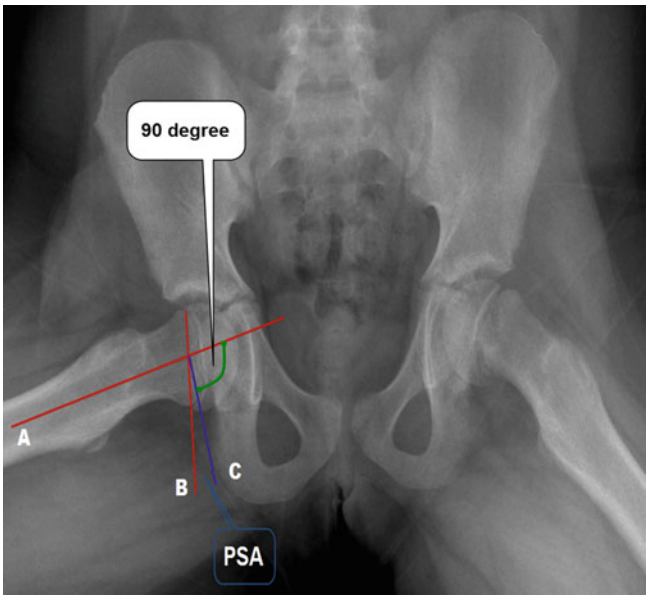

Fig. 6.4 Posterior slope angle. The posterior sloping angle (*PSA*) measured by a line (*A*) from the center of the femoral shaft through the center of the metaphysis. A second line (*B*) is drawn from one edge of the physis to the other, which represents the angle of the physis. Where lines *A* and *B* intersect, a line (*C*) is drawn perpendicular to line *A*. The *PSA* is the angle formed by lines *B* and *C* posteriorly as illustrated

Prophylactic pinning is not devoid of risk and it should be weighed against the benefit. The proponents and opponents have some evidence to support their views (Jerre et al. [67]; Sankar et al. [68]; Clement et al. [69]). Most studies showed that the average risk of contralateral lateral slip is around 18 % (Larson et al. [70]; Baghdadi et al. [71]). Most were mild slips and when treated they rarely went to develop AVN. Risk of prophylactic pinning is in the region of 5 % including AVN and peri-prosthetic fractures (Sankar et al. [68]; Baghdadi et al. [71]; Kroin et al. [72]).

We recommend a pragmatic approach for contralateral pinning where the following factors play a role in decision making:

1. Age of the child (<10 years is associated with a higher risk of bilaterality).
2. Slips associated with renal osteodystrophy and endocrine disorders (a high incidence of bilaterality)

3. Poor compliance of the child and family.
4. The nature of current slip (very bad slip occurred over a very short period of time may justify pinning the other side)

What Are the Natural History and Long Term Outcomes of Slipped Upper Femoral Epiphysis?

Natural history is the usual course of development of a disease or condition, especially in the absence of treatment. This is difficult to establish in SUFE simply because most published series reported patients who were treated. There were a few cross sectional studies that reported on the outcomes of what were presumed as untreated slips. Even if these were true slips, they were probably mild stable slips that would pursue a different natural course from most other slips. Most studies used AVN as a surrogate for bad outcome. Although AVN is rare in stable slips, bad outcome is not uncommon in severe stable slips. Larson et al. [73] reviewed 33,000 hip replacement performed in their centre between 1954 and 2007 and found SUFE was the indication in 38 hips (in 33 patients). The main reasons for hip replacement in this subset were AVN or chondrolysis in 25 hips and degenerative changes and/or impingement in 13 hips. All slips underwent either pin fixation (27) or primary osteotomy (9). Mean time from slip to hip replacement was 7.4 years in patients with AVN or chondrolysis and 23.6 years in patients with degenerative change ($P < 0.0002$). Mean age at arthroplasty was 20 years in the AVN or chondrolysis group and 38 years in the degenerative group ($P < 0.0001$). Sixteen hips (42 %) required revision arthroplasty at a mean of 11.6 years postoperatively, most commonly for component loosening and/or polyethylene wear. Kaplan Meier 5-year survival free from revision for all causes was 87 % overall and 95 % in the total hip arthroplasty subset.

Carney [74] published a series of 31 untreated chronic SUFE with a long term follow-up (ranged from 26 to 54 years). Authors stated the reasons for no treatments were not always clear from the medical records but included family refusal, delayed presentation or treating the more serious side. There were 17 mild, 11 moderate and 3 severe. The mean IHS was 89 points (92 points in mild slips, 87 points in moderate slips and 75 points in severe slips). All severe and moderate slips showed radiographic features of OA in contrast to 13 % of those with mild slip. Complications were occurred in 4 slips (1 AVN and 2 further displacements developed 3 severe slips and 1 chondrolysis in 1 mild slip.

In another series, Carney (Carney et al. [13]) reported on 155 SUFEs in 124 patients after 41 year follow up. Forty-two percent of the slips were mild; 32 % were moderate; and 26 % were severe. Various treatments methods were used (see Table 6.4). They found that there is mild deterioration that is related to the severity of the slip and complications of treatment (Fig. 6.5). Realignment was associated with a risk of substantial complications and adversely affects the natural course of the disease (Fig. 6.6).

A summary of recommendations is given in Table 6.5.

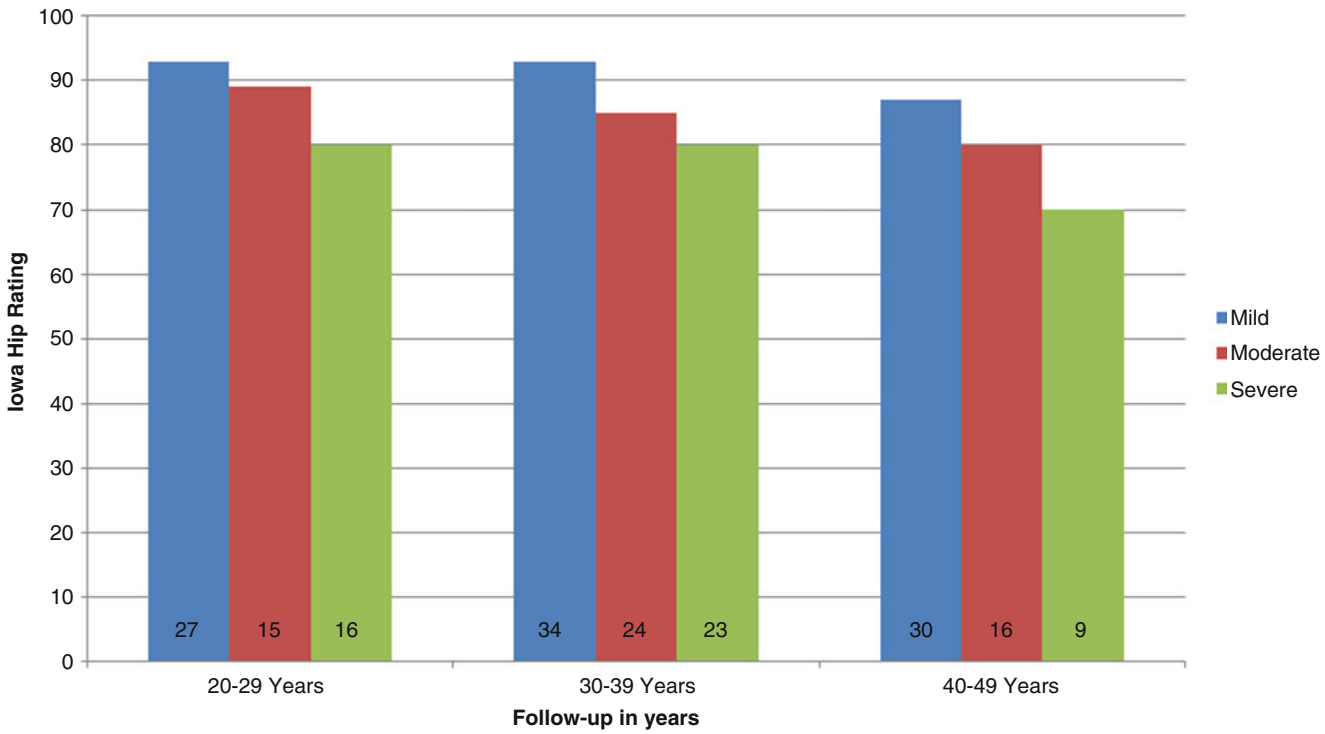

Fig. 6.5 Long-term follow-up of treated slipped upper femoral epiphysis (Adapted from Carney (Carney et al. [13]))

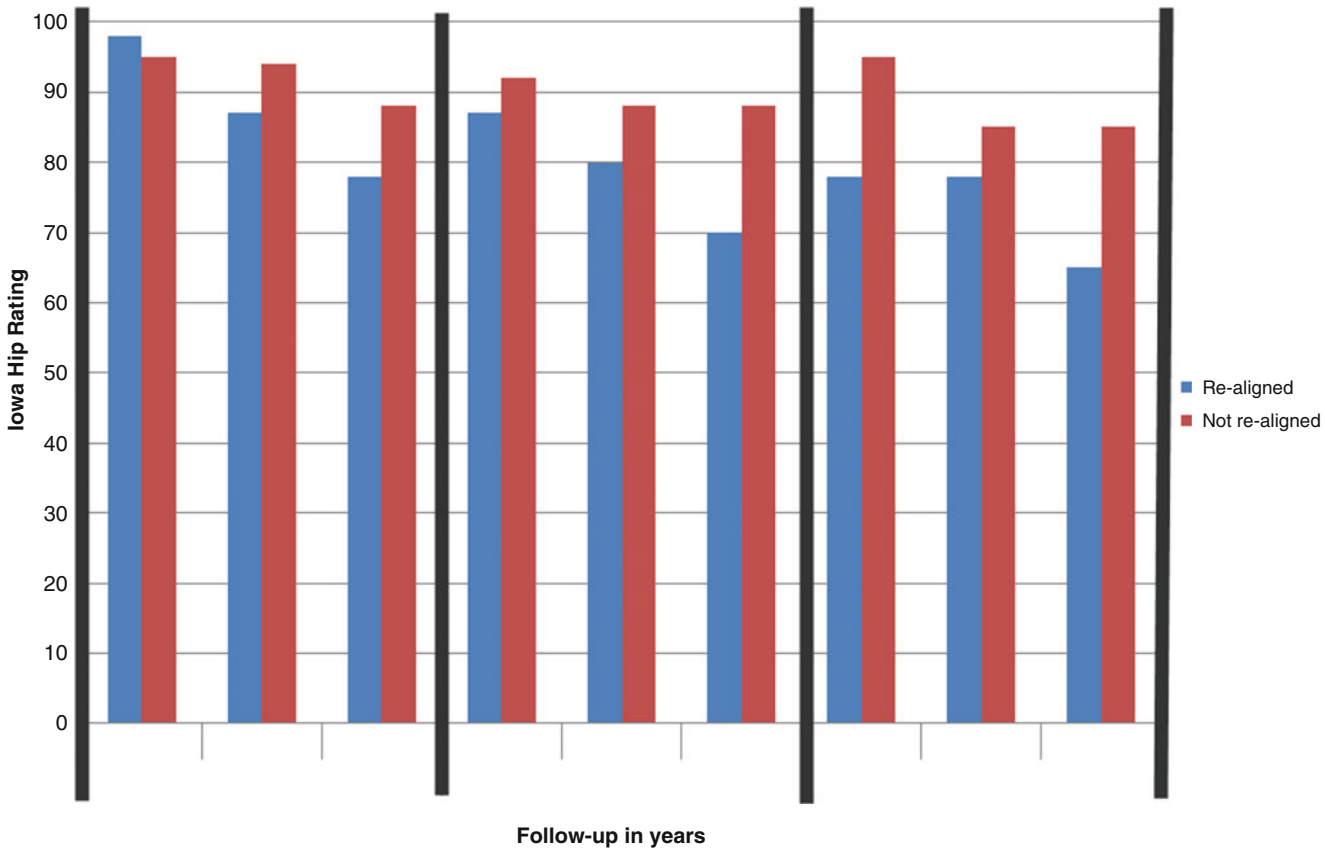

Fig. 6.6 Long-term follow-up of treated slipped upper femoral epiphysis (Adapted from Carney (Carney et al. [13]))

Table 6.5 Summary of recommendations

Clinical questions	Grade of Recommendation
Pinning in situ is the best treatment for mild and moderate stable slip.	B
Surgical dislocation may give a better results than pinning in situ for severe stable slip	C
Urgent gentle reduction, capsulotomy and fixation is the best current treatment for unstable slip	C
Routine prophylactic pinning of the contralateral asymptomatic side is not recommended	C

References

1. Loder RT, Greenfield ML. Clinical characteristics of children with atypical and idiopathic slipped capital femoral epiphysis: description of the age-weight test and implications for further diagnostic investigation. J Pediatr Orthop. 2001;21(4):481–7.
2. Alshryda S, Jones S, et al. Postgraduate paediatric orthopaedics: the candidate's guide to the FRCS (Tr and Orth) examination. Cambridge: Cambridge University Press; 2014.
3. Fahey JJ, O'Brien ET. Acute slipped capital femoral epiphysis: review of the literature and report of ten cases. J Bone Joint Surg Am. 1965;47:1105–27.
4. Alshryda, S. and J. Wright (2014). Acute slipped capital femoral epiphysis: the importance of physeal stability. Classic Papers in Orthopaedics. London: Springer. 2014; 547–8.
5. Loder RT, Richards BS, et al. Acute slipped capital femoral epiphysis: the importance of physeal stability. J Bone Joint Surg Am. 1993;75(8):1134–40.
6. Lim YJ, Lam KS, et al. Review of the management outcome of slipped capital femoral epiphysis and the role of prophylactic contra-lateral pinning re-examined. Ann Acad Med Singapore. 2008;37(3):184–7.
7. Alshryda S, Tsang K, et al. Severe slipped upper femoral epiphysis; fish osteotomy versus pinning-in-situ: an eleven year perspective. Surgeon. 2013;12(5):244–8.
8. Lowndes S, K. A, Emery D, Sim J, Maffulli N. Management of unstable slipped upper femoral epiphysis: a meta-analysis. Br Med Bull. 2009;90:133–46.
9. Alshryda S, Tsang K, Al-Shryda J, Blenkinsopp J, Adedapo A, Montgomery R, et al. Interventions for treating slipped upper femoral epiphysis (SUFE). Cochrane Database of Systematic Reviews. 2013; Issue 2. Art. No.: CD010397. DOI: 101002/14651858CD010397.
10. Southwick WO. Slipped capital femoral epiphysis. J Bone Joint Surg Am. 1984;66(8):1151–2.
11. Loder RT, Dietz FR. What is the best evidence for the treatment of slipped capital femoral epiphysis? J Pediatr Orthop. 2012;32(Suppl 2):S158–65.
12. Betz RR. Treatment of slipped capital femoral epiphysis with a spina cast. J Bone Joint Surg Am. 1995;77(3):489–90.
13. Carney BT, Weinstein SL, et al. Long-term follow-up of slipped capital femoral epiphysis. J Bone Joint Surg Am. 1991;73(5):667–74.

14. Meier MC, Meyer LC, et al. Treatment of slipped capital femoral epiphysis with a spica cast. J Bone Joint Surg Am. 1992;74(10):1522–9.
15. Adamczyk MJ, Weiner DS, et al. A 50-year experience with bone graft epiphysiodesis in the treatment of slipped capital femoral epiphysis. J Pediatr Orthop. 2003;23(5):578–83.
16. Rao SB, Crawford AH, et al. Open bone peg epiphysiodesis for slipped capital femoral epiphysis. J Pediatr Orthop. 1996;16(1):37–48.
17. Schmidt TL, Cimino WG, et al. Allograft epiphysiodesis for slipped capital femoral epiphysis. Clin Orthop Relat Res. 1996;322:61–76.
18. Szypryt EP, Clement DA, et al. Open reduction or epiphysiodesis for slipped upper femoral epiphysis. A comparison of Dunn's operation and the Heyman-Herndon procedure. J Bone Joint Surg Br. 1987;69(5):737–42.
19. Zahrawi FB, Stephens TL, et al. Comparative study of pinning in situ and open epiphysiodesis in 105 patients with slipped capital femoral epiphyses. Clin Orthop Relat Res. 1983;177:160–8.
20. Aronson DD, Loder RT. Slipped capital femoral epiphysis in black children. J Pediatr Orthop. 1992;12(1):74–9.
21. Aronson DD, Carlson WE. Slipped capital femoral epiphysis. A prospective study of fixation with a single screw. J Bone Joint Surg Am. 1992;74(6):810–9.
22. Blanco JS, Taylor B, et al. Comparison of single pin versus multiple pin fixation in treatment of slipped capital femoral epiphysis. J Pediatr Orthop. 1992;12(3):384–9.
23. Gonzalez-Moran G, Carsi B, et al. Results after preoperative traction and pinning in slipped capital femoral epiphysis: K wires versus cannulated screws. J Pediatr Orthop B. 1998;7(1):53–8.
24. Herman MJ, Dormans JP, et al. Screw fixation of Grade III slipped capital femoral epiphysis. Clin Orthop Relat Res. 1996;322:77–85.
25. Kenny P, H. T, Sedhom M, Dowling F, Moore DP, Fogarty EE. Slipped upper femoral epiphysis. a retrospective, clinical and radiological study of fixation with a single screw. J Pediatr Orthop B. 2003;12(2):97–9.
26. Koval KJ, L. WB, Rose D, Koval RP, Grant A, Strongwater A. Treatment of slipped capital femoral epiphysis with a cannulated-screw technique. J Bone Joint Surg Am. 1989;71(9):1370–7.
27. Lim YJ, Lam KS, et al. Management outcome and the role of manipulation in slipped capital femoral epiphysis. J Orthop Surg (Hong Kong). 2007;15(3):334–8.
28. Novais EN, Hill MK, et al. Modified Dunn procedure is superior to in Situ pinning for short-term clinical and radiographic improvement in severe stable SCFE. Clin Orthop Relat Res. 2014;473(6):2108–17.
29. Souder CD, Bomar JD, et al. The role of capital realignment versus in situ stabilization for the treatment of slipped capital femoral epiphysis. J Pediatr Orthop. 2014;34(8):791–8.
30. Ward WT, Stefko J, et al. Fixation with a single screw for slipped capital femoral epiphysis. J Bone Joint Surg Am. 1992;74(6):799–809.
31. Dreghorn CR, Knight D, et al. Slipped upper femoral epiphysis–a review of 12 years of experience in Glasgow (1972–1983). J Pediatr Orthop. 1987;7(3):283–7.
32. Barros JW, Tukiama. G, Fontoura C, Barsam NH, Pereira ES. Trapezoid osteotomy for slipped capital femoral epiphysis. Int Orthop. 2000;24(2):83–7.
33. Broughton NS, Todd RC, et al. Open reduction of the severely slipped upper femoral epiphysis. J Bone Joint Surg Br. 1988;70(3):435–9.
34. DeRosa GP, Mullins RC, et al. Cuneiform osteotomy of the femoral neck in severe slipped capital femoral epiphysis. Clin Orthop Relat Res. 1996;322:48–60.
35. Diab M, Hresko MT, et al. Intertrochanteric versus subcapital osteotomy in slipped capital femoral epiphysis. Clin Orthop Relat Res. 2004;427:204–12.
36. Dunn DM, Angel JC. Replacement of the femoral head by open operation in severe adolescent slipping of the upper femoral epiphysis. J Bone Joint Surg Br. 1978;60-B(3):394–403.
37. Fish JB. Cuneiform osteotomy of the femoral neck in the treatment of slipped capital femoral epiphysis. A follow-up note. J Bone Joint Surg Am. 1994;76(1):46–59.
38. Fron D, Forgues D, et al. Follow-up study of severe slipped capital femoral epiphysis treated with Dunn's osteotomy. J Pediatr Orthop. 2000;20(3):320–5.
39. Jerre R, Billing L, et al. Bilaterality in slipped capital femoral epiphysis: importance of a reliable radiographic method. J Pediatr Orthop B. 1996;5(2):80–4.
40. Nishiyama K, Sakamaki. T, Ishii Y. Follow-up study of the subcapital wedge osteotomy for severe chronic slipped capital femoral epiphysis. J Pediatr Orthop. 1989;9(4):412–6.
41. Velasco R, Schai PA, et al. Slipped capital femoral epiphysis: a long-term follow-up study after open reduction of the femoral head combined with subcapital wedge resection. J Pediatr Orthop B. 1998;7(1):43–52.
42. Madan SS, Cooper AP, et al. The treatment of severe slipped capital femoral epiphysis via the Ganz surgical dislocation and anatomical reduction: a prospective study. Bone Joint J. 2013;95-B(3):424–9.
43. Masse A, Aprato A, et al. Surgical hip dislocation for anatomic reorientation of slipped capital femoral epiphysis: preliminary results. Hip Int. 2012;22(2):137–44.
44. Ziebarth K, Zilkens C, et al. Capital realignment for moderate and severe SCFE using a modified Dunn procedure. Clin Orthop Relat Res. 2009;467(3):704–16.
45. Abraham E, Garst J, et al. Treatment of moderate to severe slipped capital femoral epiphysis with extracapsular base-of-neck osteotomy. J Pediatr Orthop. 1993;13(3):294–302.
46. Kramer WG, Craig WA, et al. Compensating osteotomy at the base of the femoral neck for slipped capital femoral epiphysis. J Bone Joint Surg Am. 1976;58(6):796–800.
47. Ireland J, Newman PH. Triplane osteotomy for severely slipped upper femoral epiphysis. J Bone Joint Surg Br. 1978;60-B(3):390–3.
48. Kartenbender K, Cordier W, et al. Long-term follow-up study after corrective Imhauser osteotomy for severe slipped capital femoral epiphysis. J Pediatr Orthop. 2000;20(6):749–56.
49. Parsch K, Zehender H, et al. Intertrochanteric corrective osteotomy for moderate and severe chronic slipped capital femoral epiphysis. J Pediatr Orthop B. 1999;8(3):223–30.
50. Rao JP, Francis AM, et al. The treatment of chronic slipped capital femoral epiphysis by biplane osteotomy. J Bone Joint Surg Am. 1984;66(8):1169–75.
51. Salvati EA, Robinson Jr JH, et al. Southwick osteotomy for severe chronic slipped capital femoral epiphysis: results and complications. J Bone Joint Surg Am. 1980;62(4):561–70.
52. Schai PA, Exner GU, et al. Prevention of secondary coxarthrosis in slipped capital femoral epiphysis: a long-term follow-up study after corrective intertrochanteric osteotomy. J Pediatr Orthop B. 1996;5(3):135–43.
53. Southwick WO. Osteotomy through the lesser trochanter for slipped capital femoral epiphysis. J Bone Joint Surg Am. 1967;49(5):807–35.
54. Ganz R, Gill TJ, et al. Surgical dislocation of the adult hip a technique with full access to the femoral head and acetabulum without the risk of avascular necrosis. J Bone Joint Surg Br. 2001;83(8):1119–24.
55. Ganz R, Parvizi. J, Beck M, Leunig M, Notzli H, Siebenrock KA. Femoroacetabular impingement: a cause for osteoarthritis of the hip. Clin Orthop Relat Res. 2003;417:112–20.
56. Harris WH. Traumatic arthritis of the hip after dislocation and acetabular fractures: treatment by mold arthroplasty. An end-result

study using a new method of result evaluation. J Bone Joint Surg Am. 1969;51(4):737–55.
57. Alves C, Steele M, et al. Open reduction and internal fixation of unstable slipped capital femoral epiphysis by means of surgical dislocation does not decrease the rate of avascular necrosis: a preliminary study. J Child Orthop. 2013;6(4):277–83.
58. Parsch K, Weller S, et al. Open reduction and smooth Kirschner wire fixation for unstable slipped capital femoral epiphysis. J Pediatr Orthop. 2009;29(1):1–8.
59. Ziebarth K, Domayer S, et al. Clinical stability of slipped capital femoral epiphysis does not correlate with intraoperative stability. Clin Orthop Relat Res. 2012;470(8):2274–9.
60. Kallio PE, Paterson DC, et al. Classification in slipped capital femoral epiphysis. Sonographic assessment of stability and remodeling. Clin Orthop Relat Res. 1993;294:196–203.
61. Kallio PE, Mah ET, et al. Slipped capital femoral epiphysis. Incidence and clinical assessment of physeal instability. J Bone Joint Surg Br. 1995;77(5):752–5.
62. Peterson MD, Weiner DS, et al. Acute slipped capital femoral epiphysis: the value and safety of urgent manipulative reduction. J Pediatr Orthop. 1997;17(5):648–54.
63. Kalogrianitis S, Tan CK, et al. Does unstable slipped capital femoral epiphysis require urgent stabilization? J Pediatr Orthop B. 2007;16(1):6–9.
64. Stasikelis PJ, Sullivan CM, et al. Slipped capital femoral epiphysis. Prediction of contralateral involvement. J Bone Joint Surg Am. 1996;78(8):1149–55.
65. Nicholson AD, Huez CM, Sanders JO, Liu RW, Cooperman DR. Calcaneal Scoring as an Adjunct to Modified Oxford Hip Scores: Prediction of Contralateral Slipped Capital Femoral Epiphysis. J Pediatr Orthop. 2016;36(2):132–8.
66. Phillips PM, Phadnis J, et al. Posterior sloping angle as a predictor of contralateral slip in slipped capital femoral epiphysis. J Bone Joint Surg Am. 2013;95(2):146–50.
67. Jerre R, Billing L, et al. The contralateral hip in patients primarily treated for unilateral slipped upper femoral epiphysis. Long-term follow-up of 61 hips. J Bone Joint Surg Br. 1994;76(4):563–7.
68. Sankar WN, Novais EN, et al. What are the risks of prophylactic pinning to prevent contralateral slipped capital femoral epiphysis? Clin Orthop Relat Res. 2012;471(7):2118–23.
69. Clement ND, Vats A, et al. Slipped capital femoral epiphysis: is it worth the risk and cost not to offer prophylactic fixation of the contralateral hip? Bone Joint J. 2015;97-B(10):1428–34.
70. Larson AN, Yu EM, et al. Incidence of slipped capital femoral epiphysis: a population-based study. J Pediatr Orthop B. 2009;19(1):9–12.
71. Baghdadi YM, Larson AN, et al. The fate of hips that are not prophylactically pinned after unilateral slipped capital femoral epiphysis. Clin Orthop Relat Res. 2013;471(7):2124–31.
72. Kroin E, Frank JM, et al. Two cases of avascular necrosis after prophylactic pinning of the asymptomatic, contralateral femoral head for slipped capital femoral epiphysis: case report and review of the literature. J Pediatr Orthop. 2015;35(4):363–6.
73. Larson AN, McIntosh AL, et al. Avascular necrosis most common indication for hip arthroplasty in patients with slipped capital femoral epiphysis. J Pediatr Orthop. 2010;30(8):767–73.
74. Carney BT, Weinstein SL. Natural history of untreated chronic slipped capital femoral epiphysis. Clin Orthop Relat Res. 1996;322:43–7.
75. Aronson J, Tursky EA. The torsional basis for slipped capital femoral epiphysis. Clin Orthop Relat Res. 1996;322:37–42.
76. Biring GS, Hashemi-Nejad A, et al. Outcomes of subcapital cuneiform osteotomy for the treatment of severe slipped capital femoral epiphysis after skeletal maturity. J Bone Joint Surg Br. 2006;88(10):1379–84.
77. Chen RC, Schoenecker PL, et al. Urgent reduction, fixation, and arthrotomy for unstable slipped capital femoral epiphysis. J Pediatr Orthop. 2009;29(7):687–94.
78. Fallath S, Letts M. Slipped capital femoral epiphysis: an analysis of treatment outcome according to physeal stability. Can J Surg. 2004;47(4):284–9.
79. Gordon JE, Abrahams MS, et al. Early reduction, arthrotomy, and cannulated screw fixation in unstable slipped capital femoral epiphysis treatment. J Pediatr Orthop. 2002;22(3):352–8.
80. Kennedy JG, Hresko MT, et al. Osteonecrosis of the femoral head associated with slipped capital femoral epiphysis. J Pediatr Orthop. 2001;21(2):189–93.
81. Palocaren T, Holmes L, et al. Outcome of in situ pinning in patients with unstable slipped capital femoral epiphysis: assessment of risk factors associated with avascular necrosis. J Pediatr Orthop. 2009;30(1):31–6.
82. Phillips SA, Griffiths WE, et al. The timing of reduction and stabilisation of the acute, unstable, slipped upper femoral epiphysis. J Bone Joint Surg Br. 2001;83(7):1046–9.
83. Rhoad RC, Davidson RS, et al. Pretreatment bone scan in SCFE: a predictor of ischemia and avascular necrosis. J Pediatr Orthop. 1999;19(2):164–8.
84. Sankar WN, McPartland TG, et al. The unstable slipped capital femoral epiphysis: risk factors for osteonecrosis. J Pediatr Orthop. 2010;30(6):544–8.
85. Sankar WN, Vanderhave KL, et al. The modified Dunn procedure for unstable slipped capital femoral epiphysis: a multicenter perspective. J Bone Joint Surg Am. 2013;95(7):585–91.
86. Seller K, Wild A, et al. Clinical outcome after transfixation of the epiphysis with Kirschner wires in unstable slipped capital femoral epiphysis. Int Orthop. 2006;30(5):342–7.
87. Tokmakova KP, Stanton RP, et al. Factors influencing the development of osteonecrosis in patients treated for slipped capital femoral epiphysis. J Bone Joint Surg Am. 2003;85-A(5):798–801.
88. Carlioz HV, Vogt JC, Barba L, Doursounian L. Treatment of slipped upper femoral epiphysis: 80 cases operated on over 10 years (1968-1978). J Pediatr Orthop. 1984;4(2):153–61.

Evidence-Based Treatment for Congenital Femoral Deficiency

7

Anthony Cooper and James A. Fernandes

Abstract

Congenital femoral deficiency is a challenging paediatric orthopaedic problem that requires a multidisciplinary approach. Patients vary significantly in term of severity from a very mild form that does not require surgical intervention to complete absence of femur. Various surgical interventions have been reported in the literatures with variable success rates. Given the rarity of the condition, the wide spectrum of severity, the many surgical interventions that have been described and the recent evolution in surgical techniques, it has been difficult to recommend "the best treatment" for this condition. In this chapter, we critically reviewed the evidence behind the core principle of treating this challenging condition.

Keywords

Short femur • PFFD • CSF • Longitudinal deficiency • Bone lengthen

Introduction

The term proximal femoral focal deficiency (PFFD), congenital short femur (CSF) and congenital femoral deficiency (CFD) are often used interchangeably. Historically congenital short femur was the first term coined to describe this deficiency. It was thought to be a simple femoral hypoplasia and a separate entity to the other forms of deficiency seen in the femur [1]. In recognition of the major deformity occurring at the proximal femur the term proximal femoral focal deficiency was described [2]. This term has become more widespread and does not always well characterize the deformities seen in this condition. The disorder should be considered as a longitudinal limb deficiency, with the majority of the shortening occurring in the femur. As such more recently congenital femoral deficiency (CFD) has been used as this term encompasses the fact that the entire femur is involved. It is also associated with a classification system which is less descriptive and more treatment based [3]. The deficiency in CFD varies substantially from a mild amount of coxa vara with minimal shortening to complete absence of the femur. Common manifestations include varying degrees of acetabular dysplasia, delayed femoral ossification, excessive external rotation of the leg, hypoplasia of the lateral femoral condyle, absent cruciate ligaments [4] as well as shortened musculature about the hip and thick, inelastic ligamentous structures.

The recent, treatment-based classification classifies CFD into [3]:

Type I: Intact femur with mobile hip and knee
- a) Normal ossification proximal femur
- b) Delayed ossification proximal femur

Type II: Mobile pseudarthrosis (hip not fully formed, a false joint) with mobile knee
- a) Femoral head mobile in acetabulum
- b) Femoral head absent or stiff in acetabulum

Type III: Diaphyseal deficiency of femur (femur does not reach the acetabulum)
- a) Knee motion >45 degrees
- b) Knee motion <45 degrees

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Type I is further sub-classified into:

0) ready for surgery; no factors to correct before lengthening
1) One factor to correct before lengthening
2) Two factors to correct before lengthening
3) Three factors to correct before lengthening
4) etc

Examples of factors requiring correction prior to lengthening of femur are coxa var., hip dysplasia, patellar subluxation etc.

The strategy of management is staged corrections of the abnormalities to reach stage I0 which is amenable to lengthening. For example, type Ia-3 is converted to Ia-2, then Ia-1 and then Ia-0 followed by lengthening. Pre-existing knee stiffness is the most functionally limiting factor and should be considered a relative indication for amputation versus reconstruction [5].

Figures 7.1, 7.2, and 7.3 illustrate the disorder in children.

A literature review of English-speaking orthopaedic literature was performed to identify areas of controversy and to review the best evidence available for treatment of patients with CFD. Given the rarity of this condition, the heterogeneity of patients with CFD and the long and complex treatment that is often required, there were only a small number of papers available. One paper with level 1 diagnostic evidence was identified.

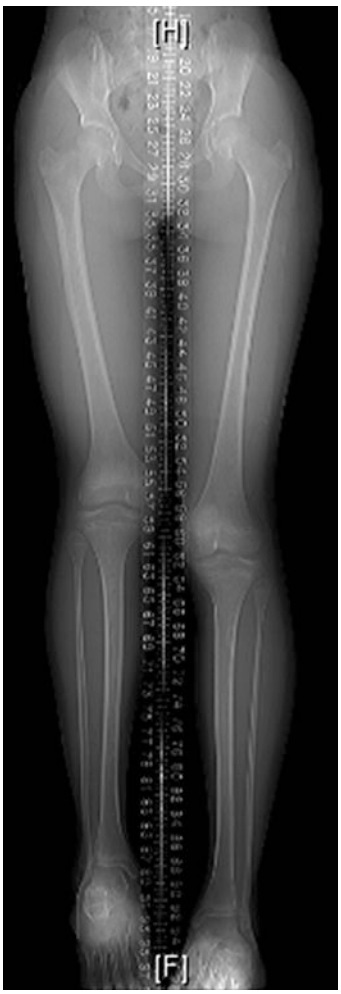

Fig. 7.1 A child with right type I CFD

What Is the Best Treatment for Congenital Femoral Deficiency?

Treatment options for CFD are individualized for each patient. The principles include assessment of final leg length discrepancy at skeletal maturity, equalization of leg length, either by epiphysiodesis of the contralateral leg or lengthening via distraction osteogenesis or ablation. Important factors to consider prior to lengthening include decisions on whether to treat acetabular dysplasia, if present, whether to reconstruct the hip and proximal femur and whether surgery to address knee instability and patella mal-tracking is indicated.

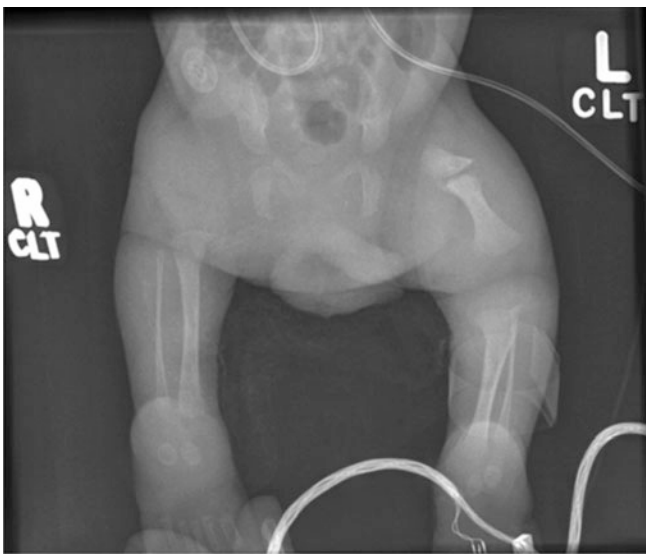

Fig. 7.2 A child with bilateral CFD, *left* IIb and *right* is IIIB

When Should Acetabular Surgery Be Performed?

Hip subluxation or dislocation is one of the most serious complications associated with femoral lengthening. The direction of dislocation is often posterior [6] Closed reduction under muscle relaxation may fail due to excessive soft tissue tension. Soft tissue releases alone may be unsuccessful

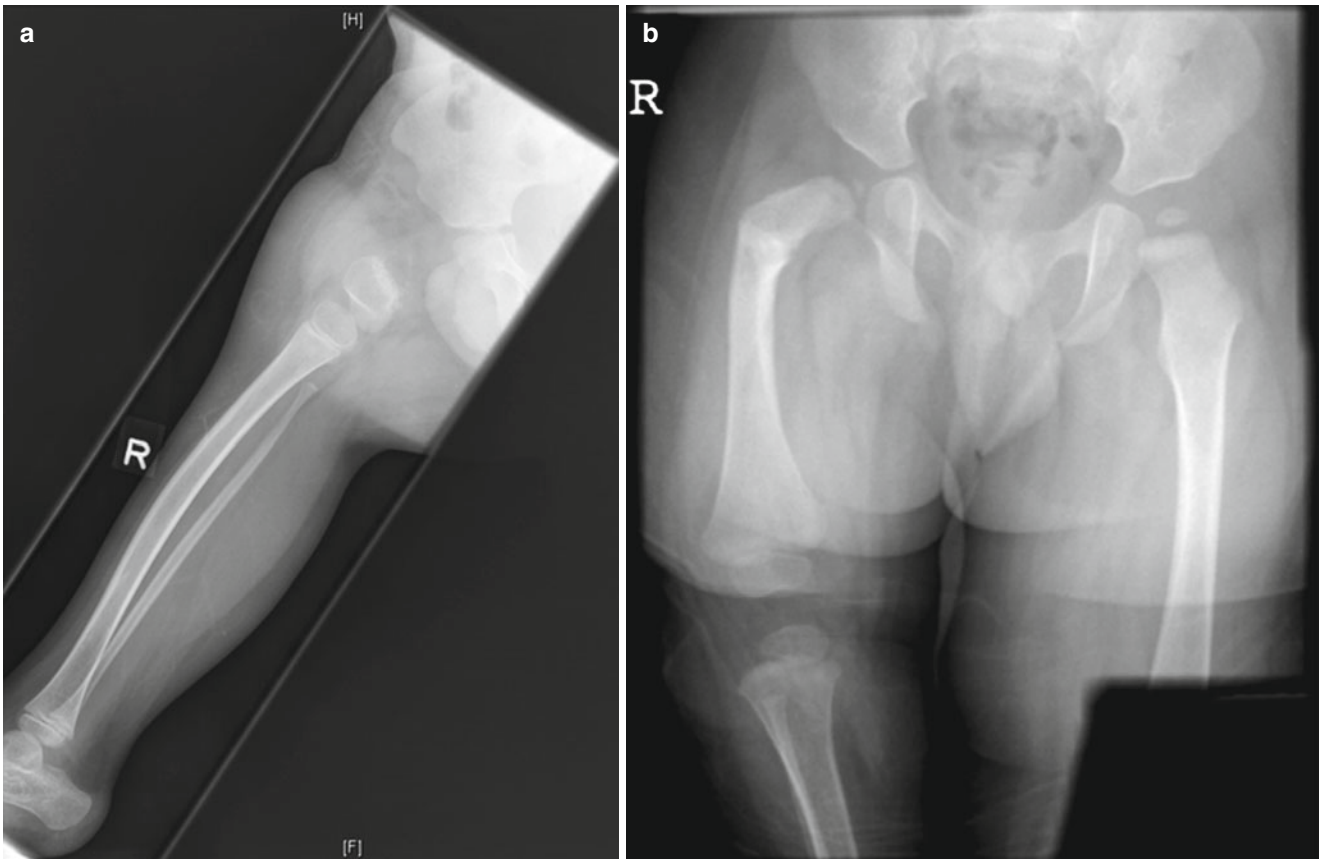

Fig. 7.3 A, B. Two other children with CFD

and femoral shortening as well as acetabular procedures such as a Dega or Shelf may be required. Therefore it is better to identify those patients in whom there is an increased risk of dislocation in order to perform preparatory surgery to protect the hip from deterioration and allow adequate lengthening to be achieved. Suzuki et al. [7] reported no hip deterioration in patients with a centre edge angle of >20 degrees in a series of patients undergoing femoral lengthening for both acquired and congenital causes compared to a 25 % rate of deterioration in hip congruency in those patients with a CEA of 20 degrees or less. Bowen et al. [8] recommends an acetabular index of 25 degrees or less as well as correction of a neck shaft angle (NSA) to 120 degrees prior to lengthening of Kalamchi Type 3a femora [9].

The choice of osteotomy type to address acetabular dysplasia should take into consideration the location of the deficiency in CFD. Suzuki et al. [7] recommended an innominate osteotomy. Millis and Hall have described a trans-iliac lengthening which is a modification of the innominate osteotomy by use of a trapezoidal wedge which both re-orientates the acetabulum and can generate between 2 and 3 cm of length [10]. However the deficiency in the acetabulum is posterior and lateral. There is relative acetabular retroversion combined with decreased femoral anteversion. This may result in posterior uncoverage if an innominate osteotomy is performed, therefore theoretically promoting rather than preventing a posterior dislocation during lengthening or increase the probability of developing femoro-acetabular impingement [11]. Paley recommends performing a Dega osteotomy in such a way to reduce the superolateral deficiency whilst also providing approximately 1 cm of length. In those patients with a closed triradiate cartilage a reorientating osteotomy such as a Ganz periacetabular osteotomy can be performed however, care should be taken to correct lateral coverage and not anterolateral coverage.

What Is the Most Appropriate Osteotomy Level for Femoral Lengthening?

Distal osteotomy sites allow for simultaneous correction of distal valgus deformity and lengthening from the same level. It also results in improved regenerate formation as the osteotomy is in the broad metaphyseal area of the bone. In addition it reduces the pressure on the hip, whilst increasing pressure on the knee joint. This can be counteracted by spanning the external fixator across the knee joint. In contrast proximal osteotomies exert less tension on the knee, but

more on the hip [3]. There is a reduction in bone regenerate healing in proximal osteotomies as compared to distal osteotomies [12]. Aston et al. [13] reported improved range of knee movement in proximal osteotomies as compared to distal osteotomies with no statistically significant difference in the healing index. The fracture rate reported in their series is reviewed later in this chapter.

Should the Knee Joint Always Be Spanned During Lengthening?

The knee in CFD is at risk of subluxation or dislocation during the lengthening phase due to the absence of either both cruciates or the anterior cruciate ligament alone [14]. During lengthening increased tension across the knee from the soft tissue structures can result in sagittal translation. Grill et al. [12] reported a 57 % incidence of knee dislocation or dislocation in patients with knee dysplasia. Gillespie and Torode [15] reported a maximum of 15–20 % increase in length of the femur before posterior subluxation of the knee occurred. Jones and Moseley [16] reported an incidence of posterior subluxation of 33 % when lengthening using the Wagner technique. In order to address this, extension of the external fixation across the knee joint can be performed. Aston et al. [13] spanned the knee in 26 out of 30 patients at the time of initial procedure and found it necessary to extend the frame across the knee in 2 of the remaining 4 patients. With careful identification of the centre of rotation of the knee joint a hinged extension can be extended across the knee, allowing for knee flexion and extension during physiotherapy. This may avoid the pitfalls associated with knee subluxation in lengthening. In a recent series one of 30 patients developed posterior subluxation only [17]. However it is difficult to determine whether knee stability in this series was conferred by a hinged external fixator, a preceding "Paley SUPERknee" procedure or more likely a combination of both.

Chomiak et al. [4] reported a level 1 diagnostic study of their arthroscopic findings of 21 patients with CFD. Only 1 patient in their series had normal cruciate ligaments. Ninety percent of patients had either an absent or hypoplastic ACL. The PCL was absent or hypoplastic in 61 % of patients. In 57 % of patients both the ACL and PCL were hypoplastic.

When Should a SUPERhip or SUPERknee Procedure Be Performed?

Prince et al. [17] reported a series of 30 cases of CFD, 24 of which (80 %) had undergone a SUPERhip procedure and 15 (63 %) had undergone a SUPERknee; however this paper was focused on outcomes following external fixation. No other series of SUPERhip or SUPERknee procedures could be identified in the literature.

Is It Necessary to Rod the Femur After Lengthening?

Fracture of the femur following removal of an external fixation device poses a significant management problem. They can occur through bone regenerate, the junction between host bone and regenerate bone, though a half pin or at a site unrelated to the previous surgery [18]. They are difficult to treat because of tight soft tissue structures due to lengthening leading to a tendency for progressive angulation, an absence of an intramedullary canal at the osteotomy site, the pin sites may produce sclerotic areas which are a block to the passage of an intramedullary nail and there is a risk of sepsis or deep infection from prior pin sites. Prince et al. [17] reported a 7 % fracture rate of the regenerate after Rusch rodding after surgery. Aston et al. [13] performed rodding at the time of the index procedure and reported an overall 30 % rate of femur fracture. There was no difference in fracture rate between proximal and distal osteotomy sites in their study when comparing proximal osteotomy and simultaneous rodding with distal osteotomy and no rodding. The addition of rodding reduced their fracture rate in proximal osteotomies from 100 % without a nail to 0 % with a nail in their small subseries of 13 patients.

What Is the Most Appropriate Ablative Procedure?

In patients in whom reconstructive surgery is not possible prosthetic fitting is required to resolve postural imbalance. Non surgical treatment involves the use of an extension prosthesis. However, these can be bulky due to the contour of the foot and an alternative surgical procedure is ablation of the foot by either a Syme's amputation or a Boyd's amputation with or without a knee fusion. Kant [19, 20] et al. compared Locomotor Index and overall satisfaction between 2 groups; those that had undergone Syme's amputation and those that had an extension prosthesis and found higher levels of function and satisfaction in the non-surgical group 21. Rotationplasty as described by Van Ness and several modifications of the procedure involves a rotational osteotomy which replaces the knee with the ankle joint that has been externally rotated by 180° [21–24]. This allows for fitting of a below knee type prosthesis. Alman et al. [25] compared the results of Syme's amputation with knee arthrodesis with rotationplasty and found no difference in gross motor function or perceived appearance, but increased energy efficiency in patients with a rotationplasty. Fowler et al. [26, 27] also

Table 7.1 Summary of grades of recommendations

Statement	LOE/Grades	References
1 There is poor evidence supporting what degree of dysplasia should be corrected prior to lengthening	4/C	[6–8, 11]
2.There is poor evidence that a distal osteotomy is advantageous over a proximal osteotomy	4/C	[13]
3. There is good evidence that without protection of the knee, subluxation or dislocation may occur during lengthening	1/B	[4, 12, 13, 15, 17]
4. There is poor evidence to support the indications of SUPERhip or SUPERknee procedures	4/C	[17]
5. There is poor evidence to support prophylactic rodding of the femur	4/C	[13]
6. There is fair evidence to support a maximum lengthening of 6 cm	3/B	[13, 17]
7. There is fair evidence to support the use of rotationplasty over Syme's amputation for ablative surgery	3/B	[19, 27–29]

found improved energy efficiency in rotationplasty patients compared to Syme's patients, with more normal knee kinematics and enhanced prosthetic knee function in rotationplasty patients, provided the tibia had not derotated. Ackman et al. [28] examined the long term effects of rotationplasty to normal controls and found significant differences in gait and posture but no differences in health and well being.

How Much Length Should Be Attempted in a Single Lengthening?

Prince et al. [17] found a statistically significant decrease in function and increase in pain if greater than 6 cm of length was achieved as well as worse pain and comfort scores with similar global function scores if greater than 25 % of the original length of the femur was obtained. Aston et al. [13] found an increased rate of delayed ossification when greater than 6 cm of lengthening was attempted using a circular fixator and fine wire fixation as well as a significantly increased rate of regenerate fracture if greater than 20 % of the original femoral length was attempted.

Summary

Table 7.1 summarizes the literature on CFD around some of the controversies which exist in its treatment. The current literature does not allow us to confidently recommend many treatments which are routinely practiced today. This is in part due to the rarity of the condition, but also to the evolution of surgical practice, which makes it difficult to assess the effects of individual procedures as they are often combined with multiple additional procedures during the course of treatment.

References

1. Ring PA. Congenital short femur; simple femoral hypoplasia. J Bone Joint Surg Br. 1959;41-B(1):73–9.
2. Aitken GT. Proximal femoral focal deficiency-definition, classification and management. In: Aitken GT, editor. Proximal femoral focal deficiency. A congenital anomalys Symposium held in Washington, June 13, 1968. Washington, DC: National Academy of Sciences; 1969. p. 1–22.
3. Paley D, Guardo F. Lengthening reconstruction surgery for congenital femoral deficiency. In: Advanced techniques in limb reconstruction surgery. Springer Science + Business Media. 2014. p. 245–299.
4. Chomiak J et al. Cruciate ligaments in proximal femoral focal deficiency. J Pediatr Orthop. 2012;32(1):21–8.
5. Alshryda S, Jones S, Banaszkiewicz P. Postgraduate paediatric orthopaedics: the candidate's guide to the FRCS (Tr and Orth) examination. Cambridge: Cambridge University Press; 2014.
6. Dhawale AA, Johari AN, Nemade A. Hip dislocation during lengthening of congenital short femur. J Pediatr Orthop B. 2012;21(3):240–7.
7. Suzuki S et al. Dislocation and subluxation during femoral lengthening. J Pediatr Orthop. 1994;14(3):343–6.
8. Bowen JR et al. Factors leading to hip subluxation and dislocation in femoral lengthening of unilateral congenital short femur. J Pediatr Orthop. 2001;21(3):354–9.
9. Kalamchi A, Cowell HR, Kim KI. Congenital deficiency of the femur. J Pediatr Orthop. 1985;5(2):129–34.
10. Millis MB, Ahearn MC, Hall JE. Transiliac leg lengthening: experiences with a modified Salter osteotomy. Orthopade. 1990;19(5):283–91.
11. Dora C et al. Morphologic characteristics of acetabular dysplasia in proximal femoral focal deficiency. J Pediatr Orthop B. 2004;13(2):81–7.
12. Grill F, Dungl P. Lengthening for congenital short femur. Results of different methods. J Pediatr Orthop. 1991;11(6):812.
13. Aston WJS et al. Lengthening of the congenital short femur using the Ilizarov technique: a single-surgeon series. J Bone Joint Surg. 2009;91-B(7):962–7.
14. Johansson E, Aparisi T. Missing cruciate ligament in congenital short femur. J Bone Joint Surg Am. 1983;65(8):1109–15.
15. Gillespie R, Torode IP. Classification and management of congenital abnormalities of the femur. J Bone Joint Surg Br. 1983;65(5):557–68.
16. Jones DC, Moseley CF. Subluxation of the knee as a complication of femoral lengthening by the Wagner technique. J Bone Joint Surg Br. 1985;67(1):33–5.
17. Prince, D.E., et al., Lengthening with external fixation is effective in congenital femoral deficiency. Clinical Orthopaedics and Related Research®. 2015.
18. O'Carrigan T, Paley D, Herzenberg JE. Obstacles in limb lengthening: fractures. In: Rozbruch SR, Ilizarov S, editors. Limb Lengthening and Reconstruction Surgery. New York: Informa Healthcare; 2007. p. 675–9.
19. Kant P et al. Treatment of longitudinal deficiency affecting the femur: Comparing patient mobility and satisfaction outcomes of syme amputation against extension prosthesis. J Pediatr Orthop. 2003;23(2):236–42.
20. Kantor R, Salai M, Ganel A. Orthopaedic long term aspects of bladder exstrophy. Clin Orthop Relat Res. 1997;335:240–5.

21. Brown KL. Resection, rotationplasty, and femoropelvic arthrodesis in severe congenital femoral deficiency. A report of the surgical technique and three cases. J Bone Joint Surg Am. 2001;83-A(1):78–85.
22. Van Nes CP. Rotation-plasty for congenital defects of the femur. J Bone Joint Surg B. 1950;32-B:12–6.
23. Hamel J, Winkelmann W, Becker W. A new modification of rotationplasty in a patient with proximal femoral focal deficiency Pappas type II. J Pediatr Orthop B. 1999;8(3):200–2.
24. Steel HH, Lin PS, Betz RR. Iliofemoral fusion for proximal femoral focal deficiency. J Pediatr Orthop. 1988;8(1):120.
25. Alman BA, Krajbich JI, Hubbard S. Proximal femoral focal deficiency: results of rotationplasty and Syme amputation. J Bone Joint Surg Am. 1995;77(12):1876–82.
26. Fowler EG et al. Contrasts in Gait mechanics of individuals with proximal femoral focal deficiency: syme amputation versus Van Nes rotational osteotomy. J Pediatr Orthop. 1999; 19(6):720.
27. Fowler E et al. Energy expenditure during walking by children who have proximal femoral focal deficiency. J Bone Joint Surg Am. 1996;78(12):1857–62.
28. Ackman J et al. Long-term follow-up of Van Nes rotationplasty in patients with congenital proximal focal femoral deficiency. The Bone & Joint Journal. 2013;95-B(2):192–8.
29. Fowler EG et al. Contrasts in gait mechanics of individuals with proximal femoral focal deficiency: Syme amputation versus Van Nes rotational osteotomy. J Pediatr Orthop. 1999;19(6):720–31.

Evidence-Based Treatments for Coxa Vara

8

Claire F. Murnaghan and Kim Ferguson

Abstract

Coxa Vara is a deformity of the femoral neck resulting in a neck shaft angle of less than 110°. It may be congenital, acquired or developmental. Developmental coxa vara has been distinguished from the other types as it occurs at the level of the proximal femoral physis. In this chapter we discuss normal proximal femoral development and the pathophysiology of coxa vara. The radiological and clinical features are discussed and illustrated along with evidence based guidance for treatment, including proposed surgical management.

Keywords

Coxa Vara • Joint reaction force • Proximal femoral deformity • Neck shaft angle • Head shaft angle • Hilgenreiner-epiphyseal angle (HEA) • Proximal femoral ossification

Introduction

Coxa Vara is a deformity of the femoral neck, resulting in an abnormal neck-shaft angle of less than 110° (Fig. 8.1).

It has been described in many different ways over the years, but we shall use the following classification [1]:

1) Developmental (previously named Infantile or Cervical)
2) Congenital (associated with congenital short femur)
3) Acquired (encompassing traumatic and those associated with skeletal dysplasias)

For the purpose of this chapter, we shall concentrate on Developmental coxa vara, as treatment of the various other types of coxa vara will depend on the heterogeneous underlying causes of the deformity.

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Developmental coxa vara in this sense occurs at the level of the physis, whereas the other causes occur distal to the physis.

Background

The finding of Developmental coxa vara (DCV) was first described by the Italian Fiorani in 1881 as a "bending" of the femoral neck [2]. He believed that the origin was rachitic. In 1881 Muller described a similar appearance in adolescents, but the terminology coxa vara was attributed to the condition described by Hofmeister in 1894 [3, 4].

Hoffa in 1905 examined pathological specimens of the proximal femur in affected individuals and concluded that the aetiology was unlikely to be either rickets or trauma and instead introduced the idea of a developmental condition [5].

In 1960 Pylkkanen performed a detailed literature search of the available information relating to coxa vara as well as adding weight to the pathophysiological knowledge of the influencing processes [6]. In Scandinavia, Johanning reported an incidence of 1 in 25,000 live births in the early 1950s [6]. Males and females are equally affected by this condition and there appears to be no difference in the frequency of the side involved. It may be bilateral in up to one third of individuals.

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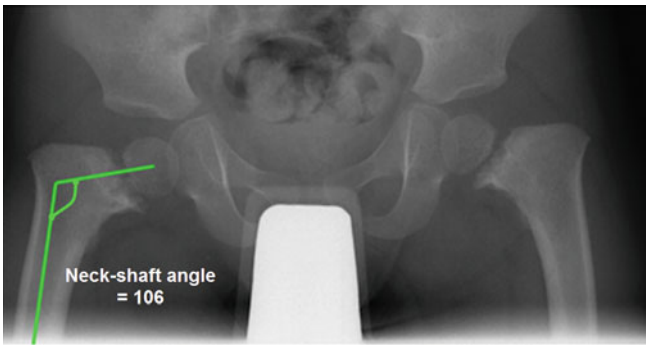

Fig. 8.1 Bilateral coxa vara with neck shaft angle approaching 106°

Normal Femoral Development

The proximal femur in the neonate is completely cartilaginous, with the recognisable shape of the femoral head and the trochanters determined by the enchondral ossification at the junctions between the metaphysis and the epiphyseal plates. The physeal plate gradually migrates proximally and divides into 2 separate plates, one for the femoral head and the second one for the greater trochanter. The medial portion of the cervical physis progresses early, resulting in elongation of the femoral neck. An ossification centre appears within the femoral head somewhere between 4 and 7 months after birth and can then be seen on x-ray. The more lateral greater trochanteric apophysis starts to ossify at about 4 years of age. The proximal femoral growth plate accounts for one third of the longitudinal growth of the femur.

Relative growth of these physes results in the changing morphology of the proximal femur. At birth, the normal neck-shaft angle is about 150° and in adulthood it is approximately 127° (often decreasing to 120° by the 7th decade). In a similar fashion, anteversion of the femoral neck may be as much as 40° at birth. By adulthood the normal femoral anteversion ranges from 4° to 25°.

Pathophysiology

Over the past century, many different theories have been suggested as causes of coxa vara, including Staphylococcal Albus infections, bending of the neck secondary to pressure within the uterus, rickets and AVN-like changes. In 1928 Fairbank described a triangular piece of bone in the distal portion of the medial femoral neck as a potential cause of the problem [7].

Following work by Zimmerman (1938) and others, it has become clear that the condition does not exist at birth, but develops after the perinatal period as a result of a defect in the enchondral ossification of the medial femoral neck [8]. Hence the nomenclature of "developmental" coxa vara.

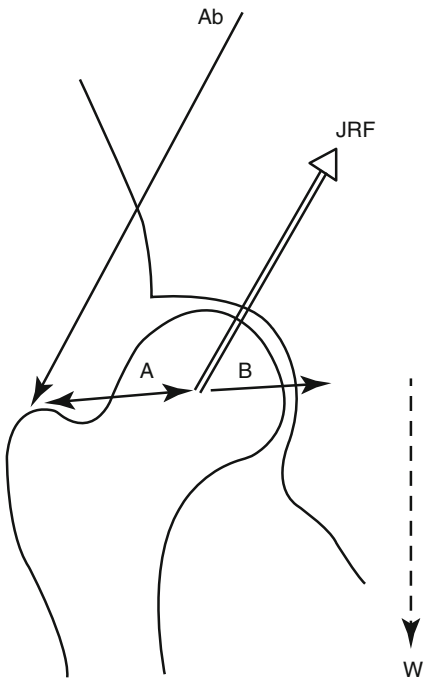

Fig. 8.2 Free body diagram of the hip in coxa vara. Joint reaction force is the force generated within a normal hip joint. This is a balance between the body weight moment arm (*B*) and the body weight (*W*), and the abductor moment arm (*A*) and abductor tension (*Ab*)

On histopathological examination of specimens obtained in Helsinki and compared to normal individuals, there were abnormalities of varying severity found in 3 main areas [6]:

a) Changes in ossification at the physeal cartilaginous plate, with disorganisation and irregularity of columns of cells
b) Changes in the adjacent metaphysis, including thin, irregular trabeculae and lack of proper ossification
c) Fibrous connective tissue infiltrating the bone marrow of the metaphysis and within the cartilaginous tissue

Deformation of the femoral neck can occur because of reduced strength in the region of the medial femoral neck because of bony and cartilaginous abnormalities. Once the neck-shaft angle reduces, the greater trochanter migrates towards the ilium and abduction is reduced. The higher level of the greater trochanter increases the abductor lever arm, reducing the joint reaction force crossing the hip in single leg stance. The joint reaction force required to maintain a horizontal pelvis during single stance gait decreases (Fig. 8.2).

In the normal hip, the force transmitted to the proximal femoral neck would include a net tension force at the superior or lateral cortex and a net compressive force at the inferior or medial cortex.

However, the shearing and bending forces on the neck will increase with increasing varus deformity as the physis

changes its position from horizontal to more vertical, with restriction of growth on the medial physis relative to the lateral physis. The more vertical position of the proximal femoral physis would increase not only the sheer component of the hip force but also the net medial compressive force on the metaphyseal bone of the femoral neck. These forces overwhelm the mechanical strength of the abnormally ossified bone in this area. This may lead to a relentless and progressive cycle of deformity.

In summary, developmental coxa vara is caused by a disturbance in ossification of the medial femoral neck, resulting in delayed growth and irregularity of bone structure, and is compounded by mechanical factors, i.e. weight-bearing through this region of abnormal bone, and attains its final stage at skeletal maturity.

Presentation

Children tend to present after walking age with an abnormal gait. In unilateral cases, a painless limp is noted and there may be a leg length discrepancy, with the affected side shorter, and increased girth of the thigh. In a young child, a late presentation of developmental hip dysplasia is a more common differential diagnosis.

In bilateral cases there is a waddling gait secondary to weakness of hip abductors as already described (a Trendelenberg gait). In cases of coxa vara, there is reduced anteversion or even some retroversion of the neck and so the femur is relatively externally rotated, resulting in reduced internal rotation. The iliofemoral ligament is under increased tension due to the relative retroversion and so complete extension of the hips may not be possible, with resultant fixed flexion leading to an exaggerated lumbar lordosis, especially if the disease is bilateral.

The clinician should always check for signs of a more generalised skeletal dysplasia such as short stature, limb length discrepancies and disproportionate growth of limbs compared with trunk. Generalised rotational profile of the lower limbs should be performed as well as a full hip examination. Other causes of coxa vara should also be considered such infection, metabolic bone disease, trauma and the aforementioned skeletal dysplasias. It is worth noting that developmental coxa vara is associated with other forms of lower limb deficiencies.

Radiological Findings and Measurements

As the femoral head begins to ossify the radiological appearances become apparent. The neck-shaft angle of the hip measures less than 110° on an AP radiograph of the pelvis, with a possible triangular fragment seen in the region of the calcar of the femoral neck (Fairbank's triangle), up until the age of about 10 years. Acetabular dysplasia seems to develop due to a change in normal weight-bearing, but is much more evident in adults than in children (Fig. 8.3).

The head-shaft angle is formed between the axis of the femoral shaft and a line drawn perpendicular to the base or the femoral capital epiphysis. The Hilgenreiner-epiphyseal angle is formed by Hilgenreiner's line and the physis [9, 10]. In normal hips it would measure approximately 25° [9]. Note that an ossified femoral epiphysis is required for all these measurements to be made (Fig. 8.4).

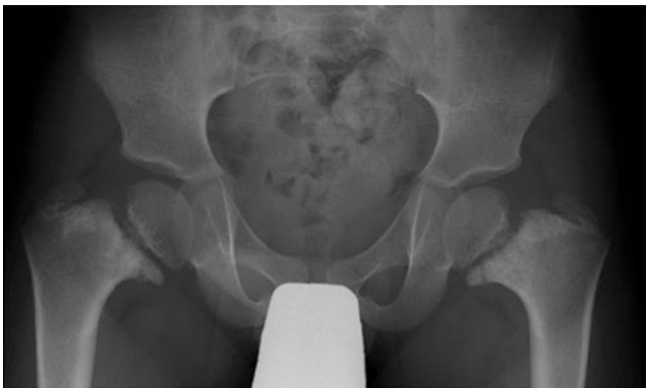

Fig. 8.3 Plain radiograph showing coxa vara with Fairbank's triangles in both hips

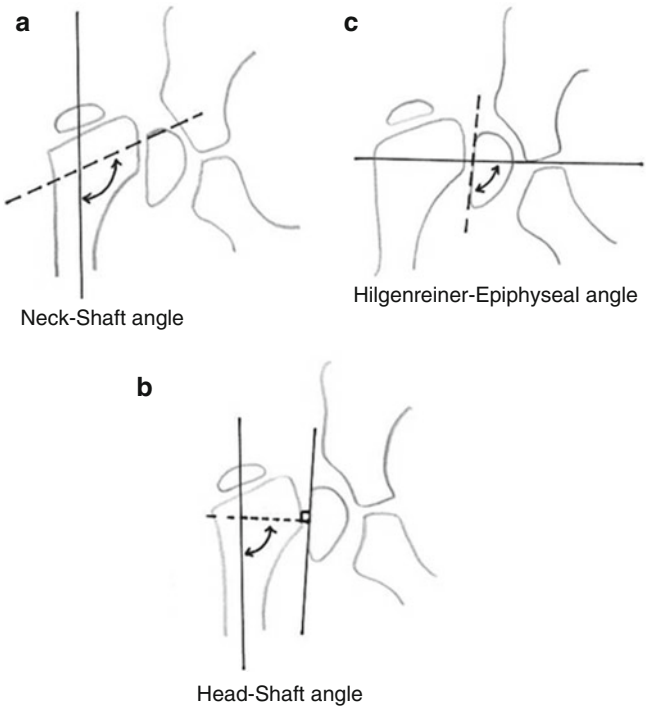

Fig. 8.4 Radiological measurement in coxa vara

Treatment

The aims of treatment for coxa vara centre on restoring normal angles (either neck-shaft or HEA), restoring normal mechanical function of the hip abductors, and attempted stimulation of more normal bone formation. Non-surgical and conservative measures have limited benefit in patients with developmental coxa vara. Surgical correction of the abnormal neck shaft angle is the mainstay of treatment. Weinstein and colleagues performed a retrospective review of children with coxa vara in the late 1980s and suggested quantification of the deformity by serial measurement of the HEA to be one of the most important indications for intervention [10].

- HEA > 60°
- Painful limp
- Neck shaft angle <110°
- Associated leg length discrepancy
- Progressive deformity

A HEA of less than 45° does not usually suggest the need for surgical intervention but between 45° and 59° the child must be reviewed and imaged regularly, in case one needs to act.

The aim of operative intervention is to prevent secondary complications. The anatomical aims of correction can mainly be achieved by a proximal femoral valgus osteotomy. Many different proximal valgus osteotomies have been described and numerous types of fixation devices proposed. In 1997 Carroll and Stevens suggested that no one type of osteotomy or fixation was better than another [11]. They also showed that children who underwent valgus osteotomy before the age of 8 years old had a 60 % chance of requiring revision surgery due to recurrence. Those over 8 years of age at the index procedure demonstrated a 50 % recurrence rate however, leading to the conclusion that age at time of operation is not predictive of final outcome.

The preferred method of valgus osteotomy has yet to be agreed upon [12, 13]. The first intertrochanteric valgus osteotomy was performed by Keetley in the late 1800s and futher modifications of this have been described [14]. Pauwel then described a complex and technically-demanding intertrochanteric osteotomy using tension band fixation of the femoral neck [15]. Epiphyseodesis of the greater trochanter has also been described [16]. Unreliable by itself, Desai then described valgus osteotomies and trochanteric osteotomies together [17].

Rather than the type of proximal femoral valgus osteotomy performed, more important is the understanding of the correction required to prevent recurrence of the varus deformity. Rates of stated recurrence of the varus deformity vary widely, but are mainly 30–70 %. Correction of the head-shaft angle to 135° did not prevent recurrence, in fact 63 % recurred but the most consistent and reliable predictor of success is reported to be a post-operative HEA of <38° (95 % success) whereas 93 % require revision surgery if the HEA remains >40° [18, 19].

Conclusions

When Should I Operate?

Surgical intervention should be considered when:

- HEA greater than 60°
- Symptomatic
- Increasing deformity
- Increasing leg length deformity

What Operation Should I Perform?

A valgus osteotomy that you are familiar with, using an implant that you use regularly and which maintains valgus inclination with a HEA less than 38°.

What Is the Evidence?

As yet, the evidence does not recommend one osteotomy type or a particular fixation device. Timing is not particularly important, in relation to age of the child. One should aim to reduce the risk of recurrence by correcting the HEA to less than 38°.

What Are the Pitfalls?

The pitfalls are under correction, which leads to a higher risk of recurrence, residual leg length discrepancy, significant growth arrest and changes in the limb more distally (e.g.: genu valgum after correction.) The neck shaft angle should not be used alone to judge correction; it should be used in combination with HEA in order to select the correct patients for surgery and to ensure that correction is complete.

References

1. Beals RK. Coxa vara in childhood: evaluation and management. J Am Acad Orthop Surg. 1998;6(2):93–9.
2. Fiorani G. Sopra una forma speciale di zoppicamento. GazzOsp. 1881;2:717.
3. Müller E. Ueber die Verbiegung des Schenkelhalses im Wachstumsalter. Beitr Klin Chir. 1888;4:137.
4. Hofmeister F. Coxa vara. Eine typische Form der Schenkelhalsverbiegung. Beitr Klin Chir. 1894;12:245–318.

5. Hoffa A. Congenital coxa vara. Deutsche.med.Wchnschr 1905;31:1257.
6. Pylkkanen PV. Coxa vara infantum. Acta Orthop Scand Suppl. 1960;48:1–120.
7. Fairbank HA. Coxa Vara due to congenital defect of the neck of the femur. J Anat. 1928;62(Pt 2):232–7.
8. Zimmermann M. Untersuchungen über Krankheitsbild und Ätiologie der „sogenannten Coxa vara congenita "oder „Coxa vara infantum ". ZOrthop. 1938;68:389.
9. Herring JA. Tachdjian's pediatric orthopaedics: from the Texas Scottish Rite Hospital for children. : Elsevier Health Sciences; 2013.
10. Weinstein JN, Kuo KN, Millar EA. Congenital coxa vara. A retrospective review. J Pediatr Orthop. 1984;4(1):70–7.
11. Carroll K, Coleman S, Stevens PM. Coxa vara: surgical outcomes of valgus osteotomies. J Pediatr Orthop. 1997;17(2):220–4.
12. Gunther CM, Komm M, Jansson V, Heimkes B. Midterm results after subtrochanteric end-to-side valgization osteotomy in severe infantile coxa vara. J Pediatr Orthop. 2013;33(4):353–60.
13. Barden B, Wesskamp B, Fitzek JG, Loer F. Curved osteotomy technique for intertrochanteric varus surgery–an experimental and computer-assisted surgery. Z Orthop Ihre Grenzgeb. 2001;139(4):311–6.
14. Keetley CB. Osteotomy of the Hip. Br Med J. 1884;1(1206):261–3.
15. Pauwels F. Biomechanics of the normal and diseased hip. : Springer Science & Business Media; 1976.
16. Langenskiold A, Salenius P. Epiphyseodesis of the greater trochanter. Acta Orthop Scand. 1967;38(2):199–219.
17. Desai SS, Johnson LO. Long-term results of valgus osteotomy for congenital coxa vara. Clin Orthop Relat Res 1993 Sep;(294) (294):204–10.
18. Amstutz HC, PD Jr W. Dysgenesis of the proximal femur (coxa vara) and its surgical management. J Bone Joint Surg Am. 1962;44-A:1–24.
19. Amstutz HC, Freiberger RH. Coxa vara in children. Clin Orthop. 1962;22:73–92.

Femoro-Acetabular Impingement in Children

9

Sarah Rubin and Manoj Ramachandran

Abstract

Ganz described the modern concept of femoroacetabular impingement (FAI) in 2003. FAI can lead to pain and reduced range of movement in teenagers and young adults. It may be primary or secondary to causes such as slipped upper femoral epiphysis (SUFE) or conditions contributing to femoral head or acetabular deformity such as Perthes' disease or developmental dysplasia of the hip (DDH). FAI can be cam, pincer or mixed form. Diagnostic signs include pistol grip deformity and crossover sign on plain radiographs or cam deformities on 3D imaging such as computer tomography (CT) or magnetic resonance imaging (MRI). Osteoarthritis (OA) and labral tears may be seen in conjunction with FAI but it is important to exclude other differentials. Treatment strategies include activity modification, arthroscopic or open surgery depending on the type of FAI and symptoms.

Keywords

Femoroacetabular impingement • Hip impingement • FAI • Cam • Pincer • Hip arthroscopy • Pistol grip deformity • Crossover sign • Alpha angle • Periacetabular osteotomy • Labral tear • Young adult hip

Introduction

Femoroacetabular Impingement (FAI) is a hip condition that typically causes pain and reduced range of movement in teenagers and young adults. The causes of impingement may be complex but usually stem from a lack of symmetry or congruity of the femoral head within the acetabulum. FAI is present in 10–15 % of all people; however at least 25 % of people with radiographic FAI may be asymptomatic. Recently the connection between FAI and early osteoarthritis (OA) has been documented.

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How Can FAI Be Subdivided?

Although FAI was described earlier in the twentieth century [1], Reinhold Ganz introduce the modern concept and treatment in 2003 [2]. He divided FAI into three types: cam, pincer and mixed. Cam type is caused by an abnormal offset between the femoral head and neck producing cartilage delamination of the acetabulum. Pincer type can be caused by over coverage due to protrusio or acetabular retroversion. This causes damage to the rim and a slower process of degeneration than cam deformity.

The most common form however is mixed FAI, which usually has a cam predominance (Fig. 9.1).

Acetabular Deformity

An unusually deep acetabulum can cause FAI because the femoral neck is excessively covered. Alternatively a retroverted acetabulum will cause impingement against the femoral neck during hip flexion and abduction.

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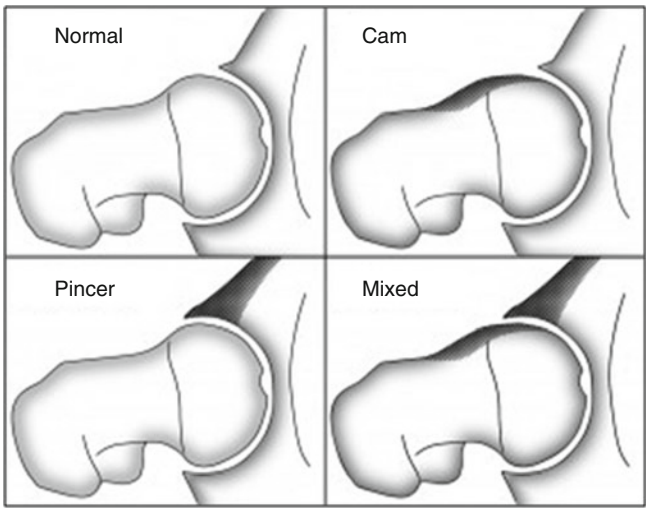

Fig. 9.1 Types of FAI

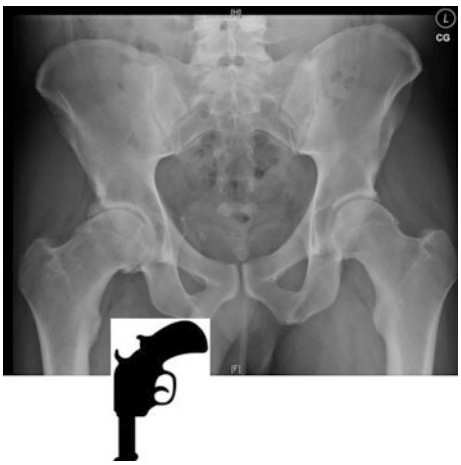

Fig. 9.2 Diagram of pistol grip deformity

Acetabular Dysplasia Classification

A. Classical: Classical DDH involves reduced coverage of the femoral head and is therefore not usually a type of dysplasia that contributes to FAI, but has been included for completion.
   a. Deficient head coverage
   b. Decreased acetabular depth
B. Acetabular retroversion: anterior wall is rotated forward or posterior wall is rotated backward
C. Acetabular over coverage: anterior and lateral wall over coverage of the femoral head.

Femoral Head Deformity

This can take two forms. Pistol grip deformity occurs when the femoral head loses sphericity and the head neck junction is less pronounced. The appearance is likened the handle of a pistol as shown in Fig. 9.2.

The femoral head can also develop a tilt deformity that is apparent superomedial migration of the femoral head.

Femoral Neck Deformity

This involves a shortened or widened femoral neck, which reduces the head-neck ratio.

What Are the Predisposing Factors for FAI?

Epidemiology

Males are affected more commonly than females. Men are more likely to have cam deformities and women are more likely to have acetabular retroversion or protrusio causing pincer type FAI. The typical age range is 14–45.

Childhood hip disorders such as slipped upper femoral epiphysis (SUFE), developmental hip dysplasia (DDH) and Perthes' disease can lead to secondary FAI.

SUFE is the leading cause of FAI contributing to its development in up to 50 % of cases [3]. Any degree of displacement of the capital femoral epiphysis can cause the femoral head to lose its spherical shape or affect the femoral head-neck ratio; however, the most common deformity caused by SUFE is a cam deformity. FAI can occur whether SUFE has been diagnosed and managed appropriately or missed. Even mild cases of SUFE that have been treated with percutaneous pinning show FAI changes on imaging in around 50 % of patients [4].

DDH can lead to FAI due to the potential for permanent deformity of the femoral head, acetabulum or head-neck shape. This can occur whether DDH is missed or managed with non-operatively or operatively. Following femoral varus osteotomy or pelvic osteotomy, it is possible for iatrogenic FAI to occur. Similarly, the incongruity and cam deformity associated with Perthes' can cause FAI and OA in later life.

Sporting activities that force the hip through repetitive or a high range of movement can cause impingement at the extremes of movement. Specific sports associated with FAI include gymnastics, dance, hockey and football. Elite athletes have a higher incidence of FAI than the general population.

Trauma may cause an acquired deformity around the hip contributing to FAI.

How Does FAI Present?

Patients typically present with symptoms of pain that can be localised to the groin, greater trochanter and sacroiliac joints.

It is unusual for pain from FAI to radiate distal to the knee; however FAI can be associated with lumbar pain that may resolve with treatment. Pain can be associated with specific activities such as prolonged sitting, walking uphill or performing sports. Other symptoms include the sensation or sound of clicking around the hip or a feeling of stiffness in the hip.

What Imaging Is Appropriate for Assessment of FAI?

The followings modalities are of value in assessing a child with FAI:

1. Plain radiographs or arthrograms (Fig. 9.3)
2. CT
3. MRI including MR arthrogram
4. Arthrogram
5. Hip arthroscopy

Figure 9.3 shows acetabular crossover sign at intersection of the anterior rim (*red line*) and posterior rim (*blue line*) and ischial spine sign. The ischial spine is visible due to retroversion of the acetabulum. The overall radiographic impression is of focal pincer FAI from a retroverted acetabulum. In addition a pistol-grip deformity (arrow) confirms cam-type FAI morphology. There is a further finding of a superolateral femoral head defect present in this radiograph.

Figure 9.4 demonstrates measurement of the alpha angle. On CT or MRI axial images a line is drawn from the centre of the femoral head through the middle of the femoral neck and a second line is drawn through a point where the contour of the femoral head-neck junction exceeds the radius of the femoral head. The angle between the lines is calculated. The image on the left shows a normal hip with an angle of under 55°. The image on the right shows a hip with FAI and an angle of 64°.

CT and in particular high resolution CT with 3D reconstruction can give a clear picture of the bony anatomy as shown in figure below where a cam lesion is evident. However, MR imaging is preferred because of the lower radiation dose (Fig. 9.5).

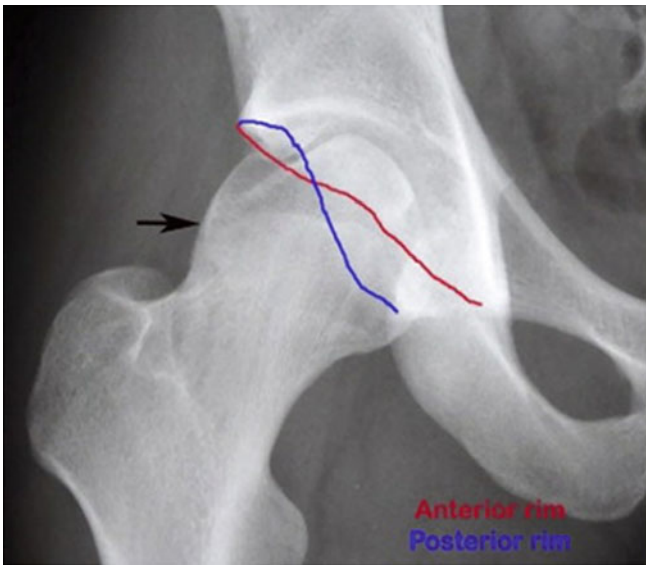

Fig. 9.3 AP radiograph of a hip

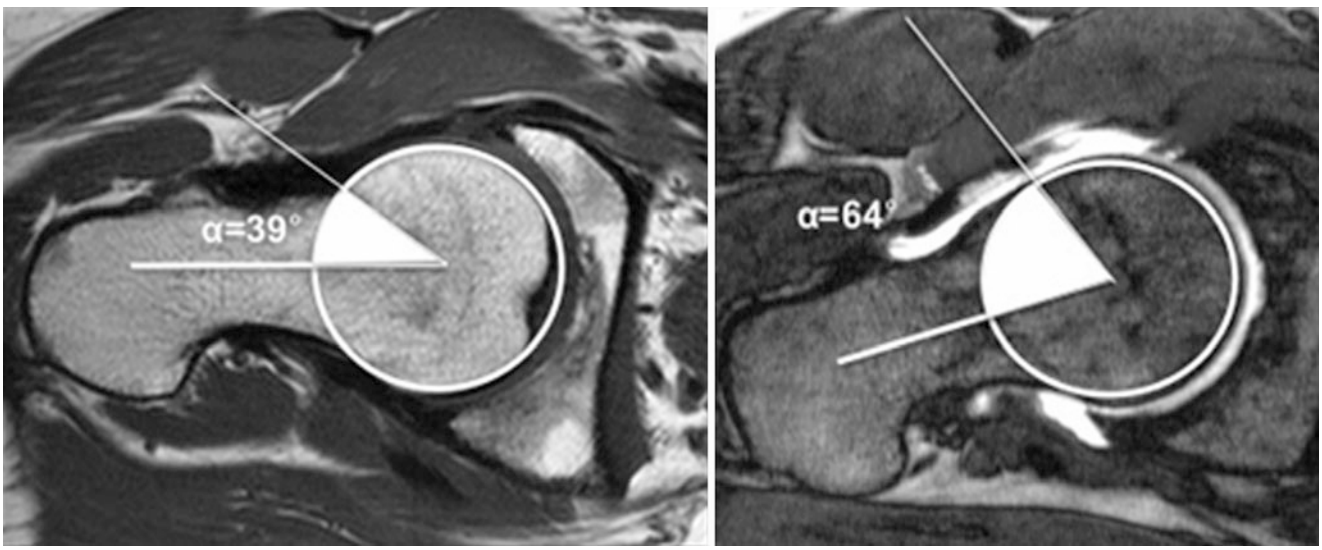

Fig. 9.4 Axial MRI of normal (*left*) and FAI hip (*right*) to demonstrate measurement of the alpha angle

Plain MRI is a useful investigation to assess the bony and soft tissue anatomy. It has the advantage of avoiding radiation exposure and is non-invasive. Soft tissue inflammation from impingement, tendonitis and muscle hernias will be evident on MRI. Alternatively, an MRI arthrogram can give more detail, assesses for associated problems and differential diagnoses such damage to articular cartilage and labral tears and is considered to be a more useful investigation.

How Should FAI Be Managed?

Non-operative management can be appropriate for those with mild symptoms or activity specific symptoms. This includes physiotherapy, activity modification and analgesia.

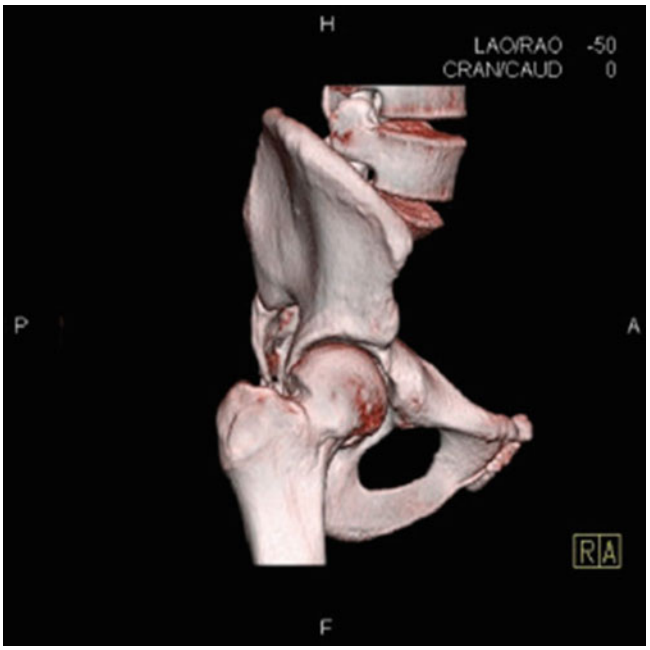

Fig. 9.5 3D CT reconstructions demonstrating a cam lesion

Physiotherapy should focus on strengthening the hip rather than range of movement and activities that involve stretching such as yoga should be avoided.

Intra-articular steroid and local anaesthetic injection can offer temporary relief from pain.

Surgical management options can be divided into:

- Arthroscopic
- Mini-open surgery
- Open surgery

With improving techniques and equipment, arthroscopy offers increasingly greater access to various pathologies within the hip (Fig. 9.6). This includes:

- Anterior cam lesions
- Anterosuperior pincer deformity
- Anterior inferior iliac spine deformity

During hip arthroscopy there is a chance the labrum will have to be detached, the pincer removed and the labrum may then be either repaired with suture anchors or debrided. Arthroscopy carries specific risks including damage from traction/counter traction such as sciatic and pudendal neurapraxia.

Traditionally arthroscopy has not been able to access medial to the retinacular vessels or the medial retinacular fold but with improved instruments, this is becoming possible (Fig. 9.7).

The mini-open approach can be used in combination with arthroscopy. For protrusio and posterior lesions, an open approach with hip dislocation and trochanteric osteotomy is often required. If impingement is due to a retroverted acetabulum, a periacetabular osteotomy may be indicated. Proximal femoral osteotomy may be performed if the proximal femur deformity is the source of FAI (Fig. 9.8).

If severe OA is associated with FAI, then the primary surgical management may involve arthroplasty.

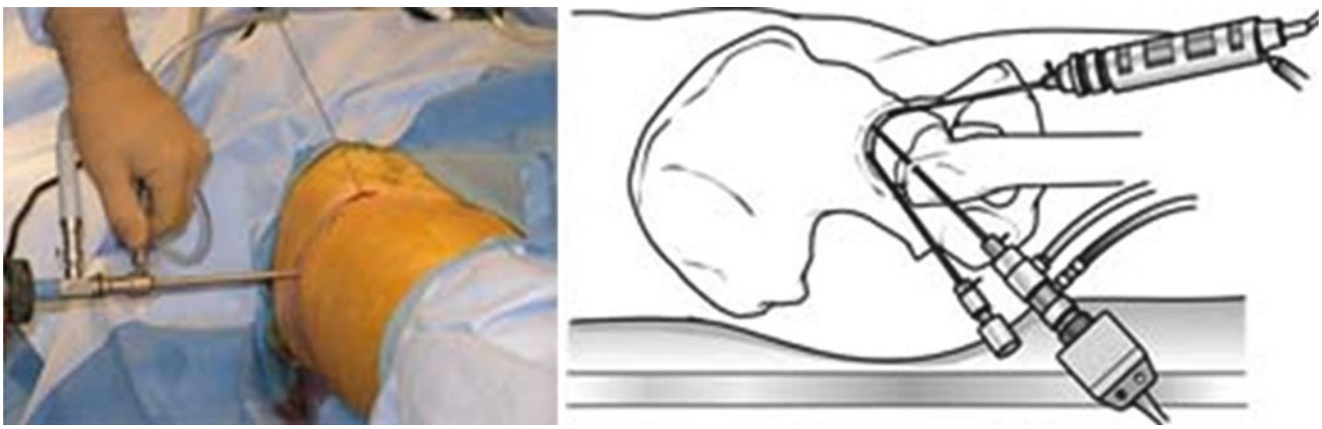

Fig. 9.6 Intraoperative photograph showing a hip arthroscopy and diagram showing port sites

Do Outcomes Differ Between Open and Arthroscopic Management of FAI?

Two systematic reviews assessing outcomes of surgical management of FAI are summarized below.

In 2015, Nwachukwu et al. [5] reviewed 16 studies; all were level II – IV evidence. Nine papers covered 600 open surgeries with a mean follow up of 57.6 months, the other 7 papers reviewed 1484 arthroscopic surgeries with a mean follow up of 50.8 months. Hip arthroscopy studies showed higher quality of life scores. Survivorship at follow up was assessed by need for hip arthroplasty and was 93 % for open and 91.5 % for arthroscopic procedures. Risk factors for hip arthroplasty were previous chondral injury and increased age.

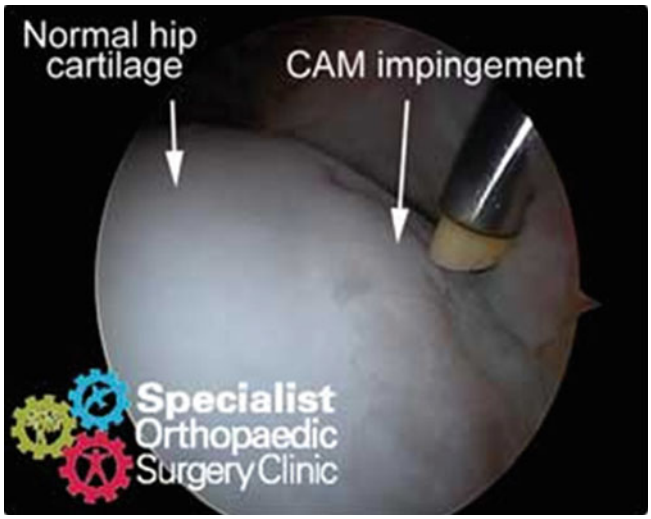

Fig. 9.7 Arthroscopy image demonstrating the femoral head and debridement of a cam lesion

In 2013 Harris et al. [6] performed a thoutough meta-analysis of 29 papers (2369 participants) on FAI management. One paper was level I evidence [7] and the rest were mainly level IV evidence. The mean follow up was 3.09 years. The followings were concluded:

- Improved outcome measures with surgical vs. non-operative management.
- No statistical difference in functional outcomes between outcomes of arthroscopic vs. open surgical techniques, however complication rates were significantly higher in dislocation techniques when compared to arthroscopic.
- 3.1 % overall rate of conversion to THR.
- Significantly better results in labral repair than debridement.

In summary outcome measures and re-operation rates are improved with arthroscopic surgery.

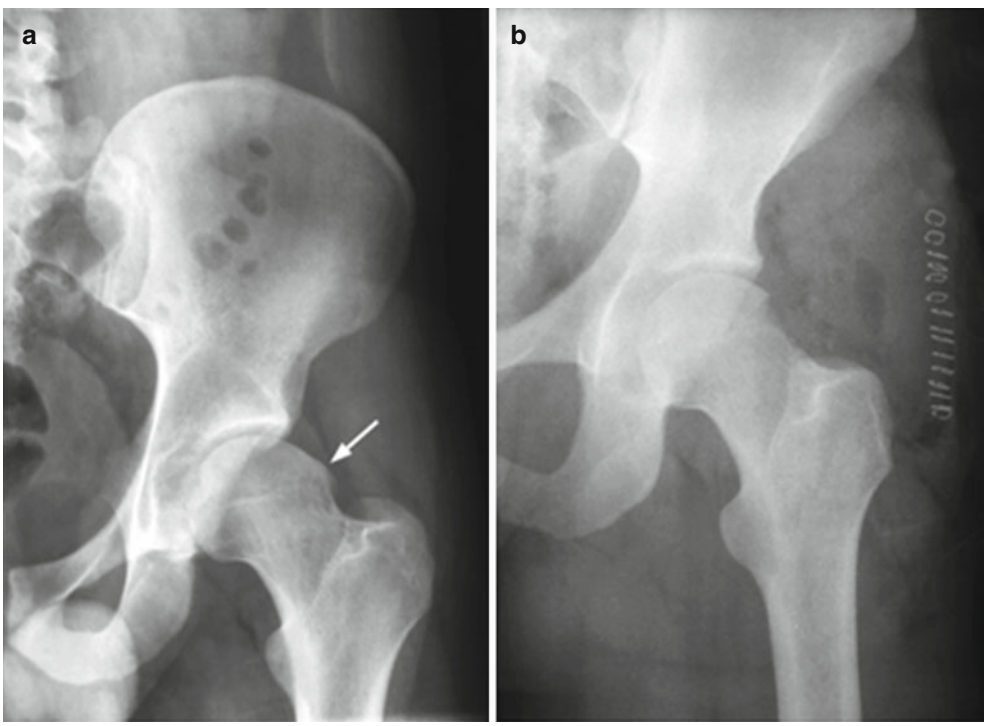

Fig. 9.8 Radiographs of a 21-year-old male. These pre and postoperative images demonstrate a femoral head cam deformity that has been resected via an open approach

This review is high quality; it addressed four main questions from a large number of studies and utilized a variety of statistical tests. The review was disadvantaged by the duration of follow up preventing long term assessment, the relatively low overall level of evidence and that studies were directly compared despite differing greatly in the assessments used to measure outcomes.

In Arthroscopic Surgery for FAI, Should a Detached Labrum Be Fixed or Debrided?

In 2012 Tibor and Leunig [8] performed a systematic review which evaluated 5 papers comparing labral resection versus preservation. It was found that arthroscopic FAI surgery improved outcome scores. Labral repair was associated with improved outcomes when compared to debridement; the improvement was small but statistically significant. Table 9.1 illustrates a summary of the review findings.

Criticisms of this review include a short follow up period of only 1 year for one paper and 2 years for the other 4 papers. As FAI treatment is relatively new and evolving studies of this type can quickly become out of date; this 2012 review found huge improvements in data quality when compared to a similar review done in 2007.

However, the review was valuable as studies included were all level III or higher and were directly comparable and also included detailed complications of surgery (Table 9.1).

What Is the Prognosis for FAI?

For any surgical management labral repair rather than debridement is associated with improved results. Following arthroscopic management of FAI, at least 50 % of patients have a reduction in pain at 3 months and 95 % of patients at 1 year. The approximate likelihood of progression to OA requiring arthroplasty within 2 years is 2 %.

Edwards et al. [14] performed a 213 patient study published in JBJS in 2009. This followed open surgery for FAI found good or excellent results with improved pain and range of movement for the majority of patients at 2 years. However up to a third of patients required further surgery within 3 years.

There is no overall consensus on a specific time when return to sport is considered safe following arthroscopic surgery. A 2015 systematic review of post hip arthroscopy rehabilitation by Grzybowski et al. reviewed 18 papers; 7 concerned FAI specifically, 4 papers focused on elite athletes and there was a minimum of 2 years follow up. Evidence was level III–IV [15]. Timing of return to sport in athletes following FAI surgery was at least 12 weeks and 96 % of athletes were able to return to sport. Overall the studies recommended following a progressive rehabilitation program with milestones to progress through prior to increasing activity, similar to following a rehabilitation program post anterior cruciate ligament reconstruction. It was recommended that a test of overall function be performed prior to return to sporting activities. The review was limited by the heterogenity of the papers and outcome measures used.

A 2010 study by Philippon et al. [16] followed up 28 skaters and hockey players. Subjects had a mean return to sporting drills of 3.4 months (range 1–5 months) following arthroscopic surgery compared with double this time following open surgery. Other studies assessing the timing of return to sport in athletes found similar results although did not

Table 9.1 Illustrates the summary of findings of a systematic review by Tibor and Leunig [8]

Study and Year	Pre op scores	Final follow up scores	OA progression and complications
Espinosa et al. [9].	F 17 (13–18) D 12 (8–13)	F 17 (13–18) D 15 (10–18)	F Tönnis grade 0.5 to 0.8 D 0.5 to 1.3
Larson and Giveans [10]	F mHHS 62 D mHHS 63	F 94.3 D 88.9	Tönnis grade trending towards higher scores in D. 3 cases of HO in debridement patients (prior to routine NSAID prophylaxis) 1 conversion to THA, 1 revision arthroscopy for debridement
Laude et al. [11]	NAHS 54.8 ± 12 (no distinction between F & D groups)	F 86 ± 11 F 82 ± 19	8 failed refixation, 1 femoral neck fracture, 2 deep infections, 1 case of HO, 11 converted to THA
Philippon et al. [12]	mHHS 58 (no distinction between F & D groups)	F 87 D 81	10 converted to THA
Schilders et al. [13]	mHHS F 60.2 (24–85) D 62.8 (29–96)	F 93.6 D 88.9	Not discussed

F fixation of labrum, *D* debridement of labrum, *mHHS* Modified Harris Hip Score, *THA* total hip arthroplasty, *HO* heterotrophic ossification, *NSAID* non-steroidal anti-inflammatory

share any common scoring systems and are not directly comparable.

Conclusion

FAI is a spectrum of symptoms arising from impingement around the hip, usually presenting in young adults and affecting males more than females. Athletes are more commonly affected. Impingement can occur secondarily to SUFE, DDH or Perthes' disease. Diagnosis is based on clinical findings combined with imaging studies. Specific signs on imaging such as crossover sign and pistol grip deformity are pathognomonic for FAI. The management can be symptomatic with activity modification but the evidence suggests patients who undergo arthroscopic or open surgery to offload the impinging areas had improved outcome scores. In athletes return to sport can be achieved within a few months of surgery. There is an association between FAI and OA, approximately 2 % of patients will go on to require arthroplasty within 2 years of any surgery. Evidence is constantly expanding in this area.

References

1. Brand RA. Femoroacetabular impingement: current status of diagnosis and treatment: Marius Nygaard Smith-Petersen, 1886–1953. Clin Orthop Relat Res. 2009;467(3):605–7.
2. Ganz R et al. Surgical dislocation of the adult hip a technique with full access to the femoral head and acetabulum without the risk of avascular necrosis. J Bone Joint Surg Br. 2001;83(8):1119–24.
3. Murgier J et al. The frequency of sequelae of slipped upper femoral epiphysis in cam-type femoroacetabular impingement. The Bone & Joint Journal. 2014;96-B(6):724–9.
4. Ganz R et al. The etiology of osteoarthritis of the hip. Clin Orthop Relat Res. 2008;466(2):264–72.
5. Nwachukwu BU et al. Arthroscopic versus open treatment of femoroacetabular impingement: a Systematic review of medium – to long-term outcomes. The American Journal of Sports Medicine. 2015.
6. Harris JD et al. Treatment of femoroacetabular impingement: a systematic review. Curr Rev Musculoskelet Med. 2013;6(3):207–18.
7. Krych AJ et al. Arthroscopic labral repair versus selective labral debridement in female patients with femoroacetabular impingement: a prospective randomized study. Arthrosc J Arthrosc Relat Surg. 2013;29(1):46–53.
8. Tibor LM, Leunig M. Labral resection or preservation during FAI treatment? A Systematic Review. HSS J. 2012;8(3):225–9.
9. Espinosa N. Treatment of femoro-acetabular impingement: preliminary results of labral refixation. J Bone Joint Surg (Am). 2006;88(5):925.
10. Larson CM, Giveans MR. Arthroscopic debridement versus refixation of the acetabular labrum associated with femoroacetabular impingement. Arthrosc J Arthrosc Relat Surg. 2009;25(4):369–76.
11. Laude F, Sariali E, Nogier A. Femoroacetabular impingement treatment using arthroscopy and anterior approach. Clin Orthop Relat Res. 2008;467(3):747–52.
12. Philippon MJ et al. Outcomes following hip arthroscopy for femoroacetabular impingement with associated chondrolabral dysfunction: minimum two-year follow-up. J Bone Joint Surg. 2009;91-B(1):16–23.
13. Schilders E et al. Arthroscopic treatment of labral tears in femoroacetabular impingement: a comparative study of refixation and resection with a minimum two-year follow-up. Bone Joint J. 2011;93-B(8):1027–32.
14. Edwards CJ et al. Early experience and results for 148 surgical hip dislocation performed For impingement. J Bone Joint Surg Br. 2009;91-B(Supp III 404):2009.
15. Grzybowski JS et al. Rehabilitation following hip arthroscopy – a systematic review. Front Surg. 2015;2:21.
16. Philippon MJ et al. Arthroscopic labral repair and treatment of femoroacetabular impingement in professional hockey players. Am J Sports Med. 2009;38(1):99–104.

Evidence-Based Treatment for Paediatric Diaphyseal Femoral Fractures

10

Mohamed Kenawey and Hossam Hosny

Abstract

Femoral fracture in children is one of the commonest fractures encountered by paediatric orthopaedic surgeons. Treatment options vary with age of the child, the type of the fracture and available resources and expertise. In this chapter we have explored the evidence that underpin the current practice, highlighting the evidence in some details and providing graded recommendations for treatment options in various scenarios.

Keywords

Paediatric femoral fractures • Nancy nails • ESINM • Rigid intramedullary nails

Introduction

Femoral fractures in children are common and depending on the age of the child, type of the Fracture and available resources and expertise, treatment may vary. In younger children (birth to 5 years) non-operative treatment is recommended [1]. The currently practiced non-operative options include Pavlik harness, early or immediate hip spica casting (including pins and plaster casts) and traction (skin or skeletal traction) with or without delayed spica casting [2].

External fixation is usually recommended in patients with head injury, in multiply injured patients, open fractures with significant soft tissue damage, some benign pathological fractures, and in certain types of fractures when the site of fracture, comminution and proximity to the growth plate are not suitable for other types of fixation [3, 4]. Delayed healing due to stress shielding and the high re-fracture rate following fixator removal have been widely regarded as major disadvantage.

Internal fixation has gained popularity in the treatment of paediatric femoral fractures and is recommended in children between 5 years to skeletal maturity [5]. Available internal fixation devices are Elastic Stable Intramedullary Nails (ESIN), compression plating or trochanteric entry rigid intramedullary nails [6].

The relevance of this topic has prompted the American Academy of Orthopaedic Surgeons in 2009 to publish evidence backed guidelines to inform choices of femoral fractures treatment and those guidelines were updated in 2015. In this chapter we explored and expanded on the current available evidence for treating femoral fractures in children.

Nonoperative Treatments

Pavlik Harness

The Pavlik harness is usually indicated in the treatment of femoral shaft fractures in young infants (birth to 6 months) [7]. This approach allows better skin care and perineal hygiene, minimal hospitalisation, ease of application and reduction by adjusting the harness with minimal cost and yields acceptable alignment with very high safety [8]. In a retrospective comparative study by Podeszwa et al. [9], two groups of infants under 1 year of age who had diaphyseal femoral fractures were treated with either Pavlik harness or the application of immediate hip spica (24 and 16 infants respectively). Infants in the Pavlik harness group had higher pain scores but there were no differences in radiographic outcomes. Skin problems were encountered in one third of the spica group and none

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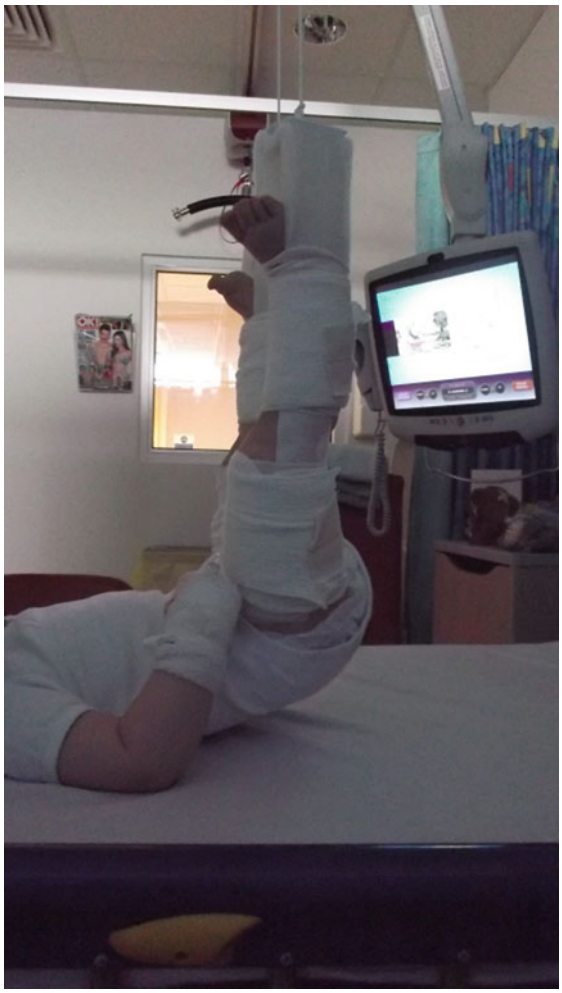

Fig. 10.1 Bryant traction for femoral shaft fracture

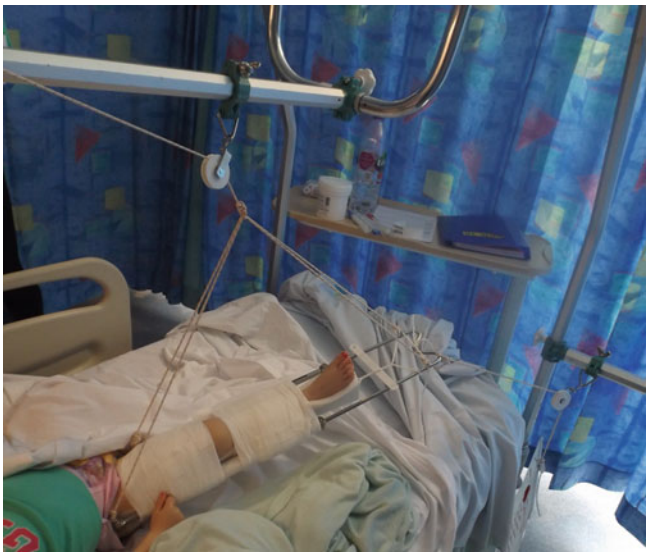

Fig. 10.2 Balanced traction as a temporary or definitive treatment for femoral fracture

were recorded with the Pavlik harness. Traditionally, Bryant's vertical overhead traction (Fig. 10.1) with the hips flexed 90 degrees and the knees straight might be used instead of a Pavlik harness in young infants [10, 11]. Vascular insufficiency and skin problems are the major risks with Bryant's traction. Wang et al. [11] compared retrospectively two cohorts of infants with femoral shaft fractures who were treated with either Pavlik harness or Bryant's traction (21 and 17 patients respectively) (level III evidence). Four of the 17 patients treated with Bryant's traction had skin complications that required treatment. Furthermore, hospital stay and charges were significantly lower for the Pavlik harness group.

Early or Immediate Hip Spica Casting and Traction

Early or immediate hip spica casting is the main treatment for isolated femoral shaft fractures in children 6 months to 5 years with shortening <2 cm [1, 5, 7, 12]. Some form of traction may be used in this age group for 3–4 weeks with or without delayed hip spica cast application in children with excessive shortening or unacceptable angulation in cast [7, 12] (Fig. 10.2). The advantages of hip spica casting are the low cost and high safety with high rate of good results [7, 13].

In a prospective study by Infante et al. [13], immediate closed reduction and hip spica application either in the emergency room under conscious sedation or in the operating room under general anaesthesia was a safe and effective treatment for isolated femoral shaft fractures in children from birth to 10 years of age who weigh <80 pounds (Level II evidence). After a follow up of at least 2 years, the only complication in 175 children was a re-fracture in a child who fell a week after cast removal. All fractures united within 8 weeks without significant residual angular deformity or limb length discrepancy. In another prospective study by Buehler et al. [14], they tried to identify and predict children who would develop excessive and unacceptable shortening (>25 mm according to their definition) following application of early hip spica cast for isolated femoral fractures (Level II evidence). Fifty children (2–10 years old) were included and 82 % (41 children) had an acceptable outcome. They introduced what they called "the telescope test" in which gentle axial compression was applied under fluoroscopy at the time of reduction and casting. If more than 3 cm of shortening could be demonstrated, traction was used rather than immediate spica casting.

The upper age limit for the use of spica for treating femoral fracture is usually decided on pragmatic basis rather than evidence basis. Although level II evidence does exist to support the use of hip spica casting up to the age of 10 years,

patients' and parent's convenience is a major drawback. Therefore, the current recommendation of hip spica treatment is limited to children between 6 months and 5 years [5].

Cassinelli et al. [15] retrospectively reviewed the results of immediate spica cast application in the emergency room in 145 children younger than 7 years (level IV evidence). All children younger than 2 years and 85 % of children 2–5 years had acceptable radiographic alignment. Re-reduction in the operating room was required in 11 % of the patients. They found that initial shortening was the only independent risk factor associated with loss of reduction. Mansour et al. [16] retrospectively reviewed two cohorts of children 6 months to 5 years of age who underwent either emergency department (ED) or operating room (OR) application of immediate spica cast (79 children versus 21 respectively) (Level III evidence). There was no significant difference regarding the quality of reduction or the rate of complications between the 2 groups. The hospital charges were significantly higher for the operating room spica casting, $15,983 versus $5150 for the ED casting.

The position of the hip and knee in the spica cast is also controversial [7]. Spica cast can be applied with the hip and knee at 90° flexion (the 90/90 sitting spica cast) [17] or in a more extended hip and knee (about 45° for both) [18, 19]. Illgen et al. [17] advocated the use of early sitting spica cast in a series of 114 femoral fractures in children under 6 years of age (Level III evidence). Loss of reduction was encountered in 20 % of patients and they proposed that a knee flexion angle <50 degrees as a predictive factor for loss of reduction and that >2 cm initial shortening was not a contraindication for early spica casting. However, due to the risk of compartment syndrome and Volkmann's ischemic contracture after 90/90 spica casting, most orthopaedic surgeons have moved to single or double leg spica casts in a more extended hip and knee position [19, 20]. Flynn et al. [19] studied prospectively the results of using single leg spica cast (walking spica cast) versus traditional spica cast in the treatment of low energy femoral shaft fractures in children 1–6 years of age (Level II evidence). The malunion rate was similar in both groups of children. However, the walking spica cast group were more likely to need cast wedging to treat fracture malalignment. All children with walking spica casts were able at least to crawl and 71 % were able to walk which significantly decreased the care burden for the family. In a more recent prospective randomized controlled trial by Leu et al. [18], 52 children 2–6 years old with diaphyseal femoral fractures were randomly assigned for either single or double leg spica cast (Level I evidence). The orthopaedic outcome was similar in both groups. However, children treated with the single spica cast were more likely to fit in regular car seats and fit comfortably in chairs. Furthermore, the parents of children treated with single leg spica cast took less time off work, which decreases the socioeconomic burden on the family (Fig. 10.3).

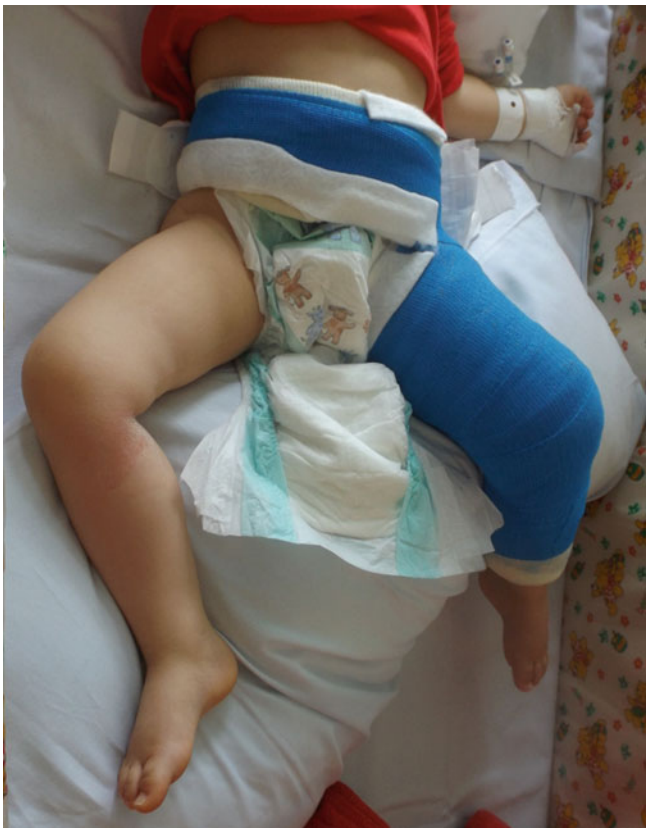

Fig. 10.3 Single leg spica for treating femoral fracture

Operative Treatments

External Fixation

External fixation use in paediatric diaphyseal femoral fractures is a minimally invasive treatment option with no soft tissue dissection and little scarring (Fig. 10.4). Blasier et al. [21] have used external fixation in the treatment of 139 fractures in 132 children with an average age of 8.9 years. The average external fixation time was 11.4 weeks and no cases of nonunion were recorded. The rate of pin tract infection, which required intravenous antibiotic treatment, was 4.5 %. Two refractures and one fracture through pin tract were encountered (Level IV evidence). In a randomised controlled trial by Wright et al. [22], external fixation was compared to early spica cast application in children aged 4–10 years (56 patients versus 45 respectively). At 2-years follow up, the malunion rate was significantly higher in the hip spica group [45 % versus 16 %]. Both treatment groups had similar means of RAND physical function health questionnaire, post-hospitalization questionnaire, and for patients satisfaction and for children happiness with treatment (Level I evidence).

In another randomized controlled trial by Bar-On et al. [23], the use of flexible intramedullary nailing (FIN) was compared to external fixation for the treatment of 20 femur

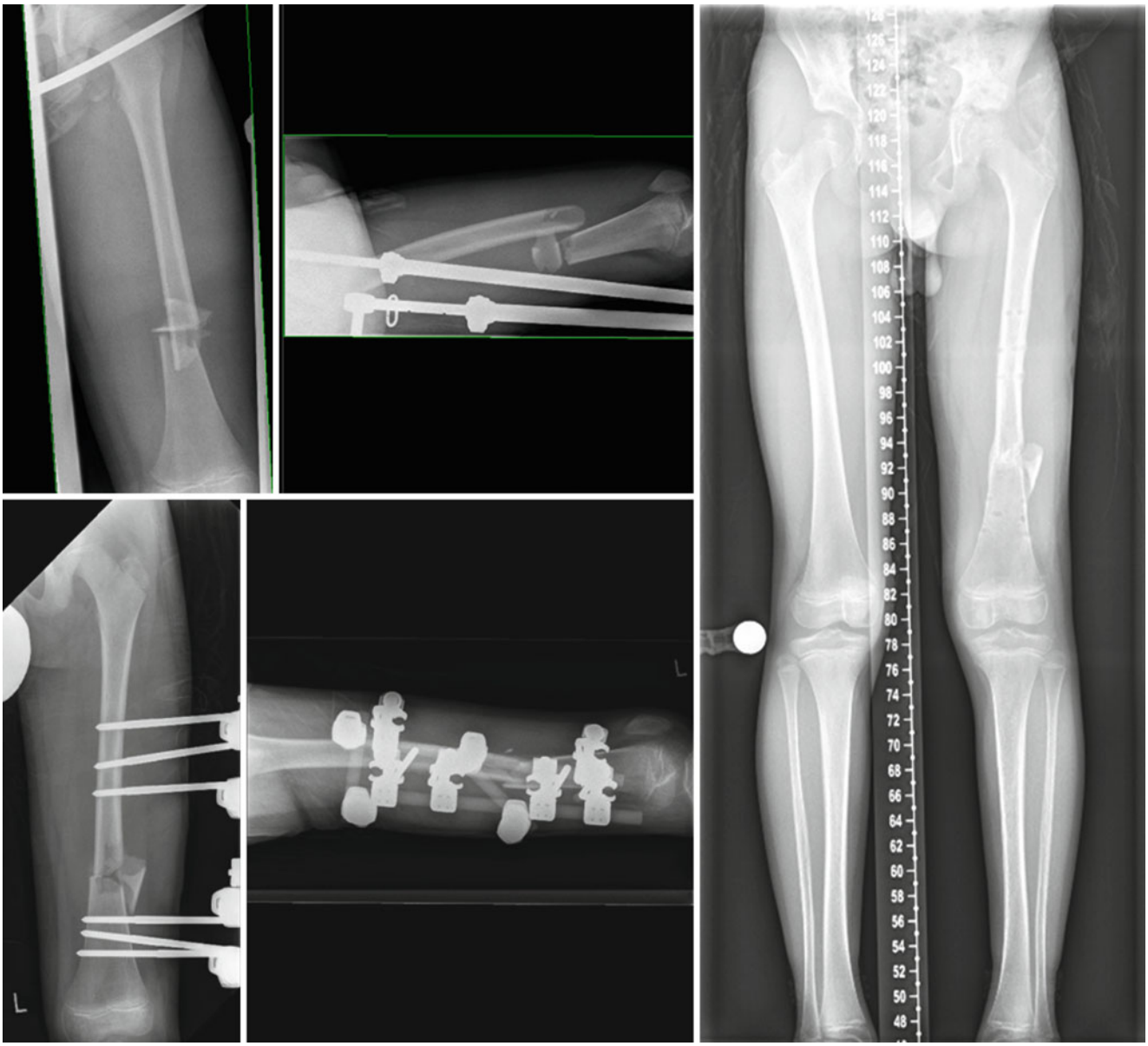

Fig. 10.4 External fixator treatment comminuted femoral shaft fracture

fractures in 19 children. The external fixator group had significantly less callus formation, full weight bearing at a mean of 10 weeks, full range of movement in 16 weeks with return to school within 13 weeks. In the FIN group, full weight bearing was possible in 7 weeks, full range of movement in 9 weeks and return to school at a mean of 5 weeks. Dynamization of the external fixator was thought to decrease stress shielding and promote callus formation with improved healing rates [24]. To study the effect of dynamization on callus formation and healing time, Domb et al. [24] randomized 52 patients to either a dynamic or a static external fixator for treatment of femoral shaft fractures. They found that axial dynamization of the external fixator had no significant impact on healing time and rate of complications (Level II evidence).

Internal Fixation

Implant Choice and Patient Characteristics – Age and Weight

Several implants have been used to stabilise femoral shaft fractures including ESIN, rigid nailing, plates and screws. ESIN is commonly considered for fixing femoral shaft fractures in children 5 years to skeletal maturity [5]. ESIN has also been used in children younger than 2 years [25–27] and patients up to the age of 18 years [25, 28]. Several studies have analyzed the effect of age and weight of patients on complication rates and final outcome using of titanium elastic nails (TEN) [28–31].

Sink et al. [29] compared two cohorts of patients with different surgical treatment algorithms over two different

periods of time (Level III evidence). The first period of 2 years represented their earlier experience during which TEN was used regardless of the age of the child or fracture stability (46 children). The higher complication rate (76 %) encountered with this cohort [32] led to a change of practice so that TEN would be used only in children younger than 11 years with a stable fracture. A second cohort of 95 children over the next 3-year period showed a significant decrease in complication rate to 12 %. They treated children 11 years and older who had unstable femoral fractures with submuscular bridge plating or rigid femoral nailing if the femoral canal size was adequate.

In a retrospective study of 234 fractures, (age range 3–18 years) (Level IV evidence), Moroz et al. [28] identified predictors of complications and poor outcome when using TEN. Age above 11 years was associated with poorer outcome (odds ratio = 3.86; P = 0.003). Furthermore, children who weighed >49 kg were 5 times more likely to have a poor outcome. There was a significant association between weight and poor outcome (P = 0.003).

Sagan et al. [33] found that weight was a significant predictor for anterior bow deformity greater than 15° with the use of TEN (Level III evidence). The mean weight of patients with this deformity was 46.5 kg ± 13.5 (n = 11), whereas the mean for those with no deformity was 36.8 kg ± 18.5 (n = 58). On the other hand, they found a trend of increasing frequency of malunion with increasing age, however this was not statistically significant.

Luhmann et al. [30] reported on the complications of TEN in a retrospective series of 43 femoral fractures in 39 children aged 3.8–9.3 years (Level IV evidence). They calculated weight/nail ratio to assess the effect of the patients' weight on the ability of TEN to control angulation. They found no association between weight/nail ratio and coronal (varus/valgus) angulation, while sagittal (apex anterior/posterior) angulation increased with increasing weight/nail ratio.

Implant Choice and Fracture Characteristics

Fracture Stability

Loss of reduction is a well-known complication following the use of ESIN in unstable femoral fractures causing unacceptable angulation or limb length discrepancy. Length unstable fracture patterns are either spiral or long oblique fractures (the length of the fracture is twice the diameter of the femur at the level of the fracture) or comminuted fractures (Winquist grades III and IV) [29, 32, 34, 35].

Sink et al. [32], in a retrospective comparative analysis (Level III evidence) of complication rates after using TEN in stable and unstable paediatric femoral fractures, found 8 patients (21 %) required further "unplanned" surgery before healing. Six of these eight had unstable fractures and underwent further surgery to shorten or remove extremely prominent or exposed nails or loss of reduction. Two had stable femoral fractures and underwent unplanned surgery to release a thigh compartment syndrome in one and to correct early coronal plane deformity by adding an external fixator in the second. Moreover, fracture shortening and angulation were significantly more prevalent in unstable fracture (10 out of 15 vs. 3 out of 24).

In a study by Lohiya et al. [36], stainless steel Ender's rods were compared to TENs in 73 children aged 4–15 years (Level III evidence). They reported a significant relationship between angular malalignment and the severity of fracture comminution. Eleven out of 14 patients (71.5 %) with Winquist grades III or IV fractures had malalignment as compared to 21 out of 59 with grades I or II.

In another study by Narayanan et al. [27], 79 femoral fractures in 78 children were stabilized using ESIN (age range 2 to 15 years). Five patients had loss of reduction that required operative correction (n = 2) or resulted in malunion (n = 3). The only two variables that were significantly associated with malunion and loss of reduction were the use of mismatched nail diameters and comminution >25 % of the shaft diameter.

Fracture Site

In a multi-centre North American study of 58 consecutive paediatric femoral shaft fractures (57 children) studying the early results of using TENs, 3 cases lost reduction in the early postoperative period (2 proximal third and one distal third fractures) while 5 of the 9 proximal third fractures healed with more than 5° of angulation. This indicates proximal and distal sites fractures may predict unfavourable outcomes [37].

Pombo et al. [38] reported the results of using TENs in the treatment of subtrochanteric fractures in 13 children 4–17 years (Level IV evidence). They defined subtrochanteric fractures as those within 10 % of the total femur length below the lesser trochanter. All patients had less than 5° of angulation. Eleven patients had excellent results. Two had satisfactory outcome due to limb length discrepancy of 1–2 cm with the fractured limb being longer than the unaffected side. They explained the limb length discrepancy as due to either intra-operative over-distraction across the fracture site or postoperative overgrowth of the femur. They stressed in their technique the importance of advancing the lateral nail into or just distal to the greater trochanter apophysis and advancing the medial nail into the femoral neck until it lies just short of the proximal femoral physis.

Elastic Nailing

Elastic Stable Intramedullary Nailing (ESIN or Nancy nails) were developed and first used at Nancy, France by Ligier et al. [39]. The principle of elastic stable nailing differs from the use of other flexible rods as Ender's rods, which are

stacked to fill the medullary canal. ESIN technique requires balancing forces between two opposing flexible nails of the same diameter. Therefore, it is important to select nails 40 % of the narrowest diaphyseal diameter; contour the nails with a similar gentle curvature, and use medial and lateral starting points that are at the same level in the metaphysis (Fig. 10.5) [27, 37]. Reported advantages are the minimally invasive approach, significantly reduced hospital stay, earlier mobilisation and functional recovery, avoid disruption of family life with its psychological impact on the child and earlier return to school and activities [25, 37, 39–41]. Different names were given for this type of fixation including ESIN (Elastic Stable Intramedullary Nailing), FIN (Flexible Intramedullary Nailing), FTN (Flexible Titanium Nails), TEN (Titanium Elastic Nails), SSEN (Stainless Steel Elastic Nailing) & Nancy nails [25, 33, 37, 39, 42].

Locked Versus Non Locked ESIN and the Use of End Caps

The advantage of using Ender's rods over ESIN is that the eyelets of the Ender's rods add locking capability to the end of the rods. This would in turn decrease symptomatic nail tip irritation and might be beneficial in case of length unstable diaphyseal fractures [34, 43].

Ellis et al. [34] retrospectively compared two groups of paediatric femoral diaphyseal fractures that had either locked Ender's nail fixation (n = 37) or unlocked Ender's rods (n = 70) (Level III evidence). They locked the end of the nail to the distal femoral metaphyses using 2.7 mm cortical screws 24 mm in length. Fracture shortening was significantly ($p < 0.001$) more in the unlocked group (12 mm versus 4.7 mm respectively) and this had caused implant migration, palpable lower ends, knee stiffness and deep infection. Eight patients in the unlocked group had a malalignment in either the sagittal or coronal plane of more than 10° but this was not statistically significant. However, the malalignment was considered unacceptable in only one patient in the locked group and revision was performed.

Self anchoring end caps to lock ESINs have been developed to provide a similar locking principle to the eyelets of the Ender's rods and tried in 8 femoral fractures (average age 10 years, range 7–15) [44]. The use of end caps eased the removal of nails as they prevented formation of excess bone around nail tips. Furthermore, they protected the skin very well and no nail tip irritation was recorded.

Titanium Rods Versus Stainless Elastic Nails or Rods

ESINs can be made of either titanium or stainless steel alloys. The French originators of this technique, Ligier et al. [39] reported the use of both materials. With the widespread adoption of this technique in North America, there has been a transition towards the exclusive use of titanium nails and several advantages such as closer elasticity to bone, better biocompatibility, better osteointegration and magnetic resonance imaging compatibility have been cited [31, 37]. The modulus of elasticity of 316L stainless steel is 187 GPa, making it 80 % stiffer than the titanium alloy (105 GPa). The lower modulus of elasticity of titanium would ease its intramedullary insertion and limit its permanent deformation. Increased elasticity and better compatibility would also decrease stress shielding and in turn would theoretically increase callus formation and healing rates [37]. The paediatric AO group has shown that a stainless steel nail has the strength of a titanium nail one size larger [45].

Wall et al. [31] compared retrospectively two groups of children (age 4–15, mean 9.4 years) (Level III evidence) who had either titanium ESIN (n = 56) or stainless steel ESIN (n = 48) fixation of femoral shaft fractures. The malunion rate was significantly higher in the titanium group (23.2 %) than in the stainless steel group (6.3 %). The increased flexibility of titanium compared to the stiffer stainless steel has been suggested as the cause of this increased deformation during fracture healing and the higher malunion rate. Furthermore, from an economic point of view, implant price by supplier in 2008 according to Wall et al. [31] was $259–$328 for titanium elastic nails depending on size of the nail and $78 for stainless steel flexible rods. In another retrospective study by Lohiya et al. [36], TENs used in 43 fractures were compared to stainless steel Ender's nails in 30 fractures (mean age 8.3, range 4–15 years) and the follow up was at least 5 years (Level III evidence). At the final follow up, significant malalignment in the sagittal plane (>15°) was found in 3 patients and minor malalignment in 29 cases and no significant relation could be found between malalignment rates and the material of the nail used. A recent randomized controlled trial was published by Goyal et al. [42] comparing TENs to stainless steel ESIN. Thirty-five children were included, 18 children in the TENs group and 17 children had stainless steel nails (age range 6–12 years). Fracture comminution grades III and IV according to the Winquist Hansen classification [35] were excluded from this comparison. There was no significant difference between both groups regarding degree of fracture site angulation at 6 months follow up (Level I evidence).

Mini-Open Versus Closed Reduction Technique

ESIN are inserted under fluoroscopic assistance with closed reduction of the fracture fragments or with the mini-open reduction technique (small incision blind hand technique). In a retrospective comparative study by Altay et al. [46] (Level III evidence), two groups of children who had TEN insertion

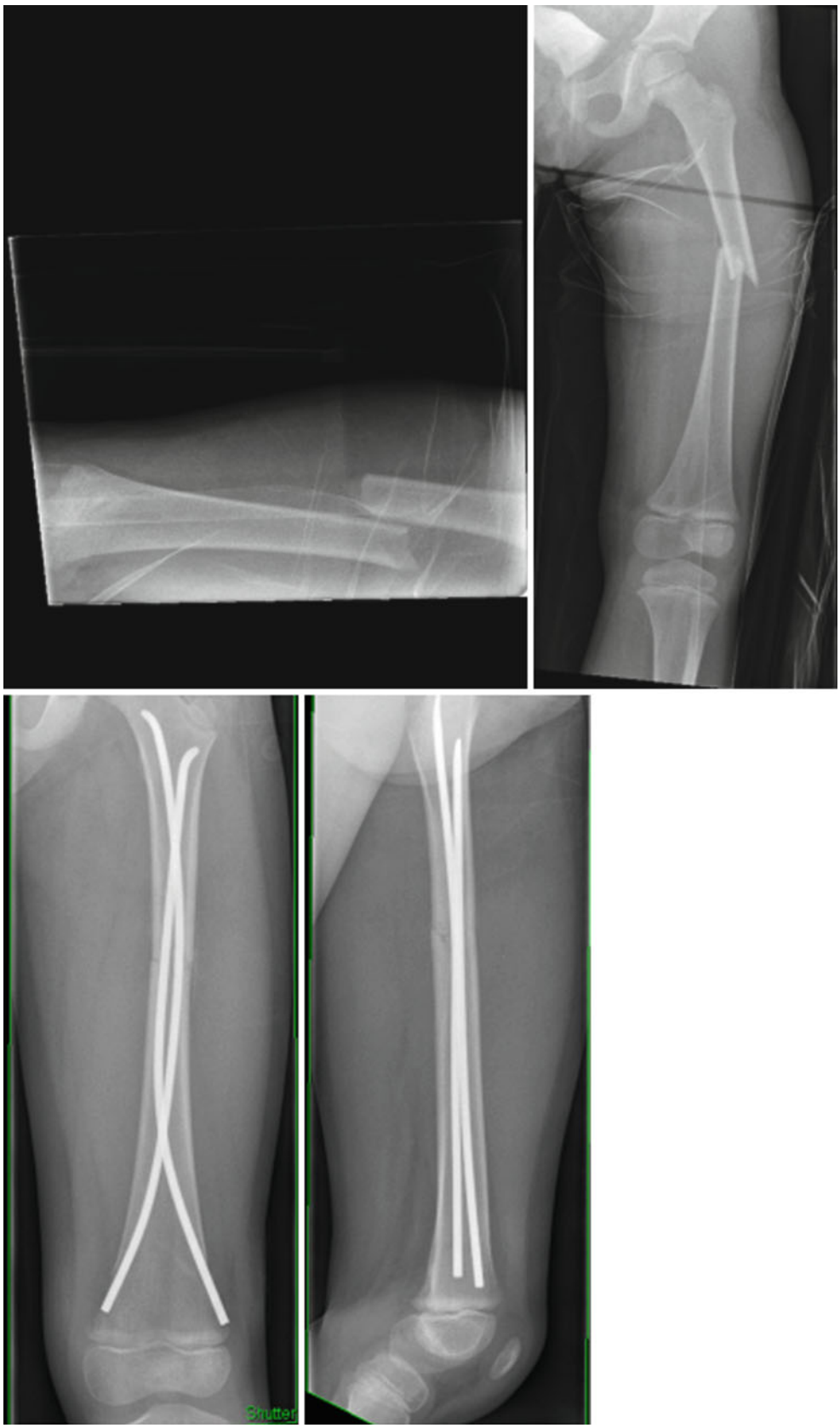

Fig. 10.5 Flexible nailing in femoral fracture

with mini-open reduction (n = 42, mean age 8.3 ± 2.7 years) or closed reduction (n = 45 children, mean age 8.8 ± 2.6 years) were compared. Both surgical and fluoroscopy times were significantly longer in the closed reduction group, while there was no significant difference between the two groups in terms of clinical and radiological results. Similar results were reported by Sun et al. [47] who retrospectively compared two groups of children (age range 4–15 years) who had TEN fixation of femoral shaft fractures (Level III evidence). The first group (n = 34, mean age 8.2 ± 2.5 years) had small incision blind hand reduction technique, while the second group (n = 34, mean age 8.6 ± 2.6 years) had closed reduction. Operative time and fluoroscopy time were significantly longer in the closed reduction group. No significant difference between the groups could be found in the complication rates or final outcome using TEN scoring system.

Complications and Nail Tip Irritation

Complication rates with the use of ESIN in the treatment of paediatric femoral shaft fractures might be seen in as many as 62 % of the patients [32]. They can be classified as minor, which resolve without additional surgery or don't result in long-term morbidity or major when unplanned surgical intervention is required or long-term morbidity ensues [28, 32, 34].

The majority of encountered complications are minor problems and are mostly related to nail tip irritation and pain around the nail insertion sites [28]. The resultant soft tissue irritation around the knee causes delayed recovery of the knee range of motion and might increase the risk of deep infections [27, 28, 32, 34, 37]. Ligier et al. [39] assumed that pin site irritation is mainly caused by early knee motion following surgery.

Narayanan et al. [27] studied specifically the factors that might increase the incidence of pain at the nail insertion sites (Level IV evidence). They found significant correlation with bent nail ends and nails that were prominent more than 10 mm outside the distal femoral cortex. Children who reported pain at nail insertion sites regained functional knee range of motion at a mean of 3 weeks later than patients who didn't report any symptoms. Their recommendation was not to bend the ends of the nails which should be trimmed short and advanced further with a hollow tamp, so that the unbent nail end lies in close apposition to the supracondylar flare of the distal femoral metaphysis. Ellis et al. [34] compared the results of using flexible stainless steel Ender's rods in either locked or unlocked configurations to the distal femoral metaphysis (Level III evidence). They found significantly more minor complications in the unlocked group, mostly due to prominent and painful implants. Two of their patients had deep infection which required drainage and both of them were associated with nail migration and prominence >13 mm.

The most commonly reported major complications were loss of reduction, unacceptable angulation exceeding the guidelines [7] and backing out of nails requiring another surgical intervention for trimming. Major complications were more common with the use of ESIN in length unstable femoral fractures [28, 29, 32, 34].

To Remove or Not to Remove ESIN Nails After Fracture Healing

Many authors including the originators of this technique recommended routine removal of ESINs. This can be performed between 3rd and 6th month postoperatively, when solid healing and circumferential callus are evident in plain radiographs [37, 39, 47]. However, whether it is necessary to routinely remove flexible nails implanted in children is still unclear. In a retrospective case series by Narayanan et al. [27], 25 children out of 78 with femoral shaft fracture treated with TEN retained their nails and they reported no symptoms attributable to the implants at the final follow up (mean 3.6 years, range 2–6 years) (Level IV evidence). In another case series by Levy et al. [26], acute complications of femoral flexible nail removal in 163 children were analyzed. The nails were removed due to pain at insertion site in only 54 % (88 children) of the cases. The remainders were removed at the recommendation of the treating surgeon or the request of the family, but not due to symptomatic implants. They couldn't remove one Ender's rod in 1 patient in this series as the rod was overgrown with bone and it lied completely within the intramedullary canal. In another 3 children, the insertion sites of 3 additional rods were also overgrown with bone, which resulted in a prolonged extraction time (mean 71 min).

Plating

Submuscular Versus Open Plating

The minimally invasive, submuscular plate osteosynthesis is a favored technique for plating in skeletally immature patients by many surgeons (Fig. 10.6). Abdelgawad et al. [48] reviewed retrospectively 60 femoral shaft fractures in 58 patients with an mean age of 9 years, which were treated with submuscular bridge plating (Level IV evidence). Forty (67 %) were length unstable or complex fracture pattern. Two major complications that required further unplanned surgery were encountered. The first was a deep infection following open femoral fracture, which was managed primarily with debridement and external fixation. The second was a broken 3.5 mm titanium plate, which was replaced by a stronger 4.5 mm stainless steel plate. Minor complications included symptomatic hardware (n = 3), superficial wound infection (n = 2), and one temporary peroneal nerve palsy. In

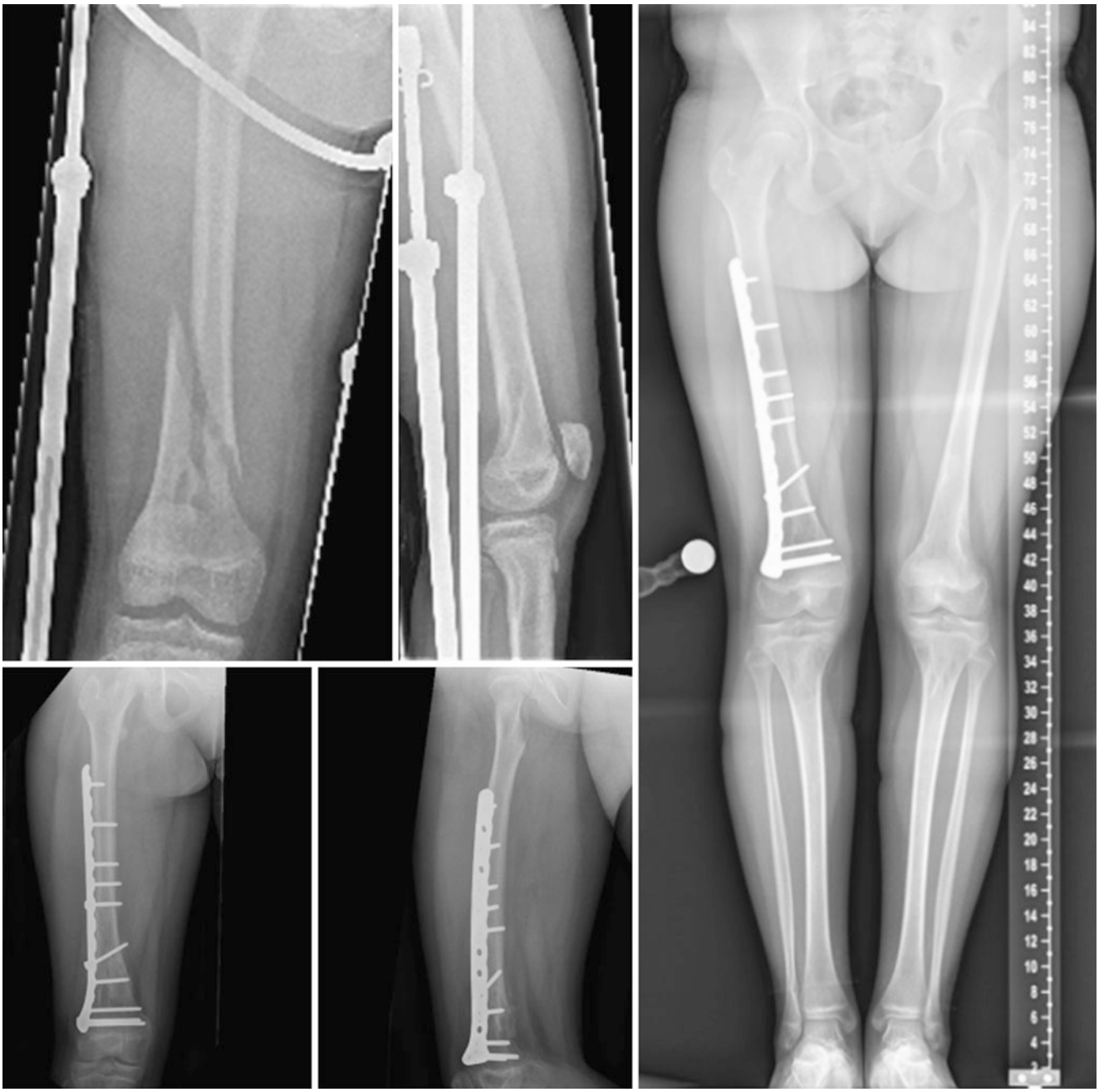

Fig. 10.6 Submuscular plating in a child with femoral fracture

one non-compliant patient there was mild loss of fixation 2 weeks postoperatively which was managed with a long leg cast for 5 weeks. Ten patients had a mean of 9.9 mm LLD (10 mm short to 20 mm long). Abbott et al. [49] compared retrospectively two groups of children who underwent either open (n = 58) or submuscular (n = 22) plating for femoral shaft fractures (Level III evidence). The mean age of both groups was 7.9 ± 3.5 and 8.5 ± 2.4 years respectively. There were no statistically significant differences between both groups regarding operative times, time from operating room to discharge, time to union (full weight bearing), the incidence of LLD of >2 cm or the presence of deep infection (1 case in the open group). On the other hand, estimated blood loss was significantly higher for open plating; however the increased blood loss was not clinically relevant as there was no difference in the need for blood transfusions between groups. The incidence of rotational asymmetry was significantly different with no cases of rotational asymmetry in the open group while 4 out of the 22 submuscular plating patients had clinically detectable rotational differences. Five patients

in the open group had unplanned return to the operating room (of no statistical significance), one patient for serial debridement of deep infection, one for periprosthetic femoral fracture 1 year postoperatively and 3 patients for revision of metal failure.

Locked Plating for Paediatric Femoral Shaft Fractures

Hedequist et al. [50] retrospectively reviewed 32 patients with locked plate fixation for femoral fractures (Level IV evidence). Their mean age was 11 years (range 6–15 years). The indications for locking plate fixation were either the presence of comminution (n = 13), pathologic fracture (n = 9), poor bone quality (n = 3) and fracture location (n = 7). Twenty-four patients had submuscular insertion of the plate and 8 patients had open plating. All fractures united uneventfully with anatomic alignment except in one patient who had a severely comminuted distal femoral fracture with development of 12° valgus angulation and 1.5 cm shortening. One patient who had locked plate fixation for osteopenia suffered from fracture at the distal end of the plate 11 months after the index surgery. Plates were removed in 7 patients on discretion of the treating surgeon without complications.

Complications of Plating

Complication rates with the use of plating in paediatric diaphyseal femoral fractures range from 0 % to 13 % [51, 52]. They can be grouped into either major or minor complications according to the need for unplanned surgical intervention [51]. The most frequently reported complications are deep infection, the development of distal femoral valgus deformity (DFVD), limb length discrepancy (LLD), bone overgrowth over the plate, stress shielding, healing problems and symptomatic screw prominence [51, 53, 54].

The development of DFVD following paediatric femoral plating is due to contouring the distal plate to accommodate for the metaphyseal flare of the femur. With growth from the distal femoral physis, the plates would migrate proximally with their distal bent causing mechanical remodeling, this being the driving force behind the DFVD and resultant LLD. Proximal plate migration would also account for the increasing medial screw prominence with growth [54].

Routine Removal of Paediatric Femoral Shaft Plating

Although there are no evidence-based studies supporting routine removal of paediatric long bone implants and femoral plates, from a clinical standpoint, their removal inherently makes sense given the plate length in the growing child [55]. The idea that larger plates should be removed routinely has been supported for many reasons. First, large implants and in particular dynamic compression plates are associated with stress shielding of the bone (about 40 % indentation and atrophy of the underlying cortex) and subsequent risk of fracture at the end of the plate (Fig. 10.7) [56, 57]. Second, longer plates spanning the length of the femur when left in place may make it more difficult for arthroplasty about the hip and knee if required in adulthood. Third, implants such as femoral plates spanning the length of the femur would require significant exposure for removal if delayed infection or repeat trauma occurred [55].

Kelly et al. [55] published a single surgeon series on 3 patients out of 33 children who had submuscular plating for femoral fractures and returned unexpectedly for follow up after 3–7 years (Level IV evidence). One patient returned due to LLD, one for DFVD and the third due to medial thigh pain from symptomatic screw prominence. Their recommendation was to remove the plate in a child with growth remaining if there is a distal contour in the plate. Pate et al. [56] reported a retrospective case series of 22 children who had femoral submuscular plate removal (Level IV evidence). The plates were removed mainly due to either implant prominence or family/treating surgeon preference. The mean time from plate insertion to plate removal was 9 months (range 5–27 months). Complications were encountered in 7 patients who had more extensive exposure to remove the plate compared to plate insertion with one patient having a broken screw head and the shaft of the screw was left inside the bone. The need for more exposure was mainly to remove bony overgrowth over either the proximal or the distal plate edges. Four patients out of the 7 had their plates removed relatively early and therefore the advice that plates should be removed in the first 6–9 months is not justified. No correlation was found between the need for more extensive exposure and fracture pattern, length of plate used, or age of patient at the time of fracture fixation. There were no postoperative complications such as infection, limitation of motion, refracture or fracture through a screw hole.

Trochanteric Entry Rigid Intramedullary Femoral Nailing

Reported advantages of using rigid intramedullary nails in older children and adolescents are mainly stable fixation, earlier mobilization and high union rate with low complication rate (Fig. 10.8) [58]. Crosby et al. [59] retrospectively reviewed a single institutions 20-year experience (1987–2009) with the use of trochanteric entry rigid intramedullary nailing for femoral shaft fractures in skeletally immature patients (Level IV evidence). A total 241 patients, 8–17 years old, with 246 fractures were included in this study. Growth

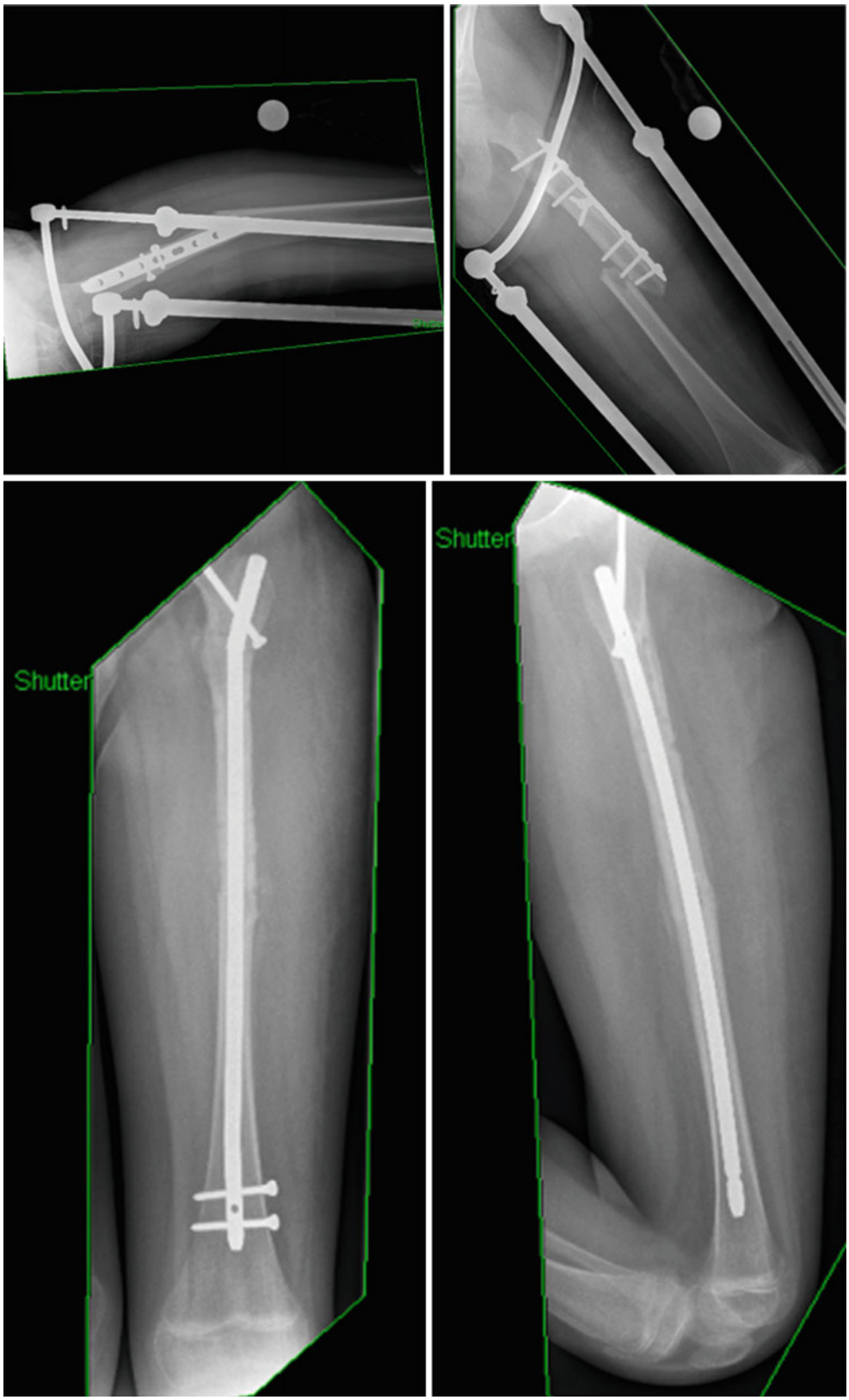

Fig. 10.7 Periprosthetic fracture of the femur

disturbance of the proximal femur was further analysed in a subcohort of patients who were followed up for at least 2 years (94 fractures). Twenty-four complications (9.8 %) were reported including: asymptomatic Brooker class-I heterotopic ossifications (n = 11) [60], delayed union requiring dynamization (n = 3), malunion with >10° of deformity (n = 3), interlocking screw migration requiring removal (n = 3), one deep infection requiring nail removal,

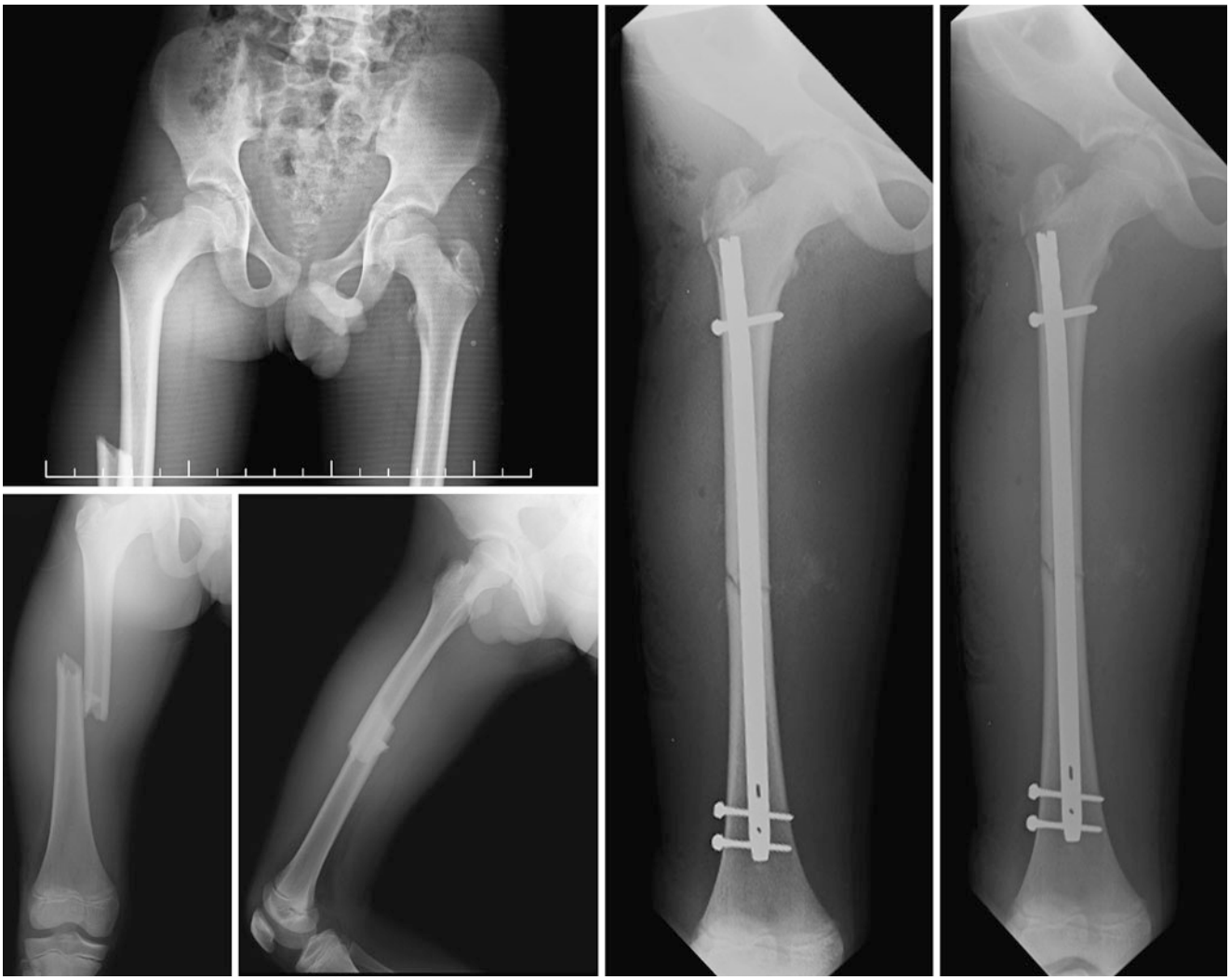

Fig. 10.8 Rigid trochanteric entry intramedullary femoral nail in an adolescent

sequestrectomy and prophylactic external fixation, and one malrotation requiring reoperation. Two patients (2.2 %) out of 94 who were followed up for at least 2 years, developed asymptomatic coxa valga and no further treatment was required. No cases with evidence of osteonecrosis were reported in this study.

Complications of Rigid Femoral Nails in Children

Rigid femoral intramedullary nails in the treatment of femoral diaphyseal fractures in skeletally immature patients must be used with caution. This is because of the risk of injury to the blood supply of the femoral head and the risk of proximal femoral growth disturbances as a result of damage to the trochanteric apophysis particularly in younger children [59, 61–64].

Growth Disturbance of the Proximal Femur and the Development of Coxa Valga

Growth disturbance of the proximal femur manifested by coxa valga and narrowing of the femoral neck might be caused by growth stimulation of the proximal femoral physis or growth retardation of the trochanteric apophysis. In the study by Crosby et al. [59], a >5 mm increase of articulotrochanteric distance was noted in 15.1 % (14 patients) at 2 year follow up. However, there were only 2 cases of proximal femoral growth disturbance manifested as asymptomatic coxa valga (defined as a 10° increase in the femoral neck-shaft angle or a femoral neck-shaft angle of >145° at the time of the latest follow-up compared to the other side) (Level IV evidence).

Gonzalez-Herranz et al. [62] have studied the long-term effect of using rigid femoral intramedullary nails in skeletally immature patients on the anatomy of the proximal

femur in 34 children with mean age of 10.8 years (3–14 years). The mean follow up was 6.2 years (5–13 years). The Kuntscher nail 7–14 mm in diameter was used in all patients except a 3-year-old boy who had a 3 mm rush pin fixation. These nails were inserted either for femoral shaft fractures or during limb preservation surgery for femoral sarcomas. They found an increase in the cervico-diaphyseal angle (CDA) of 10–25° in 30 % of patients, an increase in the articulo-trochanteric distance (ATD) of 10–29 mm in six patients (18 %) and reduction of the femoral neck diameter in eight patients (23 %). Changes in CDA and ATD were more significant in children under 13 years of age compared to older children. They found also that the changes were twice as common in patients with piriformis fossa insertion compared to trochanteric entry, which is consistent with damage of the medial portion of the greater trochanteric apophysis. Gage and Cary [61] found that trochanteric epiphyseodesis in children eight years of age or older has minimal effect on trochanteric growth and suggested that trochanteric growth is mostly appositional in children 8 years or older. Regardless of the age of the patient, trochanteric eiphysiodesis would stop no more than 50 % of the trochanteric growth and that overcorrection and coxa valaga would not be encountered as long as the tip of the greater trochanter is not violated. In agreement with these reports, Gordon et al. and Keeler et al. [64, 66] proposed that an insertion point through the lateral part of the greater trochanteric apophysis, rather than the tip of the greater trochanter, would not affect the growing proximal femoral anatomy. They postulated that the main cause of the developmental coxa valga is the violation of the medial greater trochanteric physeal plate with its remnant extending to the lateral aspect of the femoral neck ***(the femoral neck isthmic physis)***. This damage can arise from using piriformis fossa or the tip of the greater trochanter as starting points for nail insertion [63]. In their retrospective study, Keeler et al. [63] didn't find any significant change in the proximal femoral growth following the treatment of 80 femoral shaft fractures in 78 patients using rigid paediatric femoral nails inserted through a lateral trochanteric entry point (Level IV evidence).

Avascular Necrosis of the Femoral Head

There are concerns about the risk of avascular necrosis (AVN) of the femoral head with the use of rigid femoral intramedullary nails in children, in particular those inserted through the piriformis fossa [63]. MacNeil et al. [64] published a systematic review of studies reporting the use of rigid femoral intramedullary interlocking nails and the association of nail insertion sites and the incidence of AVN of the femoral head in skeletally immature patients (Level IV evidence). Nineteen relevant articles were analysed by the investigators and they were grouped into 3 groups according to the nail insertion point: (1) the piriformis fossa entry (PF), (2) insertion through the tip of the greater trochanter (TGT) or (3) through the lateral greater trochanter (LGT). Eleven articles reported the use of the PF insertion site with a total of 239 patients. The AVN rate within this group was 2 % (5 patients). The AVN rate in case of intramedullary nails inserted through the TGT was 1.4 % (2 out of 139 patients in 8 articles). Only one article was available describing the use of LGT [63] and they reported no cases of AVN in 80 operated fractures. The odds ratio of PF AVN rate versus TGT was 1.45 (0.28–7.59 95 % CI), which was not statistically significant. In conclusion, the LGT insertion point appears to be the safest as it avoids both the blood supply of the femoral head and the medial trochanteric apophysis and therefore avoids the risk of both AVN and proximal femoral growth disturbance.

Comparison Between the Use of Rigid Intramedullary Nailing and Submuscular Plating in Paediatric Femoral Shaft Fractures

Park et al. [52] published a prospective comparative study comparing the results of using rigid intramedullary nailing versus submuscular plating in adolescent femoral shaft fractures (Level II evidence). Twenty-two patients had intramedullary nails while 23 patients had plating. The mean age was 13.9 years (11–17.4). Operative time as well as fluoroscopy time was significantly shorter with intramedullary nails compared to plating. The time to full weight bearing was shorter with intramedullary nails while time to union was similar in both groups. None showed malunion over 10° or limb length discrepancy over 1 cm. Two patients in the intramedullary group had revision surgeries, one for deep infection and non-union and the other for rotational malalignment. Both patients healed uneventfully following revision surgeries. According to the authors, both rigid intramedullary nails and submuscular plating yielded good results with minimal complication rate in adolescent femoral fractures. However nails may be advantageous, as they need less fluoroscopy, easy to use in reduction and allow early weight bearing.

Recommendations

The treatment of choice for femoral fractures in infants from birth to 6 months is the Pavlik harness, which facilitates child care and hygiene with very high safety and good outcome. In children 6 months to 5 years, early or immediate hip spica cast is the standard of care for isolated femoral fractures with shortening <2 cm. In the case of excessive shortening or unacceptable angulation, other forms of treatment like

traction with delayed spica cast or operative treatment might be indicated. External fixation is an appealing option in children with femoral shaft fracture due its minimally invasive and biologic insertion. It is a viable option particularly in fractures with significant soft tissue injury, in multiply injured or head injured patients. The main disadvantages of external fixation are delayed healing and the risk of refracture. Internal fixation is indicated in diaphyseal femur fractures of children 5 years to skeletal maturity. Implant choice is mainly dependent on the age of the child and fracture stability. ESIN are the implant of choice in children 5–11 years with length stable fracture patterns. Children and adolescents 11 years to skeletal maturity with length unstable fractures are best treated with submuscular bridge plates or trochanteric entry rigid intramedullary nails according to the medullary canal diameter. Length unstable fractures are either long oblique fractures where the length of the fracture is twice the diameter of the femur at the fracture level, spiral fractures or comminuted fractures Winquist – Hansen grades III and IV. Both submuscular bridge plating and rigid intramedullary nails are viable options for treatment of adolescent diaphyseal femur fractures with low complication rates. However, nails may be advantageous as they need less fluoroscopy, technically easy to use in reduction and allow early weight bearing. The safest insertion point for the rigid femoral nails is through the lateral greater trochanter, not the tip of the greater trochanter, in order to avoid the medial trochanteric apophysis and therefore avoid the risk of both AVN of the femoral head and growth disturbances of the proximal femur and the development of coxa valga and narrowed femoral neck. Table 10.1 provides a summary of these recommendations.

Table 10.1 Summary of recommendations

Statement		Level of evidence	References
1.	Pavlik harness is indicated in the treatment of femoral shaft fractures in infants from birth to 6 months of age and is comparable to spica cast.	III/B	[9, 11]
2.	Immediate closed reduction and hip spica application is safe and effective treatment for isolated femoral fractures in children 6 months to 5 years.	II/B	[13, 14]
3.	There is no significant difference regarding the quality of reduction or the rate of complications when immediate hip spica cast was applied in the emergency room under conscious sedation compared to operating room under general anaesthesia.	III/B	[13, 16]
4.	Children treated with single leg spica cast were more likely to fit in regular car seats and fit comfortably in chairs and the parents took less time off work compared to children with double leg spica cast while final outcome was similar.	I/B	[18]
5.	External fixation yields better results in terms of malunion rate when compared to hip spica casting.	I/B	[22]
6.	Flexible intramedullary nails appear to be advantageous over external fixators in terms of healing rates, full weight bearing and return to school.	I/B	[23]
7.	There is statistically significant increased risk of unplanned surgery with the treatment of length unstable femoral fracture using ESIN.	III/B	[32, 36]
8.	ESIN should be reserved for children <11 years with length stable fractures while those with unstable fracture pattern or children >11 years should be treated by submuscular bridge plating or ALFN according to medullary canal diameter.	III/C	[29]
9.	Children weighing >49 Kg are more likely to have poor outcome following use of ESIN.	IV/I	[28]
10.	Weight of the patient is a significant predictor for malunion in the sagittal plane and anterior bow deformity >15°.	III/C	[33]
11.	There is significant relation between angular malalignment and severity of comminution Winquist grades III and IV when ESIN or stainless steel Ender's rods were used for fixation.	III/B	[32, 36]
12.	ESIN is a safe and effective treatment option for paediatric subtrochanteric femoral fractures stressing the importance of advancing the lateral nail into or just distal to the greater trochanter apophysis and advancing the medial nail into the femoral neck until it lies just short of the proximal femoral physis.	IV/C	[37, 38]
13.	Femoral fracture location appears not to significantly affect final outcome of ESIN fixation.	IV/C	[28, 37, 38]
14.	Locking of the ends of Ender's rods in the distal femoral metaphyses using 2.7 mm cortical screw 24 mm in length can significantly decrease fracture shortening by reducing distal implant migration and significantly decreasing complications like palpable implants, knee stiffness and deep infection.	III/C	[34]
15.	There is no significant relation between malalignment rates and the material of the ESINs, whether titanium or stainless steel.	I/B	[42]
16.	In case of ESIN, mini-open blind hand reduction technique significantly reduced the operative and fluoroscopy times compared to closed reduction without significant difference in complication rates or final outcome.	III/B	[46, 65]

Table 10.1 (continued)

Statement		Level of evidence	References
17.	There might be a correlation with bent nail ends and nails that were prominent more than 10 mm outside the distal femoral cortex and the incidence of nail tip irritation and pain which in turn affects the regain of functional knee range of motion.	IV/I	[27]
18.	The results of submuscular plating in complex paediatric femoral shaft fractures were not found to be affected by patients' age, weight or fracture site.	IV/C	[48]
19.	Submuscular bridge plating and open plating seem to be equally viable options for the management of pediatric diaphyseal femur fractures. However, open plating seems to have increased estimated blood loss while submuscular plating have more asymptomatic rotational asymmetry.	III/C	[49]
20.	The development of the distal femoral valgus deformity following paediatric femoral plating is explained by contouring the distal plate to accommodate the distal metaphyseal flare and with bony growth from the distal femoral physis, the plates would migrate proximally with their distal bent causing the valgus deformity.	IV/C	[53, 54]
21.	The need for more extensive exposure to remove femoral plates is caused by bony overgrowth at plate ends. No correlation was found between the need for more extensive exposure and fracture pattern, length of plate used, time from plate insertion or age of patient at the time of fracture fixation.	IV/I	[55]
22.	Growth disturbance of the proximal femur following insertion of rigid intramedullary nails is more common with piriformis fossa insertion compared to lateral trochanteric entry.	IV/C	[62, 66]
23.	The AVN rate with piriformis fossa insertion is about 2 % while with tip of the greater trochanter insertion is 1.4 %.	III/C	[64]
24.	Lateral greater trochanteric insertion point appears to be the safest as it avoids both the blood supply of the femoral head and the medial trochanteric apophysis and therefore avoids the risk of both AVN and proximal femoral growth disturbance.	IV/C	[63, 64]
25.	Both rigid intramedullary nails and submuscular plating yielded good results with minimal complications in the treatment of adolescent femoral fractures, however nails may be advantageous as they need less fluoroscopy, they are technically easier to use in reduction and allow early weight bearing.	II/C	[52]

References

1. Kocher MS et al. Treatment of pediatric diaphyseal femur fractures. J Am Acad Orthop Surg. 2009;17(11):718–25.
2. Wright JG. The treatment of femoral shaft fractures in children: a systematic overview and critical appraisal of the literature. Can J Surg. 2000;43(3):180–9.
3. Kong H, Sabharwal S. External fixation for closed pediatric femoral shaft fractures: where are we now? Clin Orthop Relat Res. 2014;472(12):3814–22.
4. Mooney JF. The use of 'damage control orthopedics' techniques in children with segmental open femur fractures. J Pediatr Orthop B. 2012;21(5):400–3.
5. AAOS. Treatment of paediatric diaphyseal femur fractures. Evidence based clinical practice guidelines. Am Acad Orthop Surg. 2015;23(12):e101.
6. Madhuri V et al. Interventions for treating femoral shaft fractures in children and adolescents. Cochrane Database Syst Rev. 2014;7:CD009076.
7. Flynn JM, Skaggs DL. Femoral shaft fractures. In: Flynn JM, Skaggs DL, Waters PM, editors. Rockwood and Wilkins' Fractures in Children. Philadelphia: Lippincott Williams and Wilkins; 2015. p. 987–1023.
8. Stannard JP, Christensen KP, Wilkins KE. Femur fractures in infants: a new therapeutic approach. J Pediatr Orthop. 1995;15(4):461–6.
9. Podeszwa DA et al. Comparison of Pavlik harness application and immediate spica casting for femur fractures in infants. J Pediatr Orthop. 2004;24(5):460–2.
10. Givon U et al. Treatment of femoral fractures in neonates. Isr Med Assoc J. 2007;9(1):28–9.
11. Wang CN et al. Femoral fractures in infants: a comparison of Bryant traction and modified Pavlik harness. Acta Orthop Belg. 2014;80(1):63–8.
12. Goebel J, Flynn JM. What is the best treatment for femoral fractures? In: Wright JG, editor. Evidence based orthopaedics: the best answers to clinical questinos, vol. 195–200. Philadelphia: Saunders, Elsevier; 2009.
13. Infante Jr AF et al. Immediate hip spica casting for femur fractures in pediatric patients. A review of 175 patients. Clin Orthop Relat Res. 2000;376:106–12.
14. Buehler KC et al. A prospective study of early spica casting outcomes in the treatment of femoral shaft fractures in children. J Pediatr Orthop. 1995;15(1):30–5.
15. Cassinelli EH et al. Spica cast application in the emergency room for select pediatric femur fractures. J Orthop Trauma. 2005;19(10):709–16.
16. Mansour 3rd AA et al. Immediate spica casting of pediatric femoral fractures in the operating room versus the emergency department: comparison of reduction, complications, and hospital charges. J Pediatr Orthop. 2010;30(8):813–7.
17. Illgen 2nd R et al. Femur fractures in children: treatment with early sitting spica casting. J Pediatr Orthop. 1998;18(4):481–7.
18. Leu D et al. Spica casting for pediatric femoral fractures: a prospective, randomized controlled study of single-leg versus double-leg spica casts. J Bone Joint Surg Am. 2012;94(14):1259–64.
19. Flynn JM et al. The treatment of low-energy femoral shaft fractures: a prospective study comparing the "walking spica" with the traditional spica cast. J Bone Joint Surg Am. 2011;93(23):2196–202.

20. Mubarak SJ et al. Volkmann contracture and compartment syndromes after femur fractures in children treated with 90/90 spica casts. J Pediatr Orthop. 2006;26(5):567–72.
21. Blasier RD, Aronson J, Tursky EA. External fixation of pediatric femur fractures. J Pediatr Orthop. 1997;17(3):342–6.
22. Wright JG et al. Treatments for paediatric femoral fractures: a randomised trial. Lancet. 2005;365(9465):1153–8.
23. Bar-On E, Sagiv S, Porat S. External fixation or flexible intramedullary nailing for femoral shaft fractures in children. A prospective, randomised study. J Bone Joint Surg Br. 1997;79(6):975–8.
24. Domb BG et al. Comparison of dynamic versus static external fixation for pediatric femur fractures. J Pediatr Orthop. 2002;22(4):428–30.
25. Heinrich SD et al. The operative stabilization of pediatric diaphyseal femur fractures with flexible intramedullary nails: a prospective analysis. J Pediatr Orthop. 1994;14(4):501–7.
26. Levy JA et al. Acute complications associated with removal of flexible intramedullary femoral rods placed for pediatric femoral shaft fractures. J Pediatr Orthop. 2013;33(1):43–7.
27. Narayanan UG et al. Complications of elastic stable intramedullary nail fixation of pediatric femoral fractures, and how to avoid them. J Pediatr Orthop. 2004;24(4):363–9.
28. Moroz LA et al. Titanium elastic nailing of fractures of the femur in children. Predictors of complications and poor outcome. J Bone Joint Surg Br. 2006;88(10):1361–6.
29. Sink EL et al. Decreased complications of pediatric femur fractures with a change in management. J Pediatr Orthop. 2010;30(7):633–7.
30. Luhmann SJ et al. Complications of titanium elastic nails for pediatric femoral shaft fractures. J Pediatr Orthop. 2003;23(4):443–7.
31. Wall EJ et al. Complications of titanium and stainless steel elastic nail fixation of pediatric femoral fractures. J Bone Joint Surg Am. 2008;90(6):1305–13.
32. Sink EL, Gralla J, Repine M. Complications of pediatric femur fractures treated with titanium elastic nails: a comparison of fracture types. J Pediatr Orthop. 2005;25(5):577–80.
33. Sagan ML et al. Residual deformity after treatment of pediatric femur fractures with flexible titanium nails. J Pediatr Orthop. 2010;30(7):638–43.
34. Ellis HB et al. A comparison of locked versus nonlocked Enders rods for length unstable pediatric femoral shaft fractures. J Pediatr Orthop. 2011;31(8):825–33.
35. Winquist RA, Hansen Jr ST. Comminuted fractures of the femoral shaft treated by intramedullary nailing. Orthop Clin North Am. 1980;11(3):633–48.
36. Lohiya R et al. Flexible intramedullary nailing in paediatric femoral fractures. A report of 73 cases. J Orthop Surg Res. 2011;6:64.
37. Flynn JM et al. Titanium elastic nails for pediatric femur fractures: a multicenter study of early results with analysis of complications. J Pediatr Orthop. 2001;21(1):4–8.
38. Pombo MW, Shilt JS. The definition and treatment of pediatric subtrochanteric femur fractures with titanium elastic nails. J Pediatr Orthop. 2006;26(3):364–70.
39. Ligier JN et al. Elastic stable intramedullary nailing of femoral shaft fractures in children. J Bone Joint Surg Br. 1988;70(1):74–7.
40. Flynn JM et al. Comparison of titanium elastic nails with traction and a spica cast to treat femoral fractures in children. J Bone Joint Surg Am. 2004;86-A(4):770–7.
41. Nascimento FP et al. Flexible intramedullary nails with traction versus plaster cast for treating femoral shaft fractures in children: comparative retrospective study. Sao Paulo Med J. 2013;131(1):5–12.
42. Goyal N et al. Randomized controlled trial comparing stabilization of fresh close femoral shaft fractures in children with titanium elastic nail system versus stainless steel elastic nail system. Acta Orthop Belg. 2014;80(1):69–75.
43. Cramer KE et al. Ender rod fixation of femoral shaft fractures in children. Clin Orthop Relat Res. 2000;376:119–23.
44. Nectoux E et al. Use of end caps in elastic stable intramedullary nailing of femoral and tibial unstable fractures in children: preliminary results in 11 fractures. J Child Orthop. 2008;2(4):309–14.
45. Hunter JB. The principles of elastic stable intramedullary nailing in children. Injury. 2005;36(Suppl 1):A20–4.
46. Altay MA et al. Mini-open versus closed reduction in titanium elastic nailing of paediatric femoral shaft fractures: a comparative study. Acta Orthop Belg. 2011;77(2):211–7.
47. Metaizeau JP. Stable elastic intramedullary nailing for fractures of the femur in children. J Bone Joint Surg Br. 2004;86(7):954–7.
48. Abdelgawad AA et al. Submuscular bridge plating for complex pediatric femur fractures is reliable. Clin Orthop Relat Res. 2013;471(9):2797–807.
49. Abbott MD, Loder RT, Anglen JO. Comparison of submuscular and open plating of pediatric femur fractures: a retrospective review. J Pediatr Orthop. 2013;33(5):519–23.
50. Hedequist D, Bishop J, Hresko T. Locking plate fixation for pediatric femur fractures. J Pediatr Orthop. 2008;28(1):6–9.
51. May C et al. Complications of plate fixation of femoral shaft fractures in children and adolescents. J Child Orthop. 2013;7(3):235–43.
52. Park KC et al. Intramedullary nailing versus submuscular plating in adolescent femoral fracture. Injury. 2012;43(6):870–5.
53. Heyworth BE et al. Distal femoral valgus deformity following plate fixation of pediatric femoral shaft fractures. J Bone Joint Surg Am. 2013;95(6):526–33.
54. Kelly B et al. Adverse sequelae due to plate retention following submuscular plating for pediatric femur fractures. J Orthop Trauma. 2013;27(12):726–9.
55. Pate O et al. Implant removal after submuscular plating for pediatric femur fractures. J Pediatr Orthop. 2009;29(7):709–12.
56. Lovell ME, Galasko CS, Wright NB. Removal of orthopedic implants in children: morbidity and postoperative radiologic changes. J Pediatr Orthop B. 1999;8(2):144–6.
57. Stromberg L, Dalen N. Atrophy of cortical bone caused by rigid internal fixation plates. An experimental study in the dog. Acta Orthop Scand. 1978;49(5):448–56.
58. Angadi DS et al. Rigid intramedullary nail fixation of femoral fractures in adolescents: what evidence is available? J Orthop Traumatol. 2014;15(3):147–53.
59. Crosby Jr SN et al. Twenty-year xperience with rigid intramedullary nailing of femoral shaft fractures in skeletally immature patients. J Bone Joint Surg Am. 2014;96(13):1080–9.
60. Brooker AF et al. Ectopic ossification following total hip replacement. Incidence and a method of classification. J Bone Joint Surg Am. 1973;55(8):1629–32.
61. Gage JR, Cary JM. The effects of trochanteric epiphyseodesis on growth of the proximal end of the femur following necrosis of the capital femoral epiphysis. J Bone Joint Surg Am. 1980;62(5):785–94.
62. Gonzalez-Herranz P et al. Intramedullary nailing of the femur in children. Effects on its proximal end. J Bone Joint Surg Br. 1995;77(2):262–6.
63. Keeler KA et al. Antegrade intramedullary nailing of pediatric femoral fractures using an interlocking pediatric femoral nail and a lateral trochanteric entry point. J Pediatr Orthop. 2009;29(4):345–51.
64. MacNeil JA, Francis A, El-Hawary R. A systematic review of rigid, locked, intramedullary nail insertion sites and avascular necrosis of the femoral head in the skeletally immature. J Pediatr Orthop. 2011;31(4):377–80.
65. Sun LJ et al. Pediatric femoral shaft fractures treated with titanium elastic nailing. Orthopedics. 2014;37(11):e1021–6.
66. Gordon JE et al. Intramedullary nailing of femoral fractures in children through the lateral aspect of the greater trochanter using a modified rigid humeral intramedullary nail: preliminary results of a new technique in 15 children. J Orthop Trauma. 2004;18(7):416–22 .discussion 423–4.

Part III

The Knee

Evidence-Based Treatment for Congenital Dislocation of the Knee

11

Abdelsalam Hegazy and Talal Ibrahim

Abstract

Various treatment modalities have been proposed for the management of congenital dislocation of the knee, while non-surgical treatment consists of traction, manipulation, serial casting and Pavlik harness application, surgical options include minimally invasive quadriceps tenotomy, open quadricepsplasty and femoral shortening. Most of the reports advocate non-surgical management initially for the reduction of the knee especially in early days after birth. However, there is no consensus regarding the best surgical treatment for the non-responsive or late presented cases. The paucity of the disease and the frequently associated problems beside the severity of knee dislocation are important factors affecting the treatment outcomes. This is an overview of the available treatment options reported to guide decision-making for the management of congenital dislocation of the knee.

Keywords

Congenital dislocation of the knee • Knee hyperextension • Genu recurvatum • Quadricepsplasty • Femoral shortening • Percutaneous tenotomy

Introduction

Congenital Dislocation of the Knee (CDK) is a rare spectrum of deformities ranging from simple knee hyperextension (genu recurvatum) to complete knee dislocation, first described by Chatelaine in 1822 [1]. The estimated incidence of CDK is approximately 1 per 100,000 live births [2]. CDK may be an isolated deformity or associated with other musculoskeletal diseases such as developmental dysplasia of the hip and clubfoot. Furthermore, CDK may be part of a syndrome such as Larsen's syndrome, Arthrogryposis Multiplex Congenita (AMC) or associated with paralytic conditions such as Meningomyelocele (MMC) (Fig. 11.1) [3].

The pathology of CDK is similar in most cases with short quadriceps tendon, tight anterior knee joint capsule and hypoplastic suprapatellar pouch with anteriorly subluxed hamstrings. However, the severity of these changes is exaggerated in complex cases and associated with secondary changes in bones and ligaments in long standing deformities [4].

The clinical presentation of knee recurvatum leads easily to early diagnosis and can be confirmed by radiography [5, 6]. The relation between the tibia and femur clinically and radiographically allows for a simple classification of CDK into recurvatum, subluxation and dislocation (Leveuf's Classification). The simple form of genu recurvatum is usually related to fetal molding due to oligohydramnios or breech position as been suggested by Haga et al. [7].

The other classification system was proposed by Finder in 1964 dividing CDK into five types according to the severity of the dislocation and its complexity. Type 1 being physiologic hyperextension up to 20° and type 5 being a complex variant including mixed categories of congenital diseases such as Ehlers-Danlos syndrome and Arthrogryposis [8] (Table 11.1, Fig. 11.2).

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How and When Should We Treat Congenital Dislocation of the Knee?

CDK is a rare entity and the existing studies in the literature are case series and case reports (Level IV and V). Most of the studies initiate management with non-surgical treatment followed by surgical intervention for failed non-surgical cases. Many modalities can be used for non-surgical treatment including the use of serial casting (Fig. 11.3), pavlik harness, skin and skeletal traction or observation. Surgical interventions include minimally invasive quadriceps tenotomy, open quadricepsplasty with joint capsule release and femoral shortening.

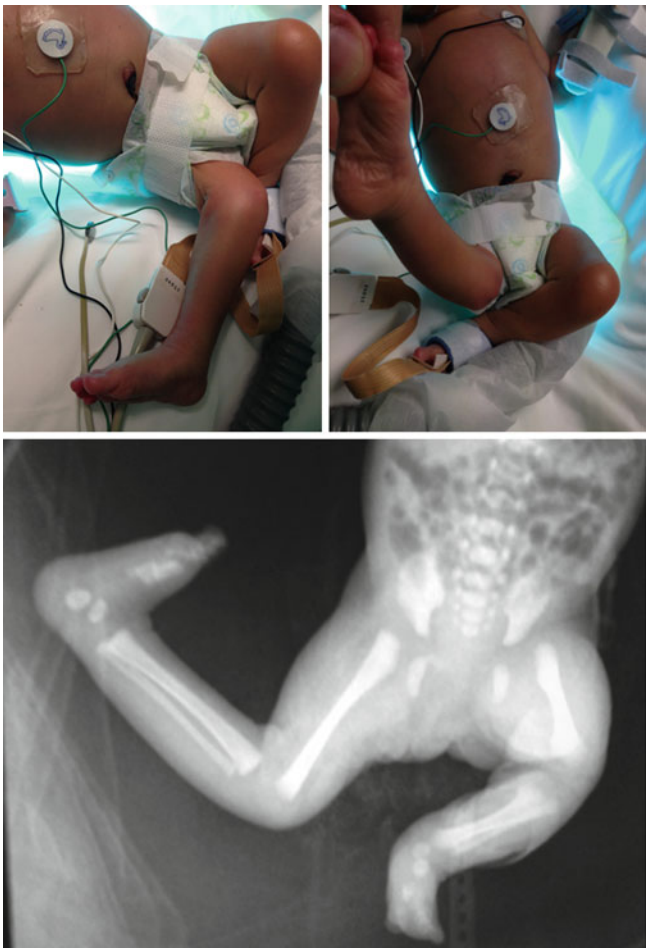

Fig. 11.1 Syndromic knees and hips dislocations

Early Versus Late Reduction

Different authors have reported variable success rates following non-surgical treatment of CDK. However, most authors agree that early treatment has an impact on the outcome in non-syndromic cases of CDK [9, 10]. Haga et al. suggested waiting 1 month for spontaneous reduction of CDK not associated with clubfoot, AMC or Larsen's syndrome [7].

Johnson et al. reviewed 17 patients with a follow-up of 11 years. Nine patients were treated non-surgically with good and fair results in 7 patients; all but 2 were less than 2 years of age when treatment was started with unilateral involvement. The role of arthrography was investigated in this study and found to be questionable as a technique to predict the response to close reduction because it was done usually before open reduction. Furthermore, a constant finding was ablation of the suprapatellar pouch in all surgical cases [11].

Meyer reviewed 68 patients and found that treatment was successful in 81 % of patients if performed before the age of 3 months. The rate of success dropped to 33 % if treatment was initiated between the ages of 3 and 6 months. [12].

Nogi and MacEwen successfully treated all but three of seventeen patients (excluding those with arthrogryposis or neuromuscular disorders) with manipulation and serial casting or Pavlik harness immediately after birth. The three failures occurred in two of delayed treatment cases and one with pseudo-reduction of the knee joint [13].

Ferris and Aichroth treated 19 CDK cases, nine of them treated non-surgically. Excellent and good results were achieved in five of the nine patients when the treatment started before 3 months of age and poor outcomes in syndromic patients with late treatment [6].

Isolated Versus Syndromic CDK

Ooishi et al. reviewed the results of 19 patients treated between 1972 and 1990. Twelve patients with isolated CDK and the remaining were syndromic patients (one with Larsen's syndrome and the other six with AMC). Non-surgical treatment started after birth in all patients. The isolated CDK cases reduced and all of the knees except one showed normal knee joint development. In contrast, the same treatment resulted in limited effect and residual subluxation in all the AMC and Larsen's syndrome patients. Three of the

Table 11.1 Finder classification

Type I	Physiologic hyperextension up to 20° considered within normal limits. Usually disappears by age 8 years (Fig. 11.2)
Type II	Simple hyperextension, a continuation of type I into adult life
Type III	Anterior subluxation with knee hyperextension up to 90° and resisted flexion beyond neutral
Type IV	Dislocation of the knee with the proximal tibia migrated upward and anteriorly
Type V	Complex variants including a mixed category of congenital diseases such as Ehlers-Danlos syndrome and arthrogryposis

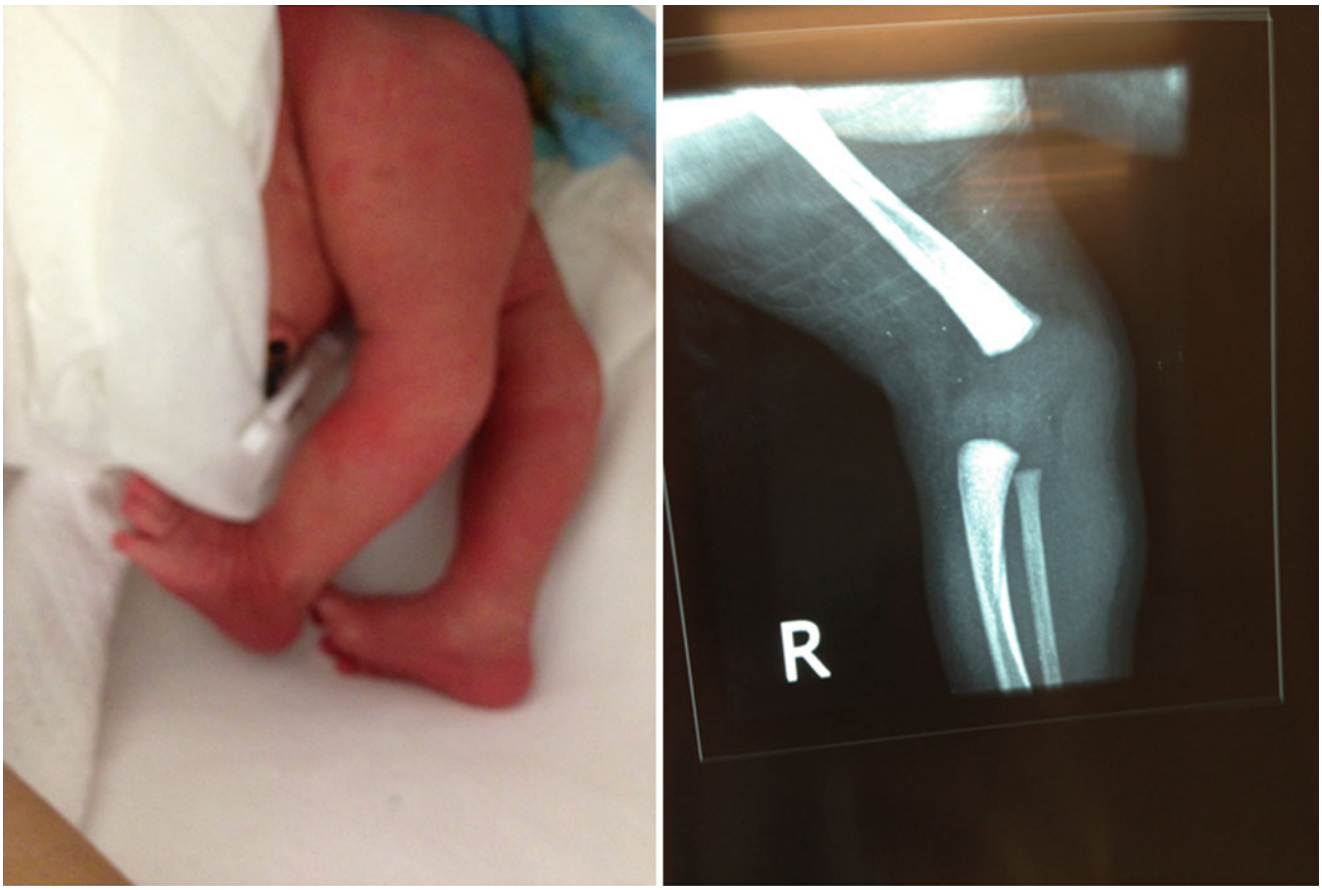

Fig. 11.2 Congenital knee dislocation, Finder type I

AMC patients underwent open reduction with quadricepsplasty [4]. Although treatment started early, non-surgical treatment failed in the syndromic and complicated cases in this study. Roy and Crawford described a percutaneous recession of the quadriceps mechanism through three stab incisions for Finder type 5 cases that are syndromic and associated with multiple anomalies. The advantage of such an approach is the avoidance of scar tissue and other complications potentially caused with more extensive surgery. The mean age of the patients at the time of surgery was 18 days, which further emphasizes the importance of early intervention for complex cases [14].

Percutaneous Tenotomy Versus Open Quadricepsplasty

Despite the general consensus on the necessity of surgical correction of CDK for unsuccessful cases to non-surgical treatment, there is no consensus regarding the type of surgical intervention. The fundamental pathological feature in CDK involves the quadriceps tendon and anterior joint capsule. Hence, most of the surgical procedures address these pathological changes to facilitate a joint reduction of the knee.

Curtis and Fisher described a long anterolateral approach with extensive mobilization of the quadriceps muscle, tendon lengthening via Z-plasty or an inverted V-incision, and an anterior capsulotomy of the knee [5]. Although successful in reducing the dislocated knee, this extensive approach unsurprisingly causes extensive scarring of the extensor mechanism; adhesions and wound complications. These complications often result in unsatisfactory outcomes, therefore this procedure was recommended for recurrence or severe cases with failed previous treatment [15].

Roy and Crawford described a percutaneous recession of the quadriceps mechanism through three stab incisions, targeting the fascia over the rectus femoris and the medial and lateral retinaculum [14]. All the patients were Finder type 5 and were treated at a mean age of 18 days. The advantage of this approach was the avoidance of scar tissue and other complications caused with more extensive surgery.

Shah et al. described a mini open quadriceps tenotomy in almost the same group of patients of Roy and Crawford and achieved 88 % of excellent and good results [16]. Patwardhan et al. showed that percutaneous needle quadriceps tenotomy in non-syndromic CDK patients is safe and less invasive than the other surgical modalities and requires less immobilization following reduction [17].

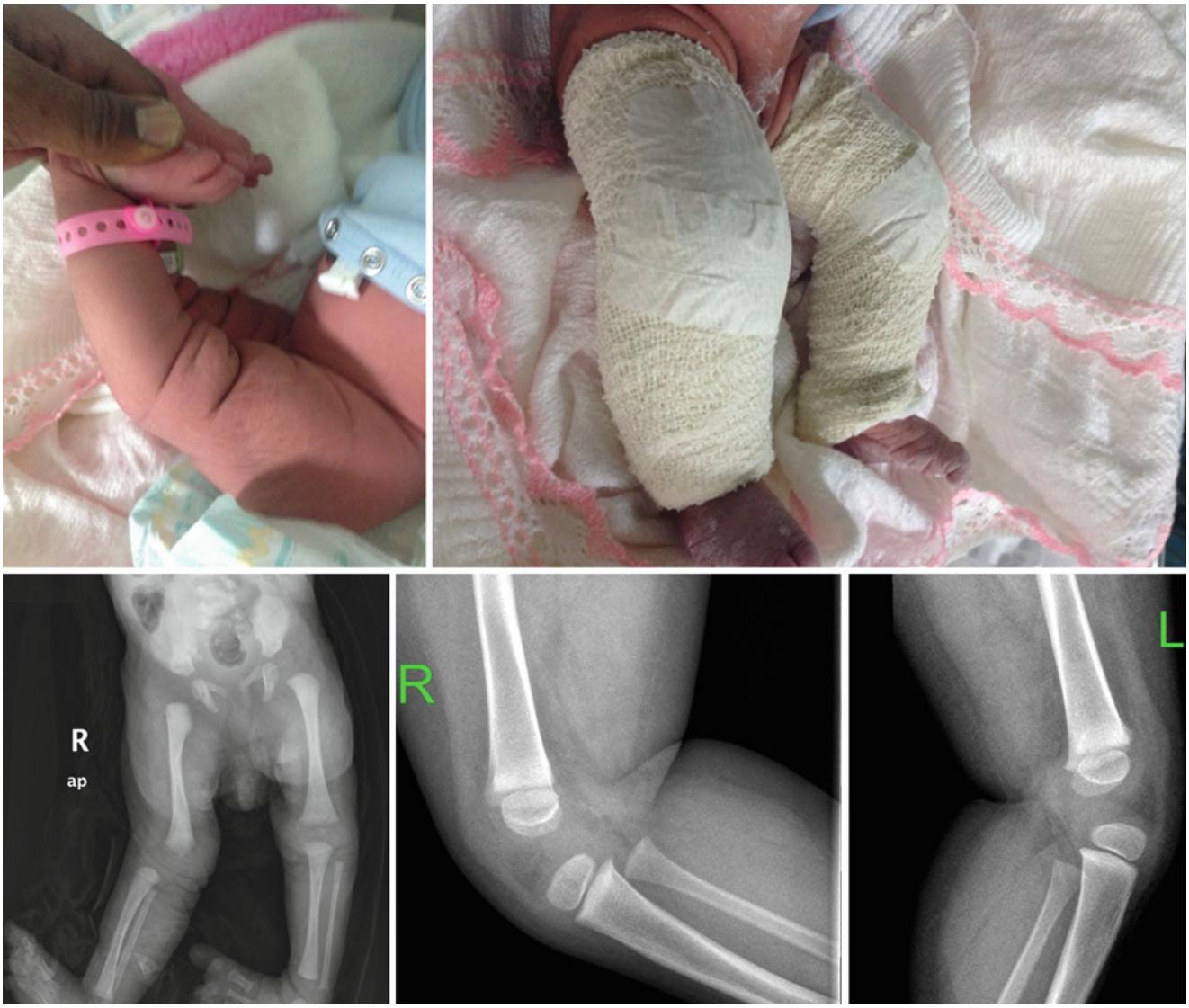

Fig. 11.3 Congenital knee dislocation treated with serial casting

The results of the above three studies confirm that percutaneous quadriceps tenotomy in the early ages of life even for complex and syndromic CDK cases is an effective method of treatment with less complications compared to open quadricepsplasty.

Femoral Shortening Versus Open Quadricepsplasty

Femoral shortening has been used successfully in the management of congenital anomalies such as the treatment of knee flexion deformities with popliteal webbing [18] and the reduction of dislocated hip joints especially after the age of 2 years [19]. A recent report by Johnston [20] described the use of a single diaphyseal femoral shortening to aid simultaneous reduction of ipsilateral hip and knee joints in teratologic joint dislocations. Johnston reported better knee range of motion, extensor mechanism function, and stability with the simultaneous reduction of the hip and knee achieved with femoral shortening compared with those who had staged reduction with conventional procedures including quadricepsplasty. However, the number of combined teratologic hip and knee dislocations was small to substantiate this procedure.

Oetgen et al. [21] compared the results of traditional quadricepsplasty with femoral shortening. The mean age of the patients at the time of surgery was 14 months. Five patients had an underlying diagnosis of Larsen's syndrome and the other two with non-syndromic CDK. All patients were treated non-surgically initially; three patients were then treated with femoral shortening and the other four with quadricepsplasty. The femoral shortening patients achieved more flexion and total range of motion than the quadricepsplasty patients, but this was statistically not significant. Furthermore,

no difference was found in the stability between the two patient groups.

Sud et al. [22] reported the mid-term follow-up results for femoral shortening in six CDK patients. Two of them were AMC patients while the others had combined anomalies. Five of the patients received non-surgical treatment for 4–6 weeks between 2 and 5 months of age. However, all the patients were treated with femoral shortening (mean: 2.35 cm) between 5 and 8 months of age without arthrotomy of the knee joint and followed for a minimum of 3 years. At final follow-up, all the patients achieved functional and pain free range of motion, satisfactory muscle power in knee flexors and extensors, no extensor lag or flexion contractures, no significant mediolateral instability, no leg length discrepancy and none of the patients required braces or walking aids for ambulation. The same author in a previous study reported an extensor lag in seven out of eleven AMC patients treated with quadricepsplasy [23] and concluded that femoral shortening for CDK conserves the quadriceps muscle and prevents extensor weakness by avoiding its release. Although the results are encouraging, long-term studies in a larger number of patients are required especially for syndromic CDK patients.

What Are the Factors that Affect the Treatment of CDK?

1. Better outcomes with early treatment of CDK even with surgical treatment.
2. Diagnosed such as Larsen's syndrome and AMC are associated with worse outcomes.
3. Femoral shortening is as effective as quadricepsplasty with no extension lag.
4. When multiple deformities are present in the same extremity, treatment should be directed at the knee first because of the positive effect of knee flexion on further foot and/or hip treatment.

Conclusion

Regardless of the severity and classification of CDK, treatment should be initiated as early as possible with gentle traction, manipulation and gradual correction with above knee plaster application. Maintaining knee flexion requires close follow-up in resistant cases with radiographs to check reduction. In cases of failure of knee reduction, minimally invasive quadriceps tenotomy with anterior capsule release should be considered in the first 4–8 weeks.

Femoral shortening and open quadricepsplasty should be used in older children with failed treatment with an inclination to femoral shortening because of the potential complications of open quadricepsplasty such as quadriceps weakness, adhesions and the wound problems.

References

1. Shattock SG. Geno recurvatum in a foetus at term. Trans Pathol Soc Lond. 1891;41:280–92.
2. Drennan JC. Congenital dislocation of the knee and patella. Instr Course Lect. 1993;42:517–24.
3. Jacobsen K, Vopalecky F. Congenital dislocation of the knee. Acta Orthop Scand. 1985;56:1–7.
4. Ooishi T, Sugioka Y, Matsumoto S, Fujii T. Congenital dislocation of the Knee. Its pathologic features and treatment. Clin Orthop Relat Res. 1993;287:187–92.
5. Curtis BH, Fisher RL. Congenital hyperextension with anterior subluxation of the knee. J Bone Joint Surg Am. 1969;51:255–69.
6. Ferris B, Aichroth P. The treatment of congenital knee dislocation: a review of nineteen knees. Clin Orthop Relat Res. 1987;216:135–40.
7. Haga N, Nakamura S, Sakaguchi R, Yanagisako Y, Taniguchi K, Iwaya T. Congenital dislocation of the knee reduced spontaneously or with minimal treatment. J Pediatr Orthop. 1997;17:59–62.
8. Finder JG. Congenital hyperextension of the knee. J Bone Joint Surg Br. 1964;46:783.
9. Laurence M. Genu recurvatum congenitum. J Bone Joint Surg Br. 1967;49:121–34.
10. Ko JY, Shih CH, Wenger DR. Congenital dislocation of the knee. J Pediatr Orthop. 1999;19:252–9.
11. Johnson E, Audell R, Opppenheim WL. Congenital dislocation of the knee. J Pediatr Orthop. 1987;7:194–200.
12. Meyer L. Congenital anterior subluxation of the knee: description of a new specimen, summary of the pathology of the deformity and discussion of its treatment. Am J Orthop Surg. 1913;10:411–37.
13. Nogi J, MacEwen GD. Congenital dislocation of the knee. J Pediatr Orthop. 1982;2:509–13.
14. Roy DR, Crawford AH. Percutaneous quadriceps recession: a technique for management of congenital hyperextension deformities of the knee in the neonate. J Pediatr Orthop. 1989;9:717–9.
15. Abdelaziz TH, Samir S. Congenital dislocation of the knee: a protocol for management based on degree of knee flexion. J Child Orthop. 2011;5:143–9.
16. Shah NR, Limpaphayom N, Dobbs MB. A minimally invasive treatment protocol for the congenital dislocation of the knee. J Pediatr Orthop. 2009;29:720–5.
17. Patwardhan S, Shah K, Shyam A, Sancheti P. Assesment of clinical outcome of percutaneous needle quadriceps tenotomy in the treatment of congenital knee dislocation. Int Orthop. 2015;39:1587–92.
18. Saleh M, Gibson MF, Sharrard WJ. Femoral shortening in correction of congenital knee flexion deformity with popliteal webbing. J Pediatr Orthop. 1989;9:609–11.
19. Herring JA. (2008) Developmental dysplasia of the hip. Tachdjian's pediatric orthopedics from the Texas Scottish Rite Hospital. 4th ed. Philadelphia: Saunders Elsevier. pp. 637–770.
20. Johnston CE. Simultaneous open reduction of ipsilateral congenital dislocation of the hip and knee assisted by femoral diaphyseal shortening. J Pediatr Orthop. 2011;31:732–40.
21. Oetgen ME, Walick KS, Tulchin K, Karol LA, Johnston CE. Functional results after surgical treatment for congenital knee dislocation. J Pediatr Orthop. 2010;30:216–23.
22. Sud A, Kumar N, Mehtani A. Femoral shortening in the congenital dislocation of the knee joint: results of the mid term follow up. J Pediatr Orthop B. 2013;22:440–4.
23. Sud A, Chaudhry A, Mehtani A, Tiwari A, Sharma D. Functional outcome following quadriceps tendon lengthening in congenital dislocation of the knee, with special reference to extensor weakness. Strateg Trauma Limb Reconstr. 2009;4:123–7.

Patello-Femoral Instability in Children

12

Andrew Wheelton, Farhan Ali, and Paul A. Banaszkiewicz

Abstract

Patello-femoral instability in the paediatric population is a common presentation with multiple treatment options and no definitive agreed management stratagem. Whilst research into this condition is abundant in the adult literature, the paediatric evidence base is less developed.

This chapter identifies important treatment questions and explores the existing available evidence. References to the adult literature are made in the absence of high level paediatric evidence and readers should be mindful of this.

Keywords

Patellar dislocations • Patellar instability • Knee cap instability

Background

Patello-femoral instability represents an intricate and multifarious condition affecting the knee extensor mechanism which is contributed to by bony morphology, soft tissue laxity and muscle balance. Patellar dislocation is defined by complete displacement of the patella out of the trochlea groove.

Episodic acute dislocation represents one end of a continuum ranging to congenital dislocation – an uncommon disorder with a dislocated patella presenting at birth due to failure of internal rotation of the myotome which forms the quadriceps muscle. Classifications representing this spectrum have been suggested by Garin [1] and Chotel [2]. The idea of a spectrum whilst helpful conceptually has lead to inconsistencies in the classification of this disorder in the literature with disagreement surrounding the term 'habitual dislocation'. Resultantly this chapter will divide patello-femoral instability into episodic acute dislocation and congenital dislocation.

A Finnish study [3] reported the annual incidence of acute patellar dislocation in children under 16 to be 0.04 % or 43 per 10,000. This represents one of the most common acute knee presentations in children with peak incidence between 13 and 15 years of age, with girls being more commonly affected than boys.

Acute dislocation can be a single event but commonly recurrence occurs and management decisions are often influenced by whether the event is a first time dislocation or not. The literature suggests that recurrence is more common in the paediatric population with Buchner [4] demonstrating recurrence of 52 % in children under 16 compared to an overall recurrence rate of 26 % for adults and Cash [5] finding a 60 % re-dislocation rate in children aged 11–14 compared to 33 % in those above 15.

As the understanding of the patho-anatomy of patella-femoral joint has increased so to have the surgical options with over 100 stabilisation procedures described in the adult and paediatric literature.

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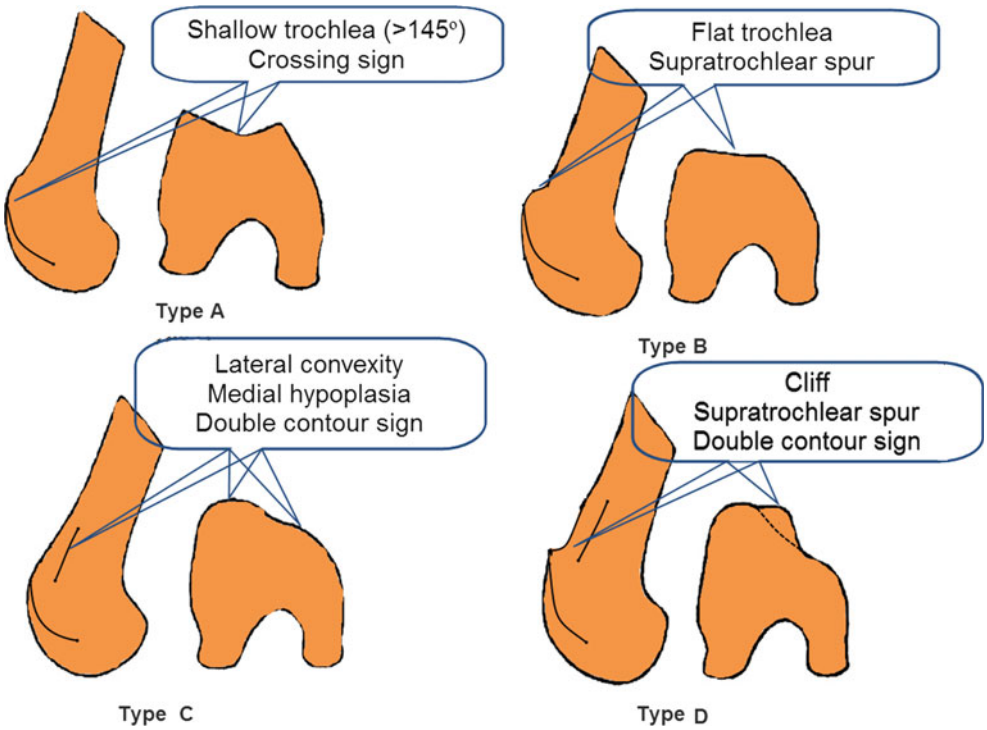

Fig. 12.1 Dejour classification of trochlear dysplasia. The Dejour classification: Grade A (54 %), Trochlear morphology is preserved but fairly shallow; Grade B (17 %), Flat or convex trochlea; Grade C (9 %), Asymmetry of trochlear facets: lateral facet is convex and medial facet is hypoplastic and Grade D (11 %), Asymmetry of trochlear facets, vertical joint (*cliff pattern*)

Are There Reliable Risk Factors to Predict Recurrence?

Multiple radiological indices have been developed to assess anatomic 'abnormalities' around the patella femoral joint. These include the Dejour [6] classification of trochlea dysplasia, the Caton-Deschamps (CD) [7] and Insall-Slavati (IS) [8] indices to measure patella alta, the tibial tuberosity – trochlea groove (TT-TG) distance [9], and rotational deformity (see Figs. 12.1, 12.2, 12.3, 12.4). Each of these have been suggested as possible predictors of recurrent patella femoral instability. Lewellan [10] retrospectively reviewed radiographs in 222 first time patella femoral dislocations in under 18 s assessing trochlea dysplasia (Dejour classification) and patella alta (CD,IS indices). Recurrent instability was significantly associated with trochlea dysplasia and the risk of recurrence was highest for those with open physes and trochlea dysplasia. Sex, age, body mass index, and patella alta were not statistically associated with recurrent instability. Jaquith [11] retrospectively reviewed 266 knees in under 18 patients presenting with a first time patella dislocation and assessed radiographs for increased patella height (CD), trochlea dysplasia (Dejour classification) and skeletal immaturity (physes open, closing, closed). Multivariate analysis identified that trochlear dysplasia, skeletal immaturity, CD index >1.45 and history of contralateral dislocation were significant risk factors for dislocation with a predicted risk recurrence of 88 % if all 4 were present, 75 % for 3 and 55 % for 2.

In the literature for adult studies Kohlitz et al. [12] retrospectively assessed MRI imaging of 186 acute lateral patella dislocations and 186 age and sex matched controls and identified that the incidence of trochlea dysplasia in the dislocation group was 66 %, of these 36 % additionally had patella alta and 9 % an abnormal TT-TG, with only 15 % of dislocators having no anatomical risk factors. They inferred a 37-fold increased risk of dislocation for individuals with trochlea dysplasia and abnormal TT-TG and a 41-fold higher risk if trochlea dysplasia and patella alta were present – whilst suggesting most dislocators have anatomical risk factors this study does not investigate risk of recurrence. Steenson [13] compared MRI imaging of 60 knees with and 120 knees without recurrent patellar instability and found the recurrent dislocation group had a higher incidence of trochlear dysplasia (68.3 % vs. 5.8 %), patella alta (60.0 % vs. 20.8 %), increased TT-TG distance (42.0 % vs. 3.2 %) and rotational deformity (26.7 % vs. 2.5 %). Furthermore, multiple anatomic risk factors were identified in 58.3 % of patients with recurrent dislocation compared to only 1.7 % of controls.

In summary whilst there has clearly been an association demonstrated between these radiological anatomical indices and recurrent patella dislocation the strength of evidence available in the literature is limited with only level 3 evidence available to support them as reliable predictors of recurrence. Further studies are required to convincingly demonstrate a causative relationship. Available data suggests they can be used help to guide treatment choice and tailor

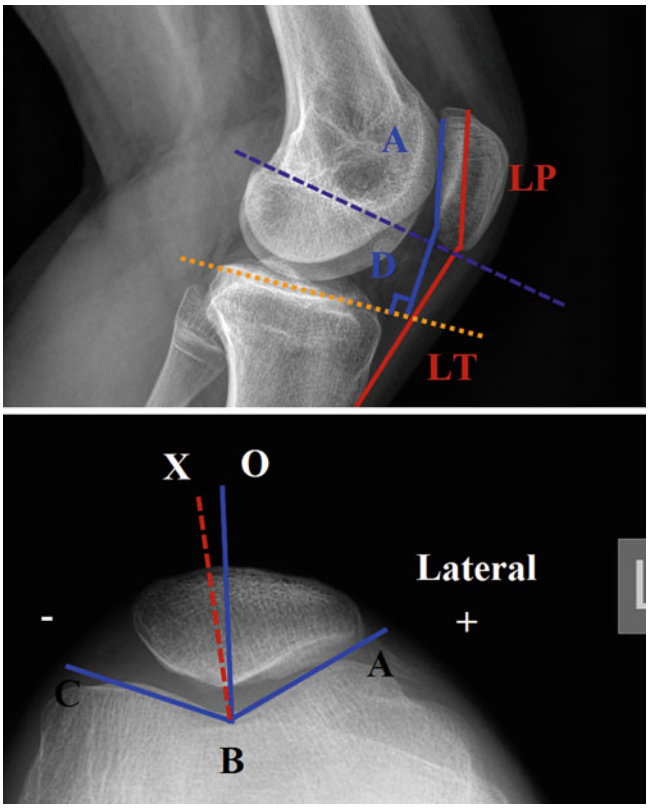

Fig. 12.2 Radiological indices around the knee. Top radiograph (*knee lateral*) shows the Blumensaat's line (*blue dashed*) touches the lower border of the patella. Insall-Salvati index: Ratio, or index, of patella tendon length (LT) to patella length (LP) should be 1.0. An index of 1.2 is alta and 0.8 is baja. Blackburne-Peel index: Ratio of the distance from the tibial plateau to the inferior articular surface of the patella (D) to the length of the articular surface of the patella (A) should be 0.8. An index of 1.0 is alta
Bottom **radiograph shows the sulcus angle ABC, line BO is the bisector and line BX passes through the lowest point of the patella. Angle OXB is the congruence angle of Merchant. A patella-femoral sulcus angle (ABC) > 144° is abnormal. Congruence angle of Merchant (OBX normally −6°–−8°). Abnormal if it is more than +16°. Positive (+) means Lateral) while (−) means medial.**

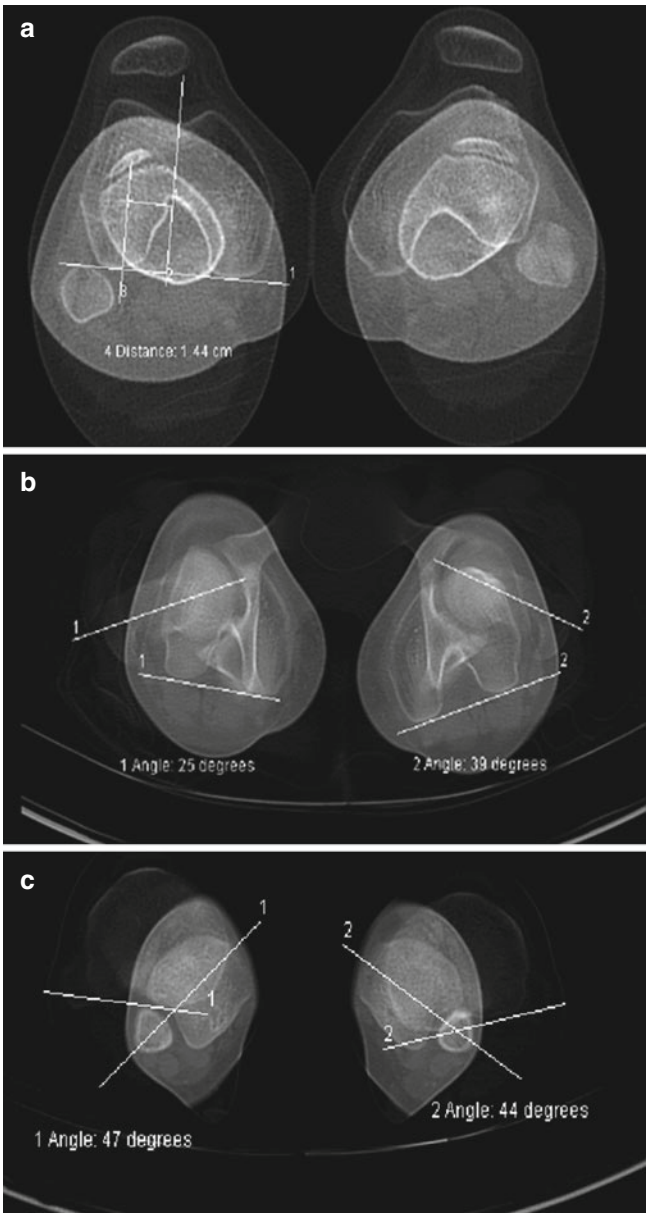

Fig. 12.3 CT scan of the lower limbs assessing the TGTT distance (**a**), femoral torsion (**b**) and tibial torsion (**c**)

surgical plan to address abnormal anatomy, and the presence of multiple abnormal anatomical indices does suggest an increased risk of recurrence.

What's the Best Treatment for Acute First Time Dislocation?

Currently there is no unanimously accepted stratagem for the treatment of first time patella dislocation, either in the paediatric or adult settings with both conservative and surgical management being popular, in no small part due to the lack of convincing evidence for favouring either approach with several papers giving conflicting evidence and outcomes. The Cochrane review [14] analysed five randomised studies and one quasi-randomised study with a total of 344 patients with first time patella dislocation. Four of the studies included paediatric patients. There was consistent evidence that the surgical intervention group had a significantly lower risk of recurrent dislocation following first time dislocation at 2–5 years follow-up with a relative risk of 0.53 favouring surgery. Given a demonstrative risk of recurrent dislocation in 222 people per 1000 in the non-surgical group, the data equates to 104 fewer people per 1000 having recurrent dislocation after surgical management. Furthermore there was (weaker) evidence of a lower risk of recurrent dislocation after surgery at 6–9 years with 110 fewer patients per 1000 having recurrent dislocation after surgery. However the

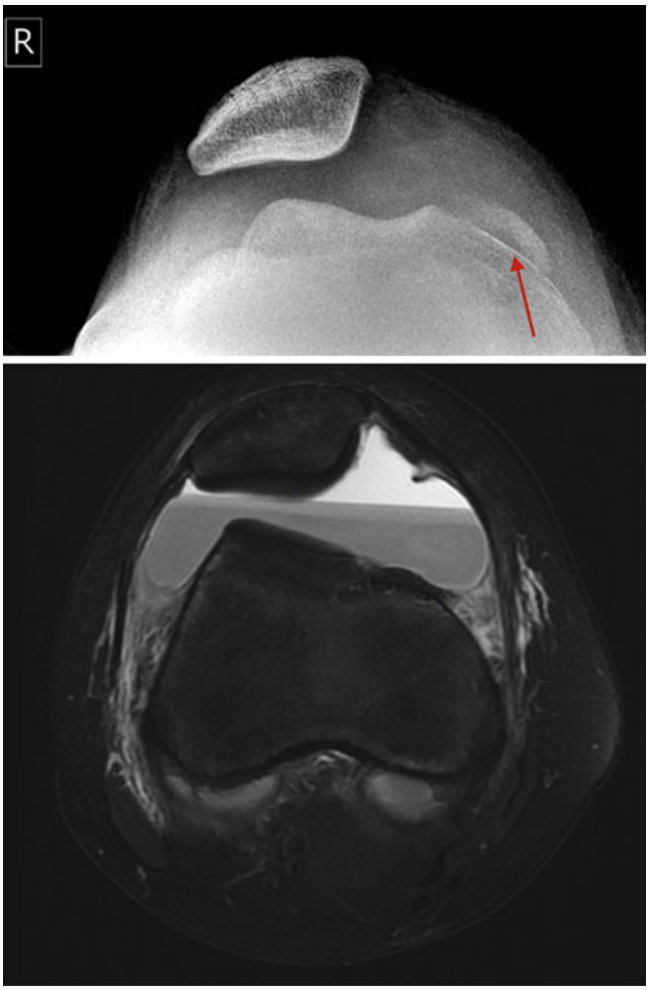

Fig. 12.4 Patellar dislocation with OCD fragment

authors of the review explained that although there was evidence in favour of surgical management in the short term the quality of the evidence was very low as the studies had a high risk of bias due to a lack of blinding and issues with selection and subsequently concluded that 'the evidence is not of sufficient quality to confirm a significant difference in outcome between surgical or non-surgical initial management' for first time acute patella dislocation. It is also difficult to generalise this review to the paediatric population as only one of the studies has a paediatric only subgroup [15] and this is specifically acknowledged in the review.

The Nikku [15] study (included in the Cochrane review) child only cohort has subsequently been published as a separate paper by Palmu [16]. This is the only randomised trial in the paediatric literature which compares conservative and surgical treatment in acute patella dislocation. In this study patients under 16 presenting with acute patella dislocation were randomised to conservative management (28 knees) or surgical intervention (36 knees). Patients were followed up for a mean of 14 years with a 94 % follow up rate. The rate of recurrent dislocation was high in both groups (71 % vs 67 %) despite this subjective outcome reporting was favourable with 75 % of conservatively treated patients and 66 % of surgically treated patients reporting a good or excellent outcome at the final follow up – suggesting disconnection between recurrence and patient reported outcomes. There was no significant difference identified between the two groups in subjective reporting, functional assessment or re – dislocation rate at either the 6 year or 14 year follow up points and the authors concluded that surgery does not improve long term outcome and therefore is not advocated for acute patellar dislocation in children and adolescents.

As the Palmu study is the only randomised trial in the paediatric literature there have understandably been no systematic reviews of conservative vs surgical treatment in acute patella dislocation undertaken, however in the adult literature four have been published since 2011 [17], [18] – updated to [19–21]. Mirroring the broader literature on the topic the results are at odds with each other, a recent met-analysis of these papers [22] attempted to assess which of these meta-analyses provides the current best available evidence. They used the Oxman-Guyattn and Quality of Reporting of Meta-analyses systems to assess the quality of the reviews and applied the Jadad algorithm to determine which offered the highest level of evidence. The four studies included 1984 patients with 997 undergoing surgery and the 987 being treated with conservative management. Three studies found that surgically treated patients had a lower re-dislocation rate whereas the other found no difference between the groups. None of the studies found a difference in functional outcome scores between the two treatment groups. The Jadad algorithm determined that the Hing and the Zheng papers offered the highest level of currently available evidence. From this they concluded that although operative treatment may offer a lower rate of re-dislocation than conservative treatment this does not correlate with functional outcome measures.

The majority of the literature assessing the comparison of operative versus non-operative management excludes patients with significant osteochondral lesions and the presence of these represent an indication for surgical management of first time dislocation. This is broadly accepted throughout the literature and is particularly supported in the paediatric population by a French multi-centre study reporting good outcomes for surgical fixation of osteochondral injuries in patella dislocation [23]. Similarly concomitant severe ligamentous injury represent an indication for surgical management and these situations represent a caveat to the approach for simple patella dislocation.

In conclusion the paediatric literature is sparse regarding evidence base for deciding conservative or operative management for acute first time patella dislocation, the only randomised study advocates a conservative approach. The data available in the adult literature whilst suggesting recurrent dislocation is

reduced by operative management does not indicate this improves functional outcome and as yet clinical equipoise remains as to the gold standard approach in simple first time acute patella dislocation. A review of anatomical risk factors should be considered to help guide management decisions.

What Is the Best Conservative Management Strategy?

The evidence base behind conservative management is deficient, particularly for the paediatric population. The broad treatment goals are relief of symptoms, preservation of range of motion, enhancement of quadriceps strength and return to previous activity. Classically a short period of immobilisation is instigated to relieve symptoms, followed by formal physiotherapy. There is no firm evidence in the literature to support immobilisation and studies have shown no difference in outcome when comparing immobilisation to none [24]. There have been no randomised trials into physiotherapy regimes [25] and no consensus has been reached on best rehabilitation schedule. Vastus medialis obliquus (VMO) has been suggested as an important target for strengthening after patella dislocation, however a systematic review has found little evidence to suggest that VMO activity can be preferentially enhanced in comparison to vastus lateralis by physiotherapy [26]. It is broadly accepted in the literature that the aim of the conservative treatment whatever form it takes is to avoid chronic muscle weakness and imbalance.

What Surgical Option Is the Best for Paediatric Patella Dislocation?

A key consideration in the skeletally immature patient is the proximity of the open physes of the distal femur and apophyses of the tibial tubercle to the site of intervention. Procedures in the armamentarium of the adult surgeon are either not recommended (eg tibial tubercle osteotomies) or modified (MPFL reconstruction) to avoid future growth disturbance.

A multitude of surgical procedures have been described for the treatment of paediatric patella dislocation. These can be divided into lateral release procedures, distal alignment procedures, MPFL reconstructions/medial reefing and trochleoplasty procedures, furthermore combinations of the above can be employed.

Lateral Release

This has historically been a first line intervention either in isolation or often combined with medial capsule reefing and other procedures [27–38]. It has been shown in the adult literature that this does not prevent recurrence [39] and improvement in outcome scores are not maintained after more than 4 years when performed in isolation for patella femoral instability [40]. There are no studies in the paediatric literature comparing lateral release with other surgical treatments, only level 4 evidence supporting its use in combination with other procedures.

Distal Realignment Procedures

As discussed previously an increased TT-TG distance has been suggested as a marker of recurrent instability [13] and this explains the rationale for adjusting the alignment of the extensor mechanism. Avoiding growth disturbance by leaving the tibial tubercle apopyhsis intact has formed the basis of the surgical techniques developed in this category. The 'Grammont (soft rod)' procedure comprises removing the deep patellar tendon fibres off the cartilaginous tibial tuberosity without severing the distal tendon attachment to periosteum and re-attaching it medially. A study by Garin [32] retrospectively assessed outcome with this procedure in 50 paediatric knees with a recurrent dislocation rate of 16 % and a 'good' satisfaction score in over 75 %, however the group is heterogeneous both in terms of degree of instability pre operatively and treatment given (procedure combined with others) and this is grade 4 evidence. Kraus [41] assessed the outcome in 65 paediatric knees – the outcome scores achieved were fair but the re-dislocation rate was high with 8 knees having a single dislocation within 3 months and 3 having recurrent late dislocations. The re-dislocations were associated with trochlea dysplasia suggesting this should also be addressed at the time of surgery.

The Roux-Gouldthwait procedure involves detachment of the lateral half of the patella tendon, guiding it under the intact medial half and reattaching it to the medial epiphysis. Nelitz [34] conducted a level 3 retrospective case controlled study comparing a cohort with unfavourable outcome and recurrent instability (37 children) following this procedure with a cohort with favourable outcome. The incidence of trochlea dysplasia was increased in the study group (89 % vs 21 %) and it was concluded that failure to address this was the likely cause of poor outcome and recurrent instability – supporting a tailored surgical approach addressing all elements of altered anatomy.

There are no studies comparing distal realignment with other procedures. In the adult literature Silanpaa [42] compared MPFL reconstruction (with adductor magnus tenodesis – 18 knees) with Roux-Gouldthwait distal realignment (29 knees) and identified a lower dislocation rate (7 % vs 14 %) and higher rate of osteoarthritis in the MPFL reconstruction group – suggesting a superiority for MPFL reconstruction.

MPFL Repair, Reconstruction and Medial Reefing

Biomechanical studies have shown that the MPFL is the major soft tissue restraint to lateral patellar dislocation [43, 44] and it is commonly injured when the patella dislocates [45], this can be identified with MRI [46]. Resultantly the MPFL has become the focus of surgical treatment for primary and recurrent patella dislocation in both paediatric and adult populations.

Repair of the acutely injured MPFL has been reported in the paediatric literature [16] and was found to be no better than conservative treatment. In the adult literature two prospective randomised studies have demonstrated improved patella stability with MPFL repair compared to conservative management [47, 48] however only one demonstrated a significant improvement in subjective outcome [48]. Delayed repair has not been shown to improve stability or outcome [49]. It has been suggested that repair of the MPFL may be insufficient on its own in the paediatric population particularly when anatomic abnormality is present [50].

MPFL reconstruction has become the main stay of surgical treatment for primary and recurrent patella-femoral dislocation in the adult population with numerous studies demonstrating achievement of good stability with few complications [51], however most of these are level 4. Bitar [52] performed a randomised controlled trial with 41 knees with acute patellar dislocation randomised into two groups – MPFL reconstruction and non-operative treatment. The surgical group had a lower recurrence rate (0 % vs 35 %) and a higher (better) Kujala outcome score at 2 years.

In the paediatric literature a myriad of techniques have been described – with the specific aim of avoiding physes. There have been several case series highlighting satisfactory outcomes with MPFL reconstruction [29, 53–57] but as yet there is no higher evidence than level 4 available.

Medial plication has a similar standing to lateral release procedures, having been historically popular and often used in combination with it [27, 29, 30, 32, 34, 37]. The adult literature suggests MPFL reconstruction is superior to medial plication resulting in better static patellar position and functional outcome [58].

Trochleoplasty

The use of trochleoplasty in skeletally immature patients is contentious, Beals [59] reports a case series of 6 knees in paediatric patients who successfully underwent trochleoplasty for patellofemoral instability. There were no redislocations and no skeletal growth complications, however it should be noted that these patients all had chromosomal abnormalities with resultant limited motor demands. A prospective study by Utting [60] followed up 59 knees treated with trochleoplasty for at least 1 year and showed a statistically significant improvement in outcome from pre-op scores – with 92.6 % of patients satisfied with the outcome. Smith [61] performed a systematic review of the literature on the procedure including the Beals paper and concluded that trochleoplasty is a safe and effective procedure for patella femoral instability in trochlea dysplasia patients but that the evidence had significant methodological limitations. A further systematic review by Bollier [62] suggested trochleoplasty 'should be reserved for sever dysplasia in which patellofemoral stability cannot otherwise be obtained' and advocated a tailored approach to address abnormal anatomy.

In summary it has not been shown which surgical option or combination is the optimum treatment. Emerging evidence suggests that if surgery is indicated then a tailored approach should be adopted to address significant anatomical abnormality.

What Is the Best Treatment for Congenital Dislocation?

Congenital dislocation is a rare entity and this is reflected in the literature with only 2 series addressing its management (Table 12.1). Gordon [36] reported a series of 17 knees with an average age at presentation of 7 years and 9 months. All patients had a fixed unreducible dislocation and all underwent surgery which entailed lateral release, advancement of VMO and in skeletally immature patients medial transfer of the patella tendon, skeletally mature patients had transfer of the tibial tubercle. At follow up all patients had 'a marked improvement' in pain and activity tolerance, there was one redislocation. Wada [63] retrospectively reviewed 7 knees in a younger cohort (average age 2.1 years), all knees were treated operatively in combination with lateral release, medial plication, V-Y lengthening of the quadriceps, medial transfer of the lateral patellar tendon and posterior release of the knee. Range of movement was improved in all knees and there was one redislocation. The authors advocate operating at a younger age to achieve the best outcomes.

A summary of recommendations is provided in Table 12.2.

Table 12.1 Of papers for outcomes of surgical techniques in paediatric literature

Author	No	Type of surgery	Age	Follow-up	Radiolocation	Outcome Score
Niedzielski [35]	11	Vastus medialis advancement, lateral release, partial patellar ligament transposition and Galeazzi semitendinosus tenodesis	NR	8.1	1	NR
Malecki [57]	39	MPFL reconstruction using Adductor magnus tendon				Kujala – statistically significant improvement Lysholm – statistically significant improvement
Nelitz [56]	21	MPFL reconstruction preserving distal femoral physis	12.2	2.8 years	0 2 had apprehension sign	Kujala improved by average of 20
ZHU [55]	29	Soft tissue transplantation	NR	NR	1 recurrent 1 medial	Lysholm Improved by 46
Kraus [41]	65	Grammont procedure (medialisation of patella tendon)	NR	8 years	8	Mean Lysholm 82
Nelitz [34]	37 study 23 control	stabilization (Roux-Gouldthwaite procedure, lateral release, medial reefing or in combination			Recurrent dislocation found in study group – more severe trochlea dysplasia	Consider tailored approach including trochleoplasty
MA [38]	61	32 – medial patella retinaculoplasty (GROUP 1) 29-medialcapsular reefing (GROUP 2)	NR	50 months	Group 1–9 Group 2–2	Kujala – improvement of 26 – group 1 29 – group 2
Ronga [33]	25	3-in-1 procedure (lateral release, vmo advancement, transfer of medial 1/3 patella tendon to mcl)	13.5	3.8 years	1 redislocation	Cincinnati score increased from 51.7 to 94.3 Kujala score increased from 52.4 to 93.8
Yercan [54]	4	MPFL reconstruction Using semitendinosus autograft with tenodesis to adductor magnus tendon (protecting physis)	NR	17.7 months	0 redislocation	NR
Luhman [64]	27	Patella realignment surgery	14.1	5.1 years	2 re-dislocations	Lysholm score 69.3 IKDC score 65.6
Wada (congenital dislocation) [63]	7	lateral release, medial plication, V-Y lengthening of the quadriceps, medial transfer of the lateral patellar tendon and posterior release of the knee	2.1 years	NR	0 redislocations	NR
Joo [37]	6	4 – in – 1	6.1	55.4 months	0	Kujala – mean 95.3
Garin [32]	50	'soft rod' patella re-alignment plus lateral release and medial reefing(insall procedure)	11 years	7.5 years	8 recurrent dislocation	IKDC score Group1(permanent dislocation) 76 % good Group 2 (episodic dislocation) 86 % good
Benoit [31]	8	distal advancement of the patella by complete mobilisation of the patellar tendon, lateral release and advancement of vastus medialis obliquus	10.3 years	13.5 years	1 re-dislocation	Lysholm score 98

(continued)

Table 12.1 (continued)

Author	No	Type of surgery	Age	Follow-up	Radiolocation	Outcome Score
Deie [53]	6	transfer of the tendon of semitendinosus to the patella using the posterior one-third of the femoral insertion of the medial collateral ligament as a pulley (MPFL reconstruction)		4 years	0 redislocations	Kaula score 96.3
Guo [30]	45	lateral retinacular releasing, medial retinacular tightening of the knee, vastus medialis muscle transfer to the patella, medial and distal transfer of the half patellar tendon	9.1 years	4.4 years	1 redislocation	NR
Gordon (congenital dislocation) [36]	17	Lateral release, advancement of vmo Transfer of patella tendon	7.9	5.1	1	NR
Letts [29]	26 children	semitendinosus transfer to the patella (MPFL reconstruction), medial retinaculum reefing and lateral release	14.4 years	3.2 years	2 redislocations (1 medial)	NR
Lipensky [28]	89	42 – lateral release 49 – combination of Roux-Goldthwaite and Campbell's procedures	NR	NR	0	NR
Beals [59]	6	trochloeplasty	NR	11 years	0	NR
LeFort [27]	85	14 – leavering of vastus medialis with a lateral capsular release and medial capsularrophy 71– medial transfer of patella ligament	9–20 years	6 years	6 redislocated	NR

NR Not Reported, *IKDC* International Knee Documentation Committee

Table 12.2 Summary of recommendations

Clinical situations	Grades
The presence of more than one abnormal radiological anatomical indices is a risk factor for recurrent patella-femoral instability	B
In the paediatric literature there is insufficient evidence to recommend surgical or conservative treatment for first time acute patella femoral dislocation	I
The adult literature suggests surgical treatment for first time acute patello-femoral dislocation reduces redislocation however this is not associated with improved subjective outcome measures	B
There is insufficient evidence to recommend what conservative therapy is best for patello-femoral instability	I
There is insufficient evidence to recommend which surgical option is best for patello-femoral instability. A tailored approach to correct anatomical abnormality should be considered	I
There is limited level 4 evidence available for treatment of congenital patella-femoral dislocation. This suggests an approach addressing all of the anatomical abnormalities present is necessary to achieve stability	C

References

1. Garin C. L'instabilité rotulienne chez l'enfant. in Cahiers d'enseignement de la SOFCOT n 52. Paris: L'expansion scientifique francaise; 1995.
2. Chotel F, Berard J, Raux S. Patellar instability in children and adolescents. Orthop Traumatol Surg Res. 2014;100:125–37.
3. Nietosvaara Y, Aalto K, Kallio PE. Acute patellar dislocation in children: incidence and associated osteochondral fractures. J Paediatr Orthop. 1994;14:513–5.
4. Buchner M, Baudendistel B, Sabo D. Acute traumatic primary patellar dislocation: long-term results comparing conservative and surgical treatment. Clin J Sport Med. 2005;15:62–6.
5. Cash JD, Hughston JC. Treatment of acute patellar dislocation. Am J Sports Med. 1988;16:244–9.
6. Dejour D, Le Coultre CB. Osteotomies in patella-femoral instabilities. Sports Med Arthrosc Rev. 2007;15:39–46.
7. Caton J, Deschamps G, Chambat P, Lerat JL, Dejour H. Patella infera. Apropos of 128 cases. Rev Chir Orthop Reparatrice Appar Mot. 1982;68(5):317–25.
8. Insall J, Salvati E. Patella position in the normal knee joint. Radiology. 1971;101(1):101–4.
9. Merchant AC, Mercer RL, Jacobsen RH, Cool CR. Roentgenographic analysis of patellofemoral congruence. J Bone Joint Surg Am. 1974;56(7):1391–6.
10. Lewellan LW, Mclntosh AL, Dahm DL. Predictors of recurrent instability after acute patellofemoral dislocation in pediatric and adolescent patients. Am J Sports Med. 2013;41(3):575–81.
11. Jaquith BP, Parikh SN. Predictors of recurrent patellar insatbility in children and adolsecents after first time – dislocation. Journal of Paediatric Orthoapedics, 2015.
12. Köhlitz T, Scheffler S, Jung T, Hoburg A, Vollnberg B, Wiener E, Diederichs G. Prevalence and patterns of anatomical risk factors in patients after patellar dislocation: a case control study using MRI. Eur Radiol. 2013;23(4):1067–74.
13. Steensen RN, Bentley JC, Trinh TQ, Backes JR, Wiltfong RE. The prevalence and combined prevalences of anatomic factors associated with recurrent patellar dislocation a magnetic resonance imaging study. Am J Sports Med. 2015;43(4):921–7.
14. Smith TO, Donell S, Song F, Hing CB. Surgical versus non-surgical interventions for treating patellar dislocation (review). Cochrane Database Syst Rev. 2015;2.
15. Nikku R, Nietosvaara Y, Kallio PE, Aalto K, Michelsson JE. Operative versus closed treatment of primary dislocation of the patella. Similar 2-year results in 125 randomized patients. Acta Orthop Scand. 1997;68(5):419–23.
16. Palmu S, Kallio PE, Donell ST, Helenius I, Nietosvaara Y. Acute patellar dislocation in children and adolescents: a randomised clinical trial. J Bone Joint Surg Am. 2008;90(3):463–70.
17. Smith TO, Song F, Donell ST, Hing CB. Operative versus non-operative management of patellar dislocation. Metaanal Knee Surg Sports Traumatol Arthrosc. 2011;19:988–98.
18. Hing CB, Smith TO, Donell S, Song F. Surgical versus non-surgical interventions for treating patellar dislocation. Cochrane Database Syst Rev. 2011;11.
19. Smith TO, Donell S, Song F, Hing CB. Surgical versus non-surgical interventions for treating patellar dislocation. Cochrane Database Syst Rev. 2015:CD008106.
20. Zheng X, Kang K, Li T, Lu B, Dong J, Gao S. Surgical versus non-surgical management for primary patellardislocations: an up-to-date meta-analysis. Eur J Orthop Surg Traumatol. 2014;24:1513–23.
21. Cheng B, Wu X, Ge H, Qing SY, Zhang Q. Operative versus conservative treatment for patellar dislocation: a meta-analysis of 7 randomized controlled trials. Diagn Pathol. 2014;18(9):60.
22. Erickson BJ, Mascarenhas R, Sayegh ET, Saltzman B, Verma NN, Bush-Joseph CA, Cole BJ, Bach Jr BR. Does operative treatment of first-time patellar dislocations lead to increased patellofemoral stability? A systematic review of overlapping meta-analyses. Arthroscopy. 2015;31(6):1207–15.
23. Chotel F, Knorr G, Simian E, Dubrana F, Versier G. Knee osteochondral fractures in skeletally immature patients: French multicenter study. Orthop Traumatol Surg Res. 2011;97(8):1877–0568.
24. Maenpaa H, Sillanpaa P, Paakkala A. A prospective,randomized trial following conservative treatment in acute primary patellar dislocation with special reference to patellar braces. Knee Surg Sports Traumatol Arthrosc. 2010;18(1):119.
25. Smith TO, Davies L, Chester R. Clinical outcomes of rehabilitation for patients following lateral patellar dislocation: a systematic review. Physiotherapy. 2010;96:269–81.
26. Smith TO, Bowyer D, Dixon J, Stephenson R, Chester R, Donell ST. Can vastus medialis oblique be preferentially activated? A systematic review of electromyographic studies. Physiother Theory Pract. 2009;25(2):69–98.
27. Lefort G, Cottalorda J, Lefebvre F, Bouche-Pillon MA, Daoud S. Femoro-patellar instability in children and adolescents. Rev Chir Orthop Reparatrice Appar Mot. 1991;77(7):491–5.
28. Lipenský R, Pavlata J, Svoboda P. Contribution to the solution of patello – femoral complaints in children and adolescents. Acta Chir Orthop Traumatol Cech. 1998;65(1):31–4.
29. Letts RM, Davidson D, Beaule P. Semitendinosus tenodesis for repair of recurrent dislocation of the patella in children. J Pediatr Orthop. 1999;19(6):742–7.

30. Guo Y, Wang C, Yi C. Surgical treatment of recurrent dislocation of the patella in children. Chin J Surg. 2000;38(12):49.
31. Benoit B, Laflamme GY, Laflamme GH, Rouleau D, Delisle J, Morin B. Long-term outcome of surgically-treated habitual patellar dislocation in children with coexistent patella alta. Minimum follow-up of 11 years. J Bone Joint Surg. 2007;89(9):1172–7.
32. Garin C, Chaker M, Dohin B, Kohler R. Permanent, habitual dislocation and recurrent dislocation of the patella in children: surgical management by patellar ligamentous transfer in 50 knees. Rev Chir Orthop Reparatrice Appar Mot. 2007;93(7):690–700.
33. Ronga M, Oliva F, Longo U, Testa V, Cherubino P, Maffulli N. The three-in-one procedure for recurrent dislocation of the patella in skeletally immature children and adolescents. J Orthop Traumatol. 2011;12(S138–S139):1590–9921.
34. Nelitz M, Theile M, Dornacher D, Wolfle J, Reichel H, Lippacher S. Analysis of failed surgery for patellar instability in children with open growth plates. Knee Surg Sports Traumatol Arthrosc. 2012;83(5):822–8.
35. Niedzielski KR, Malecki K, Flont P, Fabis J. The results of an extensive soft-tissue procedure in the treatment of obligatory patellar dislocation in children with ligamentous laxity: a post-operative isokinetic study. Bone Joint J. 2015;97B(1):129–33.
36. Gordon JE, Schoenecker PL. Surgical treatment of congenital dislocation of the patella. J Pediatr Orthop. 1999;19(2):26–264.
37. Joo SY, Park KB, Kim BR, Park HW, Kim HW. The 'four-in-one' procedure for habitual dislocation of the patella in children: early results in patients with severe generalised ligamentous laxity and aplasis of the trochlear groove. J Bone Joint Surg B. 2007;89(12):1645–9.
38. Ma L-f, Wang C-h, Chen B-c, Zhang F, Zhou J-w, Dong J-t, Wang F. Medial patellar retinaculum plasty versus medial capsule reefing for patellar dislocation in children and adolescents. Arch Orthop Trauma Surg. 2012;132(12):1773–80.
39. Lattermann C, Toth J, Bach Jr BR. The role of lateral retinacular release in the treatment of patellar instability. Sports Med Arthrosc Rev. 2007;15:57–60.
40. Panni AS, Tartarone M, Patricola A, Paxton EW, Fithian DC. Long-term results of lateral retinacular release. Athroscopy. 2005;21:526–31.
41. Kraus T, Lidder S, Svehlik M, Rippel K, Schneider F, Eberl R, Linhart W. Patella re-alignment in children with a modified Grammont technique. Acta Orthop. 2012;83(5):504–10.
42. Sillanpaa P, Mattila VM, Visuri T, Maenpaa H, Pihlajamaki H. Ligament reconstruction versus distal realignment for patellar dislocation. Clin Orthoap Relat Res. 2008;466(6):1475–84.
43. Conlan T, Garth Jr WP, JE L. Evaluation of the medial soft-tissue restraints of the extensor mechanism of the knee. J Bone Joint Surg Am. 1993;75:682–93.
44. Desio SM, Burks R, Bachus KN. Soft tissue restraints to lateral patellar translation in the human knee. Am J Sports Med. 1998;26:59–65.
45. Colvin AC, West RV. Patellar instability. J Bone Joint Surg Am. 2008;90:2751–62.
46. Elias DA, White LM, DC F. Acute lateral patellar dislocation at MR imaging: injury patterns of medial patellar soft-tissue restraints and osteochondral injuries of the inferomedial patella. Radiology. 2002;225:736–43.
47. Sillanpaa PJ, Mattila VM, Maenpaa H, Kiuru M, Visuri T, Pihlajamaki H. Treatment with and without initial stabilizing surgery for primary traumatic patellar dislocation. A prospective randomized study. J Bone Joint Surg Am. 2009;91:263–73.
48. Camanho GL, Viegas AC, Bitar AC, Demange MK, Hernandez AJ. Conservative versus surgical treatment for repair of the medial patellofemoral ligament in acute dislocations of the patella. Arthroscopy. 2009;25:620–5.
49. Christiansen SE, Jakobsen B, Lund B, Lind M. Isolated repair of the medial patellofemoral ligament in primary dislocation of the patella: a prospective randomized study. Arthroscopy. 2008;24:881–7.
50. Sillanpaa P, Mäenpää MH. First-time patellar dislocation: surgery or conservative treatment? Sports Medicine and Arthroscopy Review. 2012;20(3):128–35.
51. Smith TO, Walker J, Russell N. Outcomes of medial patellofemoral ligament reconstruction for patellar instability: a systematic review. Knee Surg Sports Traumatol Arthrosc. 2007;15:1301–14.
52. Bitar AC, Demange MK, D'Eloa CO, Camanho GL. Traumatic patellar dislocation: nonopertive treatment compared with MPFL reconstruction using patellar tendon. Am J Sports Med. 2012;40(1):114–22.
53. Deie M, Ochi M, Sumen Y, Yasumoto M, Kobayashi K, Kimura H. Reconstruction of the medial patellofemoral ligament for the treatment of habitual or recurrent dislocation of the patella in children. The Journal of bone and joint surgery. 2003;85(6):887–90.
54. Yercan HS, Erkan S, Okcu G, Ozalp RT. A novel technique for reconstruction of the medial patellofemoral ligament in skeletally immature patients. Arch Orthop Trauma Surg. 2011;131(8):1059–65.
55. Zhu J, Zhu Y-L, Wu H-S, Wu Y-L, Fu P-L. Recurrent patellar dislocation in children treated with different soft tissue transplantation: a comparative study. Chin J Tissue Eng Res. 2013;17(18):3413–20.
56. Nelitz M, Dreyhaupt J, Reichel H, Woelfle J, Lippacher S. Anatomic reconstruction of the medial patellofemoral ligament in children and adolescents with open growth plates: surgical technique and clinical outcome. Am J Sports Med. 2013;41(1):58–63.
57. Malecki K, Fabis J, Flont P, Niedzielski KR. The results of adductor magnus tenodesis in adolescents with recurrent patellar dislocation. BioMed Res Int. 2015;2015:456858.
58. Zhao J, Huangfu X, He Y. The role of medial retinaculum plicatiob versus medial patellofemoral ligament reconstruction in combined procedures for recurrent patellar instability in adults. Am J Sports Med. 2012;40(6):1355–64.
59. Beals R, Buehler K. Treatment of patellofemoral instability in childhood with creation of a femoral sulcus. J Paediatr Orthop. 1997;17(4):516–9.
60. Utting MR, Mulford JS, Eldridge JD. A prospective evaluation of trochleoplasty for the treatment of patellofemoral dislocation and instability. J Bone Joint Surg. 2008;90:180–5.
61. Smith TO, Leigh D. Outcomes following trochleoplasty for patellar instability with trochlea dysplasia: a sysematic review. Eur J Orthop Surg Traumatol. 2008;18:425–33.
62. Bollier M, Fulkerson JP. The role of trochlear dysplasia in patellofemoral instability. J Am Acad Orthop Surg. 2011;19(1):8–16.
63. Wada A, Fujii T, Takamura K, Yanagida H, Surijamorn P. Congenital dislocation of the patella. J Child Orthop. 2008;2(2):119–23.
64. Luhmann SJ, O'Donnell JC, Fuhrhop S. Outcomes after patellar realignment surgery for recurrent patellar instability dislocations: a minimum 3-year follow-up study of children and adolescents. J Pediatr Orthop. 2011;31(1):165–71.

Evidence-Based Treatment for Anterior Cruciate Ligament Tears in Children

13

Charles A. Popkin and M. Lucas Murnaghan

Abstract

Anterior cruciate ligament (ACL) tears in the skeletally immature athlete were once considered an unusual injury. However, with the sharp rise in early sports specialization and year round training, ACL tears in the developing athlete are becoming more commonplace. The treatment for this injury in the skeletally immature group is evolving. There is a increasing trend to reconstruct the ACL in the developing athlete to restore knee stability and minimize progressive chondral and meniscal injury. However, this remains a controversial topic and there remain strong advocates for nonoperative management and bracing until skeletal maturity to avoid injury to the growth plate. Surgical options include: physeal sparing (Iliotibial band and all-epiphyseal reconstructions), partial transphyseal/hybrid techniques and complete transphyseal ACL reconstructions. With proper rehabilitation after surgery, these children are returning to their sport. Despite these surgical techniques, there is good evidence that ACL injured knees are predisposed to develop early osteoarthritis. Therefore, moving forward there should be a focus on ACL prevention programs.

Keywords

ACL • Anterior cruciate ligament • Knee instability • Ligament reconstruction • All-epiphyseal • Hybrid repair

Introduction

Once considered an unusual injury, anterior cruciate ligament tears (ACL) are occurring with alarming frequency in the developing athlete [1, 2]. Recently, there has been a sharp rise in athletic activity in the pediatric and adolescent athlete. Current estimates are greater than 40 million children are playing some form of organized sports in the United States [3]. With this increase in activity and sports participation combined with a heightened awareness and recognition of ACL injury has led to a large number of ACL tears in skeletally immature athletes [3, 4]. The rate of ACL injury in this young age group is rising at a rate significantly higher than in adults [2]. Anterior cruciate ligament tears in athletes with open growth plates are a challenging problem for the patients, their parents, and their physicians alike [5].

Current literature favors early treatment for ACL injuries to return stability to the knee, but the optimal management of ACL tears in the developing athlete remains controversial [1, 6, 7]. Traditionally, these injuries were treated conservatively with activity modification and bracing. The rationale for delaying surgery originates from the concern over iatrogenic injury to the growth plate at the time of reconstruction. Furthermore, a prospective cohort study from Norway followed a non-operative treatment algorithm and found that 90 % of the children were able to participate in sports at the 2 year follow up, with a small number of surgical operations for new meniscal injuries (13 %) [8]. While nearly 40 % of the children followed in this study had to decrease activity level, this study suggests that

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ACL deficient children can be physically active and may be an adequate treatment option in some patients. However, other studies have demonstrated that activity modification and bracing are associated with poor outcomes [1, 6, 7, 9]. These ACL deficient knees place the athletes at a high risk of injury to the menisci; which are often times irreparable [9].

Operative strategies to manage ACL instability in the skeletally immature continue to evolve with time. While good results have been reported with transphyseal reconstructions, [5, 10] a very real concern for iatrogenic injury to the growth plate remains [11]. The desire to avoid growth disturbance and angular deformity, has led to the creation of new techniques to avoid the growth plate with tunnel placement [12, 13]. Early results from these techniques are proving to be superior to non-operative management. Developing athletes are returning to pre-operative activity level with good outcome scores [1]. This is relevant because many young athletes are reluctant to spend a couple seasons on the sidelines waiting for their growth plates to close [4].

Natural History

Activity modification and bracing is an appealing option for some patients and surgeons because of increased healing in children and risk of growth plate injury with surgery [8]. However, most of the results with this approach have not been favorable. Chronic ACL insufficiency leads to intra-articular damage and a high rate of irreparable meniscus tears in these developing athletes [6, 14]. One study of adolescent athletes demonstrated 50 % of ACL injuries had an associated chondral injury [15]. Furthermore, studies have shown that delaying treatment 3 months to reconstruct the ACL in skeletally immature patients can increase fourfold the chance of an irreparable medial meniscal tear and place the patient at even higher risk for a chondral injury [7]. Another study from Japan followed 18 patients aged 12 with acute ACL tears treated non-operatively and at final follow up only 1 of the 18 patients returned to sport and an alarming 11 out of 18 patients had Fairbanks changes on radiographs[16].

Work Up/Examination/Imaging

The initial presentation of an ACL tear in the skeletally immature patient is similar to the adult population. There is a high incidence in female athletes and the mechanism is usually non-contact in nature. It is common for the growing athlete to report hearing an audible 'pop' and to report a brisk onset of swelling in the knee. When a hemarthrosis is present, up to 65 % of knee injuries will include an ACL rupture [17]. Hallmark clinical tests used to detect an ACL rupture include a Lachman exam, anterior drawer testing with the knee at 90 degrees of flexion and a pivot shift test. The physician should perform a thorough examination of the knee, as meniscus tears, chondral fragments, and associated ligamentous injuries may also be present.

Imaging for a suspected ACL injury should always include plain radiographs AP, Lateral, Tunnel and Merchant views. This will allow the physician to look for an associated bony injury (tibial spine avulsion or Segond fracture) and assess the status of the distal femoral and proximal tibial physes. We recommend including a 3 ft standing film to look for any malalignment and any pre-operative leg length discrepancy [1]. MRI will allow for confirmation of the ligamentous injury as well as importantly assessing potential concomitant cartilage, ligament and/or meniscus injury (Fig. 13.1).

An essential component of treating an ACL injury in this population is developing a comfort in assessing the maturity of the young athlete [18]. This can help the physician determine how respectful to be of the growth plates around the knee when planning surgical reconstruction and discussing options. Recognizing the common discordance between chronological age and skeletal age is the first step in surgical planning [18]. Skeletal growth is usually complete by 14 in girls and 16 in boys, though significant variation does exist. The physiologic age of the patient can be assessed by a number of techniques. Commonly used methods in an orthopedic clinic are [4]:

(1) Bone age by left hand radiograph (Greulich and Pyle Atlas)
(2) Knee radiographs
(3) Menarchal status in female patients

Tanner staging is often cited as a predictive method, though this presents practical challenges in the typical environment of an orthopedic practice. It should be noted that patient or parent reported Tanner staging is an inadequate substitute for the originally described physician assessment [19].

At our institution, we recommend an abridged short hand version of the Greulich and Pyle to help with bone age without use of an atlas (See Tables 13.1, 13.2 and 13.3).

Treatment Options

What is required in these athletes is an approach to the ACL reconstruction that addresses the concern of iatrogenic injury and respects the physics, but without fearing it.

Non-operative

This approach does not lead to good results in the setting of a complete ACL rupture, unless the child is compliant with significant activity modification and abstinence from cutting

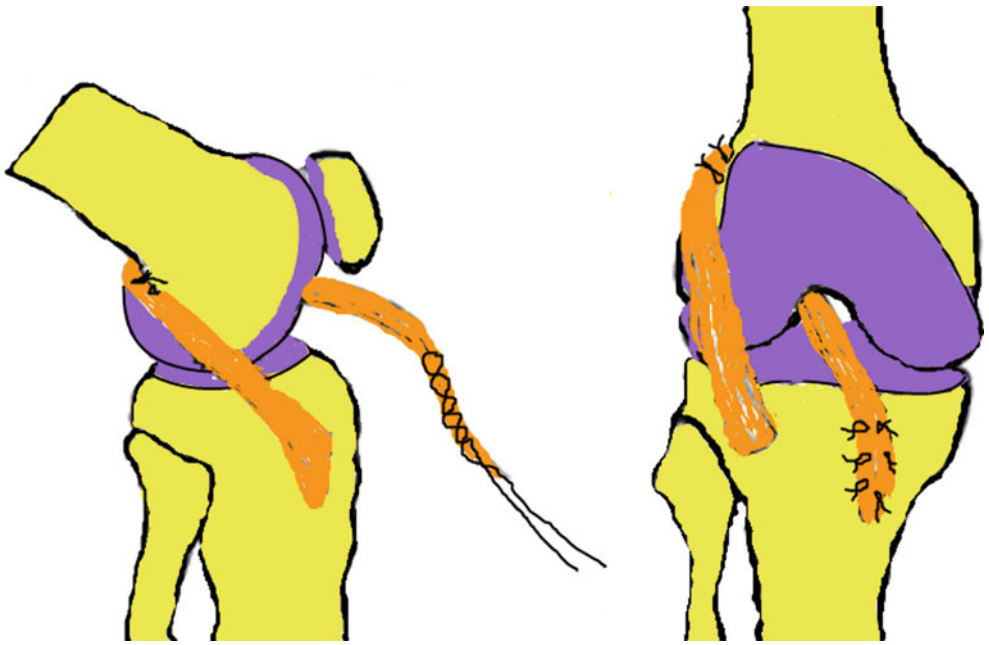

Fig. 13.1 A drawing of a physeal sparing, combined intra-articular and extra-articular ACL reconstruction

Table 13.1 The hospital for sick children cheat sheet for Greulich and Pyle

Girls	Boys	Hand radiographic finding
11.5	13	Can see thumb sesamoid
12	14	Distal phalanx growth plate fused
13	15	Middle phalanx growth plate fused
14	16	Proximal phalanx growth plate fused

and pivoting sports and activities. There is a very high rate of sport drop out [14] as well as rates of chondral and meniscal injuries. Developing athletes with partial ACL tears involving less than 50 % of the entire diameter with a clinically stable knee on examination may successfully be treated non-surgically [3, 20]. The athlete in this clinical scenario can return to sport in 6 months after the injury after a period of activity modification, therapy and bracing [3].

Operative

Iliotibial Band Reconstruction

A combined intra-articular and extra-articular reconstruction developed by MacIntosh and Darby in Toronto [21] and popularized in skeletally immature patients by Kocher and Micheli at Boston Children's [13]. The technique provides knee stability and avoids complications related to growth disturbance because there are no drill holes or tunnels (Fig. 13.2). This technique is primarily recommended for use in the prepubescent patient. To be successful, the harvest requires a length of 20 cm from the central one third of the iliotibial band. The graft is left attached to Gerdy's tubercle distally, brought into the knee in the over-the-top position and then brought under the intrameniscal ligament. The femoral side is secured by sewing the graft to the intermuscular septum and periosteum on the femur and the tibial side is secured by sewing down the graft in a trough of periosteum [1, 21].

The Level IV results of 44 children that underwent this surgery (mean age 10.3 with 5.3 years of follow up) demonstrated no growth disturbance [13]. Forty-one patients had normal or near normal Lachman examinations and 31 of 44 had no pivot shift. Subjective knee score using Lysholm was 95.7 and IKDC was 96.7. The failure rate was only 4.5 % (2 patients).

All-Epiphyseal

There are now a few different techniques that are physeal sparing and have tunnels only in the epiphysis of the femur and tibia. The Anderson technique is a freehand, two incision technique that uses quadrupled hamstring graft with a suspensory fixation on the femur and secures the tibial side by tying down on a screw and washer as the post. The mean age was 13.3 and with over 4 years of follow up the knee scores are excellent with IKDC of 96 and no reported cases of growth disturbances [13] (Figs 13.3**a** and **b**).

Modifications to the Anderson technique have been made. There is an all-inside transepiphyseal technique developed that uses pediatric knee guides to drill femoral and tibial sockets with fluoroscopic guidance. Hamstring autograft is used and the graft is secured with suspensory fixation on both sides [22].

Table 13.2 Review of literature on surgical outcomes following pediatric ACL reconstruction[a]

Technique	Studies	Number	Mean age (y)	Mean F/u (months)	Graft	Re-injury rate
Extraphyseal	Bonnard [43]	56	12.2	66	BTB	5.4 %
Physeal sparing	Kocher [13]	44	10.3	64	ITB	2/44
	Nakhostine [44]	5	14.0	24	Fascia Lata	0
	Parker [45]	6	13.3	33.	Hamstring	NR
All-epiphyseal	Cordasco [46]	23	11.8	24	Hamstring	4.3 %
	Hui [27]	16	12	24	Hamstring	NR
	Anderson [47]	12	13.3	49	Hamstring	NR
	Guzzanti [48]	8	11.2	69	Hamstring	NR
Partial transphyseal and hybrid	Emory Group [24]	NR	NR	NR	Hamstring	NR
	Lo [25]	5	12.9	89	Hamstring	NR
	Andrews [49]		13.5	58	Achilles allograft/ Fascia Lata	NR
Transphyseal	Redler [10]	16	14.2	43	Hamstring	0
	Courvoisier [50]	37	14	36	Hamstring	8.1 %
	Cohen [26]	26	13.3	45	Hamstring	6.7 %
	Liddle [28]	17	12	44	Hamstring	5.9 %
	Kocher [5]	59	14.7	43	Hamstring	NR
	McIntosh [51]	16	13.5	41	Hamstring	12.5 %
	Aronowitz [29]	15	14	25	Achilles Allograft	NR
	Lipscomb [52]	24	15	35	Hamstring	NR

[a]Adapted from Fabricant et al. [1]; with permission

Table 13.3 Levels of evidence for ACL injuries in the developing athlete

Statement	Grade of recommendation	References
Untreated ACL injuries lead to meniscal tears and chondral damage	B	[6, 7, 9]
Prevention programs can reduce ACL injuries	B	[35–37, 39]
Physeal Sparing reconstructions can restore stability to the developing knee	C	[13, 47]
Hybrid reconstruction techniques can restore knee stability with minimal risk of significant physeal arrest	C	[25, 49]
Transphyseal reconstruction can provide knee stability but some risk for physeal injury and should be reserved for children near the end of growth	C	[5, 10]

Hybrid Techniques

Hybrid techniques to reconstruct the ACL developed out of concern for physeal injury, particularly on the femur. There are a number of factors that contribute to the volume percentage of distal femoral physis that can be disrupted: drilling technique, tunnel size and tunnel inclination [23]. The group from Emory developed a technique that is transepiphyseal on the femur, avoiding this problem all together. Their technique is a hybrid and they utilize transphyseal drilling on the tibia [24]. They secure the quadrupled autologous hamstring with suspensory fixation on the femur and tie over a screw and washer on the tibia (Fig. 13.3**a** and **b**).

Lo et al., reported their results in 5 patients with wide open growth plates (mean age 12.9) [25]. Their hybrid technique involved a small central tibial with 6 mm tunnel and graft placement in the over the top position on the femur. Follow up was excellent at 7.4 years with no significant leg length difference or angular deformity. They had no re-tears. Only one patient reported poor IKDC score and they sustained subsequent patellar dislocation with osteochondral injury.

Transphyseal

Numerous different transphyseal techniques have been reported in the literature [10, 26–29]. The graft choice is predominantly autologous hamstring, but other graft choices have been reported. There remains concern about the use of use bone-patellar tendon bone. This is because placement of

a bone plug at the level of the physis can cause a local physeal arrest [30]. Furthermore, there is also concern about graft harvest as taking a bone plug from the tibial tubercle can cause anterior growth arrest leading to a recurvatum type deformity of the knee.

There are several Level IV studies that show these transphyseal techniques are producing good clinical results. Redler et al., looked at 18 transphyseal reconstructions using autologous quadrupled hamstring with mean age of 14.2 years [10]. They had mean follow up of 43 months with high scores for outcomes using the IKDC with a mean of 92, Lysholm mean of 94 and Tegner mean of 8.5. They had no re-tears during the study period, but three of the athletes tore their contralateral ACL. Furthermore they reported no leg length differences or angular deformities. (Fig. 13.4) Other groups have reported similar results. Cohen et al., reported

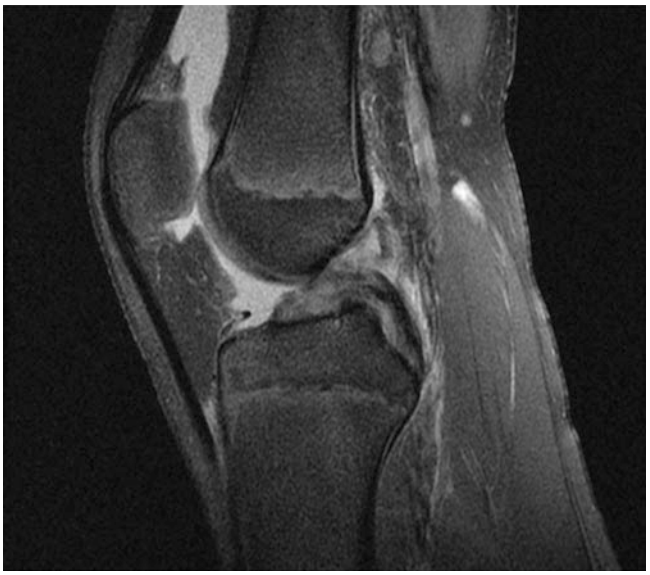

Fig. 13.2 Torn ACL Sagittal T2 image 12 year old male

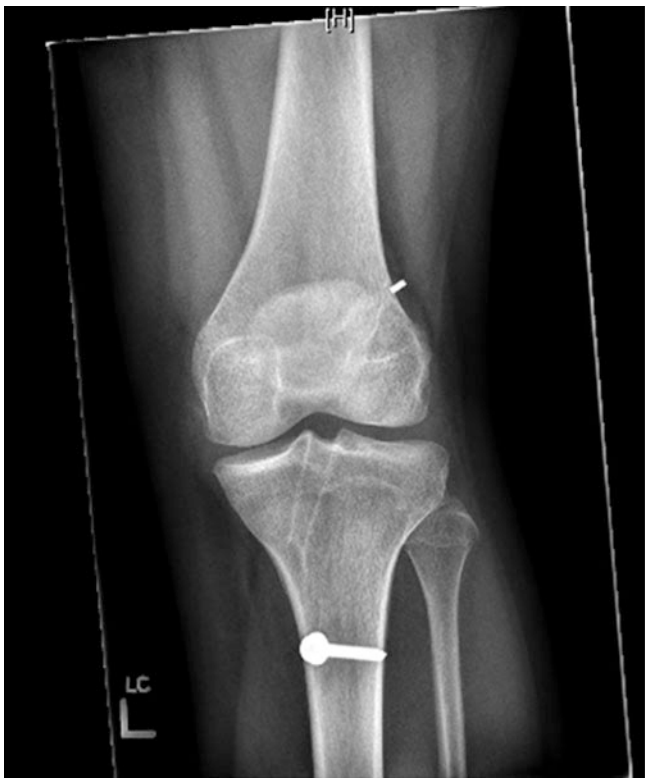

Fig. 13.4 Transphyseal ACL reconstruction images

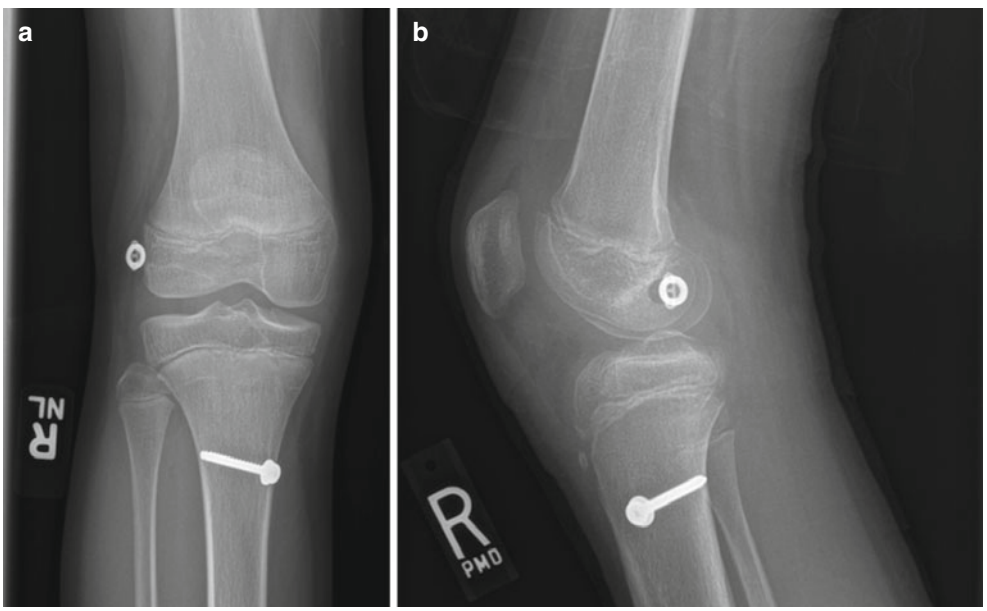

Fig. 13.3 Images from an All-Epiphyseal case

their outcomes on 26 patients that underwent transphyseal reconstructions [26]. Eighty-eight percent of their patients were able to return to same level of activity and they had mean IKDC of 91.5 and Lysholm of 93.5. The group out of Australia uses parental donor tissue for their transphyseal ACL reconstructions in children with open physes. They have youngest mean age for this type of technique [27]. They reported no growth arrest, angular deformities and IKDC knee score of 96.

Rehabilitation

Rehabilitation is an important part of reproducibly obtaining consistent results after surgical reconstruction of the anterior cruciate ligament. Postoperative stiffness in the pediatric and adolescent knee is not as significant a concern as with the adult patient. Immediate range of motion after surgery is not a requirement. Noncompliance is a more serious concern and because of this we recommend a brace for the first month after surgery [24]. Other goals for the first month after surgery include 90 degrees of flexion, regaining full extension, quadriceps strengthening and patellar mobility [1, 24].

The next phase of therapy should emphasize regaining full motion in flexion and extension, normalizing gait and performing a single leg squat with no pain and usually takes place up to 2 months after surgery. Goals in the next phase are from 2 to 4 month period after surgery and emphasis should be on core strength, quadriceps control and multiplane functional movements without unloading the affected leg or experiencing pain.

Running is allowed at 4 months as the patient continues to work on strength and flexibility. Sport specific exercises are initiated at 6–8 months. Our goal is to have the athlete back to sport by 9–12 months after the surgery. At our institution before clearance the athlete has to pass a functional ACL test.

With the patient's follow up, it is recommended to obtain a 3 ft standing films at 6 and 12 months from the date of surgery to monitor the lower extremity alignment and make sure there is no significant leg length difference [11, 31].

ACL surgery in the developing athlete can have high re-rupture rates and contralateral knee ACL injury rates. A recent paper presentation at AAOS meeting by Pinczewski et al. reported 15 year follow up results on ACL results in patients under 18 years of age and reported a 31 % re-tear rate for their ACL reconstructions. Of note, their contralateral ACL tear rate was 19 % [32]. The results of this demonstrate the importance of counseling young patients on the high rate of potential injury to their operative and non-operative knee. It also emphasizes the need for additional work and study on why the re-tear rate in this younger patient population is so high.

Prevention

There are many limitations to reconstructing the ACL in the developing athlete. The rehabilitation can take 9 months, there is risk of injury to the growth plate (11) and despite new techniques, as many as 30–35 % of middle and high school athletes choose not to return to their original sport [33]. In addition, a study highlighted that 42 % of young female soccer players, developed osteoarthritis signs on radiograph 10 years after ACL reconstruction [34]. Not surprisingly, with all the negative knee joint issues after ACL rupture, there has been significant focus lately on prevention of ACL injuries [35–37].

Neuromuscular Training programs (NMT) have been shown to work and are more effective the younger the athletes start [38]. Studies have identified modifiable risk factors with the landing patterns of athletes with noncontact ACL injuries such as: increased knee abduction moments, decreased knee flexion, poor trunk control and asymmetrical landing patterns [36, 39, 40]. Screening jump tests in the office can be used by physicians to identify high risk athletes [41]. These athletes with poor neuromuscular control can then be sent to physical therapy or to work with a school ATC or trainer to improve landing and jumping mechanics. ACL prevention programs such as the FIFA-11 (www.fifa-11.org) and PEP (Performance Enhancement and Prevention from the Santa Monica Sports Foundation; www.smsmf.org/smsf-programs/pep-program) have been shown to be effective in reducing ACL injuries and are cost effective [42].

Conclusion

As young athletes are spending more time on the field training with professional type schedules, specializing in one sport at an earlier age and diminishing time with free play, anterior cruciate ligaments in the developing athlete are becoming a more common occurrence. The pediatric orthopedic surgeon moving forward will need to properly diagnose and manage this injury. Proper assessment of the patient's chronologic and skeletal age, amount of growth remaining and demands will need to be taken into consideration to come up with the best treatment option. There are several physeal sparing and transphyseal techniques that can restore stability to the developing athlete's knee. With proper rehabilitation, these children are returning to their sport and activity. It should be emphasized that ACL re-rupture rates are higher in the younger athlete then patients over the age of eighteen. Furthermore, these athletes also are at higher risk then the general population to tear their contralateral knee ACL. Despite these new surgical techniques, there is good evidence that these reconstructed knees are predisposed to develop early osteoarthritis. Therefore, moving forward there should be a focus on ACL prevention neuromuscular training programs to help protect and prevent injury to healthy, young knees.

References

1. Fabricant PD, Jones KJ, Delos D, Cordasco FA, Marx RG, Pearle AD, et al. Reconstruction of the anterior cruciate ligament in the skeletally immature athlete: a review of current concepts: AAOS exhibit selection. J Bone Joint Surg Am. 2013;95(5):e28.
2. Werner BC, Yang S, Looney AM, Gwathmey FW, Jr. Trends in pediatric and adolescent anterior cruciate ligament injury and reconstruction. J Pediatr Orthop. 2015. Epub 2015/05/20.
3. Frank JS, Gambacorta PL. Anterior cruciate ligament injuries in the skeletally immature athlete: diagnosis and management. J Am Acad Orthop Surg. 2013;21(2):78–87 .Epub 2013/02/05
4. Dodwell ER, Lamont LE. Green DW 20 years of pediatric anterior cruciate ligament reconstruction in New York State. Am J Sports Med. 2014;42:675–80.
5. Kocher MS, Smith JT, Zoric BJ, Lee B, Micheli LJ. Transphyseal anterior cruciate ligament reconstruction in skeletally immature pubescent adolescents. J Bone Joint Surg Am. 2007;89(12):2632–9.
6. Aichroth PM, Patel DV, Zorrilla P. The natural history and treatment of rupture of the anterior cruciate ligament in children and adolescents. A prospective review. J Bone Joint Surg Br. 2002;84(1): 38–41.
7. Lawrence JT, Argawal N, Ganley TJ. Degeneration of the knee joint in skeletally immature patients with a diagnosis of an anterior cruciate ligament tear: is there harm in delay of treatment? Am J Sports Med. 2011;39(12):2582–7.
8. Moksnes H, Engebretsen L, Eitzen I, Risberg MA. Functional outcomes following a non-operative treatment algorithm for anterior cruciate ligament injuries in skeletally immature children 12 years and younger. A prospective cohort with 2 years follow-up. Br J Sports Med. 2013;47(8):488–94.
9. Millett PJ, Willis AA, Warren RF. Associated injuries in pediatric and adolescent anterior cruciate ligament tears: does a delay in treatment increase the risk of meniscal tear? Arthroscopy. 2002; 18(9):955–9.
10. Redler LH, Brafman RT, Trentacosta N, Ahmad CS. Anterior cruciate ligament reconstruction in skeletally immature patients with transphyseal tunnels. Arthroscopy. 2012;28(11):1710–7.
11. Shifflett GD, Green DW, Widmann RF, Marx RG. Growth arrest following ACL reconstruction with Hamstring autograft in skeletally immature patients: a review of 4 cases. J Pediatr Orthop. 2015.
12. Anderson AF. Transepiphyseal replacement of the anterior cruciate ligament in skeletally immature patients. A preliminary report. J Bone Jt Surg Am. 2003;85-A(7):1255–63.
13. Kocher MS, Garg S, Micheli LJ. Physeal sparing reconstruction of the anterior cruciate ligament in skeletally immature prepubescent children and adolescents. J Bone Joint Surg Am. 2005;87(11): 2371–9.
14. McCarroll JR, Rettig AC, Shelbourne KD. Anterior cruciate ligament injuries in the young athlete with open physes. Am J Sports Med. 1988;16(1):44–7.
15. Piasecki DP, Spindler KP, Warren TA, Andrish JT, Parker RD. Intraarticular injuries associated with anterior cruciate ligament tear: findings at ligament reconstruction in high school and recreational athletes. An analysis of sex-based differences. Am J Sports Med. 2003;31(4):601–5.
16. Mizuta H, Kubota K, Shiraishi M, Otsuka Y, Nagamoto N, Takagi K. The conservative treatment of complete tears of the anterior cruciate ligament in skeletally immature patients. J Bone Joint Surg Br. 1995;77(6):890–4.
17. Stanitski CL, Harvell JC, Fu F. Observations on acute knee hemarthrosis in children and adolescents. J Pediatr Orthop. 1993;13(4): 506–10.
18. Popkin CA, Murnaghan ML. Knee injuries in the growing athlete. Orthop Traumatol. 2012;26:12–9.
19. Azevedo JCBL, Macedo TB, Pedrosa LF, Arrais RF. Comparison between objective assesment and self-assesment of sexual maturation in children and adolescents. J Pediatr. 2009;85:135–42.
20. Kocher MS, Micheli LJ, Zurakowski D, Luke A. Partial tears of the anterior cruciate ligament in children and adolescents. Am J Sports Med. 2002;30(5):697–703.
21. Macintosh D, TA D. Lateral substiution reconstruction [abstract]. In: Proceedings and reports of universities, colleges, councils and associations. J Bone Joint Surg Br. 1976;58:142.
22. Fabricant PD, McCarthy MM, Cordasco FA, Green DW. All-inside, all-epiphyseal autograft reconstruction of the anterior cruciate ligament in the skeletally immature athlete: a surgical technique. J Bone Joint Surg Am Surgical Techniques. 2013;May 08;3(2):1–13.
23. Guzzanti V, Falciglia F, Gigante A, Fabbriciani C. The effect of intra-articular ACL reconstruction on the growth plates of rabbits. J Bone Joint Surg Br. 1994;76(6):960–3.
24. Hammond KE, Xerogeanes JW, Todd DC. Anatomic, transepiphyseal anterior cruciate ligament reconstruction. J Bone Joint Surg Tech. 2013;3(1):1–11.
25. Lo IK, Kirkley A, Fowler PJ, Miniaci A. The outcome of operatively treated anterior cruciate ligament disruptions in the skeletally immature child. Arthroscopy. 1997;13(5):627–34.
26. Cohen M, Ferretti M, Quarteiro M, Marcondes FB, de Hollanda JP, Amaro JT, et al. Transphyseal anterior cruciate ligament reconstruction in patients with open physes. Arthroscopy. 2009;25(8):831–8.
27. Hui C, Roe J, Ferguson D, Waller A, Salmon L, Pinczewski L. Outcome of anatomic transphyseal anterior cruciate ligament reconstruction in Tanner stage 1 and 2 patients with open physes. Am J Sports Med. 2012;40(5):1093–8.
28. Liddle AD, Imbuldeniya AM, Hunt DM. Transphyseal reconstruction of the anterior cruciate ligament in prepubescent children. J Bone Joint Surg Br. 2008;90(10):1317–22.
29. Aronowitz ER, Ganley TJ, Goode JR, Gregg JR, Meyer JS. Anterior cruciate ligament reconstruction in adolescents with open physes. Am J Sports Med. 2000;28(2):168–75.
30. Janarv PM, Wikstrom B, Hirsch G. The influence of transphyseal drilling and tendon grafting on bone growth: an experimental study in the rabbit. J Pediatr Orthop. 1998;18(2):149–54.
31. Kocher MS, Saxon HS, Hovis WD, Hawkins RJ. Management and complications of anterior cruciate ligament injuries in skeletally immature patients: survey of the Herodicus Society and The ACL Study Group. J Pediatr Orthop. 2002;22(4):452–7.
32. Roe J, Morgan M, Salmon L, Waller A, Thompson S, Pinczewski L. 15 year survival of endoscopic anterior cruciate ligament reconstruction in patients aged 18 and younger. 2015.Presented AAOS Annual Meeting 2015 Las Vegas.
33. McCullough KA, Phelps KD, Spindler KP, Matava MJ, Dunn WR, Parker RD, et al. Return to high school- and college-level football after anterior cruciate ligament reconstruction: a Multicenter Orthopaedic Outcomes Network (MOON) cohort study. Am J Sports Med. 2012;40(11):2523–9.
34. Lohmander LS, Ostenberg A, Englund M, Roos H. High prevalence of knee osteoarthritis, pain, and functional limitations in female soccer players twelve years after anterior cruciate ligament injury. Arthritis Rheum. 2004;50(10):3145–52.
35. Sugimoto D, Myer GD, Micheli LJ, Hewett TE. ABCs of evidence-based anterior cruciate ligament injury prevention strategies in female athletes. Curr Sports Med Rep. 2015;3(1):43–9.
36. Hewett TE, Myer GD, Ford KR, Heidt Jr RS, Colosimo AJ, McLean SG, et al. Biomechanical measures of neuromuscular control and valgus loading of the knee predict anterior cruciate ligament injury risk in female athletes: a prospective study. Am J Sports Med. 2005;33(4):492–501.
37. Hewett TE, Lindenfeld TN, Riccobene JV, Noyes FR. The effect of neuromuscular training on the incidence of knee injury in female athletes. A prospective study. Am J Sports Med. 1999;27(6):699–706.

38. Myer GD, Sugimoto D, Thomas S, Hewett TE. The influence of age on the effectiveness of neuromuscular training to reduce anterior cruciate ligament injury in female athletes: a meta-analysis. Am J Sports Med. 2013;41(1):203–15.
39. Hewett TE, Torg JS, Boden BP. Video analysis of trunk and knee motion during non-contact anterior cruciate ligament injury in female athletes: lateral trunk and knee abduction motion are combined components of the injury mechanism. Br J Sports Med. 2009;43(6):417–22.
40. Zazulak BT, Hewett TE, Reeves NP, Goldberg B, Cholewicki J. Deficits in neuromuscular control of the trunk predict knee injury risk: a prospective biomechanical-epidemiologic study. Am J Sports Med. 2007;35(7):1123–30.
41. Redler L, JP W, Dennis E, EF S, Ahmad CS. Physician and non-physician inter and intra-observer reliability of a field-based drop vertical jump screening test for ACL injury risk. 2015. Presented AAOS Annual Meeting 2015 Las Vegas.
42. Swart E, Redler L, Fabricant PD, Mandelbaum BR, Ahmad CS, Wang YC. Prevention and screening programs for anterior cruciate ligament injuries in young athletes: a cost-effectiveness analysis. J Bone Joint Surg Am. 2014;96(9):705–11.
43. Bonnard C, Fournier J, Babusiaux D, Planchenault M, Bergerault F, de Courtivron B. Physeal-sparing reconstruction of anterior cruciate ligament tears in children: results of 57 cases using patellar tendon. J Bone Joint Surg Br. 2011;93(4):542–7.
44. Nakhostine M, Bollen SR, Cross MJ. Reconstruction of mid-substance anterior cruciate rupture in adolescents with open physes. J Pediatr Orthop. 1995;15(3):286–7.
45. Parker AW, Drez Jr D, Cooper JL. Anterior cruciate ligament injuries in patients with open physes. Am J Sports Med. 1994;22(1):44–7.
46. Cordasco FA, Mayer SW, Graziano J, deMille P, Chiaia T, McCarthy M, et al. All-inside, all-epiphyseal ACL reconstruction in skeletally immature athletes: incidence of second surgery and two-year clinical outcomes. Orthop J Sports Med. 2015;3(7(suppl 2)).
47. Anderson AF. Transepiphyseal replacement of the anterior cruciate ligament using quadruple hamstring grafts in skeletally immature patients. J Bone Joint Surg Am. 2004;86-A Suppl 1(Pt 2):201–209. Epub 2004/10/07.
48. Guzzanti V, Falciglia F, Stanitski CL. Physeal-sparing intraarticular anterior cruciate ligament reconstruction in preadolescents. Am J Sports Med 2003;31(6):949–953. Epub 2003/11/19.
49. Andrews M, Noyes FR, Barber-Westin SD. Anterior cruciate ligament allograft reconstruction in the skeletally immature athlete. Am J Sports Med. 1994;22(1):48–54.
50. Courvoisier A, Grimaldi M, Plaweski S. Good surgical outcome of transphyseal ACL reconstruction in skeletally immature patients using four-strand hamstring graft. Knee Surg Sports Traumatol Arthrosc. 2011;19(4):588–91.
51. McIntosh AL, Dahm DL, Stuart MJ. Anterior cruciate ligament reconstruction in the skeletally immature patient. Arthroscopy. 2006;22(12):1325–30.
52. Lipscomb AB, Anderson AF. Tears of the anterior cruciate ligament in adolescents. J Bone Joint Surg. 1986;68(1):19–28.

What Is the Best Treatment for Blount's Disease?

14

Sonia Chaudhry and Paul A. Banaszkiewicz

Abstract

Bowed legs are a common appearance in children. Physiologic bowing is distinct from infantile Blounts and adolescent tibia vara, both of which are likely mechanical phenomena resulting from obesity, but have separate findings and treatment algorithms. Infantile Blounts is suspected with progressive or asymmetrical bowing with a lateral thrust during ambulation. Once confirmed with radiographs demonstrating a beaked medial proximal tibial metaphysis, treatment is initiated with bracing and/or surgical correction. Adolescent, or late-onset tibia vara, is almost always treated surgically. Treatment is aimed at both restoring knee joint line orientation and overall mechanical axis of the lower extremities with equal limb lengths.

Keywords

Tibia vara • Blount's Disease • Bowed legs

Background

The first challenge in treating bowed legs is differentiating "Blount's Disease" from the more common physiologic varus present in most children under 2 years of age. The average 15° varus tibiofemoral angle at birth is visually exacerbated once standing and walking around age 12 months improves the external rotation contracture of the hip joint (resulting from intrauterine positioning) that previously masked the increased femoral anteversion and internal tibial torsion of newborns. In these cases of physiologic genu varum, both distal femur and proximal tibial varus angulation contribute to an overall bowed appearance despite normal physeal morphology.

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Salenius and Vankka's classic 1975 study of 979 patients (1480 knee x rays) demonstrated a consistent pattern of tibio-femoral angle changes through childhood. Most new born babies have an average knee varus of 10° to 15°. As children start standing and walking, these changes to neutral between 18 and 24 months. This is followed by a steady progression towards maximum genu valgum (around 10°) achieved around age 4, and then correction into the adult valgus of about 6° over the next several years (Fig. 14.1). The standard deviation (SD) of the above quoted angle is 8° (more in the boys 10° and less in the girls 7°).

Toddlers under age 2 years should be followed clinically until resolution as long as alignment is improving, even if not on the expected schedule.

Indications for radiographs include asymmetry between right and left sides, lack of improvement with time, or a varus thrust with ambulation. After age 18 months, we obtain a digital mechanical axis view, which consists of a standing anteroposterior radiograph of the bilateral lower extremities from the hips to the ankles with the patellae facing forward. This is used to measure the tibiofemoral (TFA), mechanical axis deviation (MAD) metadiaphyseal of Drennan (MDA), and epiphyseal-metaphyseal angles (EDA) of the tibia.

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The tibiofemoral angle (TFA) is formed by the intersection of the two mid-diaphyseal lines of the femur and the tibia. The value should be within the normal range depicted by Salenius curve. MAD is the distance between the mechanical axe and the centre of the knee (the anatomic axes cross the knee almost at the centre). The normal mechanical axes pass 8 ± 7 mm medial to the centre of the knee. It is valuable in follow up of these patients. I need to see the mechanical axes approaching its normal pass through the knee.

The metadiaphyseal angle (MDA) is the angle formed by a line connecting the most distal point on the medial and lateral beaks of the proximal tibial metaphysis and a line perpendicular to the anatomic axis (or lateral cortex) of the tibia.

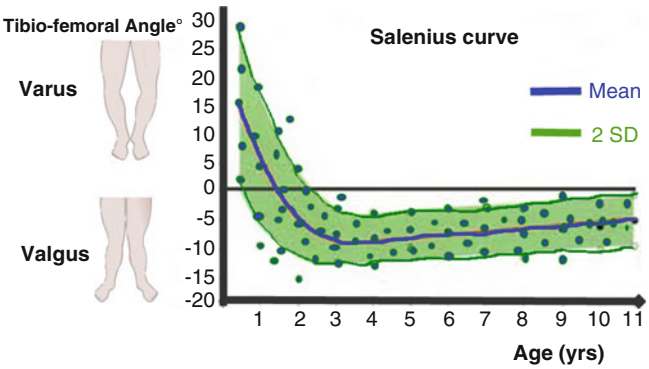

Fig. 14.1 Salenius curve

Levine and Drennan [1] found that in 29 of 30 legs with an initial MDA of more than 11°, radiographic changes of idiopathic tibia vara (Blount's disease) later developed whereas such changes developed in only 3 of 58 patients with an angulations of 11° or less.

The epiphyseal–metaphyseal angle (EMA) is determined by measuring the angle formed by a line through the proximal tibial physis parallel to the base of the epiphyseal ossification center and a line connecting the midpoint of the base of the epiphyseal ossification center with the most distal point on the medial beak of the proximal tibial metaphysis (Fig. 14.2).

Davids et al. [2] studied the knee x-rays of 80 children who were less than 3 years old. They found children with MDA >10° and EMA >20° are at greater risk for development of Blount disease and should be followed closely. In their series, none of the children with MDA <10° and EMA <20° developed Blount disease.

Angular measurements are altered by limb rotation, which is why proper positioning is paramount (Fig. 14.3**).**

Infantile Blount's is confirmed with a widened irregular medial proximal tibial physis with metaphyseal beaking (Fig. 14.4). Lateral tibial subluxation, lucent areas within the metaphyseal beak, and triangular ossification of the epiphysis may also be present.

The differential diagnosis includes skeletal dysplasia, metabolic disease, physeal disturbance from occult trauma or infection, or, rarely, focal fibrocartilaginous dysplasia. History, a skeletal survey, and lab work helps differentiate these conditions.

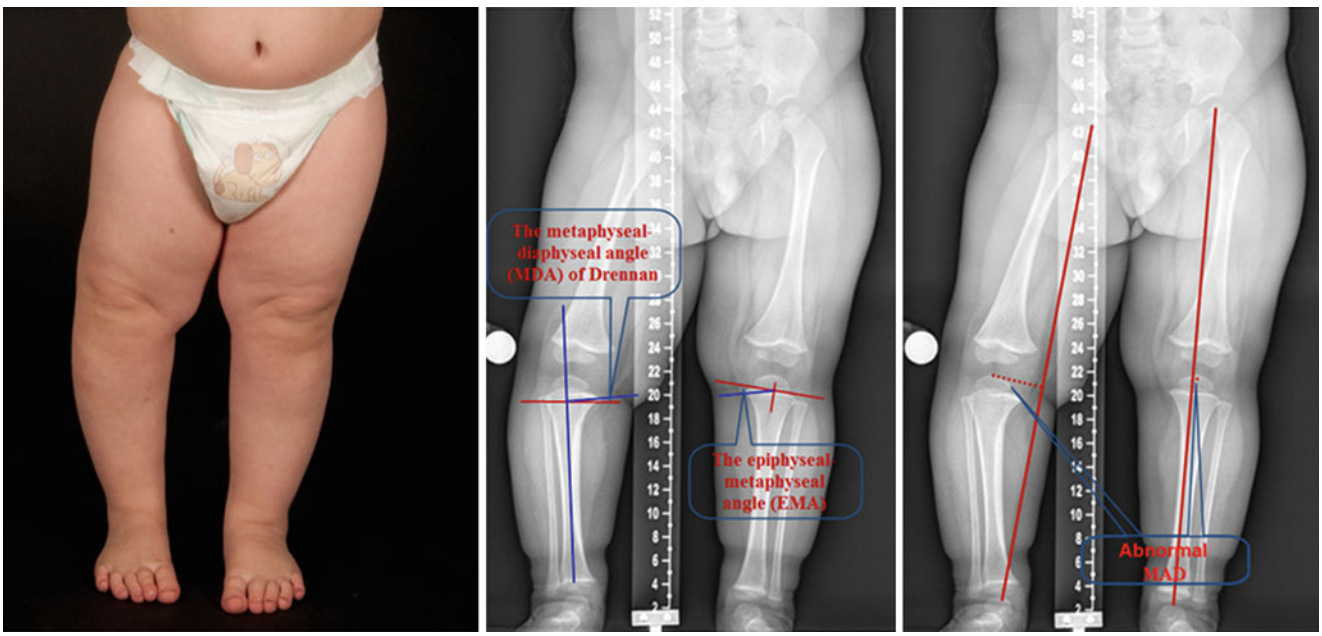

Fig. 14.2 Radiograhic measurments in Blounts diseases. The TFA is the angle the femoral and tibial mid-diaphyseal. MAD is the distance between the mechanical axe and the centre of the knee. The MDA is the angle formed by a line connecting the most distal point on the medial and lateral beaks of the proximal tibial metaphysis and a line perpendicular to the anatomic axis (or lateral cortex) of the tibia. The EMA is the angle formed by a line through the proximal tibial physis parallel to the base of the epiphyseal ossification center and a line connecting the midpoint of the base of the epiphyseal ossification center with the most distal point on the medial beak of the proximal tibial

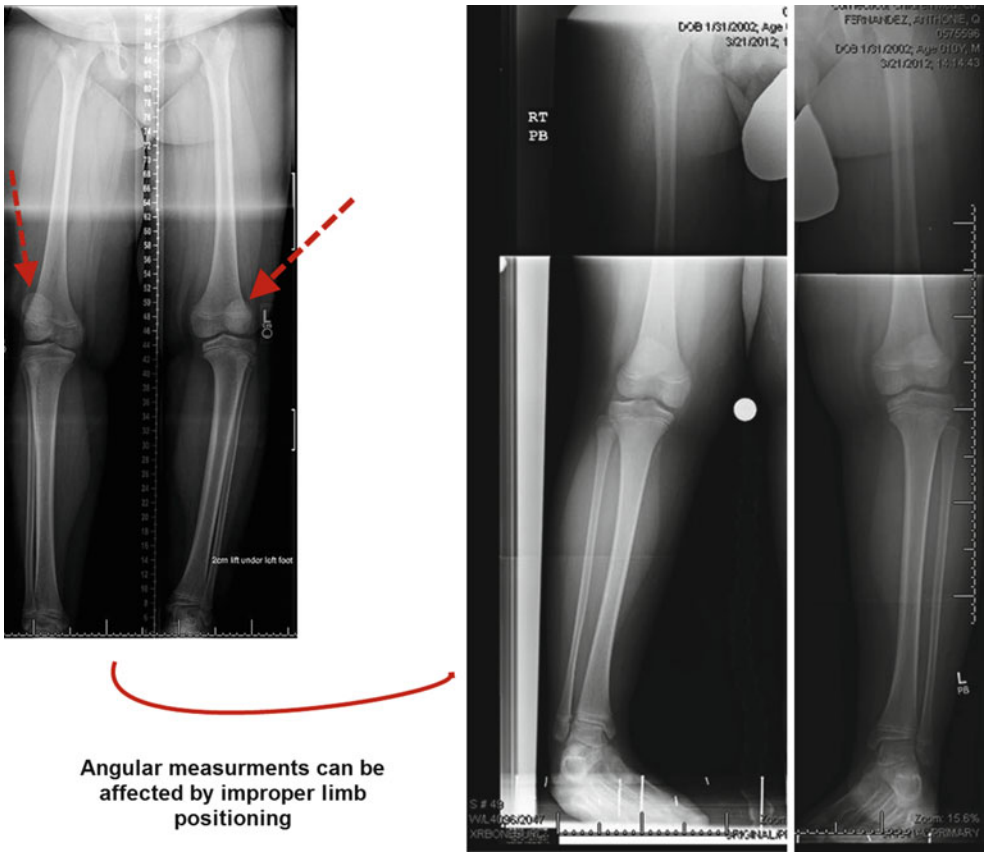

Fig. 14.3 The affect of limb position on angular meaurments. *Left*: standing limb alignment film of a 10 year old male demonstrating apparent bilateral overall varus angulation, however the patellae are not facing forward (*dashed arrows*). *Right*: the same patient was imaged on separate cassettes with the feet rotated in until the patellae faced forward, which improves the varus appearance

What Is Infantile Blount's?

When genu varum presents before age 3 and is determined to result from altered growth from the medial proximal tibial epiphysis, it is termed infantile Blount Disease [3]. Microscopically, the physis is disordered, with resultant abnormal endochondral ossification in the metaphysis causing a progressive varus as asymmetric lateral growth continues. The etiology is unknown and generally thought to be developmental, rather than congenital, as presentation before age 2 is rare [4].

Unlike physiologically bowed patients, patients with infantile Blounts tend to be obese and have a lateral thrust during gait. Each whole number increase in BMI increases the likelihood of Blount disease by 3 % [5]. Higher body mass index (BMI) has also been correlated with greater severity of varus and procurvatum deformity in this population. Additionally, patients may have marked intoeing, due to a combination of increased internal tibial torsion and femoral anteversion [6].

Langenskiold [7] divided patients into six stages according to age and radiographic metaphyseal and epiphyseal changes (Fig. 14.5). This classification has proven to be prognostic in certain populations, with resolution possible prior to stage 4, after which recurrence is likely. In nonwhite populations, however, stages can occur earlier and disease can progress despite treatment before age 4. Timing and type of treatment, therefore, must be tailored to the population being treated.

What Is the Best Nonoperative Treatment for Infantile Blounts?

A knee ankle foot orthosis (KAFO) or hip knee ankle foot orthosis (HKAFO) provides 3-point valgus pressure and can be considered in younger than age 3 or prior to Langenskiold stage II. An elastic Blount brace is a low profile option that utilizes an elastic band distally to provide the valgus force in conjunction with a medial upright, with drop locks to increase corrective force during weight bearing [4]. Bracing schedules vary from full time to day or night only, with frequent adjustment of the medial upright every couple months to provide a continuous valgus force.

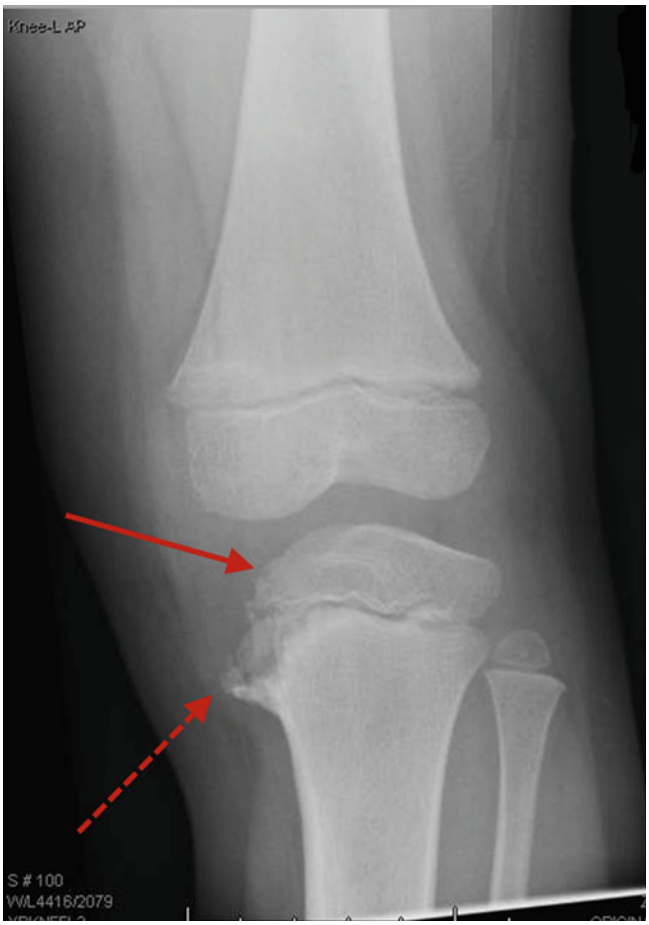

Fig. 14.4 Radiographic features of infantile Blounts. An AP of the left knee demonstrates a widened proximal tibial physis, sloped epiphysis (*solid arrow*), metaphyseal beaking with lucent areas (*dashed arrow*), and subluxation of the tibia indicating lateral instability

There is a narrow window for nonoperative treatment, as diagnosis is often made after age two, and to allow a 1-year trial without delaying surgery requires initiation before age 3. If neutral mechanical axis and healing of the radiographic Blounts lesion do not occur, corrective osteotomy should be performed prior to age 4. Surgery at earlier stage of disease is prognostic of higher success. At stage 4 and beyond, permanent physeal arrest is more likely.

The success of true bracing effect is confounded by the benign natural history of physiologic bowed legs treated as Blounts. One study limited to patients with Drennan's angles over 16° showed 86 % success. Bracing failure was more likely with ligamentous instability, body weight exceeding the 90th percentile, or late initiation [8]. Another study demonstrated 70 % success in Langenskiold stage II disease, though this was mainly in unilateral disease. Seventy percent of patients with bilateral involvement required surgical management [9].

What Is the Best Operative Treatment for Infantile Blounts?

Early Stage

The most common surgical approach is a single stage osteotomy to overcorrect the limb into about 5–10° of valgus, with concomitant lateral translation to ensure the mechanical axis passes laterally (Fig. 14.6). This removes the medial compressive forces to unload the sick medial physis and allow resumption of growth. Osteotomy can be performed with opening or closing wedges or a dome type. Staying distal to

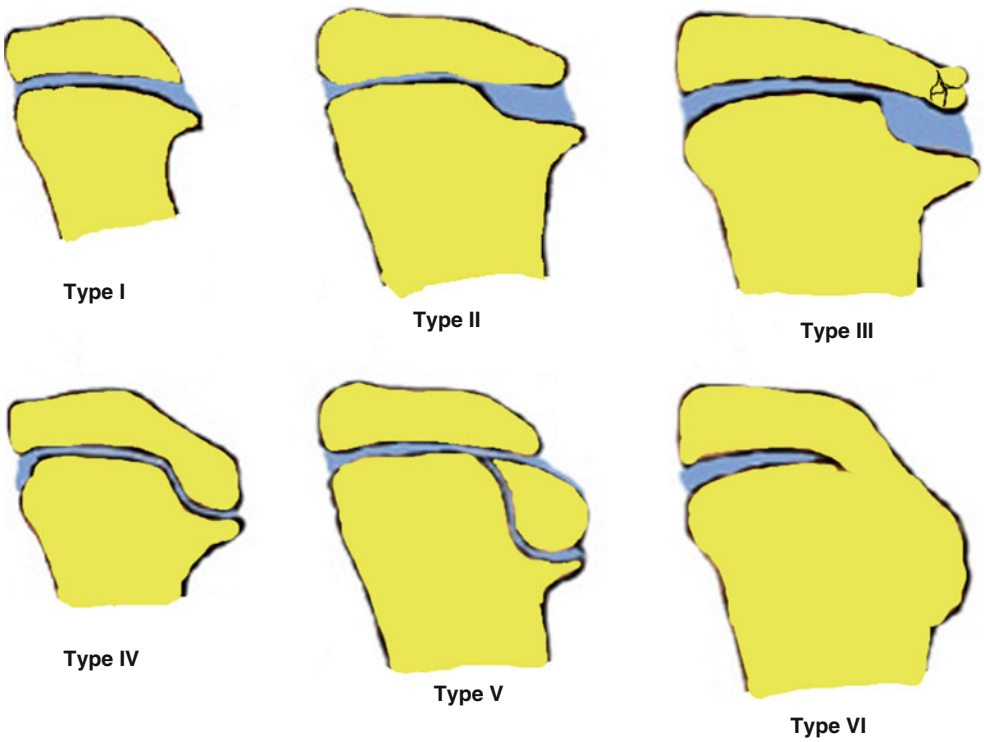

Fig. 14.5 Langenskiold staging of Blounts disease. Type I: Medial beaking, irregular medial ossification with protrusion of the metaphysis. Type II: Cartilage fills depression. Progressive depression of medial epiphysis with the epiphysis slopes medially as disease progress. Type III: Ossification of the inferomedial corner of the epiphysis. Type IV: Epiphyseal ossification filling the metaphyseal depression. Type V: Double epiphyseal plate (cleft separating two epiphyses). Type VI: Medial physeal closure

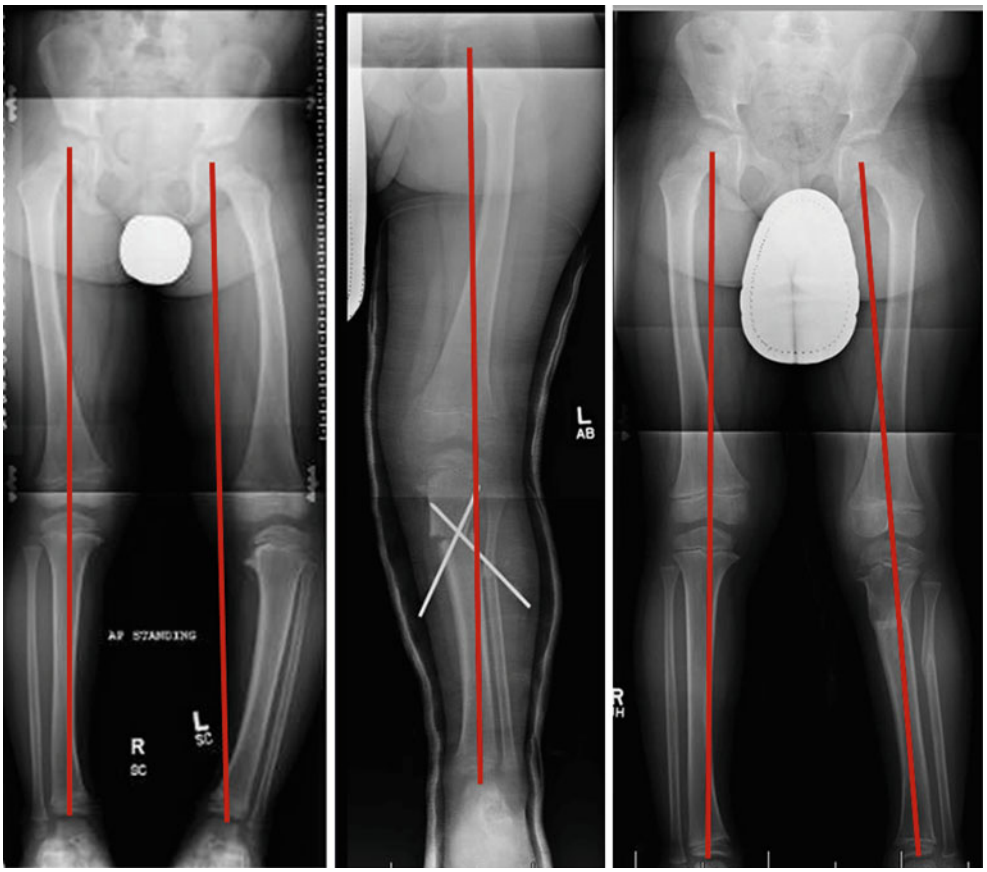

Fig. 14.6 Corrective osteotomy for infantile Blounts disease. *Left*: standing alignment film in a 4 year old male demonstrates mechanical axis going through the center of the knee joint on the right lower extremity while deviated medially on the right side. *Middle*: patient underwent acute tibial and fibular osteotomies through separate incisions to produce valgus, lateral translation, and external rotation of the distal fragment, with percutaneous K wire fixation, prophylactic anterior compartment fasciotomy, and long leg casting. *Right*: Postoperative radiographs demonstrating healed osteotomy, desired valgus overcorrection, and restoration of mechanical axis

the patellar tendon avoids physeal damage that would result in recurvatum. External rotation of the distal fragment is often needed, and judged pre and intraoperatively by assessment of bimalleolar and thigh foot (or heel thigh) axes.

Additional considerations include fibular osteotomy, fixation, and fasciotomy. Given the small but real risk of compartment syndrome, long leg casts are often bivalved to allow room for postoperative swelling. Percutaneous or internal fixation is therefore recommended, as cast loosening can result in loss of external stability. K wire fixation is most often employed. Prophylactic 3 compartment fasciotomy can be considered, and we perform this routinely. Despite these measures, subtle weakness of the extensor hallucis longus can still be observed, likely due to partial peroneal nerve palsy [4].

Another surgical option is guided growth with temporary lateral proximal tibial hemiepiphysiodesis. Lateral tension band plating has achieved 89 % success, and the distal femur can be concomitantly addressed [10]. Recurrence has been reported after implant removal, however, and patients with abnormal physes have more complications than other patients with knee deformities undergoing guided growth [11]. Screw breakage at the shaft is a significant complication that can delay or prevent correction, and patients with morbid obesity may benefit from modifications such as using double screw or H-plate configurations, solid instead of cannulated screws, or stainless steel instead of titanium implants [10].

Late Stage

The treatment for late stage disease, Langenskiold IV and V, is different as the medial physis has an effective growth arrest. Corrective osteotomy alone is likely to fail, and multiple osteotomies incur increasing surgical risk. Combining realignment with medial epiphysiolysis and interposition can prevent medial tethering and has been shown to be 80 % effective at avoiding varus recurrence when performed before age 7 for patients with stage III or greater disease [12]. In these cases, preoperative MRI is useful to determine area for physiolysis and ensure resection is performed lateral enough.

A double level osteotomy with intraarticular osteotomy for medial plateau elevation in addition to concomitant or staged high tibial osteotomy for mechanical axis correction can also be considered in older children (>7 years) with

advanced stage disease. MRI or arthrogram is important to assess the cartilaginous articular surface, which may be more congruent than radiographs would suggest. MRI has shown children with Blounts to have a thickened unossified medial epiphysis and similar tibial condylar inclination that may compensate for the sloped ossified portion [13].

Completing the growth arrest with lateral epiphysiodesis will prevent recurrence, incurs significant limb length discrepancy, and is generally reserved for patients with under 2 years of growth remaining. It should be kept in mind that Blounts patients have advanced bone age compared to chronologic age by an average of 16 months overall, 26 months in early onset group [13], as this affects prediction of length discrepancy and efficacy of guided growth. Lengthening can be performed concomitantly or at a later time.

What Is Adolescent Blounts?

adolescent blounts, or late onset tibia vara, varies in definition, but generally refers to an idiopathic progressive angulation after about age 8 in patients without concomitant pathology except for obesity [4]. Compared to infantile Blounts, it is less common, more often unilateral, affects a higher proportion of males, has less internal tibial torsion, and more often has concomitant limb length discrepancy (Table 14.1**).** Etiology is thought to be from mechanical compression caused by high body mass and repetitive microtrauma during adolescent growth spurt.

Concomitant pathology associated with late onset tibia vara includes low vitamin D levels, slipped capital femoral epiphysis, and obstructive sleep apnea (OSA). Vitamin D deficiency may be both a cause and effect, exacerbating physeal vulnerability and resulting from decreased sunlight exposure that accompanies impaired mobility and obesity. Patients with 25-vitamin D levels below 16 ng/mL are over 7 times more likely to have Blount disease than patients with higher levels [14]. Almost 2/3 of Blounts patients over age 9 have OSA, and snoring should prompt screening with polysomnography [15].

Radiographic features are distinct from infantile Blounts. The proximal metaphysis is not beaked, but the overlying physis is widened medially or all the way across. Lateral distal femoral physeal widening can be also present. One fifth of patients demonstrate distal femoral varus, contributing to 30 % (6–20°) of the overall varus. Concomitant distal tibial valgus may also develop in response to the proximal tibial varus, but is not significantly different from population normals [16, 15]. Digital imaging software is useful to measure overall mechanical axis as well as joint line angles that can be compared to reference normals [17]. We employ TraumaCad (VoyantHealth, Westchester, IL), which imports and saves angular measurements and deformity correction planning (Fig. 14.3) to our digital picture and archiving communication system (PACS) (Siemens, Erlangen, Germany) (Fig. 14.7).

What Is the Best Treatment for Adolescent Blounts?

Treatment is mainly surgical, as orthotics are both poorly tolerated and ineffective in this older obese population. The goal is correction to a neutral mechanical axis, as even with correction to less than neutral there will be apparent valgus and thigh impingement. Both internal and external fixation has been used, and simultaneous or staged correction of distal femoral deformity should be considered. Internal fixation has the advantages of shorter time to bony union and avoidance of pin-related complications, however risk of

Table 14.1 Comparison between infantile and adolescent tibia vara

Characteristic	Infantile Blounts	Adolescent Blounts
Incidence	More common	Less common
High BMI	Correlates with magnitude of deformity	Higher than early onset, correlates with deformity when BMI > 40
Low vitamin D level	Both	Both
Gender	Male predominance	Higher male predominance
Laterality	Bilateral (can be unilateral)	Unilateral (can be bilateral)
Proximal tibial varus	Higher magnitude	Lower magnitude
Distal femur varus	Not present	More often present (20 %)
Internal tibial torsion	More prominent	Less prominent
Procurvatum	More prominent	Less prominent
Radiographic Changes	Medial metaphyseal beaking	Medial proximal tibial physiolysis (widening)
Advanced bone age	Average 26 months	Average 10 months
Efficacy of Bracing	Successful in early stages when initiated before age 3	Ineffective and poorly tolerated

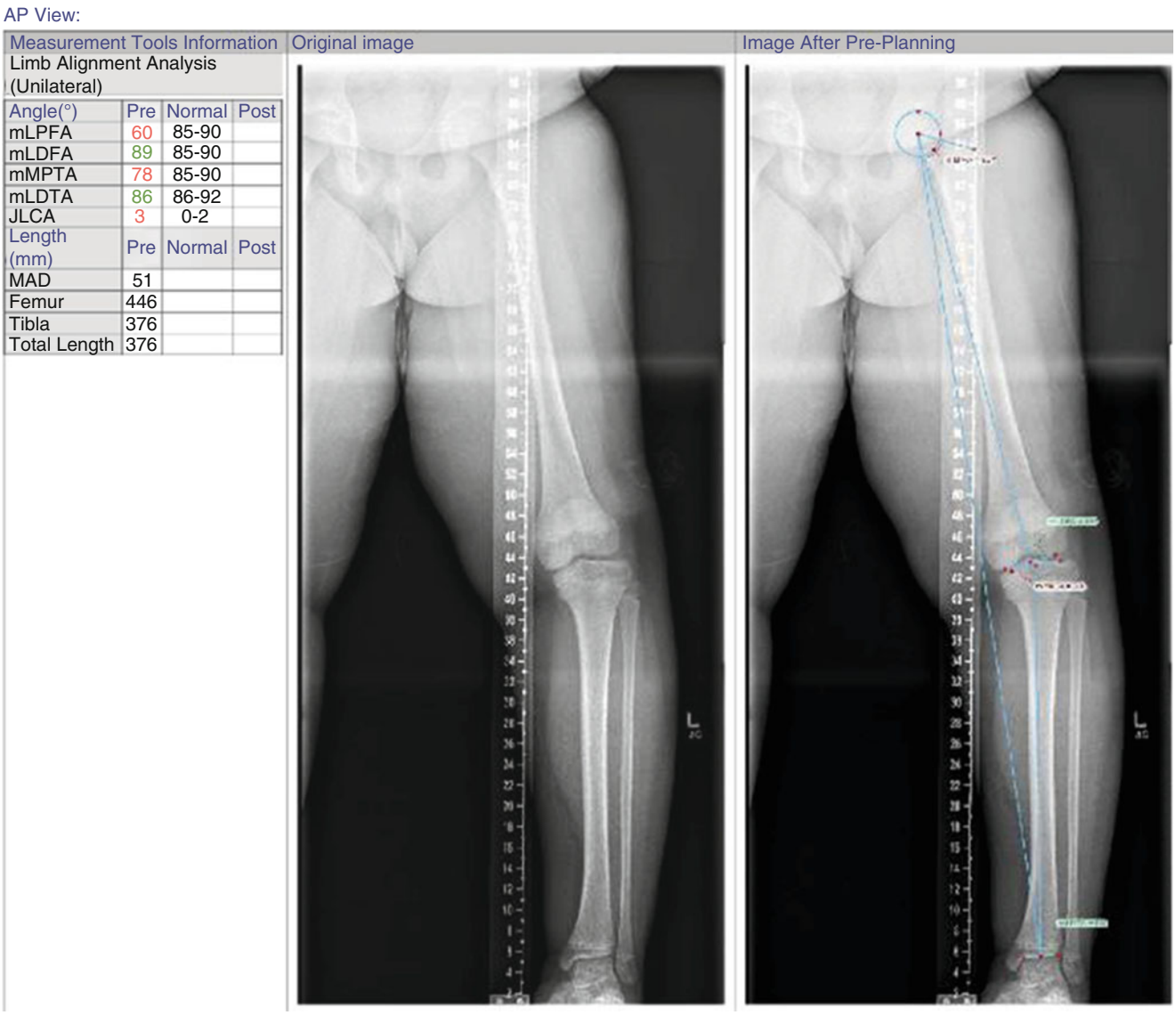

Fig. 14.7 TraumaCAD analysis of a patient with Blount disease. TraumaCAD analysis of patient from Fig. 14.3 demonstrates the varus to originate from an abnormal medial proximal tibial angle, without contribution from the distal femur

neurovascular injury, inability to address length discrepancy, and difficulty of accurate multiplanar correction has decreased its popularity.

External fixation allows more adjustability and earlier weight bearing, though patients often have difficulty with the latter and will often end up being non weight bearing. Gradual correction allows more accurate correction of mechanical axis deviation, sagittal plane deformity, and limb length than acute correction [18], with the added advantage of lower prevalence of transient peroneal nerve palsy [19]. Constructs can range from monolateral multiaxial fixators [20] to Ilizarov circular fixators, or more recently, computer-assisted correction with the Taylor Spatial Frame system (Smith and Nephew Inc., Memphis, TN) [21–23].

Lateral proximal tibial hemiepiphysiodesis is technically simpler and much less morbid than osteotomy or corticotomy, however it relies on the growth potential of the medial physis and is a uniplanar correction. Bone age should be checked, as late onset Blounts patients average 10 months of increased bone maturity by the Greulich and Pyle method [13]. Successful limb realignment with hemiepiphyseal stapling of the lateral proximal tibia, in conjunction with the distal femur when indicated, without the need for osteotomy is predicted by younger age (<10 years) at time of surgery and mild to moderate deformity [24]. Risk factors for failure include age over 14, BMI over 45 kg/m^2, and greater baseline deformity [25]. Despite two-thirds of patients failing to correct with this technique alone, it may mitigate the amount of correction subsequently needed (Fig. 14.8).

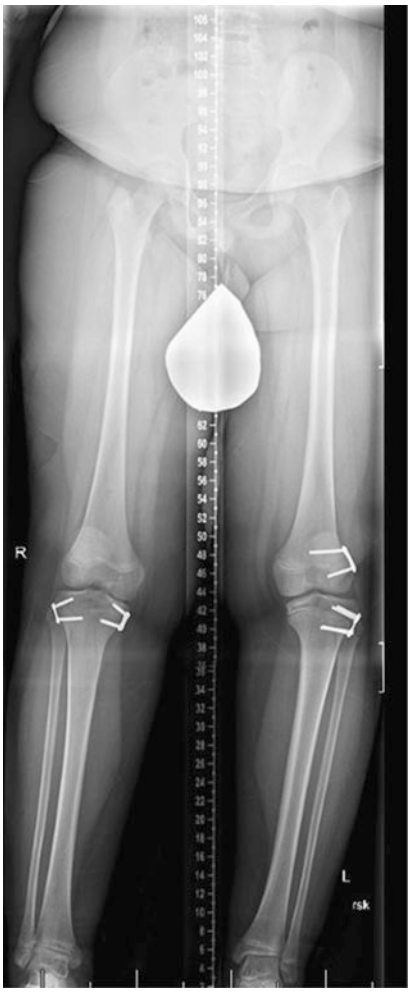

Fig. 14.8 Treatment of Blount disease using 8 plates. The above patient had a significant deformity, both distal femur and proximal tibia underwent tension band plating to maximize overall correction to avoid additional procedures with increased morbidity. Note the double screw technique used in the proximal tibia given the patient's body habitus. Patient was also underwent contralateral proximal tibial epiphysiodesis to improve limb length discrepancy

Given the role of obesity in the pathophysiology of Blounts and its associated morbidity, concomitant weight loss is an important treatment goal. Despite satisfactory radiologic outcome and nutritional counseling, however, over three-quarters of patients continue to gain weight the longer they are followed [26].

References

1. Levine AM, Drennan JC. Physiological bowing and tibia vara. The metaphyseal-diaphyseal angle in the measurement of bowleg deformities. J Bone Joint Surg Am. 1982;64(8):1158–63.
2. Davids JR, Blackhurst DW, Allen Jr BL. Radiographic evaluation of bowed legs in children. J Pediatr Orthop. 2001;21(2):257–63.
3. Blount W. Tibia vara. Osteochondrosis deformans tibiae. J Bone Joint Surg. 1937;19(1):1–29.
4. Herring JA, editor. Tachdjians, pediatric orthopaedics, vol. 1. Philadelphia: Saunders Elsevier; 2008.
5. Montgomery CO et al. Increased risk of Blount disease in obese children and adolescents with vitamin D deficiency. J Pediatr Orthop. 2010;30(8):879–82.
6. Aird JJ, Hogg A, Rollinson P. Femoral torsion in patients with Blount's disease: a previously unrecognised component. J Bone Joint Surg. 2009;91-B(10):1388–93.
7. Langenskiold A. Tibia vara; (osteochondrosis deformans tibiae); a survey of 23 cases. Acta Chir Scand. 1952;103(1):1–22.
8. Raney EM et al. Orthotic treatment of infantile tibia vara. J Pediatr Orthop. 1998;18(5):670–4.
9. Richards BS, Katz DE, Sims JB. Effectiveness of brace treatment in early infantile Blount's disease. J Pediatr Orthop. 1998;18(3):374–80.
10. Scott AC. Treatment of infantile blount disease with lateral tension band plating. J Pediatr Orthop. 2012;32(1):29–34.
11. Wiemann JM, Tryon C, Szalay EA. Physeal stapling versus 8-plate hemiepiphysiodesis for guided correction of angular deformity about the knee. J Pediatr Orthop. 2009;29(5):481–5.
12. Andrade N, Johnston CE. Medial epiphysiolysis in severe infantile tibia vara. J Pediatr Orthop. 2006;26(5):652–8.
13. Sabharwal S et al. Intra-articular morphology of the knee joint in children with Blount disease. J Bone Joint Surg (Am). 2013;94(10):883–90.
14. Montgomery CO et al. Increased risk of blount disease in obese children and adolescents with vitamin D deficiency. J Pediatr Orthop. 2010;30(8):879–82.
15. Gordon JE. Obstructive sleep apnoea syndrome in morbidly obese children with tibia vara. J Bone Joint Surg. 2006;88-B(1):100–3.
16. Myers TG et al. Incidence of distal femoral and distal ibial deformities in infantile and adolescent blount disease. J Pediatr Orthop. 2005;25(2):215–8.
17. Paley D,Tetsworth K. Mechanical axis deviation of the lower limbs. Clin Orthop Relat Res. 1992;&NA;(280):48???64.
18. Feldman DS et al. Accuracy of correction of tibia vara. J Pediatr Orthop. 2006;26(6):794–8.
19. Gilbody J, Thomas G, Ho K. Acute versus gradual correction of idiopathic tibia vara in children. J Pediatr Orthop. 2009;29(2):110–4.
20. Clarke SE, McCarthy JJ, Davidson RS. Treatment of blount disease. J Pediatr Orthop. 2009;29(2):103–9.
21. Sachs O et al. Treatment of adolescent blount disease using Taylor spatial frame with and without fibular osteotomy. J Pediatr Orthop. 2014;35(5):501–6.
22. Li Y, Spencer SA, Hedequist D. Proximal tibial osteotomy and taylor spatial frame application for correction of tibia vara in morbidly obese adolescents. J Pediatr Orthop. 2013;33(3):276–81.
23. Feldman DS et al. Correction of tibia vara with six-axis deformity analysis and the Taylor spatial frame. J Pediatr Orthop. 2003;23(3):387–91.
24. Park S-S. Outcome of hemiepiphyseal stapling for late-onset tibia vara. J Bone Joint Surg (Am). 2005;87(10):2259.
25. McIntosh AL. Treatment of adolescent tibia vara with hemiepiphysiodesis: risk factors for failure. J Bone Joint Surg (Am). 2009;91(12):2873.
26. Sabharwal S et al. Do children with blount disease have lower body mass index after lower limb realignment? J Pediatr Orthop. 2013;34(2):213–8.

Evidence-Based Treatments for Fractures Around the Knee Joint

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Abstract

Fractures around knee in children are common and pose some challenges to the treating surgeon. Two types, distal femoral physeal fractures and tibial spine fractures, attract significant controversies and variation in practice. In this chapter, we examined the evidence behind various types of treatments for these two fractures.

Keywords

Distal femur fracture • Physeal injury • Growth arrest • Leg length discrepancy • Angular deformity • Salter-Harris injury • Tibial spine injury • Tibial eminence injury • None operative management • Arthroscopic reduction • Screw fixation • Suture fixation • Arthrofibrosis

Distal Femoral Physeal Injuries

The distal femoral physis grows at a length of approximately 10 mm per year and contributes 70 % of the growth of the femur and 40 % of the growth of the lower limb [1]. Historically injuries at this location occurred with hyperextension and torsion around the knee and most commonly when trapping the foot in a cartwheel, were often complicated by vascular injury and infection, and could lead to amputation and even death [2]. In the modern era, these injuries are most often found in adolescents males [3] following motor vehicle accidents [4], or sporting activities [5], and are usually classified according to the Salter-Harris (SH) injury classification system [6]. Although uncommon and only accounting for 1 % of physeal injuries [7] they are associated with a high incidence of complications, the most serious of these include neurovascular injury [3], and growth disturbance which can be as high as 70 % [8].

Almost all the studies on this subject are retrospective studies. Without exception all the reported series have a significant number of SH type II injuries and small number of types I and III-V injuries. In addition, many studies have small numbers in their comparison groups which makes statistical analysis less robust and reliable. There is also a wide variation within the literature with regards to the definitions of both growth disturbance and limb length discrepancy (LLD), which makes direct comparisons between different studies difficult. This is further limited by the poor and subjective definitions of "good outcome" and "bad outcome" which are employed by different authors.

Should All Distal Femoral Physeal Injuries Be Treated with Reduction and Operative Fixation?

One of the challenges in understanding which injuries require surgical fixation is the diagnosis of fracture displacement and intra-articular involvement. Wall and May [9] reported difficulty in recognising displacement in type III and IV SH injuries and recommended a low threshold for both obtaining computer tomography (CT) scans prior to surgical fixation. In Arkader et al's. [5] study of 83 fractures over 10 years, 35 of 43 (82 %) displaced factures were treated with surgery. The authors reported a 49 % complication rate in displaced fractures in comparison to a 27 %

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complication rate in undisplaced fractures with no association with the degree of displacement. Hence, Arkader et al. [5] concluded that all displaced fractures should be treated with reduction and internal fixation. The authors also reported one case of an undisplaced fracture which was initially treated conservatively, which required later surgical intervention for fracture displacement. These results are similar to those reported by Stephens et al. [10] In their 15-year retrospective review of 20 cases, 17 cases required closed reduction of which seven further displaced and lost their reduction. The majority of these cases were SH II injuries (82.4 %). Four of these seven cases required further manipulation and Kirschner (K)-wiring, and one required open reduction. Edmunds and Nade [11] further support this by reporting a 64 % (7/11 patients) rate of re-displacement with closed reduction. In their series of 33 patients, all except four injuries were displaced. Ten of the thirty-three patients were lost to follow-up, and of the remaining 23, the study did not specify the number of non-displaced fractures. Edmunds and Nade defined leg length discrepancy (LLD) greater than 2 cm or any coronal angulation that caused disability as a poor outcome. Overall, 8 of the 23 patients had an open reduction and internal fixation (ORIF) with only one case of poor outcome (12.5 %); 5 of 23 patients had a closed reduction and internal fixation (CRIF) with poor results reported in 60 %, and 7 of 23 patients were treated with closed reduction and casting or traction with a poor result in 57 %. Three patients had no documented treatment for their injuries. Edmunds and Nade concluded that all displaced SH II-IV injuries should be treated with ORIF to avoid a poor outcome.

Accurate reduction is associated with a reduced incidence of LLD and coronal plane deformity. This has been shown by Lombardo and Harvey [12] in their retrospective review of 34 fractures. They reported a mean LLD of 0.81 cm and only 2 of 8 (25 %) cases with a varus or valgus deformity greater than 5° in displaced fractures that were reduced. This is compared to a mean LLD of 2.52 cm and 6 of 12 (50 %) cases with a varus or valgus deformity greater than 5° in displaced fractures that were not reduced. This finding was consistent across all SH injury types. Ilharreborde et al. [8] concluded that the degree of displacement and metaphyseal comminution were associated with growth arrest and angular deformity. On closer review of their data, the difference in rates of LLD or coronal plane deformity between cases which were completely displaced or associated with metaphsyeal comminution (66.7 %) was only slightly higher than those injuries which were not (62.5 %).

The largest case series of these injuries is the study of Eid and Hafez [3]. In their study of 151 cases, 111 patients had initial treatment with either an above knee (AK) cast or closed reduction and an AK cast or hip spica. Of these, 33 patients (29.7 %) had redisplaced at 2 weeks, and 25 of 33 were unsuccessful with repeat closed reduction and required an ORIF. All of these studies support the conclusions of Thomson et al. [13] that casting alone is not enough to maintain reduction, and that all these injuries should be treated with fixation. This recommendation is in contrast to that of Czitrom et al. [4] who reported their finding from The Hospital of Sick Children in Toronto. They treated 15 of 42 fractures with above knee casting alone (of which ten were undisplaced and required no initial reduction). Of these, four patients lost initial reduction requiring manipulation and casting. These were all SH type II injuries, and none required open reduction or fixation. Czitrom et al. concluded that all SH II injuries should be treated with closed reduction and casting, and that open reduction and fixation be reserved for Type III and IV injuries. Illharreborde et al. [8] reported no re-displacement in their retrospective series of 20 SH II injuries treated with closed reductions and above knee casts, but their numbers were small (only 4 of 20 cases), with the majority (16/20) being treated with ORIF.

Does Surgical Fixation Increase the Risk of Growth Arrest?

Physeal plate damage leading to growth arrest normally occurs 12–18 months post injury [12]. The most comprehensive summary of our understanding of growth disturbance after distal femoral physeal injuries comes from the meta-analysis performed by Basener et al. [14]. They included 16 studies with a minimum follow up of 12 months and a total of 564 cases. Basener et al. reported an overall rate of growth disturbance of 52 %, with a four times increased risk in displaced versus undisplaced injuries. As we previously described, there are variations between different studies on what is perceived as a complication or growth arrest and the exact definition of these complications. Basener et al. considered significant growth arrest to be present with either LLD of greater than 1.5 cm or a coronal plane deformity of greater than 5°, which was reported in 112 of 506 patients (22 %) (Table 15.1).

Of the cases with patient level data (151 patients), Basener et al. reported a 27 % rate of growth arrest in patients treated with surgical fixation, compared to 37 % with no surgical fixation. The surgical fixation group consisted of only 30 patients compared to 121 in the non-surgical fixation group, which may have contributed to the lower reported prevalence in the former group. This is supported by the overall incidence of growth disturbance which was greater in patients who were not treated with surgical fixation (63 % vs. 58 %). Another possible confounding factor is the selection and treatment bias of the treating surgeon in fractures that are treated with surgical fixation.

Table 15.1 Summary of recommendations

Recommendations	Grade of recommendation
Distal femoral physeal injuries	
Reduction and surgical fixation is indicated for all distal femoral physeal injuries	C
Surgical fixation is not associated with an increased risk of growth arrest	I
Transphyseal fixation does not increase the rate of growth arrest and deformity as compared with physeal sparing techniques	C
The risk of growth arrest increases with increasing Salter Harris classification	B
High energy injuries are associated with a higher risk of growth arrest	B
Anterior tibial spine fractures	
All Type II injuries should be treated by surgical reduction and internal fixation	C
Open surgery is associated with fewer complications than arthroscopic surgery	C
Screw fixation is superior to suture fixation	C
The risk of complications increases with increasing Myers and McKeever classification	B
Transphyseal screw fixation is safe	B

Does Transphyseal Pinning Increase the Risk of Growth Arrest as Compared with Physeal Sparing Techniques?

Violating physeal plates with metalwork has always had a theoretical association with increased risk of growth arrest. Growth disturbances are a recognised sequlae of paediatric distal femoral injuries [8] and for this reason understanding the effects of metalwork around the physeal plate in these injuries is of paramount importance. Arkader et al. [5] demonstrated that fractures that were internally fixed with none physeal-sparing techniques (Steinman pins; n = 20) had a higher rate of complications (65 %) compared to physeal sparing fixation techniques (n = 13; complication rate 30 %) (p = 0.06). Complications included growth arrest, loss of reduction, persistent loss of range of knee motion, malunion and peroneal nerve injury. In their retrospective study of 55 displaced fractures requiring reduction, Garret [15] et al. reviewed 44 cases treated with surgical fixation. The majority of these (40/44) were treated with two percutaneous smooth K-wires or Steinman pins (1.8–3.2 mm diameter) crossing the physis. The remaining four cases were treated with physeal sparing screws. They reported a 21.8 % overall prevalence of physeal arrest. Fractures treated with K-wires or pins that crossed the physeal plate were associated with a 17.5 % rate of growth arrest, compared to injuries treated in cast alone (27.3 %) and with screw fixation (50 %) (p = 0.2). The small number of cases in the different treatment arms prevent the demonstration of a statistically significant (p < 0.05) difference. However, the results tend to favour the argument that smooth wires or pins violating the physeal plate do not contribute to growth arrest.

What Other Factors Increase the Risk of Growth Arrest?

In the meta analysis by Basener et al. [14], the prevalence of growth disturbance was least in SH I injuries (36 %) and most in SH IV (64 %), followed by SH II (58 %) and then SH III (49 %) injuries. This is consistent with other studies that suggest increasing SH classification increases the rate of growth arrest [4, 5]. The reported increased rates of growth arrest seen in SH II injuries may be due to the large number of these cases in all studies, causing a skewing of the data. It may also be related to the fact that although injuries in the lower SH classifications (SH I & II injuries) have growth arrest, it is not as clinically significant as in the higher classifications (SH III and greater injuries). This would concur with the findings of Czitrom et al. [4]who reported that 20 of 29 SH I or II injuries had some degree of shortening but only 3 of 29 had an LLD greater than 1.5 cm.

Riseborough et al. [16] reported a rate of 57 % and 26 % for LLD and angular plane deformity respectively in their study of 66 fractures. When comparing age groups, they found 19 of 23 cases (83 %) within the juvenile age group (2–10 years old) had an LLD or angular plane deformity compared with only 18 of 36 (50 %) in the adolescent group (11 years and older). They postulated that as most juvenile injuries were associated with high energy trauma, they would lead to more significant growth sequale compared with the low energy sports injuries sustained by adolescents. Eid and Hafez [3] found similar trends in their study, with both shortening and a LLD most common in the age group 2–11 years [shortening 28 of 49 (57 %); angular plane deformity 33 of 49 (67 %)]. Garrett et al. [15] further confirmed the positive association between physeal bars and higher energy injuries (11 of 12 compared with 1 of 19 in low energy injuries) and an increasing SH classification (100 % SH IV, 50 % SH III and 17 % SH II injuries).

Anterior Tibial Spine Fractures

Fractures of the anterior tibial spine are an uncommon injury amongst the paediatric population, and were first reported in children by Pringle in 1907 [17]. They are reported to be the

cause of acute knee haemarthrosis in 5 % of cases [18], and occur most commonly from hyperextension or hyperflexion injuries when skiing, falling from bicycles [19], during field sports [20] and road traffic accidents. They are most common in the 8–14 years age group [19, 21], with a peak incidence between 11 and 13 years [22, 23]. Meyers and McKeever [19] originally classified these injuries into three types: Type I is an undisplaced injury; Type II is a displaced injury with an intact posterior cortex and Type III injuries are completely detached from the tibial plateau. They also described a Type III+ which represents a completely displaced and rotated fragment. Zaricznjy [24] later added a type IV injury which is a multifragmentory spine fracture. Historically, they were often referred to as the adolescent anterior cruciate ligament (ACL) injury and were thought to occur due to incomplete ossification of the tibial spine, requiring less tensile force to cause injury than the ACL [25]. Although a large proportion of the studies on this subject are retrospective, more recent comparative prospective studies have been published looking at specific aspects of management of this condition.

Should Type II Fractures Be Treated Operatively or None Operatively?

Over the years, there has been much debate about the optimum treatment of the type II injury. In Meyers and McKeever's [19] original description of 35 paediatric patients, type II injuries (n = 17) were treated with aspiration of haemarthrosis and cast immobilisation. Open reduction and internal fixation (ORIF) was reserved for type III injuries only (n = 8). Sixteen of the seventeen type II injuries had an excellent outcome, with the remaining one reporting a good outcome. Interestingly, only five of the eight type III injuries were reported to have an excellent outcome at average follow up of 3 years (range 0.5–20 years). These findings were confirmed by a follow up report just over a decade later by the same authors [26], when they once again demonstrated an excellent outcome with type II injuries treated with cast immobilisation (23 out of 24), and lead them to conclude that all type II injuries can be treated with closed reduction and casting. Despite this long understanding, none operative management has theoretically been associated with non anatomical reduction causing non or malunion, ACL instability and loss of knee range of motion.

Baxter and Wiley [23] reported on 13 type II injuries from a total of 42 patients in their study. Five of these were treated with cast immobilisation, and at follow up (between 3 and 10 years), there was no clinical difference in ACL laxity between the injured and the normal knee. Seven type II injuries were treated with closed reduction and one with open reduction. In both treatment modalities there was a mean difference in anterior translation of the tibia on the femur of 3.5 mm, but this was not associated with clinical instability. Eight of the thirteen (62 %) patients with type II injuries had more than 10° loss of extension, but interestingly the prevalence was greater in type III injuries (81 %), even in those that had an open reduction (52 %) or a closed reduction (41 %). Edmonds et al. [27] compared ORIF, arthroscopic reduction and internal fixation (ARIF) and closed reduction and casting in displaced type II and III injuries. They found 16.7 % of fractures that were treated with closed reduction and casting required subsequent operation for loose bodies, instability and impingement. This subset of patients that required surgery after an initial period of none operative management had a mean displacement of 6.7 mm, which led Edmonds et al. [27] to conclude that displacement of less than 5 mm may be a safe cut-off for non-operative management and a greater displacement is an indication for surgical reduction and fixation. Unfortunately, the small number of subjects involved in this subgroups analysis makes it difficult to extrapolate these results, and there is no indication from Edmonds et al. of how many of these injuries were type II and how many were not.

Another argument for surgical treatment of type II injuries is to remove any soft tissue interposition which could prevent complete fracture reduction. Falstie-Jensen and Søndergård Peterson [28] reported on four cases of meniscal incarceration, which required arthroscopic reduction, three of the four occurring in type II injuries. Other studies have reported rates of soft tissue interposition of between 32 % and 100 % [27, 29] in these injuries. Tudisco et al. [22] argue that accurate initial reduction is the key to a good prognosis, and advocate all type II injuries be treated with surgical reduction and fixation. In their study of 14 patients with mean follow up of 29 years (range 12–42), 3 (21 %) had type II injuries. Two were treated with cast immobilisation alone, and one was treated with ARIF. Post operative care included immediate weight bearing and continual passive motion for 6–10 hrs a day for the first 3 weeks. At follow up all had a full range of motion, but the two treated with cast immobilisation did have signs of clinical instability as demonstrated by the KT-1000 arthrometer. The small numbers in this study preclude any valid conclusions. Gans et al. [30] reported a total of ten non unions in 580 patients, 80 % of which occurred in type II (n = 2) and type III (n = 6) injuries that were treated non-operatively. Although small, the risk of non union seems to be more common in type II injuries treated non operatively than with surgery, and this might support the argument for operative fixation. In addition, they reported a prevalence of knee laxity of 22.2 % as demonstrated by positive anterior draw and Lachman tests, and 7.8 % as demonstrated by positive pivot shift test in subjects that had sustained type I or II injuries. This suggests that a proportion of type II injuries can have residual laxity which might be prevented by surgical fixation. This would need to be explored further through the

use of controlled trials. What is not in doubt is the significant difference in these measures of laxity that are seen in type III and type IV injuries. Gans et al. [30] reported these rates to be between 40 % and 60 %.

Is There a Difference in Outcome Between Arthroscopic and Open Surgery?

The study by McLennan [31] was one of the first published on the use of arthroscopy in the treatment of these injuries. He reported on 35 type III injuries, and used arthroscopy to reduce the fracture fragments which were then held with either cast immobilisation or percutaneous K-wires. Patients with isolated tibial spine fractures treated this way were able to return to pre injury function between 3 and 4 months after surgery. McLennan claimed that arthroscopic assisted reduction helped to reduce hospital stay and rehabilitation time, but his study had no control group to compare with. Since then, other authors have gone on to show that arthroscopic surgery for type II-IV injuries can achieve good outcomes including functional knee scores, range of motion and low rates of non union [32].

Edmonds et al. [27] reviewed the results of ORIF, ARIF and closed reduction and casting in displaced type II and III injuries in 76 children over an 8-year period. 29 children had ORIF, 19 of whom were converted from ARIF to ORIF. 28 children underwent successful ARIF and 19 had closed reduction and casting. They reported a significant difference ($p < 0.001$) in the ability to reduce the fracture fragment between the operative and non operative arms, but not between ORIF and ARIF. They also reported a similar incidence of extension lag greater than 10° between ORIF (11.1 %) and ARIF (12.5 %), and concluded that for displaced tibial spine fractures, both methods yielded excellent reduction of the fracture with no difference in long-term outcomes.

Gans et al. [30] demonstrated greater rates of loss of short term extension at the knee after arthroscopic reduction and fixation as compared with open reduction, with a rate of 9.4 % in the ARIF group versus 0 % in the ORIF group. However, they reported little difference in rates of arthrofibrosis (defined as persistent loss of 25° flexion or 10° extension 3 months after treatment and in the absence of non union, malunion, meniscal injury or bony deformity); 3.6 % in the ARIF group versus 2.7 % in the ORIF group.

Is Screw Fixation Better than Fixation with Sutures?

Laboratory based animal studies have suggested that wire fixation is stronger than fixation with screws in anterior tibial spine fractures (Fig. 15.1). Anderson et al. [33] found in skeletally immature porcine knees using physeal sparing fixation, screw fixation with cannulated 3.5 mm screws was associated with the lowest median peak failure loads (186.4 N) compared to ultra high molecular weight polyethelene (UHMWPE) suture to suture buttons (465.8 N), fibrewire suture anchors (440.5 N) and polydiaoxanone (PDS) suture buttons (404.3 N). During cyclical loading they reported percentage survival least with screws (0 %), then suture anchors and PDS buttons (78 %) and most with the UHWPE buttons (100 %). The UHMWPE button fixation demonstrated a significantly higher median peak failure load after cyclic testing than the PDS button and screw fixation. They concluded in their animal study, that physeal sparing fixation of tibial eminence fractures with UHMWPE suture buttons is biomechanically superior to both PDS suture buttons and a single screw at the time of surgery, and provides more consistent fixation than do fibrewire suture anchors.

Eggers et al. [34] found similar results in their study also on porcine knees comparing one screw, two screws, fibrewire sutures and Ethibond sutures through single and cyclic loading tests. They reported the lowest maximum load in the single cycle loading test to be in the group fixed with two 3.5 mm cannulated screws (303.4 N) and the highest with fibrewire fixation (599.6 N). After 1000 cycles of loading, the maximum load of the fibrewire fixation (582.3 N) was significantly higher than the maximum load of fixation with Ethibond (399.3 N), fixation with one screw (457.1 N), and fixation with two screws (455.2 N). They concluded that under cyclic loading conditions, suture fixation of tibial eminence fractures provides more fixation strength than screw fixation, and that a second screw has no positive effect on the biomechanical characteristics of screw fixation.

With respect to clinical signs, Gans et al. [30] reviewed studies that had used screws against those that had uses sutures and found screw usage as compared with sutures was associated with higher rates of short term loss of extension (8.3 % vs. 0 %) but less arthrofibrosis (0 % vs. 6.3 %). They also reported that screws were associated with considerably more laxity as demonstrated by positive anterior draw and Lachman's tests (82.4 % vs. 18.8 %), but almost equal positive pivot shifts (11.8 % vs. 9.4 %). From these studies we can conclude that the animal models suggest that suture fixations are stronger than those with screws, but in clinical trials sutures are associated with more long term joint stiffness.

What Factors Reduce the Risk of Arthrofibrosis of the Knee Joint Following Injury or Surgery?

Baxter and Wiley [23] found in their study that anatomical reduction alone is not protective against loss of range of motion, and there seems to be little difference in outcome

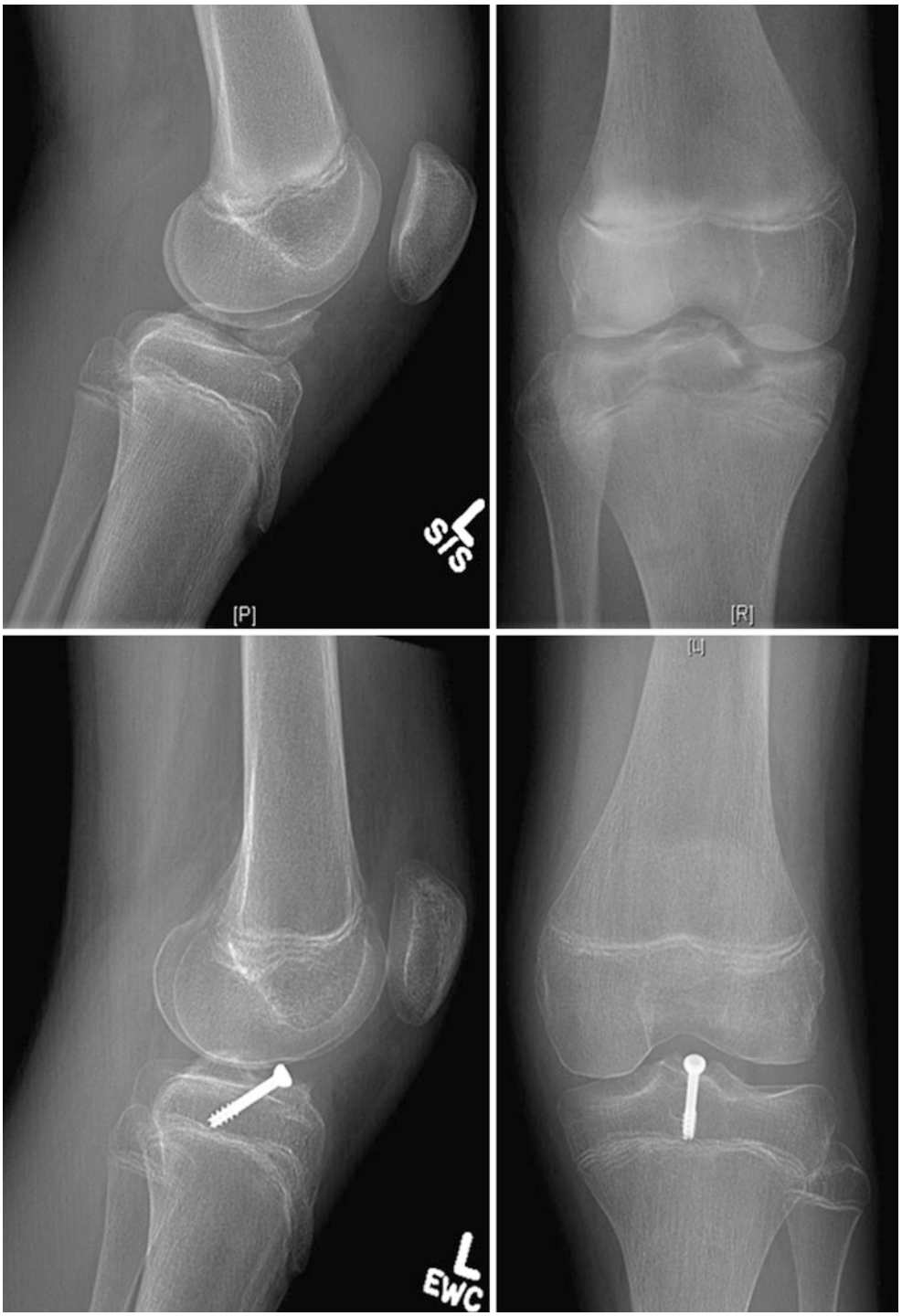

Fig. 15.1 Anterior tibial spine fracture treated with arthroscopic screw fixation

between open and closed methods of reduction. Of interest, they reported an 81 % prevalence of loss of range of motion in type III injuries as compared with 62 % in type II injuries. This supports the hypothesis, that the mechanism and energy of injury and associated soft tissue trauma has an important role to play in any subsequent joint stiffness. Parikh et al. [35] compared ARIF with epiphyseal (n = 12) versus transphyseal (n = 9) screws in 21 patients. They defined arthrofibrosis as loss of 5° of extension or 15° of flexion as compared with the normal knee, which has been described previously [36]. Eight of twelve patients (67 %) with epiphyseal screws had a pain free full range of movement at 3 months. Three of

the remaining four had removal of metalwork which resolved their symptoms. In the transphyseal group all screws were removed 3 months post surgery. Five of nine subjects (56 %) reported a pain free full range of movement at this time, and the remaining four regained full range of motion after the second procedure. Rehabilitation for both groups was dependent on post operative immobilisation. Those in casts (epiphyseal n = 6; transphyseal n = 6) started range of motion exercises after 4 weeks when the cast was removed (median time = 26 days). Patients treated with knee braces (epiphyseal n = 6; transphyseal n = 2) started exercises 1 week post operatively (median time = 7 days). Parikh et al. [35] reported no difference in final range of knee motion between the two groups, and concluded that a regime of aggressive post operative physiotherapy and repeat arthroscopy can successfully prevent loss of knee joint motion. These findings support early work on these injuries in which prolonged periods of immobilisation, sometimes up to 6 weeks in cast, were not associated with loss of joint motion [24].

The largest case series of arthrofibrosis following treatment is the study of Vander Have et al. [20]. Their study reviewed 32 children with type II (22 %) and type III (78 %) injuries. 88 % of all injuries were treated with ARIF with either transphyseal screws (n = 20) or wires (n = 8). Three of thirty-two (9 %) were treated with ORIF and one with MUA and K-wires. Following surgery 88 % were immobilised in full extension for 4–6 weeks, and 22 % were put into a knee brace and started gradual range of motion over 4–6 weeks. Twenty four of the thirty-two (75 %) required reoperation in the form of arthroscopy and adhesiolysis due to loss of full extension, full flexion, or both. All three patients that had ORIF lacked both full extension and flexion. At follow up between 6 and 24 months, 91 % of patients had regained range of motion within 5° of the unaffected knee. Of the three that did not regain range of motion, two had sustained fractures following MUA and one patient had been treated initially with ORIF. Although this study was unable to identify any causal factors to prevent arthrofibrosis, they did highlight the importance of avoiding MUA alone as a treatment for joint stiffness due to the high risk of complications.

In their meta-analysis, Gans et al. [30] identified nine studies which reported extension contractures greater than 5° or flexion contractures greater than 15°, which they termed loss of range of motion. They reported lower rates of loss of range of motion in type I/II injuries (18.2 %) as compared with type III/IV injuries (27.8 %). A similar trend was found for arthrofibrosis between the two groups (7.1 % in type I/II vs. 14.2 % in type III/IV), and confirms earlier findings that the severity of injury probably dictates the level of joint stiffness seen. More recently Watts et al. [37] reported rates of arthrofibrosis of 33.3 % in patients undergoing ARIF compared with 7.7 % in those having ORIF. They correlated these findings to time from injury to surgery and intraoperative time, and concluded that a delay of more than 7 days from injury to surgery, and an intraoperative time greater than 120 min were significant risk factors for arthrofibrosis.

References

1. Anderson M, Messner MB, Green WT. Distribution of lengths of the normal femur and tibia in children from one to eighteen years of age. J Bone Joint Surg Am. 1964;46:1197–202.
2. Aitken AP, Magill HK. Fractures involving the distal femoral epiphyseal cartilage. J Bone Joint Surg Am. 1952;34-A(1):96–108.
3. Eid AM, Hafez MA. Traumatic injuries of the distal femoral physis. Retrospective study on 151 cases. Injury. 2002;33(3):251–5.
4. Czitrom AA, Pritzker KP. Simple bone cyst causing collapse of the articular surface of the femoral head and incongruity of the hip joint. A case report. J Bone Joint Surg Am. 1980;62(5):842–5.
5. Arkader A et al. Predicting the outcome of physeal fractures of the distal femur. J Pediatr Orthop. 2007;27(6):703–8.
6. Salter R, Harris W. Injuries involving the epiphyseal plate. J Bone Joint Surg Am. 1963;45A:587–622.
7. Neer CS. Separation of the lower femoral epiphysis. Am J Surg. 1960;99(5):756–61.
8. Ilharreborde B et al. Long-term prognosis of Salter–Harris type 2 injuries of the distal femoral physis. J Pediatr Orthop B. 2006;15(6):433–8.
9. Wall EJ, May MM. Growth plate fractures of the distal femur. J Pediatr Orthop. 2012;32:S40–6.
10. Stephens DC, Louis E, Louis DS. Traumatic separation of the distal femoral epiphyseal cartilage plate. J Bone Joint Surg Am. 1974;56(7):1383–90.
11. Edmunds I, Nade S. Injuries of the distal femoral growth plate and epiphysis: should open reduction be performed? ANZ J Surg. 1993;63(3):195–9.
12. Lombardo SJ, Harvey Jr JP. Fractures of the distal femoral epiphyses. Factors influencing prognosis: a review of thirty-four cases. J Bone Joint Surg Am. 1977;59(6):742–51.
13. Thomson JD, Stricker SJ, Williams MM. Fractures of the distal femoral epiphyseal plate. J Pediatr Orthop. 1995;15(4):474–8.
14. Basener CJ, Mehlman CT, DiPasquale TG. Growth disturbance after distal femoral growth plate fractures in children: a meta-analysis. J Orthop Trauma. 2009;23(9):663–7.
15. Garrett BR, Hoffman EB, Carrara H. The effect of percutaneous pin fixation in the treatment of distal femoral physeal fractures. J Bone Joint Surg. 2011;93-B(5):689–94.
16. Riseborough EJ, Barrett IR, Shapiro F. Growth disturbances following distal femoral physeal fracture-separations. J Bone Joint Surg Am. 1983;65(7):885–93.
17. Andersen JW, Mejdahl S. Bilateral fracture of the tibial spine. Acta Orthop Belg. 1993;59(4):394–7.
18. Eiskjaer S, Larsen ST, Schmidt MB. The significance of hemarthrosis of the knee in children. Arch Orthop Trauma Surg. 1988;107(2):96–8.
19. Meyers MH, Mc KF. Fracture of the intercondylar eminence of the tibia. J Bone Joint Surg Am. 1959;41-A(2):209–20. Discussion 220–2.
20. Vander Have KL et al. Arthrofibrosis after surgical fixation of tibial eminence fractures in children and adolescents. Am J Sports Med. 2012;38(2):298–301.
21. Zionts LE. Fractures around the knee in children. J Am Acad Orthop Surg. 2002;10(5):345–55.

22. Tudisco C et al. Intercondylar eminence avulsion fracture in children: long-term follow-up of 14 cases at the end of skeletal growth. J Pediatr Orthop B. 2010;19(5):403–8.
23. Baxter MP, Wiley JJ. Fractures of the tibial spine in children. An evaluation of knee stability. J Bone Joint Surg Br. 1988;70(2): 228–30.
24. Zaricznyj B. Avulsion fracture of the tibial eminence: treatment by open reduction and pinning. J Bone Joint Surg Am. 1977;59(8): 1111–4.
25. Noyes FR, DeLucas JL, Torvik PJ. Biomechanics of anterior cruciate ligament failure: an analysis of strain-rate sensitivity and mechanisms of failure in primates. J Bone Joint Surg Am. 1974;56(2):236–53.
26. Meyers MH, McKeever FM. Fracture of the intercondylar eminence of the tibia. J Bone Joint Surg Am. 1970;52(8):1677–84.
27. Edmonds EW et al. Results of displaced pediatric tibial spine fractures: a comparison between open, arthroscopic, and closed management. J Pediatr Orthop. 2014;35(7):651–6.
28. Falstie-Jensen S, Sondergard Petersen PE. Incarceration of the meniscus in fractures of the intercondylar eminence of the tibia in children. Injury. 1984;15(4):236–8.
29. Senekovic V, Veselko M. Anterograde arthroscopic fixation of avulsion fractures of the tibial eminence with a cannulated screw: five-year results. Arthroscopy. 2003;19(1):54–61.
30. Gans I, Baldwin KD, Ganley TJ. Treatment and management outcomes of tibial eminence fractures in pediatric patients: a systematic review. Am J Sports Med. 2014;42(7):1743–50.
31. McLennan JG. The role of arthroscopic surgery in the treatment of fractures of the intercondylar eminence of the tibia. J Bone Joint Surg Br. 1982;64(4):477–80.
32. Furlan D et al. Pediatric tibial eminence fractures: arthroscopic treatment using K-wire. Scand J Surg. 2010;99(1):38–44.
33. Anderson CN et al. Biomechanical evaluation of physeal-sparing fixation methods in tibial eminence fractures. Am J Sports Med. 2013;41(7):1586–94.
34. Eggers AK et al. Biomechanical evaluation of different fixation methods for tibial eminence fractures. Am J Sports Med. 2006;35(3):404–10.
35. Parikh SN, Myer D, Eismann EA. Prevention of arthrofibrosis after arthroscopic screw fixation of tibial spine fracture in children and adolescents. Orthopedics. 2014;37(1):e58–65.
36. Nwachukwu BU et al. Arthrofibrosis after anterior cruciate ligament reconstruction in children and adolescents. J Pediatr Orthop. 2011;31(8):811–7.
37. Watts CD, Larson AN, Milbrandt TA. Open versus arthroscopic reduction for tibial eminence fracture fixation in children. J Pediatr Orthop. 2015:1.

Part IV

The Foot and Ankle

Evidence-Based Treatment for Clubfoot

16

Munier Hossain and Naomi Davis

Abstract

The management of clubfoot has been the subject of active research interest for some time and there is a large body of evidence supporting its treatment strategy. The spectrum of evidence includes randomised controlled trials (RCT), long term prospective follow up as well as systematic reviews including a recent Cochrane review. The actual quality of evidence does vary and there are no real level I studies. Besides, evidence was not always available for the most pertinent clinical questions. However, most published evidence consistently supported the superiority of the Ponseti technique for treating idiopathic clubfoot. Evidence was less clear for management of the complex idiopathic type or the non-idiopathic type clubfoot. Since the publication of results from Ponseti's team this technique has been adopted widely and results published from numerous centres from as far afield as Brazil to Bangladesh. On the basis of available publications it is estimated that the Ponseti technique is in use in at least 113 countries around the world Shabtai et al. (World J Orthop 5:585–590, 2014). The basic principle of the Ponseti technique is well established although minor variations have been attempted by different researchers. In the ensuing paragraphs we have reviewed the evidence base for management of clubfoot recommendations.

Keywords

Clubfoot • Congenital talipes equinovarus • Pirani score • Ponseti method • Kite method • French method • Tendoachilles tenotomy • Foot abduction brace • Boots and bar • Tibialis anterior tendon transfer

Background

Clubfoot, also known congenital talipes equinovarus (CTEV), is a common congenital limb deformity with an incidence of 1/1000. It involves both feet in 50 % of patients and boys are more affected than girls (3/2). It is characterised by a tight tendoachilles (equinus), an excessively turned in foot (varus) and high medial longitudinal arch (cavus) (Fig. 16.1), which if left untreated leads to long-term functional disability, deformity and pain [2].

Clubfoot can be:

- Idiopathic clubfoot which presents as an isolated anomaly in a normal child.
- Non-idiopathic clubfoot which is associated with other neuromuscular or congenital anomalies
- Complex or atypical idiopathic clubfoot which is defined as having rigid equinus, severe plantar flexion of all metatarsals, a deep crease above the heel, a transverse crease in the sole of the foot, and a short and hyperextended first toe.

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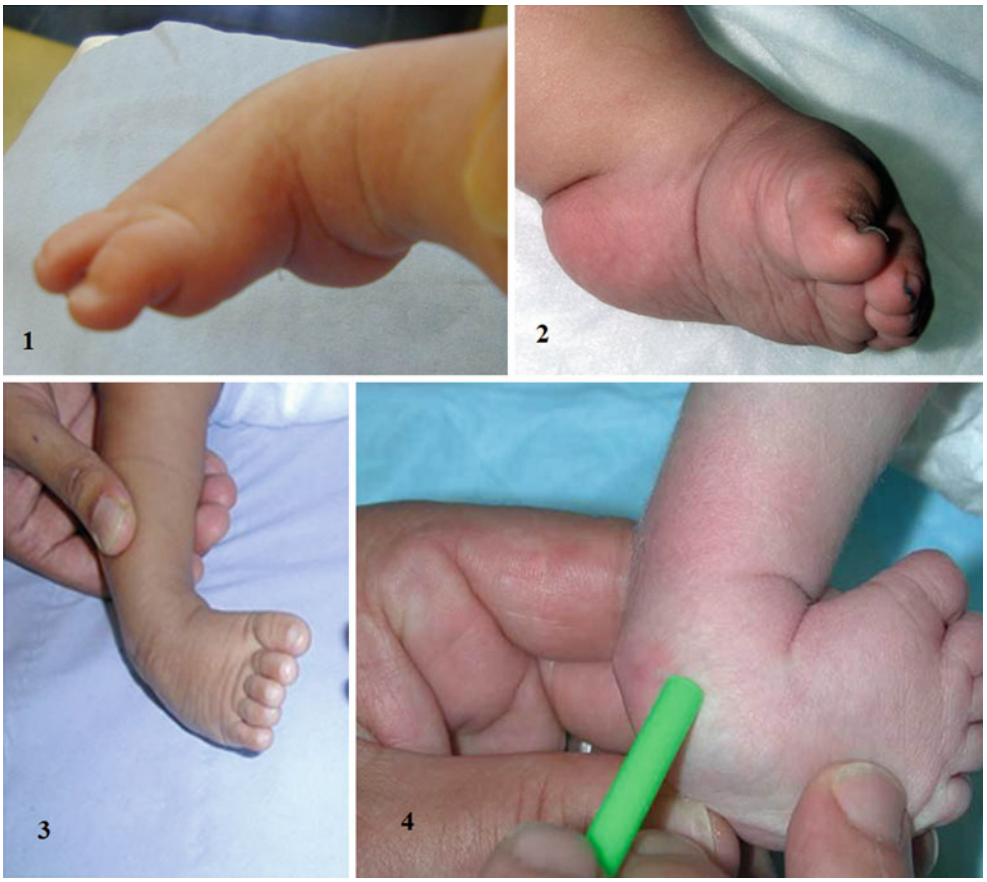

Fig. 16.1 Clinical photographs showing classical deformity of a clubfoot

Idiopathic clubfoot responds well to treatment but non-idiopathic or complex clubfoot may be amenable. Most of the evidence for management of clubfoot is available for the idiopathic type. Treatment options for the non-idiopathic or the complex idiopathic clubfoot are discussed separately in later sections.

What Is the Best Treatment for Idiopathic Clubfoot?

Management of clubfoot can be broadly divided into non operative (Ponseti, Kite, French methods) and operative types. Although several randomised controlled trials (RCT), cohorts' studies, case series have investigated various aspects of clubfoot treatments; most have limitations which leave some uncertainty about certain aspects of the treatments. In this section we endeavoured to simplify the evidence for each treatment without dismissing the limitations of the best available evidence we currently have.

Over the last 20 years, Ponseti's serial casting treatment for clubfoot has emerged as a superior treatment to other non operative and operative treatments. This has been supported by evidence from a systematic review and meta-analysis [3]; RCTs [4–6] and long term cohorts studies [7–10]. Gray [3] conducted a Cochrane review of clubfoot treatment including 14 trials (607 participants) and concluded that The Ponseti's Methods produced significantly better short term foot alignment compared to other non operative methods including the Kite methods. Relapse following the Kite's methods more often led to major surgery compared to relapse following The Ponseti's Methods.

Sud [5] conducted an RCT comparing The Ponseti Methods and Kite methods. Baseline Dimeglio scores were similar between the two groups. After an average follow-up of 27 months in the Ponseti group, correction was achieved in 33 feet (91.7 %), with only three patients requiring surgical management. There were seven relapses (21.1 %), all of which were corrected conservatively. In the Kite group, correction was achieved in 21 feet (67.7 %) after an average follow-up of 25 months, with 10 patients requiring surgical intervention. There were eight relapses of which only four could be corrected conservatively.

Sanghvi [4] compared the long-term results of the Kite and Ponseti methods of manipulation and serial casting for clubfoot in 42 patients (with 64 idiopathic clubfeet) who

were randomised to either Kite or Ponseti treatments in the early weeks of life. All the clubfeet were manipulated, casted, and followed up (for a mean of 3 years) by one experienced orthopaedic surgeon. The success rate in The Kite Method was 79 % and that in the Ponseti's method was 87 %. With The Ponseti method, the number of casts was significantly fewer (7 vs. 10); the duration of casting required to achieve full correction was significantly shorter (10 vs. 13 weeks); the maximum ankle dorsiflexion achieved was significantly greater (12° vs. 6°); and the incidence of residual deformity and recurrence was slightly lower. The study was not clear if the two groups were comparable at baseline.

Rijal [6] randomly allocated 60 feet in 38 patients to either Ponseti (30 feet) or Kite (30 feet) methods. Feet were followed up weekly for 10 weeks for change of cast and recording of hindfoot, midfoot and total Pirani scores. Correction was measured as a difference between hindfoot, mid foot and total Pirani scores weekly from weeks 1 to 10 and corresponding baseline scores. Mean Pirani scores in Ponseti feet improved much faster than Kite feet but the difference achieved statistical significance only at the 10(th) week from the start of treatment.

Richards [11, 12] compared the results of Ponseti and French methods in 386 feet in 256 children under the age of 3 months at presentation. Treatment allocation was on the basis of parental choice after the pros and cons of both techniques had been explained. Mean follow up was 4.3 years. The two groups had similar grades of severity scores before treatment. The initial correction rates were 94.4 % for the Ponseti method and 95 % for the French functional method. Relapses occurred in 37 % of the feet that had initially been successfully treated with the Ponseti method. One-third of the relapsed feet were salvaged with further non operative treatment, but the remainder required operative intervention. Relapses occurred in 29 % of the feet that had been successfully treated with the French functional method, and all required operative intervention. At the time of the latest follow-up, the outcomes for the feet treated with the Ponseti method were good for 72 %, fair for 12 %, and poor for 16 %. The outcomes for the feet treated with the French functional method were good for 67 %, fair for 17 %, and poor for 16 %.

Faulks and Gottschalk [13, 14] compared gait analysis of patients treated by the Ponseti method and feet treated by the French method. Normal ankle motion was documented more frequently in the Ponseti feet compared with the French methods. More of the children treated with the French method walked with knee hyperextension, a mild equinus gait, and mild foot drop.

Jeans [15, 16] assessed plantar pressures in feet treated with the Ponseti and the French methods at age 5 years. Twenty controls were used for comparison. Data from 164 patients (238 feet; 122 Ponseti and 116 French) showed no significant differences between the two methods, except the French method feet had a significantly less medial movement of the centre of pressure (COP) than the Ponseti feet ($P = 0.0379$). This shows that there is a mild residual deformity in these feet despite clinically successful outcomes.

Zwick [17] conducted an RCT of Ponseti versus surgical treatment. All babies were less than 2 weeks old on initiation of treatment. Surgical group (16 feet) received initial manipulation and weekly casting until 6–8 months of age followed by posteromedial release by the Cincinnati incision. Ponseti group (12 feet) received standard Ponseti treatment. Two patients from the Ponseti group crossed over to the surgical group. The authors used the Functional rating system (FRS) devised by Laaveg and Ponseti [18] as the outcome measure. The authors used a priori sample size calculation but were compelled to stop the trial for ethical reasons after interim analysis revealed that the surgical group had worse outcome. Minimum follow up was 3.3 years. The groups had similar baseline Pirani score. At final follow up the FRS score as well as the passive ankle range of motion was better in the Ponseti group. These results are supported by other long term follow up studies. Smith [9] performed a retrospective comparison of surgery (37 feet) with the Ponseti method (29 feet). Both groups were compared with healthy young adults. The authors found that compared to the control subjects both groups had reduced strength and motion of the ankle. However, the Ponseti group had better ankle motion and less pain. Similarly Graf [19] assessed the long term results of surgical release in 24 adults who underwent posteromedial release via the Cincinnati incision before the age of 18 months. The authors found that although surgery resulted in a plantigrade foot; pain, stiffness and weakness gave functional limitations. Dobbs [20] assessed the long term function of 73 feet treated with extensive soft tissue surgery. Mean follow up was 30 years. The authors found that 47 % patients had poor long term foot function as assessed by the Laaveg-Ponseti functional score. This unfavourably compared to Ponseti's long term follow up results where only 12 % patients had poor results using the same outcome instrument. In addition Dobbs et al found functional limitation to be inversely correlated to the extent of soft tissue surgery.

What Is the Best Treatment for a Late Presenting Clubfoot?

Patients who present after walking age are considered to have presented late. A number of authors from the developing countries have tried Ponseti technique in older children with varying degrees of success. In Lourenco's [21] series from Brazil, 16 feet were graded as functionally good at a mean follow up of 3.1 years. The mean age was 3.9 years at

Table 16.1 Outcome of treatment for late presenting idiopathic clubfoot

Studies	N (Feet)	Mean age in years	Follow-up in years	Cast required Mean (range)	Relapse	Surgical release
Lourenco et al. [21]	24	3.9 (1.2–9)	3.1 (2.1–5.6)	9 (7–12)	7	8
Khan et al. [22]	25	8.9 (7.5–11)	4.7	12 (10–14)	6	?
Yagmurlu et al. [23]	31	2 (1–6)	3.5	6 (4–8)	0	0
Banskota et al. [24]	55	7.4 (5–10)	2.5 (2–3.3)	9.5 (6–11)	9	28
Ayana et al. [25]	32	4.4 (2–10)	3 (2–4)	8 (6–10)	4	6

presentation. Relapses were treated with a repeat TAT but none required tibialis anterior tendon transfer (TATT). The authors suggested longer time for manipulation and less frequent cast changes to allow for remodelling, less abduction in brace and Ankle foot orthosis (AFO) instead of boots and bar brace in the older child (Table 16.1).

Khan et al [22] evaluated the efficacy of Ponseti technique in 21 children (25 feet) over 7 years of age. Eighteen feet had a full correction (85.7 %). Dimeglio score improved from 14.2 to 0.18 at final follow up.

Yagmurlu et al [23] published their results of Ponseti treatment in 27 patients (31 feet) who were treated after the walking age. Standard Ponseti technique was used. Seventeen feet required open tenotomy. The authors noticed a significant improvement in all components of the deformity but found that children older than 20 months tended to improve less.

Banskota et al [24] presented a series of 36 patients aged 5–10 on presentation. Mean Pirani score at presentation was 5.1 (3–6). The authors did not modify their standard technique and a higher number of patients in their series underwent surgery. However, 46 feet achieved a plantigrade feet without extensive soft tissue surgery. Twenty-seven children were completely satisfied with the treatment. The authors suggested that the Dimeglio scoring system was more appropriate to assess the feet of older children.

Ayana [25] from Ethiopia prospectively evaluated 22 children aged 2–10 years (32 feet) with neglected clubfoot. All were treated by the Ponseti method. The deformity was assessed using the Pirani scoring system. The average follow-up time was 3 years. A plantigrade functional foot was obtained in all patients. Two patients (four feet) had recurrent deformity. They required re-manipulation and re-tenotomy of the Achilles tendon and one other patient required TATT for dynamic supination deformity of the foot.

Singh [26, 27] presented the results of ligamentotaxis using Joshi's External Stabilisation system (JESS) in 20 patients. This is a technique of controlled differential distraction using a mini external fixation. The case series included late presenters as well as relapses and incomplete corrections following previous conservative treatment. All patients achieved good clinical results as per Pirani score and authors concluded that differential distraction by fixator was an effective and patient-friendly method of management neglected clubfoot.

When Is the Best Time to Start Treatment?

There is a general impression that Ponseti manipulation should begin in the "first week of life" [28]. However Iltar [29] found that infants in whom correction was commenced after the first month had a better Dimeglio score than those in whom the correction was commenced earlier. There were 29 patients (40 feet) with idiopathic clubfoot in the series. Eighteen patients (26 feet) had Ponseti treatment within the first month of life and the rest had treatment commenced after the first month. Median follow up was 34 months. There was no significant difference in initial Pirani and Dimeglio scores between the two groups. Both groups demonstrated significant improvement following treatment and there was a marginal difference only in Dimeglio score following treatment between the two groups. The authors suggested that the Ponseti treatment should be commenced after the first month of life or when the involved foot is ≥8 cm in length. This was a retrospective study with no control. Although the difference was statistically significant, its clinical value is doubtful. In a recent meeting of the First European consensus meeting the assembled experts agreed that "treatment for clubfoot should start not later than within the first month of life" with the caveat to defer treatment in a premature baby for several weeks to allow for the baby's foot to grow in size [30].

What Type of Casting Material Should Be Used?

Pittner [31] conducted a trial to compare the effectiveness of plaster of Paris (POP) to semi rigid fibreglass (SRF) in the management of clubfoot. Several methodological flaws have been noted in the study regarding sample size calculation, allocation, concealment and randomisation. Thirteen patients (16 feet) received SRF and 18 patients (23 feet) received POP. The authors used Dimeglio score to describe severity of clubfeet deformity. There was no difference in initial score between the two groups. Although similar number of casts were required in both groups the Dimeglio score was significantly higher in the SRF group on completion of treatment (6.4 vs. 4.1; P = 0.037). There was no difference in the rate of tendoachilles tenotomy between the two groups but two

patients in SRF group required surgical soft tissue release. The follow up period is unclear and the authors did not describe relapse rate. However they found that parents preferred SRF over POP.

Hui [32] performed a randomised trial to compare time to clubfoot correction between POP and SRF. They performed a priori sample size calculation and used sealed envelope and computerised randomisation. There were 12 patients (18 feet) in the POP group and 18 patients (26 feet) in the SRF group. There was no difference in Pirani score between the two groups (5.3 SRF, 4.9 POP). There was no significant difference in the number of casts required between the two groups. However relapse was higher in the SRF group (47 %) compared to the POP group (25 %). Need for repeat Ponseti casting and surgery was also higher in the SRF group. However, these were not statistically significant and the authors felt that the higher relapse rate in the SRF group could be due to slightly longer follow up (36 months vs. 24 months for POP group) as well as less compliance with foot abduction orthosis wear in the SRF group (71 % vs. 92 % in the POP group). The authors also noted that parents preferred SRF over POP.

It is not possible to perform a summary estimate of the two trials as they used two different rating systems and it is unclear if the population was similar, the follow up rates were clearly dissimilar and the reported primary outcomes were different. However, it appears that although POP may give better results parents tend to prefer SRF.

Should the Cast Be Applied Below or Above the Knee?

Brewster et al [33] produced a case series of 51 babies with 80 idiopathic type clubfoot treated with a modified Ponseti technique; instead of traditional plaster casts they used below the knee soft casts. Mean follow up was 27 months and mean time to TAT was 8.5 weeks. Twenty-five percent patients did not require TA tenotomy in their series and relapse rate was 6.27 %. There was a single case of cast slippage. The authors felt that their results were comparable to traditional above knee plaster cast technique.

Subsequently Maripuri et al [34] conducted an RCT of below versus above knee casting using the Ponseti technique to compare treatment times and failure rates between the two different casts. There were 17 feet in the above-knee group and 16 feet in the below knee group. Patients were randomised using opaque sealed envelope. Their primary outcome measure was time to readiness for tendo-achilles (TA) tenotomy or achievement of full correction. Two or more episodes of plaster slip or more than 8 weeks of manipulation requirement was defined at treatment failure. They undertook a priori sample size calculation and required around 30 feet in each arm of the trial. However, the authors felt compelled to stop the trial early when interim analysis showed a 37.5 % failure rate in the below-knee group. The authors concluded that below knee cast was not suitable for the Ponseti technique. Given the size of these two studies, contradicting results is not a surprise. Moreover, both studies have flaws. Experts warn against early termination of trials on the basis of interim estimation and show with simulation studies that small trials stopped early with few events are likely to result in a large overestimation of treatment effects. Moreover, the techniques may be different between the two studies. Brewster [33] used soft cast and described careful molding above the heel to prevent cast slippage. Maripuri [34] did not describe their technique of cast application. Our recommendation is to use above knee cast. The potential benefits of having a shorter cast may not outweigh the risk of failure associated with uncertainties about below the knee cast.

How Frequently Should Casts Be Changed?

Ponseti originally described weekly manipulations to correct clubfoot deformity [28]. His team subsequently investigated if cast treatment could be successfully shortened due to an increase in number of patients presenting in their centre, who often travelled a long distance and could benefit from a shortened time to correction [35]. Patients were assigned to 5 or 7 days based solely on geography. Ninety percent of patients required five or fewer casts for correction, and there was no difference between groups (P = 0.85). Average time from first cast to TAT was 16 days for the 5-day group and 24 days for the 7-day group (P = 0.001). Relapse rate for compliant patients was similar in both groups. The authors attempted cast changes more frequently at 3–4 days and found feet to be swollen and oedematous. They concluded that changing casts every 5 days was safe, effective and probably the fastest way to achieve correction with minimal side-effects.

Xu [36] published a similar study from China where patients travelling long distance were assigned to twice weekly cast changes (26 patients, 40 feet) and local Beijing patients were assigned to weekly cast changes (20 patients, 32 feet). All patients had idiopathic clubfeet. The baseline and final Pirani scores were similar between the two groups, both groups required similar number of casts (mean 5) and tenotomy requirement was also similar (87.5 %). The time to correction was 21 days in the accelerated regime and 35 days in the regular regime. Mean follow up was 4 years. There were six relapses in the accelerated group and five relapses in the regular group. Xu et al did not experience swelling or oedema with twice weekly cast changes.

Elgohary et al [37] published a quasi-randomised trial from Egypt. Twenty children (34 feet) were treated with the regular regime and 21 children (32 feet) were treated with the accelerated regime. Patients receiving the accelerated regime had cast changes twice a week. Pirani score at baseline and end of treatment were similar between the two groups. Mean number of casts and tenotomy requirement was also similar. The length of follow up was unclear but they had a higher relapse rate that was not significantly different between the two groups (14.7 % regular group vs. 15.6 % accelerated group). Authors noted that children with relapse had presented with a high Pirani score (≥5). They did not record compliance with bracing following correction. Time to correction was significantly shorter in the accelerated group 18.13 ± 3.02 days (11–22 days) compared to the regular group 33.36 ± 6.69 days (21–42 days).

Harnett et al [38] from Malawi compared a standard Ponseti regime with an accelerated Ponseti regime of thrice weekly cast changes. Following a sample size calculation they recruited 40 patients with idiopathic clubfoot (61 feet) into an RCT. Randomisation was computer generated. Following treatment patients were put in foot abduction brace and were followed up for 6 months. Nineteen patients (29 feet) were allocated to the accelerated protocol and 21 patients (32 feet) were allocated to the standard protocol. There was no significant difference in Pirani score before or after treatment between the two groups. Tenotomy requirement was similar in both groups and both required median of five cast changes. Three patients in the accelerated regime had Pirani score >1 at 21 days after treatment. They were crossed over to the standard treatment, all had tenotomy and one required tibialis anterior tendon transfer. Median number of treatment days was significantly shorter with the accelerated regime (16 days vs. 42 days for the standard regime). Thirty-six patients (55 feet) were followed up for 6 months and there was no relapse. The authors concluded that the accelerated protocol of twice weekly cast changes was equally effective as the standard protocol of weekly cast changes.

Other relevant findings were published by Tarrazas-Lafargue et al [39] which demonstrated that removing the cast the night before cast change was associated with a longer treatment duration. It was a retrospective review of 44 patients (63 clubfeet) had their cast removed the night before to allow for skin care. Forty-six patients (66 clubfeet) had cast removed immediately before change of cast. The immediate removal group required less number of casts for deformity correction (mean 5, range 4–10) compared to the early removal group (ten casts, range 4–22).

In summary: there is a reasonable body of evidence to suggest that cast changes can safely be performed less than weekly without risk of any adverse events. Studies that promoted accelerated regime did not investigate rigorously the cost effectiveness and convenience for parents and healthcare providers. Moreover, it is not advisable to remove casts the night before.

How Many Casts Are Required for Correction or When Should One Consider Treatment to Have Failed?

Ponseti had indicated that on average between five and seven casts could be required for correction of deformity, with the last cast worn for 2–3 weeks [28]. A 30-year follow up study of idiopathic clubfoot treated at Ponseti's centre indicated that between seven and nine casts were required for deformity correction. He wrote that not more than ten casts should be required for correction. Extrapolating data from the RCTs it appears that between four and eight casts may be required for correction of the foot deformity in idiopathic clubfoot. It is notable that in the trials where regular Ponseti regime was compared with the accelerated regime the number of casts required were the same even when the accelerated protocol was employed. A survey of North American surgeons found that median number of casts required for correction was seven. A number of factors may affect how many casts may be required. Dyer and Davis [40] from Manchester found that the number of casts required would depend on the severity of initial deformity as assessed using the Pirani scoring. Time to presentation is also an important factor. The older the patients at presentation the more casting is required [21, 24, 25].

In summary fewer than 8 casts should be required when treatment is started early for idiopathic clubfoot [30]. Late presenters and those with more severe deformity may require more. Casting should be discontinued when a plateau in correction is achieved.

Is Tendoachilles Tenotomy Always Required and When Should This Be Performed?

Tendoachilles tenotomy (TAT) is an integral part of the Ponseti technique [28]. Ponseti described that his technique corrected all elements of the foot deformity simultaneously except equinus deformity which was corrected last and was facilitated by the TAT. He advised that TAT should be performed to correct persistent equinus deformity when the foot was abducted to at least 60° but dorsiflexion was limited to <20°. His initial series had a tenotomy rate of 79 % [41]. Subsequent publications reported up to 100 % rate of tenotomy, especially in older children [42]. Dyer and Davis [40] found that if the hindfoot Pirani score was ≥2.5 then there was a 72 % chance of requiring TAT. However, they commented that a low Pirani score did not rule out the requirement for TAT.

A survey of North American surgeons reported a tenotomy rate of 81 % [43, 44]. Following the widespread validation of Pirani score authors advised that TAT should be performed once mid-foot score was 0. A requirement for TAT is that the sub-talar malalignment is corrected and the calcaneus swings out from underneath talus. It has been suggested that this can be clinically appreciated by palpating the anterior process of the calcaneus [32]. Ponseti advised percutaneous tenotomy under local anaesthesia. Subsequent authors have described the technique using local anaesthesia, light sedation, propofol or general anaesthesia with good results [45].

Is There a Role for Botulinum Toxin?

Alvarez [46] published the 5-year results of 44 patients (65 feet) with idiopathic clubfeet treated with Ponseti method and supplemental Botulinum toxin A (BTX-A) injection [38]. Mean baseline Pirani score was 5.1 ± 1.3. BTX-A was injected at 10 U/Kg onto the Gastrosoleus complex when forefoot was corrected to ≥60° of abduction but hindfoot was still in equinus. Following injection patients were put into above knee casts for 4 weeks with weekly cast changes in between. Patients did not receive TAT. Subsequently they went into foot abduction orthosis. Forty-four patients (65 feet) were followed up at 5 years. Recurrence was defined as ankle dorsiflexion (DF) ≤5° with knee in 90° of DF or a requirement for re-intervention that included repeat injections. Only two patients were non-compliant with foot abduction bracing. Overall recurrence rate was 52 % and 15.4 % required surgery. It does not appear that TAT was performed even though some patients received up to six injections.

A double blind RCT by Cummings [47] compared BTX-A versus placebo in 20 patients (32 feet) with Dimeglio type III clubfoot. Seventeen feet had BTX-A and 15 feet had placebo. The intervention group had BTX-A injected into the gastrosoleus and the tibialis posterior tendons at the beginning of casting. The control group had a placebo injection of human albumin in saline. Both group had standard Ponseti manipulation followed by TAT as required. TAT was performed when deformity correction reached a plateau and was followed by further manipulations. The primary outcomes were time to correction and need for TAT. The mean follow up was 2.3 years. There was no significant difference in time to correction, number of required casts or TAT requirement between the two groups. The TAT rate was low in this study (6/32 feet) and the author also appeared to have modified the technique to perform TAT earlier. Nevertheless, the author concluded that BTX-A when used as an adjunct to the Ponseti treatment did not speed correction, reduce the rate of TAT requirement nor prevent relapse.

In summary: it appears that there is no benefit in using BTX-A as an adjunct to the Ponseti technique.

What Is the Best Treatment for Relapsed Clubfoot?

Ponseti had commented that "regardless of the mode of treatment, the clubfoot has a strong tendency to relapse." There is no agreed classification for a relapse nor does there appear to be any consistent management plan [48, 49]. A survey of North American surgeons found a very wide range of reported relapse rate (mean 22 %, range 0–95 %) and the need for surgical soft tissue release following relapse (mean 7 %, range 0–90 %) [44]. Ponseti's group from Iowa defined relapse as reappearance of any of the components of the clubfoot deformity [42]. Ponseti wrote that of all components of the clubfoot deformity the hindfoot relapse is the most important, first with recurrence of equinus and then of varus. Ponseti's group have recommended repeat casting as the first line of treatment following relapse, followed by repeat TAT. TATT is recommended for dynamic forefoot supination [22].

Bhaskar [50] has proposed a classification scheme for clubfoot relapse that may be useful for planning treatment and comparing results. The authors presented a series of 91 children (164 feet) with relapse. Mean follow up was 4.9 years. They suggested three patterns of increasing severity of relapse:

- Grade I
  - Grade IA Reduced ankle dorsiflexion from 15° to neutral with knee in extension
  - Grade IB Dynamic forefoot adduction of supination of foot (flexible relapse)
- Grade II
  - Grade IIA Fixed equinus of any degree
  - Grade IIB Fixed adduction of forefoot and midfoot (fixed lateral curvature)
- Grade III Two or more fixed deformities

The authors suggested re-casting for group IA relapse. 24/30 feet with group IB relapse were treated with full time bracing (24/30) and 6/30 with TATT. The authors suggested more extensive surgery for rigid deformities although results and rationales were not clearly described.

There is consistent evidence in the literature to suggest that compliance with bracing is the single most important factor in determining relapse and it is not related to the severity of deformity, age of presentation or the number of casts required for treatment [42, 51–53]. Lovell and Morcuende [54]have suggested that patients presenting with late relapse should be investigated for undiagnosed neuromuscular disease. Morin [55] were able to reduce the rate of relapse

simply by improving communication with the family. Others have suggested that longer bracing, improved compliance, earlier identification of relapse and repeat casting can reduce the need for surgery [42].

In summary compliance with bracing is the single important factor predicting relapse following Ponseti treatment. Improved compliance can reduce the rate of relapses. Initial treatment following relapse is repeat casting and bracing but it is likely patients will require TATT±TAL. Long term functional outcome following treatment for a relapse is not known but a minority is likely to experience functional limitation.

What Is the Best Treatment for Complex Idiopathic Clubfoot?

Complex idiopathic clubfoot (CIC) is defined as a clubfoot with rigid equinus, severe plantar flexion of all metatarsals, a deep crease above the heel, a transverse crease in the sole of the foot, and a short and hyperextended first toe [56]. Ponseti et al reviewed the results of 50 patients (75 feet) presenting with such features. Thirty-one patients had previously been treated unsuccessfully, 11 had previous TAT and three had TA lengthening. Mean age at presentation was 3 months. They altered the manipulation technique to take account of the severe ankle equinus and plantar flexion of all metatarsals. There were two elements to the modified technique. First, to correct heel varus the hindfoot was abducted with counter pressure applied to the lateral malleolus in addition to the talar head and secondly, to correct flexed metatarsals and ankle equinus the ankle was held with both hands and all the metatarsals were pushed into dorsiflexion by both thumbs while the knee was stabilised in flexion. Following deformity correction above knee cast was applied with the knee in at least 110° of flexion. All patients required TAT. Mean follow up was 20 months. There was a 14 % relapse rate although the authors did not clarify what constituted "relapse." However, they indicated that all of the patients with relapse had difficulty complying with the foot abduction orthosis. These patients were treated with repeat manipulation and three patients had repeat TAT. Two patients deemed to have complex clubfoot had surgical soft tissue release. Authors commented that it was the cavus and the equinus components of the foot deformity that were the most difficult to correct in these feet. Turco commented in a presentation at the First International Congress on Clubfeet in 1990 that early surgery in these feet resulted in "grotesque overcorrected deformities with severe flatfeet" [57]. However, Turco did not publish his results, nor did he elaborate in his Congress abstract. Therefore the only evidence we have is of the modified Ponseti technique.

There is little evidence to support the treatment strategy of complex idiopathic clubfoot. However, Ponseti's group has shown that the standard technique requires modification while treating these patients. The follow up was not long and the long term functional results, relapses and further interventions are not known.

What Is the Best Treatment for Non-idiopathic Clubfoot?

A number of published case series guide treatment strategy for non-idiopathic clubfoot. Morcuende [58] published a retrospective review of 16 patients (32 feet) with non-idiopathic clubfoot due to arthrogryposis who were treated with the Ponseti technique. The authors did not indicate the severity of the deformity. Eight patients had previous unsuccessful treatment including one case of TAT. Full correction was achieved in 15 patients all of whom required TAT. On average seven casts were required for correction of foot deformity. The uncorrected patient underwent surgical soft tissue release. The authors did not modify their technique for these difficult patients but recommended that the feet should be placed in maximum 50° of abduction rather than the standard 70°. There were four cases of relapse (25 %) and all of them required surgical soft tissue release. Relapse was not related to non-compliance with bracing (Table 16.2).

Boehm [59] investigated the results of Ponseti technique for management of 12 cases (24 feet) of non-idiopathic clubfoot from distal arthrogryposis. Four patients had prior casting, none had previous tenotomy. The authors used the Dimeglio system to grade severity of the foot deformity. Eleven patients had very severe deformity (Dimeglio grade IV), the other patient had grade II deformity. The authors treated eight patients with the standard Ponseti technique and the other four with the modified technique as recommended for management of complex idiopathic clubfoot. All feet were corrected and all underwent TAT. All three cases of relapse in this series were non-complaint with bracing. Two of them were treated with repeat casting and the other patient required soft tissue surgery because of repeat relapse.

Gerlach [60] published a prospective case series of 16 patients (28 feet) with clubfoot and myelomeningocoele. Mean Dimeglio grade was 3.3 and 11 feet were of grade IV severity. Twenty four feet required TAT. A single patient who did not require TAT had a Dimeglio grade II deformity. Eleven patients (19 feet) had a relapse. The authors defined relapse as presence of ≥5° of hindfoot varus and <10° of ankle dorsiflexion. Non-compliance was the supposed cause of relapse in seven feet. Eleven feet had repeat casting, four feet required repeat TAT and four required surgical soft tissue release. Two patients sustained iatrogenic distal tibial fracture during treatment.

Janicki [61] published the results of Ponseti treatment of 23 patients (40 feet) with non idiopathic clubfoot who had either a neuromuscular or a syndromic condition. The authors did not mention severity of the foot deformity. Thirty-six feet were

Table 16.2 Outcome of Ponseti treatment of non-idiopathic clubfoot

Author	N (feet)	Diagnosis	Age (months) Mean (range)	Follow up Mean (range)	Mean cast to correct	Full correction	Relapse	Soft tissue release
Morcuende et al. [58] Retrospective	32	Arthrogryposis	3 (0–12)	4.6 years (10–12)	7 (5–12)	30	8	10
Boehm et al. [59] Retrospective	24	Distal arthrogryposis	3.7 ± 4.2	Minimum 2 years	6.9 ± 2.1	24	6	2
Gerlach et al. [60] Prospective	28	Myelomeningocoele	12.4 weeks (1.2–25.9)	33.8 months (25–42.7)	5 (4.6–5.7)	27	19	4
Janicki et al. [61] Retrospective	40	Neuromuscular or syndromic	11.4 weeks (1–52)	32.6 months (12–56)	6.4 (3–11)	36	16	14

Table 16.3 Summary of recommendations

Clinical situations	Grades
Ponseti technique gives better short term results compared to the Kite technique in the management of idiopathic clubfoot	B
Ponseti technique gives better long term results than surgery in the management of idiopathic clubfoot	B
Both the Ponseti technique and the French technique achieve good initial correction in the management of idiopathic clubfoot	B
Ponseti technique is a valid treatment option for late presenting idiopathic clubfoot	B
There is insufficient evidence to recommend ligamentotaxis as a treatment option for late presenting idiopathic clubfoot	I
Treatment should be started as soon as possible in a term baby	B
Treatment may be delayed in a small feet or in pre-term babies by several weeks to allow the feet to grow	C
Plaster of Paris used as the casting material appears to give better functional results and is easier to mold	C
Cast should be applied above the knee	B
Accelerated Ponseti technique may well be equally effective compared to standard weekly cast change regime	B
There is no benefit in using BTX-A as an adjunct to the Ponseti technique	B
Compliance with bracing is the single most important factor in preventing relapse following treatment with the Ponseti technique	B
There is insufficient evidence for recommending treatment strategy of complex idiopathic or non-idiopathic clubfoot	I

corrected. Twenty-seven feet required TAT. Following relapse nine feet were successfully treated with recasting alone, two feet required repeat TAT and the rest required surgical soft tissue release. Compliance with bracing was not mentioned.

In summary non-idiopathic clubfoot is more rigid and is likely to require more casting than idiopathic clubfoot. It also has a higher relapse rate when treated with the Ponseti technique. Repeat casting including repeat tendoachilles tenotomy is an option in treating relapse but it may necessitate surgical soft tissue release. Long term outcome of treatment is unknown (Table 16.3).

References

1. Shabtai L, Specht SC, Herzenberg JE. Worldwide spread of the Ponseti method for clubfoot. World J Orthop. 2014;5(5):585–90.
2. Ponseti I, Morcuende J, Mosca V, Pirani S, Dietz F, Herzenberg J. Clubfoot: Ponseti management. 2nd ed. Global-HELP Organization; 2005.
3. Gray K et al. Interventions for congenital talipes equinovarus (clubfoot). Cochrane Database Syst Rev. 2014;8:CD008602.
4. Sanghvi AV, Mittal VK. Conservative management of idiopathic clubfoot: Kite versus Ponseti method. J Orthop Surg (Hong Kong). 2009;17(1):67–71.
5. Sud A et al. Ponseti's vs. Kite's method in the treatment of clubfoot–a prospective randomised study. Int Orthop. 2008;32(3):409–13.
6. Rijal R et al. Comparison of Ponseti and Kite's method of treatment for idiopathic clubfoot. Indian J Orthop. 2010;44(2):202–7.
7. Cooper DM, Dietz FR. Treatment of idiopathic clubfoot. A thirty-year follow-up note. J Bone Joint Surg Am. 1995;77(10):1477–89.
8. Ippolito E et al. Long-term comparative results in patients with congenital clubfoot treated with two different protocols. J Bone Joint Surg Am. 2003;85-A(7):1286–94.
9. Smith PA et al. Long-term results of comprehensive clubfoot release versus the Ponseti method: which is better? Clin Orthop Relat Res. 2013;472(4):1281–90.
10. Alshryda S, Wright J. Congenital club foot: the result of treatment, in classic papers in Orthopaedics. London: Springer; 2014. p. 571–3.
11. Richards BS et al. A comparison of two nonoperative methods of idiopathic clubfoot correction: the Ponseti method and the French functional (physiotherapy) method. J Bone Joint Surg Am. 2008;90(11):2313–21.
12. Steinman S et al. A comparison of two nonoperative methods of idiopathic clubfoot correction: the Ponseti method and the French functional (physiotherapy) method. Surgical technique. J Bone Joint Surg Am. 2009;91(Suppl 2):299–312.
13. Faulks S, Richards BS. Clubfoot treatment: Ponseti and French functional methods are equally effective. Clin Orthop Relat Res. 2009;467(5):1278–82.
14. Gottschalk HP, Karol LA, Jeans KA. Gait analysis of children treated for moderate clubfoot with physical therapy versus the Ponseti cast technique. J Pediatr Orthop. 2010;30(3):235–9.
15. Jeans KA, Karol LA. Plantar pressures following Ponseti and French physiotherapy methods for clubfoot. J Pediatr Orthop. 30(1):82–9.
16. Jeans KA, Erdman AL, Karol LA. Plantar pressures after nonoperative treatment for clubfoot: intermediate follow-up at age 5 years. J Pediatr Orthop. 2015. (published ahead of print). doi: 10.1097/BPO.0000000000000589.
17. Zwick EB et al. Comparison of Ponseti versus surgical treatment for idiopathic clubfoot: a short-term preliminary report. Clin Orthop Relat Res. 2009;467(10):2668–76.

18. Laaveg SJ, Ponseti IV. Long-term results of treatment of congenital club foot. J Bone Joint Surg Am. 1980;62(1):23–31.
19. Graf A et al. Long-term outcome evaluation in young adults following clubfoot surgical release. J Pediatr Orthop. 2010;30(4):379–85.
20. Dobbs MB, Nunley R, Schoenecker PL. Long-term follow-up of patients with clubfeet treated with extensive soft-tissue release. J Bone Joint Surg Am. 2006;88(5):986–96.
21. Lourenco AF, Morcuende JA. Correction of neglected idiopathic club foot by the Ponseti method. J Bone Joint Surg Br. 2007;89(3):378–81.
22. Khan SA, Kumar A. Ponseti's manipulation in neglected clubfoot in children more than 7 years of age: a prospective evaluation of 25 feet with long-term follow-up. J Pediatr Orthop B. 2010;19(5):385–9.
23. Yagmurlu MF et al. Ponseti management of clubfoot after walking age. Pediatr Int. 2011;53(1):85–9.
24. Banskota B et al. The Ponseti method in the treatment of children with idiopathic clubfoot presenting between five and ten years of age. Bone Joint J. 2013;95-B(12):1721–5.
25. Ayana B, Klungsoyr PJ. Good results after Ponseti treatment for neglected congenital clubfoot in Ethiopia. A prospective study of 22 children (32 feet) from 2 to 10 years of age. Acta Orthop. 2014;85(6):641–5.
26. Singh A. Evaluation of neglected idiopathic ctev managed by ligamentotaxis using jess: a long-term followup. Adv Orthop. 2011;2011:218489.
27. Singh A et al. Ligamentotaxis for complex calcaneal fractures using Joshi's external stabilization system. Indian J Orthop. 2008;42(3):330–5.
28. Ponseti I. Congenital clubfoot: fundamentals of treatment. 2nd ed. Oxford: Oxford University Press; 1996.
29. Iltar S et al. Treatment of clubfoot with the Ponseti method: should we begin casting in the newborn period or later? J Foot Ankle Surg. 2010;49(5):426–31.
30. Bohm S, Sinclair M. Report of the 1st European consensus meeting on Ponseti clubfoot treatment. J Child Orthop. 2013. doi:10.1007/s11832-013-0497-4.
31. Pittner DE, Klingele KE, Beebe AC. Treatment of clubfoot with the Ponseti method: a comparison of casting materials. J Pediatr Orthop. 2008;28(2):250–3.
32. Hui C et al. Comparison of cast materials for the treatment of congenital idiopathic clubfoot using the Ponseti method: a prospective randomized controlled trial. Can J Surg. 2014;57(4):247–53.
33. Brewster MB et al. Ponseti casting: a new soft option. J Bone Joint Surg Br. 2008;90(11):1512–5.
34. Maripuri SN et al. Ponseti casting for club foot – above- or below-knee? A prospective randomised clinical trial. Bone Joint J. 2013;95-B(11):1570–4.
35. Morcuende JA et al. Results of an accelerated Ponseti protocol for clubfoot. J Pediatr Orthop. 2005;25(5):623–6.
36. Xu RJ. A modified Ponseti method for the treatment of idiopathic clubfoot: a preliminary report. J Pediatr Orthop. 2011;31(3):317–9.
37. Elgohary HS, Abulsaad M. Traditional and accelerated Ponseti technique: a comparative study. Eur J Orthop Surg Traumatol. 2015;25(5):949–53.
38. Harnett P et al. An accelerated Ponseti versus the standard Ponseti method: a prospective randomised controlled trial. J Bone Joint Surg Br. 2011;93(3):404–8.
39. Terrazas-Lafargue G, Morcuende JA. Effect of cast removal timing in the correction of idiopathic clubfoot by the Ponseti method. Iowa Orthop J. 2007;27:24–7.
40. Dyer PJ, Davis N. The role of the Pirani scoring system in the management of club foot by the Ponseti method. J Bone Joint Surg Br. 2006;88(8):1082–4.
41. Radler C et al. Midterm results of the Ponseti method in the treatment of congenital clubfoot. Int Orthop. 2013;37(9):1827–31.
42. Jowett CR, Morcuende JA, Ramachandran M. Management of congenital talipes equinovarus using the Ponseti method: a systematic review. J Bone Joint Surg Br. 2011;93(9):1160–4.
43. Ponseti IV, Morcuende JA. Current management of idiopathic clubfoot questionnaire: a multicenter study. J Pediatr Orthop. 2004;24(4):448.
44. Zionts LE et al. The current management of idiopathic clubfoot revisited: results of a survey of the POSNA membership. J Pediatr Orthop. 2012;32(5):515–20.
45. Radler C. The Ponseti method for the treatment of congenital club foot: review of the current literature and treatment recommendations. Int Orthop. 2013;37(9):1747–53.
46. Alvarez CM et al. The use of botulinum type A toxin in the treatment of idiopathic clubfoot: 5-year follow-up. J Pediatr Orthop. 2009;29(6):570–5.
47. Cummings RJ. The effectiveness of botulinum A toxin as an adjunct to the treatment of clubfeet by the Ponseti method: a randomized, double blind, placebo controlled study. J Pediatr Orthop. 2009;29(6):564–9.
48. Zhao D et al. Results of clubfoot management using the Ponseti method: do the details matter? A systematic review. Clin Orthop Relat Res. 2014;472(4):1329–36.
49. Zhao D et al. Relapse of clubfoot after treatment with the Ponseti method and the function of the foot abduction orthosis. Clin Orthop Surg. 2014;6(3):245–52.
50. Bhaskar A, Patni P. Classification of relapse pattern in clubfoot treated with Ponseti technique. Indian J Orthop. 2013;47(4):370–6.
51. Dobbs MB et al. Factors predictive of outcome after use of the Ponseti method for the treatment of idiopathic clubfeet. J Bone Joint Surg Am. 2004;86-A(1):22–7.
52. Haft GF, Walker CG, Crawford HA. Early clubfoot recurrence after use of the Ponseti method in a New Zealand population. J Bone Joint Surg Am. 2007;89(3):487–93.
53. Ponseti IV. Relapsing clubfoot: causes, prevention, and treatment. Iowa Orthop J. 2002;22:55–6.
54. Lovell ME, Morcuende JA. Neuromuscular disease as the cause of late clubfoot relapses: report of 4 cases. Iowa Orthop J. 2007;27:82–4.
55. Morin ML, Hoopes DM, Szalay EA. Positive communication paradigm decreases early recurrence in clubfoot treatment. J Pediatr Orthop. 2013;34(2):219–22.
56. Ponseti IV et al. Treatment of the complex idiopathic clubfoot. Clin Orthop Relat Res. 2006;451:171–6.
57. Turco V. Recognition and management of the atypical idiopathic clubfoot. Foot Ankle. 1991;11(5):328.
58. Morcuende JA, Dobbs MB, Frick SL. Results of the Ponseti method in patients with clubfoot associated with arthrogryposis. Iowa Orthop J. 2008;28:22–6.
59. Boehm S et al. Early results of the Ponseti method for the treatment of clubfoot in distal arthrogryposis. J Bone Joint Surg Am. 2008;90(7):1501–7.
60. Gerlach DJ et al. Early results of the Ponseti method for the treatment of clubfoot associated with myelomeningocele. J Bone Joint Surg Am. 2009;91(6):1350–9.
61. Janicki JA et al. Treatment of neuromuscular and syndrome-associated (nonidiopathic) clubfeet using the Ponseti method. J Pediatr Orthop. 2009;29(4):393–7.

What is the Best Treatment for Congenital Vertical Talus?

17

Munier Hossain and Naomi Davis

Abstract

Congenital vertical talus (CVT) is a rare congenital anomaly with a reported incidence of 1:100,000. The aetiology of this condition is unclear. It is thought around 50 % of cases are isolated but the rest have associated musculoskeletal, neurologic or chromosomal abnormalities. Traditional treatment involves extensive soft tissue releases. A less invasive correction according to the Ponseti principle has been recently advocated. In this chapter we explore the best available evidence for CVT management and highlight the difficulty in constructing a high grade recommendation on management.

Keywords

Congenital vertical talus • Convex pes valgus • Congenital convex foot • Rocker bottom deformity • Congenital flatfeet • Rigid flatfoot • Congenital oblique talus • Calcaneovalgus • Dobbs technique • Reverse Ponseti technique • Peritalar release

Background

Congenital vertical talus (CVT) is a rare congenital anomaly which is characterized by a fixed dorsal dislocation of the talonavicular joint in conjunction with rigid hindfoot equinus. These structural abnormalities produce a rocker bottom deformity of the foot that is noticeable at birth. The reported incidence ranged from 1:10,000 to 1:100,000 [1, 2]. The aetiology of this condition is unclear. Around 50 % of cases are thought to be isolated but the rest have associated musculoskeletal, neurologic or chromosomal abnormalities.

There are two recognised types of vertical talus: the first with isolated talonavicular dislocation and the second with concomitant calcaneocuboid dislocation[3]. Lichtblau [4] identified three groups of CVT:

1. Teratogenic CVT: a rigid, often bilateral deformity, present at birth with tight extensors and heel cords.
2. Neurogenic CVT: often associated with myelomeningocoele or neurofibromatosis. It is not as rigid as the teratogenic type and easier to correct.
3. Acquired CVT: it is unilateral, moderate in severity; the calcaneum is not necessarily fixed in equinus; and the deformity is partially correctable. This type has been attributed to intrauterine malposition.

A less severe manifestation of the deformity has been called the oblique talus. In this variant there is a rocker bottom deformity of the foot and an equinus contracture of the hindfoot, but the navicular can be reduced when the forefoot is plantar flexed. Severe flatfoot are sometimes confused with CVT because the talus is vertical on a lateral radiograph. The clinical and radiographic difference between the conditions is that in a severe flatfoot, the calcaneum can be easily dorsiflexed and there is no fixed dislocation of the navicular [5].

Several authors tried to aid differentiating between CVT and other similar conditions. Eyre-brook [6] first proposed a lateral foot x-ray in "fullest plantar flexion" to differentiate

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Table 17.1 Adelaar score of congenital convex foot

Clinical	Score	Radiological	Score
Poor cosmetic appearance		Abnormal talonavicular angle Talometatarsal axis	
Ankle+ loss of subtalar movement		Hindfoot equinus	
Prominent talar head		Abnormal talometatarsal axis	
Loss of medial longitudinal arch		Talonavicular subluxation	
Hindfoot valgus			
Abnormal shoe wear			

1 point for the absence of each item. 0 point for the presence of each item. Total score 10
Excellent 10, good 7–9, fair 4–6, poor ≤3

between true CVT and other causes of flexible vertical talus. He demonstrated how feet with obvious convexity of the sole and apparent vertical talus get fully aligned on plantar flexion view and calcaneus is restored to its normal relationship with the tibia. Subsequently Hamanishi [7] proposed the radiological angles of talar axis- metatarsal base angle (TAMBA) and calcaneal axis-first metatarsal base angle (CAMBA). He proposed the values of 60° of TAMBA and 20° of CAMBA to differentiate between flexible oblique talus and rigid CVT.

Unfortunately most authors were not explicit about their diagnostic criteria for CVT and therefore it is difficult to be certain that their series contain only cases of true CVT. Around 50 % of CVT patients have other neurological or teratological abnormalities and their CVT is non-isolated. It is well recognised that prognosis is generally worse for non-isolated CVT. However, most published series considered isolated and non-isolated CVT patients together while reporting surgical outcomes. Moreover some of the published work even did not acknowledge the various types of CVT mentioned above. This is pertinent since there is a link between various types and successful outcomes.

Most studies that looked at CVT results used non validated outcome measures. A number of papers used Adelaar score [8] to report patient outcome. This is a 10 point clinical-radiological scoring system. Absence of an item gives 1 point. Maximum point is 10 and a higher score suggests better outcome (Table 17.1). The validity and reliability of this scoring system is unknown and it is not a patient reported outcome. Duncan and Fixsen [9] used a patient reported satisfaction score for reporting their results of CVT correction. The instrument was modified from the one first proposed by Laaveg-Ponseti for reporting results of clubfoot management who attempted to correlate both patient satisfaction and surgeon assessment of functional outcome

Table 17.2 The modified Laaveg-Ponseti questionnaire

Satisfaction (20 points)
I am: (a) Very satisfied with the end result 20 (b) Satisfied with the end result 16 (c) Neither satisfied nor unsatisfied with the end result 12 (d) Unsatisfied with the end result 8 (e) Very unsatisfied with the end result 4
Function (20 points)
In my daily living, my foot: (a) Does not limit my activities 20 (b) Occasionally limits my strenuous activities 16 (c) Usually limits me in strenuous activities 12 (d) Limits me occasionally in routine activities 8 (e) Limits me in walking 4
Pain (30 points)
My foot: (a) Is never painful 30 (b) Occasionally causes mild pain during strenuous activities 24 (c) Usually is painful after strenuous activities only 18 (d) Is occasionally painful during routine activities 12 (e) Is painful during walking 6

in a single instrument [10]. The modified Laaveg-Ponseti questionnaire is reproduced in Table 17.2. Further, complications and recurrences were discussed but authors did not clarify what constituted "recurrence" although many authors did compare their results with the normative radiological values of Vanderwilde et al. [11]. The rate of revision surgery was discussed but it was not clear what the authors' indications were for revision surgery.

What Is the Natural History of Untreated CVT?

There are no studies looking at the natural history of untreated CVT but some authors have discussed the consequences of untreated or conservatively treated but uncorrected CVT. It has been claimed that if left untreated the child with CVT will have a painful and rigid flatfoot with weak push-off power and a rocker bottom deformity [8, 12, 13]. They have difficulty with footwear and have painful callosities due to limited weight bearing area [14, 15]. It is likely that such a deformity would result in significant functional deficit. Natural history of untreated CVT is also confounded by the fact that approximately 50 % of the patients will have major musculoskeletal or neurologic abnormality that may also significantly affect their walking ability. Published papers do not discuss the natural history of isolated CVT separately from the teratologic type. Authors appear to agree that untreated CVT does not affect walking and assert that in some patients the deformity may not be noted until patients start walking [15, 16].

What Is the Best Time to Start Treatment?

There is general agreement that treatment should be started as soon as possible after birth [14]. This is identical to the treatment principle followed in clubfoot management. In both cases the initial plan of treatment is to gently manipulate the foot by stretching the soft tissues and immobilise the foot in the corrected position. It is expected that correction of foot deformity will lead to reshaping of cartilage anlage. Hence early treatment is essential to take advantage of flexibility of the neonatal skeleton. Aslani et al. [17] found that patients older than 2 years of age at onset of treatment required longer casting and more manipulations for correction of CVT. Adelaar et al. [8] also commented that prognosis was better if treatment was started early and noted that result of surgical correction was poor in those aged over 3½ years. Mazzocca et al. [18] recommended that surgery should be performed at around 2 years of age, others also agree to this timeline [13, 16].

What Is the Best Treatment for CVT?

A number of treatments have been attempted for correction of CVT and treatment has evolved over the years taking into account the complications and results thereof. The goal of treatment in CVT has been to restore the anatomical relationship of the hindfoot bones and to recreate a plantigrade foot to allow pain-free weight bearing. Initial reports attested to failure of conservative treatment with manipulation followed by serial casting, boots and bars, shoe wedges, corrective night splint etc. [16] As a result surgical reduction with extensive soft tissue release became the treatment of choice. A number of reports were published attesting to the results of surgery. The cornerstone of surgery was to perform soft tissue release to restore hindfoot relationship to normal. A number of techniques were then used to keep the hindfoot joints reduced. This included re-routing of the tibialis anterior, transposition of the peroneus brevis, k wire fixation, partial or complete naviculectomy [19]. Subsequently a two stage procedure became popular. The forefoot and midfoot was initially reduced to the fixed plantar-flexed hindfoot by reducing the TNJ and maintaining the reduction with k wire through the TNJ. Six weeks later a second procedure was performed to release the tendoachilles and the posterior ankle and the sub-talar joints [20]. Coleman et al. [3] advocated that the dorsal extensors should be lengthened as part of the first procedure and the tibialis posterior tendon should be advanced to the plantar surface of the navicular as part of the second procedure. Later Ogata and Shoenecker [21] recommended a single stage correction in view of their observed complications of two stage surgery. Seimon [22] published a small series with good results from single stage surgery. Single stage correction became the standard performed procedure. However, there was considerable variation in practice among the proponents of single stage release. In addition to TNJ capsulotomy and tendoachilles lengthening surgeons variably performed dorsal extensor lengthening, peroneal lengthening, tibialis anterior tendon transfer to the neck of talus, or more extensive postero-medial and/or lateral release with or without additional bony surgery. Single stage release is the standard surgical procedure at present and results are still being published. The proponents of single stage correction can be divided into two camps: those who perform peri-talar release and those who perform mid-tarsal release. Surgery can be performed either through a Cincinnati incision or a modification thereof, multiple separate incisions or a dorsal incision. Mazzocca et al. [18] and Saini et al. [12] have claimed better results for single stage correction using the dorsal approach instead of the traditional Cincinnati approach. However, while Mazzocca essentially performed mid-tarsal release Saini appears to have performed peri-talar release with the addition of a posterior incision to approach the ankle and the sub-talar joint. Stricker-Rosen [23] and Seimon [22] also had good results using a longitudinal dorsal incision although they only performed TNJ release and Stricker-Rosen only had teratogenic CVT in their series. It appears from the limited evidence that dorsal approach might give better outcome than the Cincinnati approach especially for mid-tarsal or isolated TNJ release. Results of single stage surgical correction are presented in Table 17.3.

Dobbs et al. [24, 25] published their results of a new method of treatment of CVT in 2006. The authors extrapolated the Ponseti principle for CVT management. As in the Ponseti method the essence of this technique is to apply serial manipulation and casting using the head of the talus as the centre of rotation. Manipulation is applied in the opposite direction to that applied for clubfoot. Following midfoot alignment limited surgery with pinning of the TNJ and percutaneous TA tenotomy is performed. The authors also variably performed extensor or peroneal lengthening. The first published series was on assumed idiopathic CVT. Subsequently the same group also separately published results of treatment of non-idiopathic CVT and suggested treatment modification in this group with more extensive soft tissue surgery [26]. Subsequent reports by Eberhardt et al. [27, 28] and Wright et al. [29] evaluating the Dobbs technique have also confirmed the need for more extensive surgery in the more rigid type of CVT. Proponents of the Dobbs technique have suggested that this new method of less invasive treatment may lead to more flexible and less painful feet in the long term [30]. They rest their argument on the basis of the results of surgical management of clubfoot that showed

Table 17.3 Results of surgery for CVT correction

Studies	No (feet)	Isolated CVT	Type of surgery	Concomitant additional surgery	F/U (Y)	Age at surgery (months)	Normal footwear	Recurrence	AVN Talus	R
Adelaar et al. [8]	18	?	Single stage peri-talar release	TATT Peroneal ±extensor lengthening	3	11.4	?	? 1 excellent 12 good	1	?
Walker et al. [20]	15	6	Two stage	Nil	10.2	8.5	?	2	0	1
Dodge et al. [32]	36	13	Single stage release 6 Two stage release 10	STJ fusion 11 Naviculectomy 2 Triple fusion 1	14	?	10 needed custom footwear	?	0	4
Seimon [22]	10	3/7 patients	Single stage TNJ release	Peroneal ±extensor lengthen	5.2	5–13	10	0	0	0
Napiontek [33]	32	11	Single stage peri-talar release Pre-op MC 32	TATT Peroneal ±extensor lengthen Grice-Green 8	9.2	39	?	? 7 Overcorrected 6 Poor outcome	7	8
Stricker and Rosen [23]	20	3	Single stage TNJ release Pre-op MC	Peroneal ±extensor lengthen	3½	11.8	20 “standard shoe or brace”	5 17 had TAMBA >10°	0	0
Duncan and Fixsen [9]	10	8	Single stage peri-talar release	TATT Tib Post shortening Peroneal ±extensor lengthening	9	31	10	0 1 cavo-varus	0	0
Kodros and Dias [34]	42	5	Single stage peri-talar release	Peroneal ±extensor lengthening	7	?	42	?	0	10
Mazzocca et al. [18]	33	14	Posterior release 25 Dorsal approach 8 patients pre-op MC 14	TATT Peroneal ±extensor lengthening	Minimum 3 years	?	?	8	12[a]	11
Zorer et al. [35]	17	8	Single stage peri-talar release Pre-op MC 7	Peroneal ±extensor lengthening	3.5	28.4	17/17 10/17 also wore orthosis	0 Poor outcome 2 Fair 2	0	0
Abdel Razzak [36]	28	15	Single stage Peri-talar release Pre-op MC NK	TATT Peroneal ±extensor lengthening	9 ± 5.3 months	19.1 ± 7.1	26/28	3	0	0
Mathew et al. [37]	5	5/5	Single stage peri-talar release Pre-op MC 5/5	TATT Tib Post advancement	7.5	32	5/5	0	0	0
Saini et al. [12]	20	2/20	Single stage peri-talar release	extensor lengthening	4	16	NK	0	0	0
Ramanoudjame et al. [38]	31	15	Mid-tarsal release	Open reduction TNJ, lengthening Tib Ant, extensor, TA 23 TA transfer 5 Calcaneal osteotomy 5	11	12	31	0	9	5

F/U follow-up, *R* revision, *TATT* tibialis anterior tendon transfer, *STJ* subtalar joint, *TNJ* talonavicular joint, *MC* manipulation and casting, *NK* numbers not known

[a]12 AVN of the talus in the posterior release group

Table 17.4 Results of Dobbs technique

Authors	No (feet)	Isolated CVT	Type of treatment	Additional surgery	Mean follow up	Mean age at treatment	Normal footwear	Recurrence	AVN talus	Revision
Dobbs et al. [25]	19	19	MC followed by limited surgery	Extensor ± peroneal lengthening	Minimum follow up 2 years	8 months	19	6 None had TNJ pinning	0	6 repeat casting, and 4 TNJ pinning
Bhaskar [39]	4	4	MC followed by limited surgery	Extensor ± peroneal lengthening	8.5 months	2 weeks	0	0	0	0
Eberhardt et al. [27]	20	9	MC followed by limited surgery	Extensor ± peroneal lengthening	24 months	20 days to 14 months	NK	1/14	NK	NK
Chalayon et al. [26]	25	0	MC followed by limited surgery	TNJ and STJ capsulotomy 20 Open TNJ reduction 5	Minimum follow up 2 years	6 months	NK	5 (all had CCJ subluxation)	? 0	5 repeat MC an and pinning of CCJ and TNJ
Aslani et al. [17]	15	5	MC followed by limited surgery	nil	Mean 2 years	1 month–9 years	15	0	0	0
Wright et al. [29]	21	12	MC followed by limited surgery	Open TNJ reduction (if unreduced)+ Tib Ant transfer) Extensor ± peroneal lengthening	3 years	5 months	NK	10 (none had TNJ capsulotomy)	NK	8 2 MC only 6 mini open Reduction of TNJ

STJ subtalar joint, *TNJ* talonavicular joint, *CCJ* calcaneocuboid joint, *MC* manipulation and casting, *NK* numbers not known

poor long term function with extensive soft tissue surgery [31]. Early results of Dobbs technique have been published from only a few centres. Results are not directly comparable between the two techniques and it appears that the Dobbs technique may still evolve. Although short term results appear promising we need longer follow up especially from non-originating centres. Results of Dobbs technique are presented in Table 17.4.

In summary, CVT is a very rare condition with an unknown cause. It may well be that the majority of patients have an underlying condition that affects treatment outcomes. Surgical and non surgical interventions reported are almost as varied as the deformities and patients they treat. Long term outcomes are poorly understood, especially as the walking abilities of some patients may be limited by altered neurology from a variety of causes. Basic principles of managing foot deformities in infants seem appropriate in terms of early treatment to improve the cartilage anlage and limiting soft tissue release, but that and the use of tendon transfers remain largely unproven (Table 17.5).

Table 17.5 Summary of recommendations

Clinical questions	Grade of recommendations
Untreated CVT results in a painful rigid flatfoot	C
For best results surgery should be performed before the child is 2 years of age	C
Treatment for CVT should be started as soon as possible	C
Treatment for CVT should be tailored to the individual patient taking into account the degree of rigidity and the underlying pathoanatomy	C
Dorsal approach is better than the posterior approach for surgical correction	C
CVT with both TNJ and CCJ dislocation requires more invasive surgery than CVT with isolated TNJ dislocation	C
It is not clear if the Dobbs technique is a better alternative to the traditional soft tissue surgery	I
Prognosis is worse if treatment is started late, in patients with non-isolated CVT and CVT with combined TNJ and CCJ dislocation	C
The best treatment for the older child or for cases of recurrence is not clear	I

What Type of Patient Has Poor Prognosis?

There is accumulating evidence to suggest that prognosis is worse if treatment is started late, for non-idiopathic type CVT and for CVT with both TNJ and CCJ dislocation [8, 17, 26].

What Is the Best Treatment for the Older Child with CVT?

There is no agreement regarding a late presenter although it is understood that treatment should be started as soon as possible. Aslani et al. claimed success in treating a child aged 9 years using the Dobbs technique[17]. Naviculectomy, Grice-Green arthrodesis and triple fusion have been recommended for what authors considered to be older children but results are not known [9, 15, 32].

References

1. Jacobsen ST, Crawford AH. Congenital vertical talus. J Pediatr Orthop. 1983;3(3):306–10.
2. Khader A, Huntley JS. Congenital vertical talus in Cri du Chat Syndrome: a case report. BMC Res Notes. 2013;6:270.
3. Coleman SS, Stelling 3rd FH, Jarrett J. Pathomechanics and treatment of congenital vertical talus. Clin Orthop Relat Res. 1970;70:62–72.
4. Lichtblau S. Congenital vertical talus. Bull Hosp Joint Dis. 1978;39(2):165–79.
5. Herring JA, editor. Tachdjians, pediatric orthopaedics, vol. 1. Philadelphia: Saunders Elsevier; 2008.
6. Eyre-Brook AL. Congenital vertical talus. J Bone Joint Surg Br. 1967;49(4):618–27.
7. Hamanishi C. Congenital vertical talus: classification with 69 cases and new measurement system. J Pediatr Orthop. 1984;4(3):318–26.
8. Adelaar RS, Williams RM, Gould JS. Congenital convex pes valgus: results of an early comprehensive release and a review of congenital vertical talus at Richmond Crippled Children's Hospital and the University of Alabama in Birmingham. Foot Ankle. 1980;1(2):62–73.
9. Duncan RD, Fixsen JA. Congenital convex pes valgus. J Bone Joint Surg Br. 1999;81(2):250–4.
10. Laaveg SJ, Ponseti IV. Long-term results of treatment of congenital club foot. J Bone Joint Surg Am. 1980;62(1):23–31.
11. Vanderwilde R, et al. Measurements on radiographs of the foot in normal infants and children. J Bone Joint Surg Am. 1988;70(3):407–15.
12. Saini R, et al. Results of dorsal approach in surgical correction of congenital vertical talus: an Indian experience. J Pediatr Orthop B. 2009;18(2):63–8.
13. Bosker BH, et al. Congenital convex pes valgus (congenital vertical talus). The condition and its treatment: a review of the literature. Acta Orthop Belg. 2007;73(3):366–72.
14. Rodriguez N, Choung DJ, Dobbs MB. Rigid pediatric pes planovalgus: conservative and surgical treatment options. Clin Podiatr Med Surg. 27(1):79–92.
15. Drennan JC. Congenital vertical talus. Instr Course Lect. 1996;45:315–22.
16. Lloyd-Roberts GC, Spence AJ. Congenital vertical talus. J Bone Joint Surg Br. 1958;40-B(1):33–41.
17. Aslani H, et al. Primary outcomes of the congenital vertical talus correction using the Dobbs method of serial casting and limited surgery. J Child Orthop. 2013;6(4):307–11.
18. Mazzocca AD, et al. Comparison of the posterior approach versus the dorsal approach in the treatment of congenital vertical talus. J Pediatr Orthop. 2001;21(2):212–7.
19. Colton CL. The surgical management of congenital vertical talus. J Bone Joint Surg Br. 1973;55(3):566–74.
20. Walker AP, Ghali NN, Silk FF. Congenital vertical talus. The results of staged operative reduction. J Bone Joint Surg Br. 1985;67(1):117–21.
21. Ogata K, Schoenecker PL, Sheridan J. Congenital vertical talus and its familial occurrence: an analysis of 36 patients. Clin Orthop Relat Res. 1979;139:128–32.
22. Seimon LP. Surgical correction of congenital vertical talus under the age of 2 years. J Pediatr Orthop. 1987;7(4):405–11.
23. Stricker SJ, Rosen E. Early one-stage reconstruction of congenital vertical talus. Foot Ankle Int. 1997;18(9):535–43.
24. Dobbs MB, et al. Early results of a new method of treatment for idiopathic congenital vertical talus. Surgical technique. J Bone Joint Surg Am. 2007;89 Suppl 2 Pt.1:111–21.
25. Dobbs MB, et al. Early results of a new method of treatment for idiopathic congenital vertical talus. J Bone Joint Surg Am. 2006;88(6):1192–200.
26. Chalayon O, Adams A, Dobbs MB. Minimally invasive approach for the treatment of non-isolated congenital vertical talus. J Bone Joint Surg Am. 2012;94(11):e73.
27. Eberhardt O, Fernandez FF, Wirth T. The talar axis-first metatarsal base angle in CVT treatment: a comparison of idiopathic and non-idiopathic cases treated with the Dobbs method. J Child Orthop. 2012;6(6):491–6.
28. Eberhardt O, Fernandez FF, Wirth T. Treatment of vertical talus with the Dobbs method. Z Orthop Unfall. 2011;149(2):219–24.
29. Wright J, et al. Reverse Ponseti-type treatment for children with congenital vertical talus: comparison between idiopathic and teratological patients. Bone Joint J. 2014;96-B(2):274–8.
30. Alaee F, Boehm S, Dobbs MB. A new approach to the treatment of congenital vertical talus. J Child Orthop. 2007;1(3):165–74.
31. Dobbs MB, Nunley R, Schoenecker PL. Long-term follow-up of patients with clubfeet treated with extensive soft-tissue release. J Bone Joint Surg Am. 2006;88(5):986–96.
32. Dodge LD, Ashley RK, Gilbert RJ. Treatment of the congenital vertical talus: a retrospective review of 36 feet with long-term follow-up. Foot Ankle. 1987;7(6):326–32.
33. Napiontek M. Congenital vertical talus: a retrospective and critical review of 32 feet operated on by peritalar reduction. J Pediatr Orthop B. 1995;4(2):179–87.
34. Kodros SA, Dias L. Single stage surgical correction of congenital vertical talus. J Paediatr Orthop. 1999;19(1):42–8.
35. Zorer G, Bagatur AE, Dogan A.Single stage surgical correction of congenital vertical talus by complete subtalar release and peritalar reduction by using the Cincinnati incision. J Pediatr Orthop B. 2002;11(1):60–7.
36. Abdel-Razzak MY. Surgical correction of congenital vertical talus by one stage peritalar reduction and tibialis anterior transfer. Alex J Paediatr. 2002;16(2):309–25.
37. Mathew PG, Sponer P, Karpas K, Shaikh HH. Mid-term results of one-stage surgical correction of congenital vertical talus. Bratisl Lek Listy. 2009;110(7):390–3.
38. Ramanoudjame M, Loriaut P, Seringe R, Glorion C, Wicart P. The surgical treatment of children with congenital convex foot (vertical talus): evaluation of midtarsal surgical release and open reduction. Bone Joint J. 2014;96-B(6):837–44. doi: 10.1302/0301-620X.96B6.32313.
39. Bhaskar A. Congenital vertical talus: treatment by reverse ponseti technique. Indian J Orthop. 2008;42(3):347–50. doi: 10.4103/0019-5413.41860.

Evidence-Based Treatment for Metatarsus Adductus

18

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Abstract

Metatarsus adductus (MA) is a common congenital foot deformity; often attributed to "packaging disorders". It is usually an isolated deformity; however it can be part of a more complex foot deformity such as a club foot or skew foot. Natural history of untreated isolated MA appears favourable and treatment is often not required. Conservative treatments such as serial casting, orthosis, modified shoe wears are widely practiced. There is no evidence to show that conservative treatments are superior to no treatment. Surgical treatments should be reserved for metatarsus adductus associated with other congenital deformity such as residual club foot or skew foot.

Keywords

Metatarsus adductus • Metatarsus varus • Metatarsus varus congenitus • Metatarsus adductovarus • Hooked forefoot

Background

Metatarsus adductus (MA) is a term used when the forefoot is adducted at the tarso-metatarsal joint but the hindfoot is normal. In contrast to a club foot where the hind foot is in equinus and varus or a skew foot where the hind foot is in valgus and the midfoot is abducted [1] (Fig. 18.1). MA should be differentiated from Metatarsus primus varus where only the first metatarsal is adducted and the 1st–2nd intermetatarsal angle is increased. La Reaux and Lee [2] had described mandatory criteria for MA diagnosis which included a high arch, adduction and inversion of the forefoot, prominent lateral border and most importantly increased rigidity such that the foot cannot be abducted beyond the midline. However, these criteria have not been widely followed in published papers.

Bleck's clinical classification [3] is widely used for diagnosis and classification of MA. He advised the use of the heel bisector axis (HBA), which is a line drawn along the middle of the weight-bearing plantar surface of the heel and normally crosses the second toe. MA is categorised as "mild" if the HBA crosses the third toe, "moderate" when it crosses the fourth toe and "severe" when it crosses lateral to the fourth toe (Fig. 18.2). HBA also serves as the reference for forefoot flexibility. MA is termed "flexible" when the foot is correctable beyond the HBA, "partially flexible" when the foot is correctable up to the HBA and "rigid" when it is not correctable. However, Bleck did not find any correlation between the severity and flexibility of MA at diagnosis and the eventual outcome. Radiological assessment can be performed in the older child by measuring the MA angle (MAA). The MAA is the angle formed by the longitudinal axis of the metatarsals and that of the lesser tarsus. The axis of the second metatarsal is considered to be the axis of the metatarsal.

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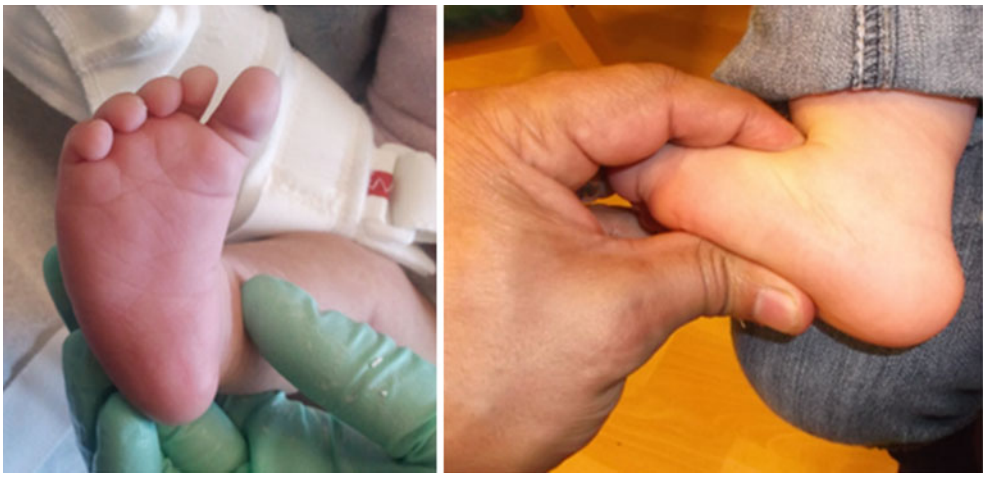

Fig. 18.1 Clinical photographs of a child with metatarsus adductus. There is no hindfoot equinus; an important differentiating sign from a clubfoot

What Is the Natural History of Metatarsus Adductus?

Ponseti and Becker [4] found that nearly 90 % of patients with MA achieve spontaneous resolution with time. The authors advised that passively correctable MA did not require any treatment but recommended casting for feet that were not passively correctable. They reported good results from casting of semi-rigid or rigid feet. Rushforth [5] followed 130 MA of all grades of severity for a mean follow up of 7 years and found 86 % were normal or mildly deformed but fully mobile; 10 % were still moderately deformed but were asymptomatic and 4 % remained deformed and stiff. It was not possible to detect these resistant cases before the age of 3 years, but the low failure rate would seem to justify a policy of expectant treatment. Ghali et al. [6] presented a small series of both untreated (31) and surgically treated (38) MA at a median follow-up of 4 years. Surgical treatment involved antero-medial release. Patients were assessed both clinically and radiologically. He reported "excellent" results in both groups. [7] presented a follow up study of 243 feet with MA. It is not clear what the length of follow up was. He treated flexible feet with stretching and semi-rigid or rigid feet with casting. He found a favourable outcome and commented that children with residual deformity had "no difficulties with function or shoe wear". However Bohne also offered surgery to 9 feet with "severe residual deformity". The Scandinavian series of [8] reported on 76 patients with MA at 6 years follow-up and found that 13 % of children had a persistent MA. Widhe subsequently published a longer follow up of the same cohort at 16 years [9]. Those with persistent MA at 6 years continued to improve and only three patients had a persistent MA deformity at 16 years. All of them were asymptomatic despite the deformity. The initial grade of severity of the MA patients in this series is not known. Widhe also compared gait analysis and dynamic foot pressure at 16 years between those with normal feet at birth and those with MA and found no difference between the two groups. Ponseti's group published a 32-year follow up of patients with MA [10]. The cohort included 16 passively correctable feet that had no treatment. The rest had semi-rigid or rigid deformity and were treated with serial manipulation and casting. The authors used a functional rating system to assess functional outcome and also performed radiological assessment. No one had poor results and none required subsequent surgery. All the feet that were initially passively correctable went on to achieve spontaneous resolution. There was only a single case of hallux valgus at final follow up. The authors advised against surgical treatment of rigid MA.

Evidence from the published literature appears to suggest a favourable outcome for untreated MA. Most will resolve without treatment by early childhood and those that persist are likely to improve with age. A small minority of MA may persist but are unlikely to give any functional problem. There is enough evidence to suggest that passively correctable MA deformity does not require any treatment apart from parental reassurances. Manipulation and casting is an option in feet that do not correct passively although no treatment is probably equally feasible.

What Is the Role of Serial Casting?

Given the favourable natural history of MA, it is difficult to ascertain the value of manipulation and serial casting. Widhe [9] showed that MA continues to improve to adulthood. There has been no comparative study of serial casting with control. Although it has been recommended for metatarsus

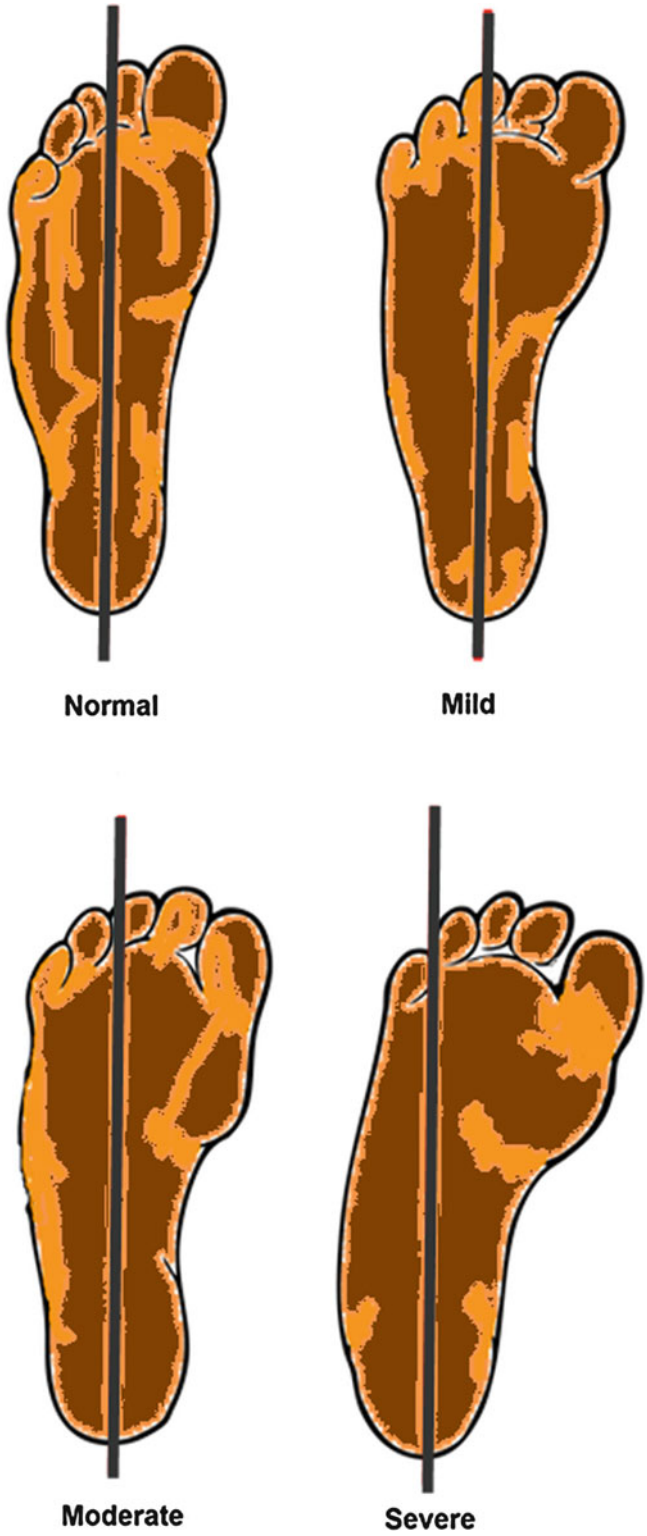

Fig. 18.2 Bleck's classification of metatarsus adductus

adductus of intermediate severity, there is strong evidence that these deformities will self-correct over time without intervention with very few exceptions [4, 5, 8, 9].

Is There a Role for Orthotics?

Pentz and Weiner [11] published their experience with 795 patients treated for MA over a 13-year period with a straight metal bar and attached reverse last shoe protocol. Nearly uniform excellent results were encountered, with a 99 % likelihood of obtaining a fully corrected foot. Surgical intervention was deemed necessary in less than 1 % of cases. Lack of control seriously undermines this paper. As stated earlier that numerous reports of high rate of natural resolution, it is not unreasonable to assume that the high success rate of 99% full correction attributed to natural history of MA and not due to orthotic support. Herzenberg and Burghardt [12] conducted a randomised trial of casting versus Bebax orthosis in 27 infants (43 feet) and found orthosis to be equally effective but cheaper [13]. However, the inclusion criterion was "infants who failed home stretching". The age range was 3–9 months and it is not clear if these patients required any treatment at all.

Is There an Association Between Metatarsus Adductus and Hallux Valgus?

Long term follow up studies of untreated MA report a low rate of hallux valgus deformity in this population [5, 8, 9]. However other studies investigating patients with hallux valgus deformity undergoing surgery have found a higher prevalence of MA. The reported prevalence is around 30 % [13]. Ferrari and Malone-Lee [14] randomly reviewed 100 x-rays and found a higher prevalence of MA in feet with hallux valgus (55 %) compared to those without (19 %). However, retrospective studies of this nature are contaminated by selection bias and are unlikely to be representative of the true association.

Is Metatarsus Adductus a Risk Factor for Developmental Dysplasia of the Hip?

The prevalence of DDH in MA on the basis of radiological evaluation is reported to be between 2 % and 10 % [15, 16]. Paton [17] investigated the relationship between neonatal foot deformities and severe developmental dysplasia of the hip (DDH) in a 11-year prospective longitudinal observational study [17]. They found a 4 % risk of type IV DDH in MA and recommended sonographic imaging and surveillance for DDH in patients presenting with MA. Two well-conducted systematic reviews and meta-analyses [18, 19] found a non-significant association between foot deformities and DDH, pooled data from five cohort studies showed an Odds ratio of 3.24 (95 % CI 0.88–11.97: P = 0.08).

Table 18.1 Summary of recommendations

Clinical questions	Grade of recommendation
1. The natural history of untreated MA is quite favourable	B
2. There is no need for treatment of passively correctable MA	B
3. Casting or orthosis are options for treating semi-rigid or rigid MA	I
4. It is not proven that persistent MA predisposes to hallux valgus later in life	C
5. MA may be a risk factor for DDH	C

What Is the Role for Surgery?

It is not clear if surgery has any role to play in primary MA. Reports from [8, 9] and the Ponseti group [4] would indicate none. Only 1 % patients in [11] series required surgery. Studies recounting results of surgical treatment for MA had heterogeneous cohorts and included primary MA, skew foot as well as residual MA from clubfoot treatment. A number of surgical techniques have been described. Cahuzac [20] described a technique of medial capsulotomy to re-orient the first cuneiform metatarsal joint, sectioning of the abductor hallucis brevis and abduction osteotomy of the metatarsals for correction of MA. Fifteen of these feet had residual deformity from clubfoot. Napiontek [21] reported their results of opening wedge osteotomy of medial cuneiform in children aged less than 4 years. Thirty-three of the thirty-seven feet in this series had MA from residual clubfoot deformity. Gordon [22] reported minimum 2-year follow up results of combined midfoot osteotomy in 50 feet with severe MA of whom only 11 feet had primary MA. Knorr [23] published their results of the Cahuzac procedure in 34 patients where 9 only had primary MA. The aetiology, pathogenesis and natural history of MA and clubfoot are different. It is misleading to recommend surgical treatment for MA based on these studies' conclusions (Table 18.1).

References

1. Herring JA, editor. Tachdjians, Pediatric orthopaedics, vol. 1. Philadelphia: Saunders Elsevier; 2008.
2. La Reaux RL, Lee BR. Metatarsus adductus and hallux abducto valgus: their correlation. J Foot Surg. 1987;26(4):304–8.
3. Bleck EE. Metatarsus adductus: classification and relationship to outcomes of treatment. J Pediatr Orthop. 1983;3(1):2–9.
4. Ponseti IV, Becker JR. Congenital metatarsus adductus: the results of treatment. J Bone Joint Surg Am. 1966;48(4):702–11.
5. Rushforth GF. The natural history of hooked forefoot. J Bone Joint Surg Br. 1978;60-B(4):530–2.
6. Ghali NN, Abberton MJ, Silk FF. The management of metatarsus adductus et supinatus. J Bone Joint Surg Br. 1984;66(3):376–80.
7. Bohne W. Metatarsus adductus. Bull N Y Acad Med. 1987;63(9):835–8.
8. Widhe T, Aaro S, Elmstedt E. Foot deformities in the newborn--incidence and prognosis. Acta Orthop Scand. 1988;59(2):176–9.
9. Widhe T. Foot deformities at birth: a longitudinal prospective study over a 16-year period. J Pediatr Orthop. 1997;17(1):20–4.
10. Farsetti P, Weinstein SL, Ponseti IV. The long-term functional and radiographic outcomes of untreated and non-operatively treated metatarsus adductus. J Bone Joint Surg Am. 1994;76(2):257–65.
11. Pentz AS, Weiner DS. Management of metatarsus adductovarus. Foot Ankle. 1993;14(5):241–6.
12. Herzenberg JE, Burghardt RD. Resistant metatarsus adductus: prospective randomized trial of casting versus orthosis. J Orthop Sci. 2013;19(2):250–6.
13. Aiyer AA, et al. Prevalence of metatarsus adductus in patients undergoing hallux valgus surgery. Foot Ankle Int. 2014;35(12):1292–7.
14. Ferrari J, Malone-Lee J. A radiographic study of the relationship between metatarsus adductus and hallux valgus. J Foot Ankle Surg. 2003;42(1):9–14.
15. Kumar SJ, MacEwen GD. The incidence of hip dysplasia with metatarsus adductus. Clin Orthop Relat Res. 1982;164:234–5.
16. Jacobs JE. Metatarsus varus and hip dysplasia. Clin Orthop. 1960;16:203–13.
17. Paton RW, Choudry Q. Neonatal foot deformities and their relationship to developmental dysplasia of the hip: an 11-year prospective, longitudinal observational study. J Bone Joint Surg Br. 2009;91(5):655–8.
18. de Hundt M, et al. Risk factors for developmental dysplasia of the hip: a meta-analysis. Eur J Obstet Gynecol Reprod Biol. 2012;165(1):8–17.
19. Ortiz-Neira CL, Paolucci EO, Donnon T. A meta-analysis of common risk factors associated with the diagnosis of developmental dysplasia of the hip in newborns. Eur J Radiol. 2011;81(3):e344–51.
20. Cahuzac JP, Laville JM, de Gauzy JS, Lebarbier P. Surgical correction of metatarsus adductus. J Pediatr Orthop. 1993;2(2):176–81.
21. Napiontek M, Kotwicki T, Tomaszewski M. Opening wedge osteotomy of the medial cuneiform before age 4 years in the treatment of forefoot adduction. J Pediatr Orthop. 2003;23(1):65–9.
22. Gordon JE, et al. Combined midfoot osteotomy for severe forefoot adductus. J Pediatr Orthop. 2003;23(1):74–8.
23. Knorr J, et al. Percutaneous correction of persistent severe metatarsus adductus in children. J Pediatr Orthop. 2013;34(4):447–52.

Evidence-Based Treatment of Flexible Flat Foot in Children

19

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Abstract

Flexible flatfoot, mobile pes planus, is one of the commonest normal variants raising concern in children. It is present in nearly all infants, is common in older children and occurs in a number of adults. It can be a major source of parental anxiety, yet is now recognised as a normal anatomical variant associated with supple joints. Accordingly, it is associated with younger age, male sex and obesity. Reassurance by education and explanation of the natural history is the mainstay of treatment. We look at the available evidence in the condition and provide treatments recommendations (Table 19.1).

Keywords

Flexible • Flatfoot • Mobile pes planus • Paediatric • Treatment • Orthotics

Background

Traditionally flatfoot has been defined as a weight bearing foot with a low or absent longitudinal arch. This definition is based on the static anatomic comparison of the height of the arch within a population. However there is a lack of agreement on a true definition of flatfoot or pes planus, and there is considerable inter-clinician variability in the visual assessment of feet for this characteristic, even for extremes of foot type [1]. Morphologically it has classically been claimed that humans possess a mid-foot stabilized in bone that allows the metatarsals to act as an efficient propulsive lever [2] and variations from this may have been misinterpreted as being pathological. Newer work points to mid-foot compliance in humans and indeed in living apes as representing a functional continuum, with a clear quantitative overlap between the albeit less flexible feet of humans and the more compliant feet of non-human apes [3]. The traditional view ignores normal variations seen in arch height among adults and children and also between different ethnic groups and failed to acknowledge a spectrum of arch height in normal individuals. In a clinical setting it is reassuring to recognise that a flexible flat foot flattens on weight bearing, but resolves when non-weight bearing. Furthermore, it resolves with the muscle action of rising onto tiptoe (Fig. 19.1). Even without a strict definition, most children and at least 20 % of adults have flat feet, which are flexible [4]. This flexibility reduces with age and ordinarily such feet in children and adults remain fully functional and asymptomatic. Generalised mobility in other joints and increased body weight may be reflected in a higher prevalence of flatfeet, independent of age.

Table 19.1 Table of recommendations

Statement	Grade of recommendation
Most flexible flat feet in children do not cause symptoms	B
Shoe-wearing in early childhood is detrimental to the development of a normal longitudinal arch	C
Wearing shoe modifications during childhood is ineffective and unnecessary	B
Wearing shoe modifications during childhood may lead long-term psychosocial effects	I

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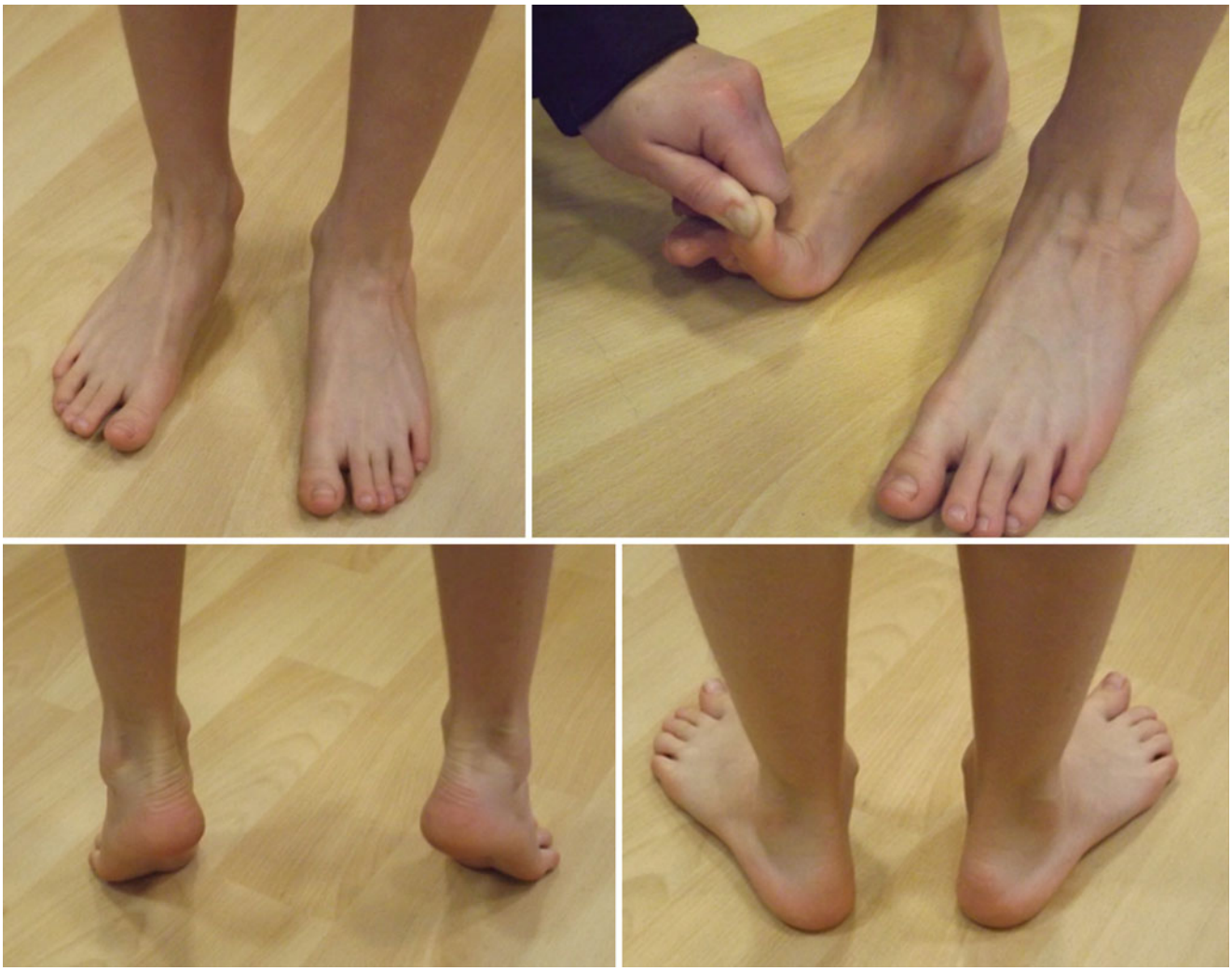

Fig. 19.1 Flexible flat feet, which resolve on tip toeing

Flexible flatfeet account for approximately two thirds of all flat feet and do not seem to cause disability. We acknowledge the importance of identifying within this normal variation any cases with pathological features for a fuller evaluation; specific attention ought to be paid to any with foot pain or stiffness, especially if the subtalar joint is involved. Investigations to identify any tarsal coalition or children with inflammatory joint conditions may be pertinent for these. Feet with these pathological features are not included in this brief review. Children ought to be specifically examined for any contracture of the Achilles tendon associated with a flat foot appearance especially in the older child. It is unclear whether the tendon contracture is primary or a secondary development but attention to improving that aspect seems reasonable.

There are a number of different aetiological causes proposed for paediatric pes planus in the literature. These include clinical pathologies such as ligamentous laxity, neurological and muscular, genetic and collagen disorders. Staheli and colleagues [5] demonstrated that in most children, the arch develops normally by the age of 5 years. They studied the feet of 441 normal subjects, who ranged in age from 1 to 80 years, to document the configuration of the longitudinal arch. They showed that flat feet are usual in infants, common in children, and within the normal range of the observations made in adult feet. Pfeifer et al. [6], reported that whilst 54 % of children aged 3 years had a flat foot appearance only 24 % of children had a flat foot at 6 years of age. These authors also found the incidence was higher in boys and found that children classed as obese were three times as likely to retain flat feet than those with a healthy weight.

There are two main theories described in the literature to explain the development of a flexible flat foot appearance. Basmajian and Stecko [7] believe that the height of the arch is determined by the bone-ligament complex and that muscles only function to maintain balance, propel the body and navigate uneven surfaces rather than determine the shape of the foot. Duchenne [8] and Jones [9] believe that the maintenance of the longitudinal arch is based on muscle strength.

The management of flexible flat foot can vary from conservative to surgical, nevertheless for the vast majority of typical cases reassurance with an explanation of the natural history and education is all that is required. Whilst selected cases may merit follow-up for confirmation, the majority do not require review. In addition to advice, a variety of conservative treatments are described: stretching, activity modification, orthotics, manipulation and casting. Surgical treatment is only selectively employed by some, generally after failure of conservative treatment, for atypical cases with indications being individualised [10].

What Is the Long-Term Effect of Flexible Flat Feet?

There is no good evidence that flexible flat feet cause symptoms. There is anecdotal evidence of an association with knee, foot and leg pain but controlled studies are lacking. The evidence for children with flat feet becoming adults with flat feet is also absent. Esterman and Piletto [2] showed that the shape of a foot in adulthood had no significant effect on pain, injury or function. This included adults with pes planus.

Hogan and Staheli [11] found no relationship between arch configuration and pain scores in adults and suggested that flexible flatfeet are not a source of disability. This study is consistent with previous studies and provides additional evidence against the practice of treating flexible flatfeet in children.

What Is the Effect of Wearing Shoes on Flexible Flat Feet?

Rao and Joseph [12] analysed 2300 children between the ages of 4–13 years and showed that the incidence of flat feet among children who used footwear was 8.6 % compared with 2.8 % in those who did not ($p < 0.001$). Significant differences between the predominance in shod and unshod children were noted in all age groups, most marked in those with generalised ligament laxity. Flat foot was most common in children who wore closed-toe shoes, less common in those who wore sandals or slippers, and least in the unshod. The authors suggested that shoe-wearing in early childhood is detrimental to the development of a normal longitudinal arch. Sachithanandam and Joseph [13] studied 1846 skeletally mature individuals to establish the influence of the age at which shoe-wearing began on the prevalence of flat foot. They showed that the incidence was higher the earlier shoe wearing started and least in those where it had not occurred until they were older: 3.24 % among those who started to wear shoes before the age of six, 3.27 % in those who began between 6–15 years of age and 1.75 % in those who first wore shoes at the age of 16 ($p < 0.001$). Flat foot was most common: in those who, as children, wore footwear for over eight hours each day; in the obese individuals ($p < 0.01$); and in those with ligament laxity ($p < 0.0001$). Even after adjusting for these two variables, significantly higher rates of prevalence were noted among those who began to wear shoes before 6 years old. All this suggests an association between the wearing of shoes in early childhood and flat foot.

What Evidence Is There for the Treatments Available for Flexible Flat Feet?

Whilst management usually consists of reassurance by advice and explanation of the natural history, other measures have been advocated including orthotics, either custom made or prefabricated. These were more frequently employed in the past however in a classic paper Wenger et al. [14] reported a single blinded randomized clinical trial in children from 1 to 6 years old with flexible flat feet, excluding children with any underlying neurological condition. The children were randomised to either: corrective orthopaedic shoes, use of a heel cup, to a custom moulded plastic insole or to a control group who had no treatment. The study looked at the radiological changes that occurred over 3 years and showed that all groups showed improvement. There was no significant difference between the treated patients and the control group. Whitford and Esterman [14, 15] similarly using a single blinded randomized clinical trial looked at a larger series of children with flexible pes planus and compared custom and prefabricated orthotics with a control group. They showed no significant difference in motor proficiency, exercise efficiency, self perception or pain relief; concluding that the use of in-shoe orthotics in the management of flexible flat feet did not improve on the natural history. The absence of benefit raised the consideration of harmful effect of wearing modified shoes. Driano and colleagues [16], in an instructive study, assessed the short – and long-term psychosocial effects of wearing modified shoes during childhood. They compared measures of self-esteem and self-image of adults who wore shoe modifications during childhood with controls who did not. Those who wore shoe modifications showed lower self-esteem than controls ($p < 0.05$) and recalled a decrease in their self-image, and the experience as negative. They recall being teased about their footwear, and their activities were limited. These findings show that wearing shoe modifications during childhood, in addition to being ineffective and unnecessary is a negative experience in childhood and associated with lower self-esteem in adult life. The cost of using unnecessary ineffective treatments comes under increasing scrutiny.

Staheli [17] concluded that the assessment of children with flatfoot aims to identify the occasional pathological foot, typified by stiffness which causes disability and merits treatment. In contrast, the physiological flatfoot is a common variant of normal, which is not associated with disability and warrants reassurance. The physiologic flatfoot is most prevalent in those who are naturally loose-jointed, are obese, or had usually worn shoes early in childhood. Orthotic or shoe modifications are not simply ineffective and an unnecessary cost but more importantly reduce footwear comfort for the child, are embarrassing for them and risk leading to lowered self-esteem in adult life.

References

1. Cowan DN, Robinson JR, Jones BH, et al. Consistency of visual assessments of arch height among clinicians. Foot Ankle Int. 1994;15:213–7.
2. Elftman H, Manter J. The Evolution of the human foot, with especial reference to the joints. J Anat. 1935;70:56–67.
3. Bates KT, Collins D, Savage R, et al. The evolution of compliance in the human lateral mid-foot. Proc Biol Sci. 2013;280:20131818. doi:10.1098/rspb.2013.1818.
4. Harris RI, Beath T. Army foot survey: an investigation of foot ailments in Canadian soldiers. Ottawa: National Research Council of Canada. 1947.
5. Staheli LT, Chew DE, Corbett M. The longitudinal arch. A survey of eight hundred and eighty-two feet in normal children and adults. J Bone Joint Surg Am. 1987;69:426–8.
6. Pfeiffer M, Kotz R, Ledl T, et al. Prevalence of flat foot in preschool-aged children. Pediatrics. 2006;118:634–639 doi:118/2/634 [pii].
7. Basmjian JV, Stecko G. The role of muscles in arch support of the foot. J Bone Joint Surg Am. 1963;45:1184–90.
8. Duchenne G, Kaplan E. Physiology of motion demonstrated by means of electrical stimulation and clinical observation and applied to the study of paralysis and deformities. Philadelphia: W.B. Saunders. 1959.
9. Jones BS. Flat foot. A preliminary report of an operation for severe cases. J Bone Joint Surg Br. 1975;57:279–82.
10. Bouchard M, Mosca VS. Flatfoot deformity in children and adolescents: surgical indications and management. J Am Acad Orthop Surg. 2014;22:623–632. doi:22/10/623 [pii].
11. Hogan MT, Staheli LT. Arch height and lower limb pain: an adult civilian study. Foot Ankle Int. 2002;23:43–7.
12. Rao UB, Joseph B. The influence of footwear on the prevalence of flat foot. A survey of 2300 children. J Bone Joint Surg Br. 1992;74:525–7.
13. Sachithanandam V, Joseph B. The influence of footwear on the prevalence of flat foot. A survey of 1846 skeletally mature persons. J Bone Joint Surg Br. 1995;77:254–7.
14. Wenger DR, Mauldin D, Speck G, et al. Corrective shoes and inserts as treatment for flexible flatfoot in infants and children. J Bone Joint Surg Am. 1989;71:800–10.
15. Whitford D, Esterman A. A randomized controlled trial of two types of in-shoe orthoses in children with flexible excess pronation of the feet. Foot Ankle Int. 2007;28:715–723 doi:958189 [pii].
16. Driano AN, Staheli L, Staheli LT. Psychosocial development and corrective shoewear use in childhood. J Pediatr Orthop. 1998;18:346–9.
17. Staheli LT. Planovalgus foot deformity. Current status. J Am Podiatr Med Assoc. 1999;89:94–9. doi:10.7547/87507315-89-2-94.

Evidence-Based Treatment for Tarsal Coalition

20

Anastasios Chytas, Samena Chaudhry, and Sattar Alshryda

Abstract

Tarsal coalition is an abnormal connection between two or more bones; commonly in the foot. The connection may be fibrous, cartilaginous or bony. The commonest types are calcaneonavicular followed by talocalcaneal. Majority are asymptomatic and require no surgery (grade B). Non-operative treatment should be tried first in symptomatic patients (grade B/C). Surgical treatments include resection, resection with graft interposition (could be tendon, fatty tissue or bone wax) or arthrodesis. Resection with or without graft indicated in persistently symptomatic patients when there are no secondary degenerative changes (grade B). Persistent symptoms after resection are correlated to the severity of associated deformity, particularly the amount of the heel valgus. Correcting associated deformity may improve symptoms (grade C). Arthrodesis is indicated when severe symptoms are caused by secondary degenerative changes (grade C) (Table 20.1).

Keywords

Tarsal coalition • Talocalcaneal coalition • Calcaneonavicular coalition • Calcaneal osteotomy

Background

Tarsal coalition is an abnormal connection between two or more tarsal bones of the foot that may produce pain and limitation of foot motion [1]. This connection may be complete or partial and involves predominantly the joints of the hindfoot and midfoot [2]. A coalition can be fibrous, cartilaginous or a bony connection as a result of a failure of differentiation and segmentation of mesenchyme [3].

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The incidence of tarsal coalitions is considered to be less than 1 % in the literature [4], but cadaveric and advanced imaging studies have increased this incidence up to 13 % [5]. The most common coalitions are calcaneonavicular (53 %) and talocalcaneal (37 %). Talonavicular, calcaneocuboid, naviculocuneiform and cubonavicular coalitions are less common [6]. Tarsal coalitions seem to be bilateral in 50–60 % of cases [3].

Tarsal coalitions can be acquired secondary to infection, trauma, surgery, arthritis or neoplasia but these are rare. Congenital coalitions have been reported to be the most prevalent in adolescents [7]. It has been found that an inherited defect in genetic coding can result in tarsal coalition. Leonard [6] described an autosomal dominant pattern of inheritance with almost full penetrance. Thirty-nine percent of first degree relatives may have tarsal coalitions [6]. Mosca has described tarsal coalition as a developmental malformation instead of congenital deformity, as it is not present at birth but it is genetically programmed to develop. Tarsal coalitions may be present with other congenital disorders

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Table 20.1 Table of recommendations

Statement	Grade of recommendation
Not all patients with tarsal coalition will develop symptoms	B
A symptomatic patients do not require any treatment	B
Non operative treatments should be tried first before considering surgical treatment	B/C
Resection is recommended in persistently symptomatic patients	B
The superiority of interposition graft materials has not been established	I
Severe associated deformity and degenerative changes are predictors for persistent symptoms	C
Severe associated deformity is an indication for corrective osteotomy	C
Secondary degenerative changes are indication for arthrodesis	C

such as fibular hemimelia, clubfoot, Apert syndrome, and Nievergelt-Pearlman syndrome, involving multiple bony connections [3].

Most patients are asymptomatic. Only 25 % will go on to develop symptoms and this usually happens between 8 and 12 years for children with calcaneonavicular coalitions [5] and between 12 and 16 years for children with talocalcaneal coalitions [7]. Progressive flattening of longitudinal arch and later onset of aching pain, are the main clinical features of tarsal coalition. Symptoms are activity related and are improving with rest. Tip toeing and Jack's test will not reconstitute the medial arch due to rigidity of subtalar joint (Fig. 20.1). The term "peroneal spastic flat foot" has been misused in the past as a synonym to tarsal coalition, but it describes a rigid flat foot with involuntary contracture of peroneal tendons that may exist without tarsal coalition [3]. Mubarak et al. [8] also noticed that plantar flexion may be decreased in calcaneonavicular coalitions apart from subtalar joint movements.

Weight bearing radiographs of the foot anteroposterior, lateral and oblique 45° can diagnose a calcaneonavicular coalition with the oblique being the most helpful view [9]. On oblique and lateral radiographs "anteater's sign" is diagnostic for calcaneonavicular coalition [10]. In addition to previous radiographs, the Harris axial view can help in identifying talocalcaneal coalitions [11]. An anteroposterior ankle view could show a ball-and-socket type joint in a long-standing tarsal coalition. A C-sign on the lateral view can be indicative of talocalcaneal coalition, but since it has also been found to be positive in flexible flat foot, it is considered neither sensitive nor specific for the diagnosis of talocalcaneal coalition [12]. Dorsal talar beaking is another obvious sign on the lateral view. This is considered a traction spur rather than degenerative disease [13]. The gold standard imaging modality to diagnose tarsal coalition is Computed

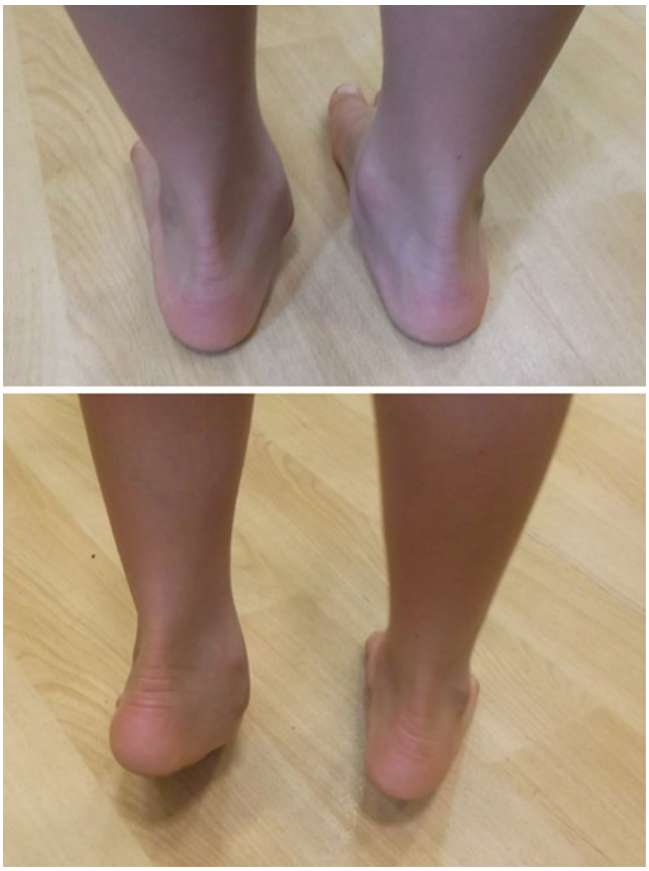

Fig. 20.1 Clinical photograph of a child with tarsal condition. Heels should move to varus and the medial arches are reconstituted on tip toeing. This does not usually happen when there is a tarsal coalition as in the left of this child

Tomography which can offer also information regarding the size and location, as well as other concurrent coalitions (Figs. 20.2 and 20.3) [14]. Upasani et al. [15] suggested that a CT scan should be performed in order to assess if there is any concurrent osteoarthritis of adjacent joints as well as for preoperative planning as persistent postoperative symptoms may relate to inadequate resection. MRI scanning has comparable results with CT scan and may be useful for early identification of fibrous coalitions [16, 17].

Should We Treat Non-symptomatic Coalitions?

Only 25 % of patients with tarsal coalitions will develop symptoms [6]. There is no published evidence to support that painless tarsal coalition will become symptomatic and disabling in the long term. On the other hand, it is unknown if resection of symptom free tarsal coalitions will lead to better outcome in comparison to natural history [3]. For this reason, treatment should be attempted only for symptomatic coalitions.

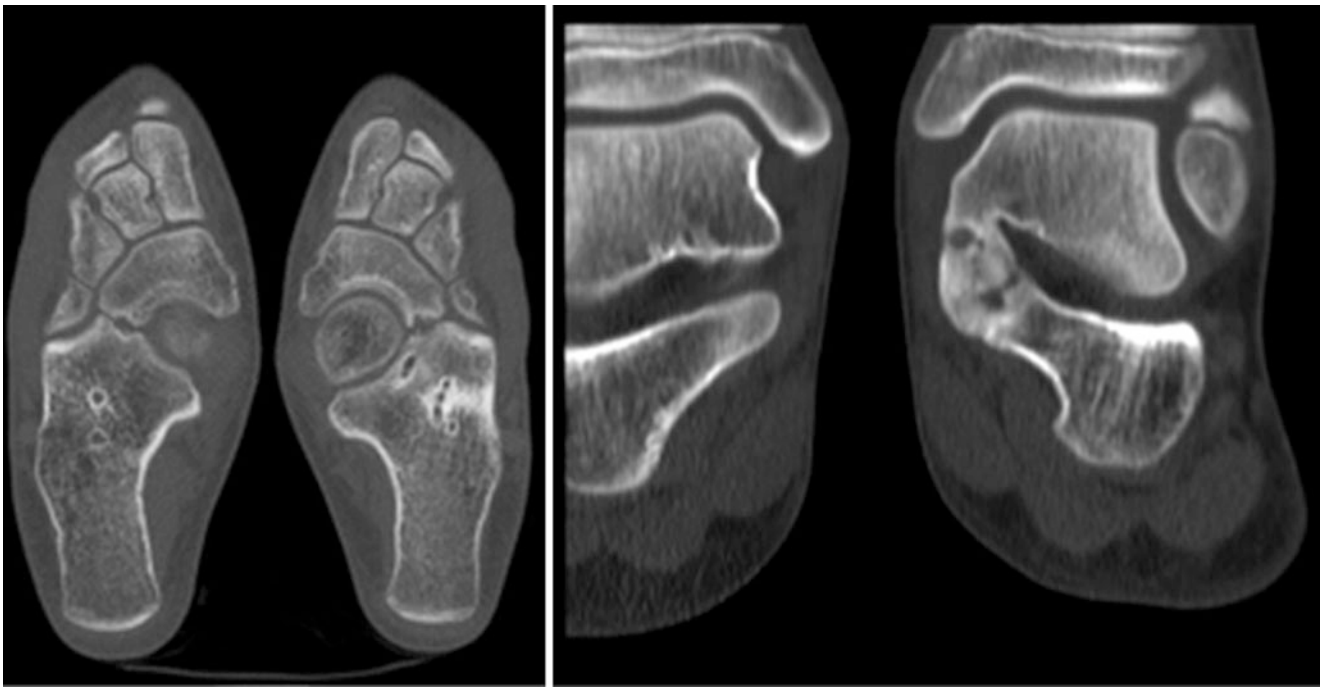

Fig. 20.2 CT scan showing bilateral calcaneonavicular tarsal coalition (*left*) and right talocalcaneal tarsal coalition (*left*)

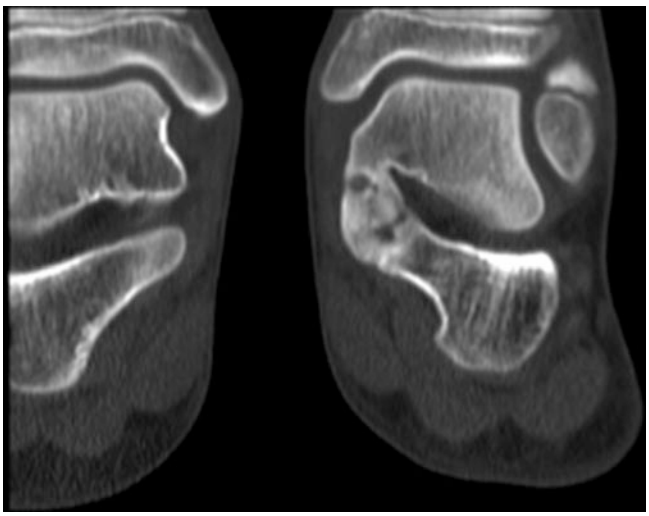

Fig. 20.3 Talocalcaneal tarsal coalition

Is There Any Role for Non-operative Treatment?

It is often suggested that non-operative management should be always used prior to surgical interventions for symptomatic tarsal coalitions. Activity modifications, non steroidal anti-inflammatory drugs and orthotics may provide symptomatic relief. Flat insoles without firm of hard arch have been suggested in order to achieve additional cushioning support for the treatment of talocalcaneal coalitions [3]. A trial of short leg walking cast with hindfoot in neutral position for 4–6 weeks can also provide with symptomatic relief. Thirty percent of patients will remain symptom free after cast immobilization [18]. Calcaneonavicular coalitions are less likely to respond to casting in comparison to talonavicular coalitions [19].

What Is the Best Operative Treatment for Calcaneonavicular Coalitions?

Those patients who have persistent and disabling pain refractory to non-operative measures are usually candidates for operative treatment. The main aim of operative treatment is pain relief [3]. Asymptomatic and minimal symptomatic patients should not be offered an operative intervention [19].

Badgley [20] was the first to describe resection of calcaneonavicular coalitions. Mitchell and Gibson [21] published a case series with simple excision of calcaneonavicular bars with full recurrence of coalition in one third of the patients and partial recurrence in another third of patients. The high recurrence rates led Cowell [22] to modify this technique with interposition of the extensor digitorum brevis (EDB) in the gap of resection. Gonzalez et al. [19] presented their experience with the above procedure and showed excellent results in 77 % with 23 % recurrence rate. Moyes et al. [23] compared their results between patients who had EDB interposition and those who didn't after calcaneonavicular coalition resection and suggested the interposition of EDB as

important part of the operation. Mubarak et al. [8] with a cadaveric study showed that EDB muscle is not enough to fill the gap after resection of calcaneonavicular coalition and presented his results with the use of free fat graft as an interposition material showing lower reossification rates (13 %) and good long term pain relief. Most of other studies have shown encouraging results with resection of calcaneonavicular coalition followed by interposition of either fat tissue or EDB [13, 24, 25].

Recently, a few case reports and small case series have been published suggesting the endoscopic resection of calcaneonavicular coalition as an alternative to open resection, resulting in good outcome with symptomatic relief and improvements in functional scores [26–31]. Larger patient series treated with arthroscopic coalition resection with long-term follow up as well as comparative studies with open resection patient series will be needed to determine the outcome of this method and the true benefit over standard of care.

Significant degenerative arthritis of talonavicular and calcaneocuboid joints are contraindications for the resection of calcaneonavicular coalition and thus preoperative CT scan is imperative [5]. In case of concurrent joint degeneration or failure of complete resection of the coalition that leads to disabling pain, triple arthrodesis has been proposed [5]. Triple arthrodesis has been used as a salvage operation after failure of coalition resection to relief symptoms with satisfactory results [13, 19, 21, 32]. Most case series with primary triple arthrodesis for calcaneonavicular coalition are involving mixed population with adults and show that this operation provides adequate pain relief but has unpredictable functional outcome [13, 14]. The good results from treating calcaneonavicular coalitions with resection and the reluctance of surgeons to perform triple arthrodesis in children due to poor long term outcome made primary triple arthrodesis a rarely used alternative [3] (Table 20.2).

What Is the Best Operative Treatment for Talocalcaneal Coalitions?

Surgery is indicated for those with persistent disabling pain despite attempts at non operative management. The main treatment options include resection of the coalition, osteotomy or arthrodesis. Currently, most symptomatic TCCs are treated with coalition resection and fat-graft interposition.

In 1994, Wilde and colleagues [33] reviewed their results from treating TCCs and found worse results in patients with a CT scan showing a coalition area of greater than 50 %. In their study, these patients were also found to have heel valgus more than 16°, narrowing of the posterior talocalcaneal joint, and impingement of the lateral talar process of the calcaneus. In contrast, patients who had coalitions with a relative area less than 50 % were associated with heel valgus less than 16°, normal thickness of the posterior talocalcaneal joint, and absence of impingement of the lateral talar process on the calcaneus and did better with resection. In the cases with a relative coalition area greater than 50 %, they recommended arthrodesis as opposed to resection.

Downey [34] published three key parameters in determining the appropriateness of a surgical resection versus arthrodesis: skeletal maturity of the patient, the site of the coalition whether intra-articular or extra-articular and the presence of secondary arthritic changes are present. In his opinion, patients that are skeletally immature are the most likely to benefit following resection. However, the presence of significant secondary arthritic changes, particularly in an adult, is a strong indication for arthrodesis.

There have been many different techniques for coalition resections that have been described. There are also many different substances available to help prevent reformation of the union, with bone wax and autogenous fat graft interposition being the most common [35, 36].

Others have reported different ways of maintaining separation between the previous coalition sites with similar rates of success. Raikin et al. [37] reported that 13 of 14 patients who had undergone middle facet talocalcaneal coalition resection had better function with split FHL interposition. A lateral arthroereisis procedure was used by Giannini et al. [38] on 14 feet with talocalcaneal coalitions in patients between 9 and 18 years of age, and also demonstrated favorable results.

Kumar et al. [39] reported satisfactory results in 88.9 % of patients who underwent resection of a talocalcaneal coalition combined with no interposition, fat graft interposition, or split FHL interposition. The patient's age, tissue type, and interposition method did not seem to influence the result. However, postoperative pain with activity was not uncommon, and would seemed to indicate that even when range of motion is restored, the quality of the subtalar joint articular surfaces plays an important role in prognosis. Therefore, arthroscopic assessment of the subtalar joint before resection of any coalition would seem appropriate.

Luhmann and Schoenecker [40] recommended that all pediatric and adolescent patients with a symptomatic TCC that failed non operative treatment and did not have an arthritic hindfoot should be treated with a talocalcaneal resection as opposed to arthrodesis. They further concluded that patients with TCCs greater than 50 % or heel valgus greater than 21° could still have a very satisfactory outcome. However, they did finally establish that those patients with severe hindfoot valgus should undergo either non operative treatment with the use of an orthosis or operative treatment of the deformity with a calcaneal osteotomy or lateral column lengthening, and that a hind foot arthrodesis can be held

Table 20.2 Treatment of calcaneonavicular coalitions in children and adolescents

Studies	No (feet)	Type of primary surgery	Additional surgery	FU(Y)	Age (Y)	Outcome	Recurrence	Complications	LOE
Mitchell and Gibson [21]	41	Open resection	Nine feet had revision to triple arthrodesis	6	10–14	68 % complete symptoms relief 58 % regained subtalar ROM	1/3 complete recurrence 1/3 incomplete		IV
Mubarak et al. [8]	96 (78 FU)	Open resection and fat graft interposition	5 % needed additional osteotomies for deformity correction	29 m	7–17	87 % excellent 8 % fair 5 % failures	13 %		IV
De Vriese et al. [24] (includes adults)	18	Open resection and EDB interposition		3 (1–10)	14.2 (9–40)	10 excellent 4 good 1 fair 3 poor	16 %		IV
	1	Open resection and sinus tarsi spacer	Removal of spacer			1 poor	100 %		
	2	Triple arthrodesis (one adult after failed resection)				?			
Gonzalez and Kumar [19] (includes Cowel's patient series)	75	Open resection and EDB interposition	Five feet had triple arthrodesis	2–23	11.2 (8–17)	77 % excellent – good 16 % fair 7 % poor	23 % partial recurrence		IV
Swiontkowski et al. [13] (includes adults)	39	Open resection and interposition of fat graft or EDB	Two had triple arthrodesis and one repeat resection	4.6	12 years (8–54)	35 improved Four deteriorated	?		IV
	5	Triple arthrodesis				All patients had pain relief			
Inglis et al. [32]	16 (11 patients)	Open resection	Three feet had triple arthrodesis and two feet had repeat resection	23		Eight patients satisfactory results Three patients unsatisfactory	25 %	?	IV
Moyes et al. [23]	10	Open resection with EDB interposition	Nil	3.4	12	Nine patients pain free with good ROM	No	One patient pain due to degeneration	IV
	7	Open resection				Four patients had no symptoms	Three patients		
Khoshbin et al. [25]	19	Open resection with EDP interposition	One concurrent calcaneal osteotomy and one TA lengthening	15.3	11.8	Median (mean) AAOS-FACS 89 (83.6)	?	No complications	III
	1	Open resection with bone wax/fat interposition					?		

(continued)

Table 20.2 (continued)

Studies	No (feet)	Type of primary surgery	Additional surgery	FU(Y)	Age (Y)	Outcome	Recurrence	Complications	LOE
Knorr et al. [28]	3	Arthroscopic resection		1	11–15	AOFAS Mean From 58 preop to 91 at 1 year	No	No	IV
Singh and Parsons [29]	2	Arthroscopic resection		6–9 m	11–14	MOXFQ mean Score from 58 preop to 17	No	No	IV
Molano-Bernardino et al. [30]	1	Arthroscopic resection		2	12	AOFAS From 55 preop to 100 at 2 years	No	No	IV
Nehme et al. [31]	2	Arthroscopic resection		5	13	AOFAS From 23 preop to 82 at 2 years	No	No	IV

as a salvage procedure if a TCC resection fails or the patient has an arthritic hindfoot.

In 1997, McCormack and colleagues [41] reviewed nine symptomatic TCCs that underwent complete resection with fat-graft interposition with a mean duration of follow-up of 11.5 years. In this study, patients had no limitation in range of motion and showed no evidence of degenerative change or joint space narrowing on radiographs indicating that resection of symptomatic TCC provides satisfactory results in the majority of patients, and its benefits are maintained 10 years after the procedure.

Mosca et al. [42] reviewed the short-term to intermediate-term results of 8 patients with 13 symptomatic TCCs that underwent calcaneal lengthening osteotomy for deformity correction with or without coalition resection. In this study, the investigators adhered to recently proposed criteria to determine if a TCC is resectable, including less than 50 % the surface area of the posterior facet, less than 16° of hind foot valgus, and with minimal or no narrowing of the posterior facet of the subtalar joint. In this series, they concluded that a calcaneal lengthening osteotomy is a desirable alternative to triple arthrodesis for a painful foot with severe hind foot valgus deformity and an unresectable, solid talocalcaneal tarsal coalition.

Many authors believe that that TCC and a valgus hindfoot are two separate conditions and recommend resection of the coalition to gain motion and then performing a calcaneal cuboid or cuneiform osteotomy 6–12 months later to correct the flatfoot deformity if painful [3].

A triple arthrodesis can be a good salvage procedure in the older patient with degenerative changes or in those who have failed a previous resection. But the evidence suggests, only a 20-year life span before ankle arthritis causes this to fail [43] (Table 20.3).

Table 20.3 Treatment of talocalcaneal coalitions in children and adolescents

Studies	No (feet)	Type of primary surgery	Additional surgery	FU (Y)	Age (Y)	Outcome	Recurrence	Complications	LOE
Olney and Asher [44]	9	Resection with bone wax and fat	None	3.5	14	5 excellent 3 good 1 fair 1 poor	One re-resection		IV
Kumar et al. [39]	18	Resection with 50 % having superior half FHL interposition	None	4	14	8 excellent 8 good 1 fair 1 poor	One re-resection		IV
Wilde et al. [33]	20	Resection and fat graft	None	12	13	10 excellent 10 fair or poor		Two inadequate resections	IV
Luhmann and Schoenecker [40]	25	Resection and fat graft	Peroneal or FHL/EDL lengthening Lateral column lengthening	15	12.5	48 % very satisfied 28 % satisfied 8 %unsat	3	Requiring triple arthrodesis	IV
McCormack et al. [41]	10	Resection and fat graft	1 had hindfoot osteotomy	10	11.5	7 excellent 2 fair 1 poor		1	IV
Comfort and Johnson [45]	23	Resection and fat graft	None	29 m	14	60 % excellent or good 40 % fair or poor	Four requiring re-resection or fusion		IV
Raikin et al. [37]	14	Resection with FHL interposition	None	4	14	11 excellent 1 good 1 fair 1 poor			IV
Scranton [46]	23	Resection	Triple arthrodesis in four	3.9	15	13 good 10 satisfactory			
Jagodzinski et al. [47]	9	Arthroscopic resection	None	12–66 m	15	Mean improvement of SAFAS 7.9–3.6 for seven patients	One recurrence	One posterior tibial nerve damage One hypertrophic scar – hypersensitivity	IV
Khoshbin et al. [25]	13	Resection	One calcaneal osteotomy	13.1	11.9	Mean AOFAS FACS score 79 and SFCS 84			IV

References

1. Herring JA, editor. Tarsal coalitions in: Tachdjian's pediatric orthopaedics: from the Texas Scottish Rite Hospital for Children. Philadelphia: Elsevier Saunders; 2014. p. 827–33.
2. Zaw H, Calder JD. Tarsal coalitions. Foot Ankle Clin. 2010;15(2):349–64.
3. Mosca V. Subtalar coalition in paediatrics. Foot Ankle Clin. 2015;20(2):265–81.
4. Stormont D, Peterson HA. The relative incidence of tarsal coalition. Clin Orthop Relat Res. 1983;181:28–36.
5. Swensen S, Otsuka NY. Tarsal coalitions – calcaneonavicular coalitions. Foot Ankle Clin. 2015;20(4):669–79.
6. Leonard M. The inheritance of tarsal coalition and its relationship to spastic flat foot. J Bone Joint Surg Br. 1974;56(3):520–6.
7. Murphy J, Mubarak SJ. Talocalcaneal coalitions. Foot Ankle Clin. 2015;20(4):681–91.
8. Mubarak S, Patel PN, Upasani VV. Calcaneonavicular coalition: treatment by excision and fat graft. J Pediatr Orthop. 2009;29(5):418–26.
9. Newman J, Newberg AH. Congenital tarsal coalition: multimodality evaluation with emphasis on CT and MRI imaging. Radiographics. 2000;20(2):321–32.
10. Crim J, Kjeldsberg KM. Radiographic diagnosis of tarsal coalition. AJR Am J Roentgenol. 2004;182(2):323–8.
11. Harris R, Beath T. Etiology of peroneal spastic flat foot. J Bone Joint Surg Br. 1948;30(4):624–34.
12. Brown R, Rosenberg ZS, Thornhill BA. The C sign: more specific for flatfoot deformity than subtalar coalition. Skeletal Radiol. 2001;2001(30):84–7.
13. Swiontkowski M, Scranton PE, Hansen S. Tarsal coalitions: long-term resuts of surgical treatment. J Pediatr Orthop. 1983;3(3):287–92.
14. Deutsch A, Resnick D, Campbell G. Computed tomography and bone scintigraphy in the evaluation of tarsal coalition. Radiology. 1982;144(1):137–40.
15. Upasani V, Chambers RC, Mubarak SJ. Analysis of calcaneonavicular coalitions using multi-planar 3-dimensional computed tomography. J Child Orthop. 2008;2(4):301–7.
16. Emery K, Bisset III GS, Johnson ND, Nunan PJ. Tarsal coalition: a blinded comparison of MRI and CT. Pediatr Radiol. 1998;28(8):612–6.
17. Guignard D, Journeau P, Mainard-Simard L, Popkov D, Haumont T, Lascombes P. Child calcaneonavicular coalitions: MRI diagnostic value in a 19-case series. Orthop Traumatol Surg Res. 2011;97(1):67–72.
18. Jayakumar S, Cowel HR. Rigid flatfoot. Clin Orthop Relat Res. 1977;122:77–84.
19. Gonzalez P, Kumar SJ. Calcaneonavicular coalition treated by resection and interposition of the extensor digitorum brevis muscle. J Bone Joint Surg Am. 1990;72(1):71–7.
20. Badgley C. Coalition of the calcaneus and the navicular. Arch Surg. 1927;15(1):75.
21. Mitchell G, Gibson JM. Excision of calcaneonavicular bar for spasmodic flat foot. J Bone Joint Surg Br. 1967;49:281–7.
22. Cowell H. Extensor brevis arthroplasty. J Bone Joint Surg Am. 1970;82:820.
23. Moyes S, Crawfurd EJ, Aichroth PM. The interposition of extensor digitorum brevis in the resection of calcaneonavicular bars. J Pediatr Orthop. 1994;14(3):387–8.
24. De Vriese L, Dereymaeker G, Molenaers G, Fabry G. Surgical treatment of tarsal coalitions. J Pediatr Orthop B. 1994;3:96–101.
25. Khoshbin A, Law PW, Caspi L, Wright JG. Long-term functional outcomes of resected tarsal coalitions. Foot Ankle Inter. 2013;34(10):1370–5.
26. Lui T. Arthroscopic resection of the calcaneonavicular coalition or the "too long" anterior process of the calcaneus. Arthroscopy. 2006;22(8):903 .e1–4
27. Bonasia D, Phisitkul P, Amendola A. Endoscopic coalition resection. Foot Ankle Clin. 2015;20(1):81–91.
28. Knorr J, Accadbled F, Abid A, Darodes P, Torres A, Cahuzac JP, Sales de Gauzy J. Arthroscopic treatment of calcaneonavicular coalition in children. Orthop Traumatol Surg Res. 2011;97(5):565–8.
29. Singh A, Parsons SW. Arthroscopic resection of calcaneonavicular coalition/malunion via a modified sinus tarsi approach: an early case series. Foot Ankle Surg. 2012;8(4):266–9.
30. Molano-Bernardino C, Colano P, Garcia MA, Lopez-Vidriero E. Experimental model in cadavera of arthroscopic resection of calcaneonavicular coalition and its first in-vivo application: preliminary communication. J Pediatr Orthop B. 2009;18(6):347–53.
31. Nehme A, Monsef JB, Ghannam AGB, Imad JP, Moucharafieh R, Wehbe J. Arthroscopic resection of a bilateral calcaneonavicular coalition in a child. J Foot Ankle Surg. 2016;55(5):1079–82.
32. Inglis G, Buxton RA, Macnicol MF. Symptomatic calcaneonavicular bars: the results 20 years after surgical excision. J Bone Joint Surg Br. 1986;68B(1):128–31.
33. Wilde P, Torode IP, Dickens DR, Cole WG. Resection for symptomatic talocalcaneal coalition. J Bone Joint Surg Br. 1994;76(5):797–801.
34. Downey M. Tarsal coalitions. A surgical classification. J Am Podiatr Med Assoc. 1991;81(4):187–97.
35. Cowell H, Elener V. Rigid painful flatfoot secondary to tarsal coalition. Clin Orthop Relat Res. 1983;177:54–60.
36. Zhou B, Tang K, Hardy M. Talocalcaneal coalition combined with flatfoot in children: diagnosis and treatment: a review. J Orthop Surg Res. 2014;9:129.
37. Raikin S, Cooperman DR, Thompson GH. Interposition of the split flexor hallucis longus tendon after resection of a coalition of the middle facet of the talocalcaneal joint. J Bone Joint Surg Am. 1999;81(1):11–9.
38. Giannini S, Ceccarelli F, Vannini F, Baldi E. Operative treatment of flatfoot with talocalcaneal coalition. Clin Orthop Relat Res. 2003;411:178–87.
39. Kumar S, Guille JT, Lee MS, Couto JC. Osseous and non-osseous coalition of the middle facet of the talocalcaneal joint. J Bone Joint Surg Am. 1992;74(4):529–35.
40. Luhmann S, Schoenecker PL. Symptomatic talocalcaneal coalition resection: indications and results. J Pediatr Orthop. 1998;18(6):748–54.
41. McCormack T, Olney B, Asher M. Talocalcaneal coalition resection: a 10-year follow-up. J Pediatr Orthop. 1997;17(1):13–5.
42. Mosca V, Bevan WP. Talocalcaneal tarsal coalitions and the calcaneal lengthening osteotomy: the role of deformity correction. J Bone Joint Surg Am. 2012;94(17):1584–94.
43. Pell RT, Myerson MS, Schon LC. Clinical outcome after primary triple arthrodesis. J Bone Joint Surg Am. 2000;82(1):47–57.
44. Olney B, Asher MA. Excision of symptomatic coalition of the middle facet of the talocalcaneal joint. J Bone Joint Surg Am. 1987;69(4):539–44.
45. Comfort T, Johnson LO. Resection for symptomatic talocalcaneal coalition. J Pediatr Orthop. 1998;18(3):283–8.
46. Scranton PJ. Treatment of symptomatic talocalcaneal coalition. J Bone Joint Surg Am. 1987;69(4):533–9.
47. Jagodzinski N, Hughes A, Davis NP, Butler M, Winson IG, Parsons SW. Arthroscopic resection of talocalcaneal coalitions – a bicentre case series of a new technique. Foot Ankle Surg. 2013;19(2):125–30.

Evidence-Based Treatment for Pes Cavus

21

Munier Hossain and James S. Huntley

Abstract

The aim of treatment of Pes cavus deformity is to achieve a plantigrade foot that is mobile and pain-free. The discerning surgeon would wish to know what treatment would achieve this outcome with the best results. Unfortunately a simplistic (evidence-informed) conclusion is not feasible for several reasons: Pes cavus is an anatomical condition with multiple aetiologies which have different natural histories and prognoses. Historically, poliomyelitis and untreated/recurrent clubfoot accounted for a substantial load of Pes cavus patients – but these are now comparatively infrequent; the largest group is 'Neurological', and here the prognosis differs markedly between those with progressive (as opposed to non-progressive) neurological conditions. Patients present at different stages of maturity, with variable severity of deformity; many have had previous surgery. There is a paucity of definitive outcome measures; surgeons have used different instruments to define their surgical outcome, making it problematic to directly compare different treatments. Surgical planning is largely individualised, taking into account the disparity of factors. Surgery can be divided into soft tissue surgery, bony surgery and fusion. Triple arthrodesis has been the mainstay of surgery in the past but has not always produced satisfactory results. Joint-sparing surgery is gaining more popularity, especially where the deformity is flexible. The evidence base is extremely limited.

Keywords

Pes Cavus • Cavo-varus • Charcot-Marie-Tooth • Orthoses • Botulinum toxin • Flexible cavus • Rigid cavus • Tendon transfer • Osteotomy • Arthrodesis

Background

"Pes cavus" means a foot with a high arch deformity. For assessment and management of this deformity it is important to appreciate its *multi-planar* nature [1]. Pes cavus can be due to a high calcaneal pitch (Calcaneocavus) or increased plantar-flexion of the first ray (± fore-/mid-foot pronation) or the forefoot in general (Plantaris); there may be associated hindfoot varus (Cavovarus)/valgus/calcaneus/equinus (Equinocavovarus), and forefoot adductus. Cavovarus deformity is the most common (Fig. 21.1).

In children, Pes Cavus is usually neuromuscular in aetiology, and half of this group has Charcot-Marie-Tooth (CMT) disease [2]; other neurologic or spinal cord conditions present similarly. These disorders give rise to muscle imbalance that produces the observed deformity. The clinical deformity in the cavovarus foot has a characteristic pattern:

- weak intrinsic muscles and relatively spared extrinsics > Claw toes
- weak anterior compartment (Tibialis anterior in particular) and strong Peroneus longus > plantar-flexed first ray > MTPJ extension contracture > contracted plantar fascia > cavus

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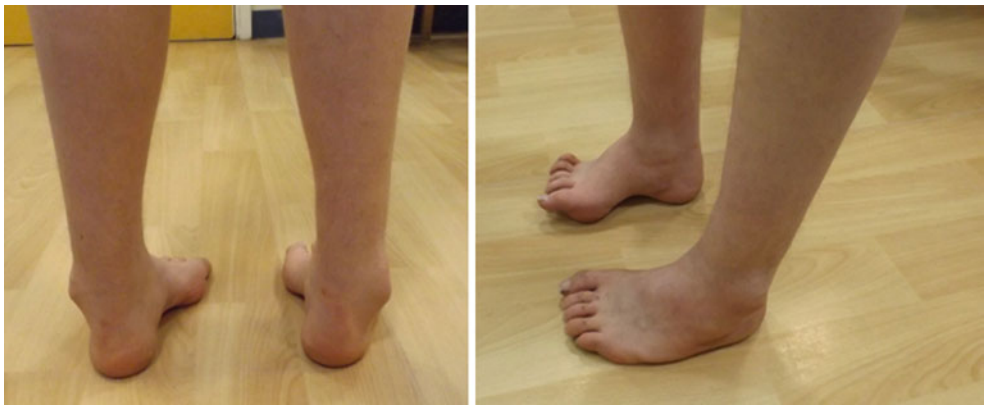

Fig. 21.1 Pes cavus foot, cavovarus type

- weak Tibialis anterior and Peroneus brevis and unopposed Tibialis posterior > varus hindfoot

The diagnosis is clinical but there are established radiological criteria on weight-bearing lateral views of the ankle and foot. The two most commonly used measurements are the *calcaneal pitch* (angle between the inferior surface of the calcaneum and the floor; greater than 30° in Pes cavus) and the *Meary angle* (angle between the long axes of the first metatarsal and the talus; more than 5° in Pes cavus).

Questions

From a clinical perspective, we wished to establish the evidence base for the following questions (the first concerning the role of conservative management, and the latter four on modes of surgical management):

1. Is there a role for conservative treatment in Pes cavus?
2. What is the evidence for the treatment options in management of flexible Pes cavus?
3. What are the results of joint-sparing surgery for management of rigid Pes cavus?
4. What are the results of triple arthrodesis in management of rigid Pes cavus?
5. What are the results of external fixation in management of Pes cavus?

Searching for Evidence

We searched MEDLINE and the Cochrane database for relevant studies and limited our search to the English language. The complete search yielded 325 references. Twelve references were deemed relevant and included for review after screening of the title and abstract. We also checked the reference list of the included articles and used the snowball method for reference harvesting. This generated nine further references for inclusion.

Search was conducted using the following strategy: (09/01/2016)

- Cochrane Database with search term "(Pes Cavus OR Cavo-varus)" three citations
- PubMed search: ("Foot Deformities/surgery"[Mesh]) AND ("Foot Deformities/therapy"[Mesh]) AND ((Randomized Controlled Trial[ptyp] OR Clinical Trial[ptyp] OR Comparative Study[ptyp] OR Observational Study[ptyp]) AND English[lang] AND (infant[MeSH] OR child[MeSH] OR adolescent[MeSH])) 251 citations
- PubMed (www.ncbi.nlm.nih.gov/pubmed/) clinical queries search/systematic reviews: Therapy/Narrow[filter] AND (("foot deformities"[MeSH Terms] OR ("foot"[All Fields] AND "deformities"[All Fields]) OR "foot deformities"[All Fields] OR ("pes"[All Fields] AND "cavus"[All Fields]) OR "pes cavus"[All Fields]) AND ("therapy"[Subheading] OR "therapy"[All Fields] OR "treatment"[All Fields] OR "therapeutics"[MeSH Terms] OR "therapeutics"[All Fields])) AND ((Randomized Controlled Trial[ptyp] OR Clinical Trial[ptyp] OR Comparative Study[ptyp] OR Observational Study[ptyp]) AND English[lang] AND ("infant"[MeSH Terms] OR "child"[MeSH Terms] OR "adolescent"[MeSH Terms])) 71 citations

Is There a Role for Conservative Treatment in Pes Cavus?

There is good quality evidence in support of conservative treatment (rather than no treatment) of Pes cavus, limited by its utility especially in a progressive deformity. In a randomised controlled trial, Burns et al. compared the effect of custom-made orthoses vs sham orthoses in 154 adults with symptomatic bilateral Pes cavus [3]. This trial was conducted on adults, so there are questions over its applicability to children. Foot pain was the primary outcome measure; at 3 months, participants using the custom-made orthoses reported significant reduction in foot pain (difference, 8.3 points; 95 % confidence interval [CI], 1.2–15.3 points;

P = .022). Plantar pressure and functional scores were also improved (although quality of life did not).

A subsequent Cochrane review suggested that off-the-shelf orthotics did not improve clinical outcomes [4]. The same group subsequently conducted a controlled trial to investigate the use of Botulinum toxin type A (BoNT-A) in preventing the progression of cavus deformity in ten children (20 feet) with CMT type 1A [5]. The control leg received no injections. After 2 years, there was no significant difference in radiological alignment, foot posture, ankle flexibility or strength between the sides. The authors concluded that BoNT-A did not affect the progression of cavus deformity in CMT 1A.

Surgical Treatment of Pes Cavus

Surgical treatment can be divided into soft tissue (contracture release, tendon lengthening/shortening/transfer), osteotomy and arthrodesis. These are not mutually exclusive but need to be combined and tailored to the needs of the individual patient. The Coleman block test [6] is an important pre-operative decision-making tool, determining whether the hindfoot varus is correctible (flexible) or fixed (rigid). Hindfoot surgery is unnecessary in a flexible, forefoot-driven Pes cavus, but essential in a rigid hindfoot varus.

A variety of procedures have been described for treatment of both flexible and rigid deformities; no single combination has gained unanimous approval. This is understandable as surgical decision-making takes into account a multiplicity of factors, including: apex of deformity, rigidity of deformity, type of cavus, hindfoot position and flexibility, static/dynamic deformity, muscle strength and joint degeneration etc. Despite the claims of various proponents, it remains unclear as to when is the best time for surgery, or indeed whether early surgery prevents later joint degeneration (Figs. 21.2 and 21.3).

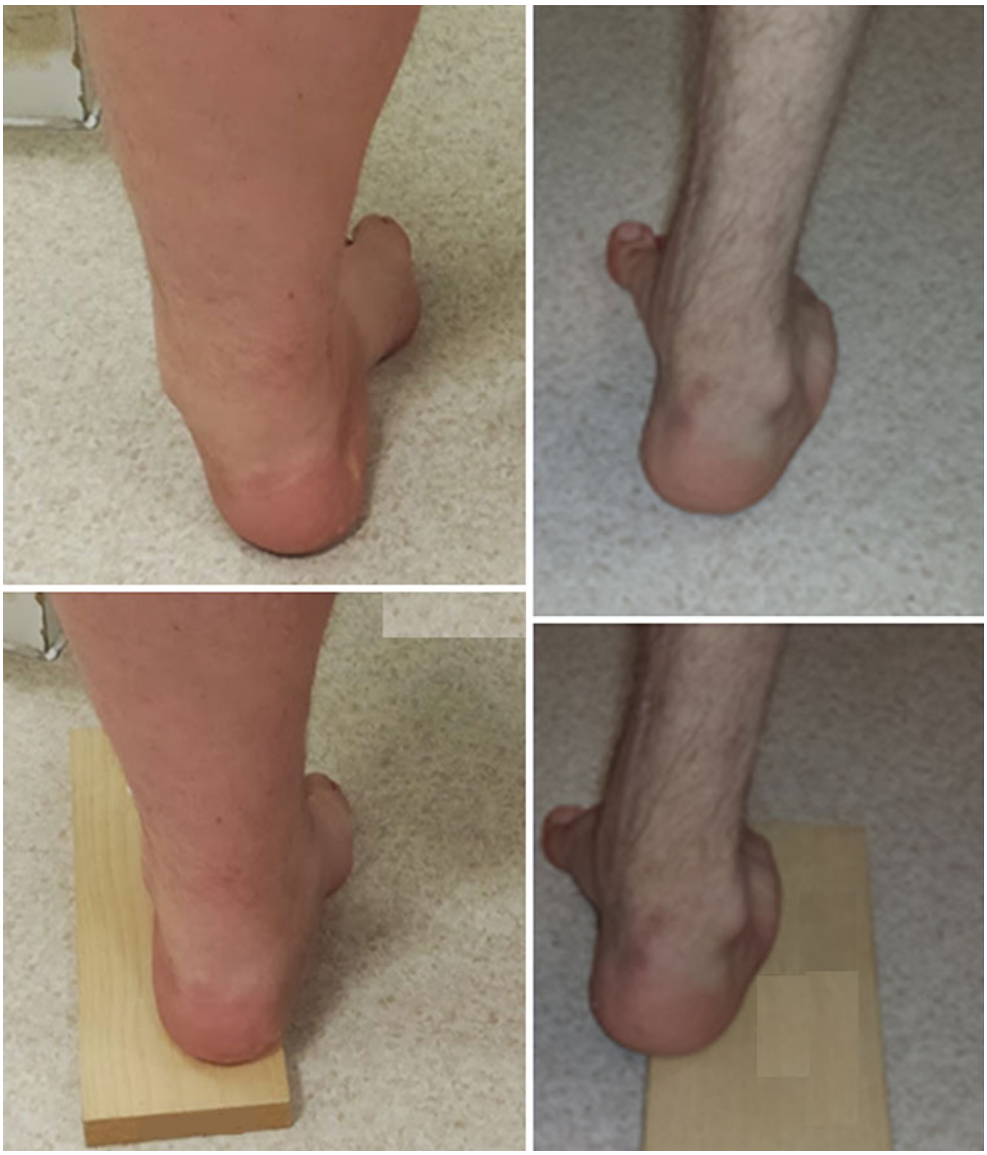

Fig. 21.2 Coleman's test; *left*: hindfoot is flexible. *Right*: hindfoot is fixed

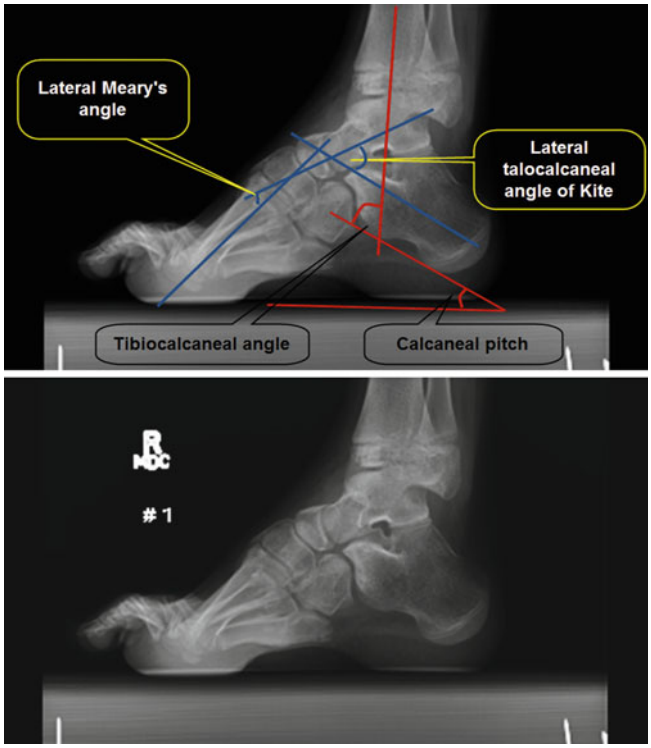

Fig. 21.3 Pes cavus: radiological assessment

What Is the Evidence for the Treatment Options for Management of Flexible Pes Cavus?

In flexible Pes cavus deformity, joint-sparing surgery (purely of soft tissues or with bony surgery also) can be performed, with the intent of preserving joint function. The primary outcome of interest is a pain-free mobile plantigrade foot and, in the longer term, preservation of joint function without degenerative change.

Chan et al. performed a useful study in this regard [7], investigating foot pressure distributions before and after surgical correction in nine children with CMT who had joint sparing surgery. Although surgery improved all radiological parameters, the pressure distributions remained abnormal. They concluded that pressure distribution normalisation depends on achieving a balanced foot and a correct heel position.

Roper and Tiberwal [8] reviewed the results of soft tissue surgery at a mean follow up of 14 years in ten CMT type I patients (mean age at surgery 14 years, but age range 5–36 years) who underwent open Tendoachilles lengthening (TAL), split transfer of Tibialis anterior tendon (TAT), plantar fasciotomy (PF), claw toe release with flexor-extensor transfer and modified Robert Jones procedure for claw hallux correction. Jones originally proposed Extensor Hallucis Longus (EHL) tendon transfer to the first metatarsal neck for claw toe correction [9]. Subsequent modification involved the fusion of the hallux IP joint [10]. Unfortunately, the authors did not indicate the severity of Pes cavus, nor the flexibility of the hindfoot varus; in fact, one patient underwent calcaneal osteotomy for "very severe varus". Outcomes of interest were subjective, including function, appearance and symptoms. Global outcomes were classified as excellent, good, fair or poor. There were no complications. Two patients had recurrent deformity that required repeat soft tissue surgery. All patients had satisfactory results and were able to walk "unlimited distances" on latest review.

In an informative level IV study, Ward et al. [11] described the results (at a mean follow up of 26 years) of joint-sparing surgery in 25 patients with CMT who had a flexible hindfoot [mean age at surgery 15.5 years (8.7–25.1)]. The authors developed an algorithmic approach to flexible Pes cavus management. All patients underwent PF (to reduce the cavus deformity) and transfer of Peroneus longus (PL) tendon to Peroneus brevis (PB) (to remove the deforming force on the first ray). Most patients also underwent first metatarsal osteotomy (DFO), if the foot was deemed not plantigrade following the initial procedures. If there was clawing of the great toe patients underwent Extensor Hallucis Longus (EHL) recession. Those with pre-operative power of at least grade IV in Tibialis anterior underwent TAT transfer to the lateral cuneiform to supplement eversion strength; the transfer of the TAT was not part of the authors' initial practice but was subsequently included, and three patients underwent secondary tendon transfer. Overall, effectively deviating from the latterly advised protocol, TAT transfer was performed to the cuboid or the middle cuneiform in nine patients (14 feet). Some patients had other additional surgery (hallux IPJ fusion – 6 feet, TAL – 1 foot). Seventeen patients (29 feet) had both clinical and radiological assessment. Seven patients (8 feet) required 11 subsequent operations of which there was one calcaneo-cuboid fusion and one ankle fusion. Eleven patients required orthosis at follow-up. General health (SF-36) score means were 49.8 (mental component score) and 37.7 (physical functioning score). Foot function was assessed using the Foot function index (FFI), having three sub-scales of pain, disability and activity limitation (maximum score is 100 with higher scores indicating worse function). Mean scores in the three sub-scales were 35, 40.5 and 22.1, respectively. Twenty-one patients had some degree of hindfoot varus although cavus correction was well maintained. Osteoarthritis (OA) was seen most commonly at the medial cuneiform-first metatarsal joint; 11 joints in 8 feet demonstrated OA.

Chatterje and Sahu reported the results of midfoot osteotomy in 18 adolescents [mean operative age 11.3 years (range: 8.6–15 years); mean follow-up 5.4 years, with no loss to follow-up] who had unilateral Pes cavus (all but one

Table 21.1 Summary of the results of surgery for management of flexible Pes cavus (all level IV case series)

Study	N (feet)	Diagnosis	Treatment	Follow up in years Mean (range)	Re-operations (triple fusion)	Outcome
Roper and Tiberwal [8]	10 (18)	CMT Type 1	TAL, split transfer of TAT, PF, claw toe release and modified Robert Jones procedure	14 (6–32)	3 (0)	All "satisfactory"
Ward et al. [11]	25 (41)	CMT	PF, Transfer to PL to PB, EHL recession, transfer of TAT, DFO 1st MT	26 (9.9–33.5)	11(0)	21 hindfoot varus Significant OA in 8 feet (11 joints)
Chatterjee and Sahu [12]	18 (18)	Post Polio 17 Meningocele 1	Japas osteotomy (included PF) Tendo Achilles lengthening [13]	5.4 (3–13)	4 (2)	Very good: 6 Good: 8 Poor: 4
Leeuwesteijn et al. [13]	33 (52)	CMT	DFO 1st MT and soft tissue surgery	4.7 (1–13)	10(2)	90 % satisfied with deformity correction
Faldini et al. [14]	12 (24)	CMT 1A	PF, MTO and NCA Jones procedure and DFO 1st MT	6 (2–13)	0	Mean MFS improvement: 14 points

following poliomyelitis; the other having meningocoele) [12]. Patients were treated with the Japas osteotomy (midfoot osteotomy with the apex placed over the navicular) after initial open PF release. Thirteen patients required additional TAL; two patients had a rigid hindfoot. No radiological parameters were presented. Outcome was subjective, being graded as "very good", "good" or "poor" based on completeness of deformity correction. Four patients had poor results necessitating further surgery: two underwent triple fusion and two underwent calcaneal osteotomy.

Leeuwesteijn et al. reported a series included 33 patients with CMT [mean operative age was 28 years (range: 13–59 years); only five patients were adolescents (<16 years); mean follow-up was only 57 months], with flexible hindfoot deformity [13]. All patients underwent DFO of the first metatarsal; additional surgery consisted of hallux IPJ arthrodesis (34 feet), percutaneous TAL (28 feet), claw toe correction (28 feet), Peroneus longus to Peroneus brevis transfer (27 feet), EHL transfer (15 feet), Tibialis posterior tendon transfer due to drop foot (six cases), PF release (1 foot). The authors chose to transfer EHL to the Tibialis anterior or Peroneus tertius tendons (rather than the neck of the first metatarsal), thinking that this transfer regime resulted in a lower incidence of hallux elevatus. There were no major complications. Outcomes were assessed using the FFI and patient satisfaction score: there was a statistically significant improvement in pain (from 29.3 to 14.8) and disability (from 37.8 to 23.5) components of the FFI. Ninety percent of respondents were satisfied with the deformity correction but even over this time frame, two patients underwent triple arthrodesis (TA) due to deformity recurrence.

The most recent evidence Faldini et al. [14], presenting the results [mean follow up 6 years (range: 2–13 years)] of 12 CMT 1A patients (24 feet) with bilateral foot deformities treated by PF release, midtarsal osteotomy (MTO), naviculocuneiform arthrodesis (NCA), Jones procedure and DFO of the first metatarsal. It is interesting that the authors do not appear to have attempted tendon transfer to balance power. They maintain that elevation of the first metatarsal head would indirectly correct the varus heel in a flexible deformity. It is notable that their pre-operative investigations reveal that 17 and 16 feet (of the 24) had 5/5 power in Peroneus brevis and Tibialis anterior, respectively; this may be why their regime was successful, in spite of the absence of tendon transfers. Five patients required additional surgery for claw toe correction. Outcomes were assessed using the Maryland foot score (MFS), rated as excellent (100–90 points), good (89–75 points), fair (74–50 points), or poor (<50 points). Mean score improved from 72 to 86 and 12 feet reported excellent results. Two feet had superficial wound dehiscence. There was no recurrence or subsequent surgery.

In summary, a sequence of level IV case series suggests joint-sparing surgery to be a viable treatment option in flexible Pes cavus. There are no universally agreed guidelines – patients need careful assessment and treatment options should be individualised. Current evidence appears to suggest that a combination of soft tissue and bony procedures are necessary. In flexible Pes cavus, there is inadequate evidence to determine if joint sparing procedures delay progression of deformity or subsequent joint degeneration. Patients should be counselled that further treatment may be necessary (Table 21.1).

What Are the Results of Joint Sparing Surgery for Management of Rigid Pes Cavus?

For a rigid deformity, earlier procedures attempted only uniplanar or biplanar correction. More recently, techniques have focussed on achieving multiplanar correction. Most of these operations involve a midfoot osteotomy that heals – either by arthrodesis, pseudoarthrodesis or bony union. Technically,

these may not be joint-sparing surgery, but most preserve the Chopart joint complex.

Sammarco and Taylor reported the results of superolateral sliding calcaneal osteotomy and dorsolateral closing wedge metatarsal osteotomy in 15 patients (mostly adults; mean age was 33 years; range, 15–61 years) with underlying neurological abnormality (ten with CMT) [15]. Patients were assessed using the Maryland foot and AOFAS scores, demonstrating improvement post-operatively. Additionally, there was radiographic improvement of both cavus and adductus. There were two delayed unions and three non-unions. One patient developed midfoot OA. The youngest patient had recurrence of heel varus and the authors felt that this procedure should be used with caution before skeletal maturity. All patients were satisfied with surgery and brace-free at final follow up.

Wicart and Seringe reported on 26 children (mean age at surgery, 10.3 years; mean follow up, 6.9 years) with an underlying neurological condition (16 had CMT) [16]. Surgery consisted of selective PF release, plantar opening wedge osteotomy of the cuneiform bones and Dwyer opening wedge osteotomy of the calcaneum, if necessary [32 feet (89 %) had hindfoot stiffness requiring Dwyer osteotomy]. Additional surgery included first metatarsal osteotomy (22 feet), medial soft tissue release (17 feet), lateral column shortening (seven cases) and tendo achilles lengthening (two cases). One patient had deep infection. Assessment was both clinical and radiological. The authors used an unvalidated, self-devised 'global score' for patient outcome assessment. Seventeen patients had recurrence of varus deformity; 11 patients were deemed to have poor results; 12 patients required TA.

Weiner et al. treated 89 patients (86 feet developing Pes cavus after clubfoot; mean operative age, 9.7 years; mean follow-up, 7.6 years) with the Akron dome multiplanar midfoot osteotomy [17]. Some patients also had PF release but the numbers were not given. There were no major complications. Outcome assessment was subjective, with the authors reporting that 106 cases (76 %) had satisfactory results. Recurrent deformity (33 patients) was treated with repeat midfoot osteotomy. Twenty-nine patients required TA. The authors performed a sub-group analysis between patients who were younger or older than 8 years at the time of surgery, finding that older children had better results, possibly because they reached skeletal maturity quicker (with less time/growth available for deformity recurrence).

Mubarak and van Valin treated 13 children with cavovarus feet [multiple aetiologies, predominantly neurological; mean operative age, 11 years (SD 3 years); mean follow-up, 4 years] with joint-sparing surgery [18]. Their rationale was to correct the deformity near its apex, and to spare transgression of the midtarsal joints. Hindfoot flexibility was assessed using the kneeling method. The authors used a sequential method to correct foot deformity, initially performing opening wedge osteotomy of the medial cuneiform and closing wedge osteotomy of the first metatarsal to correct the cavus. This was followed by a closing wedge osteotomy of the cuboid (for additional forefoot correction) and lateral displacement closing wedge calcaneal osteotomy for residual varus. They also performed Peroneus longus to Peroneus brevis transfer to balance the foot; 25 % cases also underwent PF release to correct residual tightness. There was significant improvement in both Meary and Hibb angles. A subjective grading system assessing patient outcome, based on correction of forefoot cavus, hindfoot varus and patient satisfaction suggested only one foot had poor results. The authors recommended that a balancing transfer of Peroneus longus to Peroneus brevis should be performed alongside osteotomies.

Mubarak and Dimeglio treated 11 children [mean operative age, 9.3 years (range: 4 months – 15.3 years); minimum follow-up, 1.2 years] with severe cavovarus deformity, by navicular excision and closing wedge osteotomy of the cuboid [19]. Residual varus was corrected with calcaneal osteotomy in 2 feet. Two patients required "minor" re-operation. All patients had plantigrade and pain-free feet at follow-up. The authors recommended this combination of techniques as a salvage procedure for the stiff cavus foot.

Zhou et al. claim to have undertaken a prospective study on 17 patients with rigid Pes cavus [multiple aetiologies; mean age, 16.8 years (range: 12–36 years); mean follow-up 25.3 months; range, 12–48 months] treated by midfoot osteotomy combined with joint sparing internal fixation for treatment of [20]. However, patients were "tracked after treatment" which would suggest that this was a retrospective study. It is unclear if data were recorded prospectively. Patients underwent extra-articular midfoot wedge osteotomy stabilised with cannulated screws. Additional surgery included Tibialis posterior tendon transfer, TAL and claw toe correction. Additional surgery was performed only "selectively in some patients". There were no major complications, and all patients had bony union. Outcome was assessed using AOFAS and radiological criteria. There was a statistically significant 40 point improvement in AOFAS score. There were also significant improvements in calcaneal pitch, and Meary's, tibiotalar and Hibb's angles. The majority of patients (94 %) were very satisfied or satisfied with minor reservations. There was no appreciable worsening of joint degeneration. A longer term follow-up report would be of interest.

In summary, a number of different osteotomy techniques have been described in a sequence of level IV case series. Although a multiplanar osteotomy has the potential for maximum deformity correction, it is not clear which is the best option (Table 21.2).

Table 21.2 Summary of the results of joint sparing surgery in management of rigid Pes cavus (all level IV case series)

Study	N (feet)	Diagnosis	Treatment	Follow up in years Mean (range)	Re-operations	Outcome
Sammarco and Taylor [15]	15 (21)	All neurological CMT 10	Calcaneal and metatarsal osteotomy	4 (1–8)	Revision calcaneal osteotomy: 1 Intertarsal fusion: 1 TAL: 1	Improvement of 17.8 points in Maryland foot and ankle score 42.8 points in AOFAS hindfoot score 47.9 points in AOFAS midfoot score
Wicart and Seringe [16]	26 (36)	CMT 16 All neurological	PF, osteotomy of the cuneiform and calcaneum	6.9	12 TA	Poor 11
Weiner et al. [17]	89 (139)	Variable Clubfeet 86 CMT 13	Akron dome midfoot osteotomy	7.6	33 revision midfoot osteotomy 30 TA	106 (76 %) satisfactory
Mubarak and Valin [18]	13 (20)	Variable CMT 3	Osteotomy of the 1st MT, medial cuneiform, cuboid and 2nd, 3rd MT, PR and PL to PB transfer	4 (2–8)	NR	35 % very good 60 % good 5 % poor
Mubarak and Dimeglio [19]	11 (16)	Arthrogryposis: 6 Clubfeet: 5 Neurogenic: 5	Navicular excision and cuboid osteotomy	4.9 (1.2–9.9)	Two ("minor procedure")	All plantigrade and pain-free
Zhou et al. [20]	17	Poliomyelitis: 10 Idiopathic: 4	Midfoot osteotomy PTT transfer NR TAL NR Claw toe correction NR	2 (1–4)	NR	40 points improvement in mean AOFAS score

NR non reported

What Are the Results of Triple Arthrodesis in Management of Rigid Pes Cavus?

Results of Triple arthrodesis (TA) have not been very satisfactory over the long term. However, these patients are not directly comparable to those undergoing joint-sparing surgery; a less aggressive option may not have been an option. Authors have emphasised that TA should be reserved as a last resort in severe, rigid deformity [18].

Wetmore and Drennan treated 16 adolescents with CMT (Mean operative age, 15 years; mean follow-up, 21 years) by TA [21]. Fourteen patients had poor results and were orthotic dependent. Seven feet (23 %) had recurrence of cavovarus deformity; 23 feet (77 %) had progressive degeneration of foot joints. Outcome progressively worsened with time, and six patients required ankle arthrodesis. In view of the poor results, the authors surmised that TA should only be used as a last resort in limbs with progressive peripheral neuropathy and severe, rigid deformity.

Wukich and Bowen reported the results of TA of 22 patients (mean operative age, 16.8 years; mean follow up, 12.5 years) with CMT [22]. Outcome was assessed on the basis of residual deformity, pseudoarthrosis, pain , callosity, and degenerative arthritis: 45 % of patients had residual deformity, and 60 % had claw-toe deformity. Only 11 feet (32 %) had good results. Three patients were brace dependent. Twenty-one feet (62 %) had midfoot OA and 8(24 %) had ankle OA. Seventy percent of patients complained of persistent pain. There were 12 additional surgeries, including three cases of revision TA. The authors advised concurrent PTT transfer to correct foot drop deformity and also emphasised the role of TA as a salvage procedure in severe, rigid deformity.

Mann and Hsu treated ten adolescent CMT patients (mean operative age, 13.3 years; mean follow-up, 7.5 years) with TA [23]. Three feet underwent associated Posterior tibialis tendon (PTT) transfer. Only 5 feet achieved fusion and plantigrade status. Three feet developed midfoot pseudoarthrosis but did not have functional limitation. One foot required revision TA and 3 feet required subsequent PTT transfer. Three feet had residual deformities and were asymptomatic. Because of incomplete follow up post-operative OA was not reported.

Salzman et al. treated 57 patients [multiple aetiologies – 46 patients having underlying neurological causes, the commonest being poliomyelitis (34); mean operative age, 16 years; mean follow-up, 44 years] with TA with/without associated soft tissue surgery. The commonest soft tissue procedure was tendon transfer (28), followed by TAL [6] performed within 8 weeks of TA. Eight feet (12 %) had minor complications; 55 % complained of pain of one or the other joints at final follow-up, with 16 patients (28 %) being analgesic dependent. Thirteen feet had pseudoarthrosis of which ten were painful. Eighteen patients (32 %) had

Table 21.3 Summary of the results of Triple arthrodesis in management of rigid Pes cavus (all level IV case series)

Study	N (feet)	Diagnosis	Treatment	Follow up in years Mean (range)	Re-operations	Outcome
Wetmore and Drennan [21]	16 (30)	CMT	Triple arthrodesis	21 (6–51)	Ankle arthrodesis: 6	Poor: 14 (Orthotic dependent) Persistent varus/cavus: 9 Recurrence: 7 OA: 23
Wukich and Bowen [22]	22 (34)	CMT	TA Percutaneous PF: 7	12.5 (2.5–40)	12 (Revision TA: 3)	Progressive OA: 29 Persistent pain: 70 %
Mann and Hsu [22]	10 (12)	CMT	TA PTT transfer: 3 TAL and PF: 1	7.5 (2.3–18)	Revision TA: 1 Clawtoe correction: 1 PTT transfer: 3 TAT transfer: 1	Midfoot pseudoarthrosis: 3 Residual deformity: 3 (molded AFO dependent)
Salzman et al. [24]	57 (67) (26 feet had valgus deformity)	Multiple Polio: 37 CMT: 6	TA Additionally within 8 weeks: Tendon transfer: 28 TAL: 6 Midfoot arthrodesis: 6 PF: 3	44 (29–68)	18 (revision TA: 3)	Pain in foot or ankle: 37 Ankle OA: 57 Midfoot OA: 56 Pseudoarthrosis: 13

subsequent operations (revision TA: 3). Fifty-two feet had residual hindfoot deformity. All patients had ankle OA of which 34 were moderate or severe; 31 patients had midfoot OA. Only 19 feet (28 %) had good results on final follow-up. This study had the longest follow up; unsurprisingly, the authors noted that clinical and radiological outcomes worsened with time. The authors did not find any correlation between the degree of joint space narrowing and reported pain/deformity. Fifty-four patients (95 %) were satisfied with the result of surgery at final follow-up. In view of gradual deterioration of results following TA the authors advised appropriate counselling in younger patients.

In summary, from a small cadre of level IV case series, triple arthrodesis seems to be a useful salvage procedure in patients with severe rigid Pes cavus deformity. However, tendon transfer for balancing may still be required. Patients should be counselled that results deteriorate over time (Table 21.3).

What Are the Results of External Fixation in Management of Pes Cavus?

The external fixator is an attractive option in Pes cavus correction because there can be gradual simultaneous corrections in all planes, in a minimally invasive fashion without major disruption of soft tissues. This is especially useful where soft tissue coverage is poor (eg multiple previous surgeries), in neglected or relapsed deformity, or with infection or associated limb deformity. Although it has been widely used (and reported) in neglected clubfoot deformity, there are not many reports of its use in paediatric Pes cavus per se.

Shalaby and Hefny treated 20 patients with a range of complex foot deformities [aetiology was neurological in 11 patients (two cases of Pes cavus; two cases of hindfoot varus); 12 had had previous surgery and 10 had poor soft tissue condition; mean operative age was 26 years, range: 17–46 years; mean follow-up 25 months] with a V osteotomy and Ilizarov fixation [25]. Mean time in fixator was 15 weeks, followed by 6 weeks in a short leg cast; a plantigrade foot was achieved in all but one case, resulting in "improvement" in shoe fitting. Mean improvement in Meary angle was from 20° to 4°. Four patients had mild residual deformity and two patients experienced mild recurrence. There were seven cases of pin-site infection.

Lee et al. managed 26 patients [all had an underlying neurologic condition, the commonest being lipomeningocoele (11 feet); 19 patients were children (age range of series, 5–51 years); 14 feet had undergone pervious surgery; mean follow-up, 6 years] with equinocavovarus deformity by a modified Ilizarov technique [26]. The modification consisted of combining it with conventional soft tissue and bony procedures. Nine patients had distraction histogenesis and limited

Table 21.4 Summary of management of rigid Pes cavus with external fixation (all level IV case series)

Study	N (feet)	Diagnosis	Treatment	Follow up in years Mean (range)	Re-operations or additional surgery	Outcome
Shalaby and Hefny [25]	20 (25)	Post-Polio: 7 Neglected CTEV: 4 CMT: 2 Others: 7	V osteotomy and Ilizarov technique	2 (1.5–3)	Reversal: 1 Flexor tenotomy: 2	Plantigrade: 24 Recurrence: 2 Residual deformity: 4
Lee et al. [26]	26 (29)	Lipomeningocoele: 11 Arthrogryposis: 7 Post-polio: 6 Cerebral Palsy: 4 Nerve injury: 1	Ilizarov technique, Selective soft tissue procedure and osteotomy	6 (1–13)	Combined osteotomy: 15 TA: 7 Tendon transfer: 5	Plantigrade: 24 Recurrence: 4 Residual deformity: 2 Excellent: 16 Poor: 4
Kirienko et al. [27]	27 (27)	Poliomyelitis	Ilizarov technique, selective soft tissue procedure and arthrodesis	7.17 (0.5–15)	V osteotomy and calcaneal osteotomy: 5 Supramalleolar osteotomy: 2 TA: 5 Ankle arthrodesis: 1 Chopart joint arthrodesis: 1 TAL: 8	Plantigrade: 25 Recurrence: 2 Residual deformity: 2 All satisfied with post-operative gait

soft tissue release, others required additional osteotomy (calcaneal, mid-tarsal, first metatarsal). The authors also performed tendon transfer to balance the foot in five cases and TA in seven cases. After a mean period of 23 days, the external fixator was removed and patients were cast for 2–5 weeks. Twenty-four feet were corrected to plantigrade position; 19 patients were satisfied with the results of surgery. There were four recurrences (poor outcome) and six complications.

Kirienko et al. corrected foot deformity from poliomyelitis in 27 patients (four paediatric patients aged 17–18 years; ten patients had undergone previous surgery; mean follow-up, 7 years) using the Ilizarov technique [27]. Only six patients had treatment with isolated frame application; others required additional soft tissue or bony procedures including arthrodesis. Nine patients had some degree of Pes cavus deformity. Mean time in foot frame was 4.2 months. A plantigrade foot was achieved in 25 cases without major complication and all patients were satisfied with their post-operative gait. There were two cases of residual deformity and two cases of recurrence.

In summary, on the basis of a further body of level IV evidence (case series), external fixation appears to be a useful option in treatment of Pes cavus. However, it is rarely used in isolation and surgeons have often performed additional soft tissue and bony procedures (including tendon transfer and arthrodesis) and their attempt to maintain a stable plantigrade foot (Tables 21.4 and 21.5).

Table 21.5 Summary of recommendations

Clinical situations	Grades
Custom made orthoses may be useful in reducing pain in symptomatic patients with Pes Cavus	B
Botulinum toxin A does not prevent progression of cavus deformity in CMT Type 1A	B
Joint sparing surgery is an option for management of both flexible and rigid Pes cavus	C
Tendon transfer is necessary to balance the forces in Pes cavus caused by progressive neurological conditions	C
Triple arthrodesis is an option for management of rigid Pes cavus	C
External fixation is an option for management of Pes cavus but may require to be supplemented with tendon transfer or arthrodesis	C
Surgery must be individualised to every patient taking into account the deformity parameters, degree of rigidity and disability	C
It is not possible to make an evidence informed decision regarding the best combination of soft tissue and bony procedures for joint sparing surgery	I
In Pes cavus, the optimum age/stage for surgery is unclear	I
It is not clear if joint-sparing surgery is a better option than Triple arthrodesis in rigid Pes cavus	I

References

1. Mosca VS. The cavus foot. J Pediatr Orthop. 2001;21(4):423–4. Available from: http://www.ncbi.nlm.nih.gov/pubmed/11433149. Cited 7 Jan 2016.
2. Brewerton DA, Sandifer PH, Sweetnam DR. "idiopathic" pes cavus: an investigation into its aetiology. Br Med J. 1963;2(5358):659–61. Available from: http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=1872745&tool=pmcentrez&rendertype=abstract. Cited 7 Jan 2016.
3. Burns J, Crosbie J, Ouvrier R, Hunt A. Effective orthotic therapy for the painful cavus foot: a randomized controlled trial. J Am Podiatr Med Assoc. 2006;96(3):205–11.
4. Burns J, Landorf KB, Ryan MM, Crosbie J, Ouvrier RA. Interventions for the prevention and treatment of pes cavus. Cochrane Database Syst Rev Online. 2007;(4):CD006154. Available from: http://www.ncbi.nlm.nih.gov/pubmed/17943889
5. Burns J, Scheinberg A, Ryan MM, Rose KJ, Ouvrier RA. Randomized trial of botulinum toxin to prevent pes cavus progression in pediatric Charcot-Marie-Tooth disease type 1A. Muscle Nerve. 2010;42(2):262–7. Available from: http://www.ncbi.nlm.nih.gov/pubmed/20544937
6. Coleman SS, Chesnut WJ. A simple test for hindfoot flexibility in the cavovarus foot. Clin Orthop Relat Res. 1977;(123):60–2. Available from: http://www.ncbi.nlm.nih.gov/pubmed/852192. Cited 28 Oct 2015.
7. Chan G, Sampath J, Miller F, Riddle EC, Nagai MK, Kumar SJ. The role of the dynamic pedobarograph in assessing treatment of cavovarus feet in children with Charcot-Marie-Tooth disease. J Pediatr Orthop. 2007;27(5):510–6. Available from: http://www.ncbi.nlm.nih.gov/pubmed/17585258. Cited 29 Nov 2015.
8. Roper BA, Tibrewal SB. Soft tissue surgery in Charcot-Marie-Tooth disease. J Bone Joint Surg Br. 1989;71(1):17–20. Available from: http://www.ncbi.nlm.nih.gov/pubmed/2914996. Cited 29 Nov 2015.
9. Jones R. III. The soldier's foot and the treatment of common deformities of the foot. Br Med J. 1916;1(2891):749–53. Available from: http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=2347606&tool=pmcentrez&rendertype=abstract. Cited 29 Nov 2015.
10. M'Bamali EI. Results of modified Robert Jones operation for clawed hallux. Br J Surg. 1975;62(8):647–50. Available from: http://www.ncbi.nlm.nih.gov/pubmed/1174809. Cited 29 Nov 2015.
11. Ward CM, Dolan LA, Bennett DL, Morcuende JA, Cooper RR. Long-term results of reconstruction for treatment of a flexible cavovarus foot in Charcot-Marie-Tooth disease. J Bone Joint Surg Am. 2008;90(12):2631–42. Available from: http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=2663331&tool=pmcentrez&rendertype=abstract. Cited 2 Oct 2015.
12. Chatterjee P, Sahu MK. A prospective study of Japas' osteotomy in paralytic pes cavus deformity in adolescent feet. Indian J Orthop. 2009;43(3):281–5.
13. Leeuwesteijn AEEPM, de Visser E, Louwerens JWK. Flexible cavovarus feet in Charcot-Marie-Tooth disease treated with first ray proximal dorsiflexion osteotomy combined with soft tissue surgery: a short-term to mid-term outcome study. Foot Ankle Surg. 2010;16(3):142–7. Available from: http://www.ncbi.nlm.nih.gov/pubmed/20655015. Cited 2 Oct 2015.
14. Faldini C, Traina F, Nanni M, Mazzotti A, Calamelli C, Fabbri D, et al. Surgical treatment of cavus foot in Charcot-Marie-tooth disease: a review of twenty-four cases: AAOS exhibit selection. J Bone Joint Surg Am. 2015;97(6):e30. Available from: http://www.ncbi.nlm.nih.gov/pubmed/25788311. Cited 2 Oct 2015.
15. Sammarco GJ, Taylor R. Cavovarus foot treated with combined calcaneus and metatarsal osteotomies. Foot ankle Int. 2001;22(1):19–30. Available from: http://www.ncbi.nlm.nih.gov/pubmed/11206819. Cited 10 Nov 2015.
16. Wicart P, Seringe R. Plantar opening-wedge osteotomy of cuneiform bones combined with selective plantar release and dwyer osteotomy for pes cavovarus in children. J Pediatr Orthop. 2006;26(1):100–8. Available from: http://www.ncbi.nlm.nih.gov/pubmed/16439912. Cited 14 Nov 2015.
17. Weiner DS, Morscher M, Junko JT, Jacoby J, Weiner B. The Akron dome midfoot osteotomy as a salvage procedure for the treatment of rigid pes cavus: a retrospective review. J Pediatr Orthop. 2008;28(1):68–80. Available from: http://www.ncbi.nlm.nih.gov/pubmed/18157049. Cited 5 Dec 2015.
18. Mubarak SJ, Van Valin SE. Osteotomies of the foot for cavus deformities in children. J Pediatr Orthop. 2009;29(3):294–9. Available from: http://www.ncbi.nlm.nih.gov/pubmed/19305283. Cited 5 Dec 2015.
19. Mubarak SJ, Dimeglio A. Navicular excision and cuboid closing wedge for severe cavovarus foot deformities: a salvage procedure. J Pediatr Orthop. 2011;31(5):551–6. Available from: http://www.ncbi.nlm.nih.gov/pubmed/21654465. Cited 5 Dec 2015.
20. Zhou Y, Zhou B, Liu J, Tan X, Tao X, Chen W, et al. A prospective study of midfoot osteotomy combined with adjacent joint sparing internal fixation in treatment of rigid pes cavus deformity. J Orthop Surg Res. 2014;9(1):44. Available from: http://www.josr-online.com/content/9/1/44
21. Wetmore RS, Drennan JC. Long-term results of triple arthrodesis in Charcot-Marie-Tooth disease. J Bone Joint Surg Am. 1989;71(3):417–22. Available from: http://www.ncbi.nlm.nih.gov/pubmed/2925716. Cited 6 Dec 2015.
22. Wukich DK, Bowen JR. A long-term study of triple arthrodesis for correction of pes cavovarus in Charcot-Marie-Tooth disease. J Pediatr Orthop. 1989;9(4):433–7. Available from: http://www.ncbi.nlm.nih.gov/pubmed/2732324. Cited 6 Dec 2015.
23. Mann DC, Hsu JD. Triple arthrodesis in the treatment of fixed cavovarus deformity in adolescent patients with Charcot-Marie-Tooth disease. Foot Ankle. 1992;13(1):1–6. Available from: http://www.ncbi.nlm.nih.gov/pubmed/1577335. Cited 6 Dec 2015.
24. Saltzman CL1, Fehrle MJ, Cooper RR, Spencer EC, Ponseti IV. Triple arthrodesis: twenty-five and forty-four-year average follow-up of the same patients. J Bone Joint Surg Am. 1999;81(10):1391–402
25. Shalaby H, Hefny H. Correction of complex foot deformities using the V-osteotomy and the Ilizarov technique. Strategies Trauma Limb Reconstr. 2007;2(1):21–30. Available from: http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=2321725&tool=pmcentrez&rendertype=abstract. Cited 6 Dec 2015.
26. Lee DY, Choi IH, Yoo WJ, Lee SJ, Cho T-J. Application of the Ilizarov technique to the correction of neurologic equinocavovarus foot deformity. Clin Orthop Relat Res. 2011;469(3):860–7. Available from: http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=3032860&tool=pmcentrez&rendertype=abstract. Cited 6 Dec 2015.
27. Kirienko A, Peccati A, Abdellatif I, Elbatrawy Y, Mostaf ZMA, Necci V. Correction of poliomyelitis foot deformities with Ilizarov method. Strategies Trauma Limb Reconstr. 2011;6(3):107–20. Available from: http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=3225570&tool=pmcentrez&rendertype=abstract. Cited 6 Dec 2015.

Evidence-Based Management of Ankle Fractures in Children

22

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Abstract

Ankle fractures in children are common and pose unique diagnostic and therapeutic challenges. In this chapter we investigated and summarised the strength of evidence behind methods of assessing and treating these fractures. Lateral and mortise views are indicated when Ottawa ankles rules are positive. AP view does not add more information (grade B). Computed tomography is useful adjunct for assessment and management planning in selected cases (grade B). Undisplaced ankle fractures can be effectively managed with cast immobilisation and close radiographic follow-up evaluation (grade B). Reduction (closed or open) and internal fixation should be considered when there is a displacement (grade B/C); however the cut off for what is considered displaced has not been universally agreed on. Most published studies used 2 mm displacement as a cut off for operative treatment; hence there is a lack of evidence on the outcomes of non operative treatment when the displacement is more than 2–3 mm.

Keywords

Ankle fractures • Transitional fractures • Tillaux fractures • Triplane fractures • McFarland fractures • Salter-Harris classification

Introduction

The ankle joint is a modified hinge joint between the tibial plafond, medial and lateral malleoli proximally and the talus distally. The ankle joint is stabilised by several ligaments which are essential for normal function. These ligaments include the anterior talofibular (ATFL), calcaneofibular (CFL) and posterior talofibular ligaments (PTFL) laterally, and the deltoid ligament medially. The lower ends of the tibia and fibula articulate together forming the inferior tibiofibular joint (ITFJ) which is stabilised by the anterior tibiofibular (ATiFL), interosseous ligament and posterior tibiofibular (PTiFL) ligaments [1].

The distal tibial epiphysis starts ossifying between 6 and 24 months of age and ossification extends into the medial malleolus around 7 years. In 20 % of cases there is a separate ossification centre in the medial malleolus. This should not be confused with a fracture. The distal tibial physis closes over an 18-month period starting around age 14 years in girls and 16 years in boys. The central part of the physis closes first, followed by the medial side and lastly the lateral side (Fig. 22.1).

The distal fibula epiphysis starts ossifying during the second year of life and closes 12–24 months later than the distal tibial physis. The distal tibial physis grows 4 mm a year and account for 40 % of the leg longitudinal growth.

Stability is a very important concept in the management of ankle fractures. Ligaments play a major role in ankle

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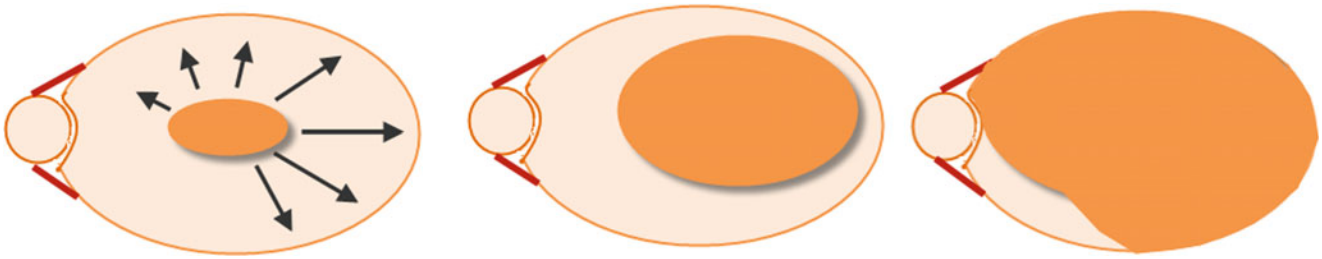

Fig. 22.1 Progression of physeal closure of the distal tibia

Table 22.1 AO ankle fracture classification

Type A	Type B	Type C
A1 Infrasyndesmotic lesion, isolated 1. Rupture of the lateral collateral ligament 2. Avulsion of the tip of the lateral malleolus 3. Transverse fracture of the lateral malleolus	B1 Transsyndesmotic fibular fracture, isolated 1. Simple 2 Simple, with rupture of the anterior syndesmosis 3 Multifragmentary	C1 Suprasyndesmotic lesion, diaphyseal fracture of the fibula, simple 1. With rupture of the medial collateral ligament 2. With fracture of the medial malleolus 3. With fracture of the medial malleolus and a Volkmann (= Dupuytren)
A2 Infrasyndesmotic lesion, with fracture of the medial malleolus 1. Rupture of the lateral collateral ligament 2. Avulsion of the tip of the lateral malleolus 3. Transverse fracture of the lateral malleolus	B2 Transsyndesmotic fibular fracture, with medial lesion 1. Simple with rupture of the medial collateral ligament and rupture of the anterior syndesmosis 2. Simple with fracture of the medial malleolus and with rupture of the anterior syndesmosis 3. Multifragmentary	C2 Suprasyndesmotic lesion, diaphyseal fracture of the fibula, multifragmentary 1. With rupture of the medial collateral ligament 2. With fracture of the medial malleolus 3. With the fracture of the medial malleolus and a Volkmann (= Dupuytren)
A3 Infrasyndesmotic lesion, with postero-medial fracture 1. Rupture of the lateral collateral ligament 2. Avulsion of the tip of the lateral malleolus 3. Transverse fracture of the lateral malleolus	B3 Transsyndesmotic fibular fracture, with medial lesion and a Volkmann (fracture of the postero-lateral rim) 1. Fibula simple, with rupture of the medial collateral ligament 2. Fibula simple, with fracture of the medial malleolus 3. Fibula multifragmentary, with fracture of the medial malleolus	C3 Suprasyndesmotic lesion, proximal fibular lesion 1. Without shortening, without Volkmann 2. With shortening, without Volkmann 3. Medial lesion and a Volkmann

fracture stability. In practice most surgeons base their decisions on clinical assessment and plain x-rays to assess ankle fracture stability. Although this may be adequate in most cases where the features of instability are obvious, this is not the case in all fractures leading to some patients undergoing unnecessary surgery. In children, this is even more complicated due to the presence of the physis which poses diagnostic and therapeutic challenges to the treating surgeons.

Stress tests, CT scan and MRI scan have been used to aid decision making.

Several classifications have been proposed to help understand the nature of ankle fractures, inform treatment choices and predict future outcomes. Most of these have been based on morphological description and their predictive values for instability and better treatment choice have been questioned [2, 3]. However, to understand the published evidence better, basic knowledge of these classifications is valuable.

In adult practice, there are two common classifications in use: Weber and Lauge-Hanson classification.

Weber classified ankle fractures according to the relation of the fibular fracture to the syndesmosis into: type A (below the syndesmosis), type B (at the level the syndesmosis) and type C (above the syndesmosis). Weber classification was subsequently adopted and incorporated in the AO ankle classification which was expanded into two sub-layers as shown in Table 22.1.

Lauge-Hansen [4] classified ankle fractures according to the position of the foot at the time of impact (supinated or pronated) and the direction of the force applied to the ankle (adduction, abduction or external rotation). A pronated foot will result in tight deltoid ligament and lax lateral ligamentous complex and vice versa for a supinated foot. Lauge-Hansen indicated that these two elements (the position of the foot and the direction of the force) determine the order in which ankle stabilising structures fail, and that these structures fail in a predictable order (Table 22.2). Several biomechanical studies failed to reproduce the work and classification of Lauge-Hansen [5, 6]). Moreover, findings from MRI

Table 22.2 Lauge-Hansen classification

Lauge-Hansen class	Sequence of structures failure with increasing force caused by an injury
Supination – external rotation (SER)(foot is supinated and the force is external rotation)	Stage I: ATiFL rupture or avulsion fracture of tibia or fibula
	Stage II: short oblique fibula fracture (anteroinferior to posterosuperior)
	Stage III: PTiFL rupture or avulsion of posterior malleolus
	Stage IV: Medial malleolus transverse fracture or disruption of deltoid ligament
Supination – adduction (SA)	Stage I: Talofibular sprain or distal fibular avulsion
	Stage II: Vertical medial malleolus and impaction of anteromedial distal tibia
Pronation – abduction (PA)	Stage I: Medial malleolus transverse fracture or disruption of deltoid ligament
	Stage II: ATiFL rupture or avulsion
	Stage III: Transverse comminuted fracture of the fibula above the level of the syndesmosis
Pronation – external rotation (PER)	Stage I: Medial malleolus transverse fracture or disruption of deltoid ligament
	Stage II: ATiFL disruption
	Stage III: Lateral short oblique or spiral fracture of fibula (anterosuperior to posteroinferior) above the level of the joint
	Stage IV: Posterior tibiofibular ligament rupture or avulsion of posterior malleolus

studies of displaced ankle fracture did not have the patterns of ligament and bony injury predicted by their apparent Lauge-Hansen type [1, 2, 7].

Salter and Harris introduced a classification system which carries their name based on the relationship of fracture lines to the growth plate [8, 9]. The classification is relatively simple and easily remembered and has been relatively successful in predicting future growth disturbance although the latter has been contested [10]. (Fig. 22.2)

Dias and Tachdjian [11] modified the Lauge-Hansen classification to include the Salter-Harris classification so that it can applied to children ankle fractures. Subsequently four other types of fractures were added, namely the Tillaux, triplane, axial compression, and miscellaneous physeal fractures [12].

Tillaux fracture and triplane fractures have been added to the classification. The juvenile Tillaux fractures (to differentiate it from adult similar avulsion fracture) are SH-III fractures involving the anterolateral aspect of the distal tibia plafond which is not fused to the metaphysis yet (Fig. 22.3). These fractures are caused by external rotation forces and can be reduced by internally rotating the foot [13].

The triplane fracture was described by Lynn in 1972 [14]. As the name implies these fractures occur in three different planes: coronal, transverse, and sagittal. On the AP radiographs the fracture appears as a SH-III (like Tillaux fracture) whilst on the lateral view it appears as a SH-II (also called two-part triplane fracture) or SH-IV (also called three-part triplane fracture). Four-part triplane was also described in the literatures [15, 16] (*see* Figs. 46.1 and 46.2 in Chap. 46).

The prognostic value of these classifications has been debated. Leary [10] retrospectively reviewed 124 children after physeal fractures of the distal end of the tibia. They defined premature physeal closure (PPC) as radiographic evidence of physeal closure as compared to the uninjured side in this patient population. Fifteen fractures (12.1 %) were complicated by PPC, 67 % of the PPC observed occurred in SH-II fractures, followed by 13 % in SH-III, 13 % in SH-IV, and 7 % in triplane fractures. They did not observe any physeal arrest in the SH-I or Tillaux fractures. They were able to demonstrate statistically significant correlations between mechanism of injury and PPC and between the amount of initial fracture displacement and the rate of PPC. For each millimetre of initial displacement, there was a relative risk of 1.15 ($P < 0.01$).

In another study of 49 children with physeal fractures of the distal tibia or fibula or both, the Salter-Harris classification system could not significantly predict the growth pattern [17] (Fig. 22.2).

Spiegel et al. [18] followed 184 distal tibia and/or fibula, fractures for an average of 28 months after injury using the Salter-Harris classification. They differentiated three groups according to their risk for shortening of the leg, angular deformity of the bone, or incongruity of the joint. The low-risk group consisted of 89 patients, 6.7 % of whom had complications; this group included all type I and type II fibula fractures, all type I tibia fractures, type III and type IV tibia fractures with less than 2 mm of displacement, and epiphyseal avulsion injuries. The high-risk group consisted of 28 patients, 32 % of whom had complications; this group included type III and type IV tibia fractures with 2 mm or more of displacement, juvenile Tillaux fractures (Fig. 22.3), triplane fractures, and comminuted tibial epiphyseal fractures (type V). The unpredictable group was made up of 66 patients, 16.7 % of whom had complications; only type II tibia fractures were included. The incidence and types of complications were correlated with the type of fracture (Salter-Harris classification), the severity of displacement or comminution, and the adequacy of reduction.

de Sanctis et al. [19]) reviewed 158 ankle fractures; 132 were treated conservatively and 26 patients underwent surgical

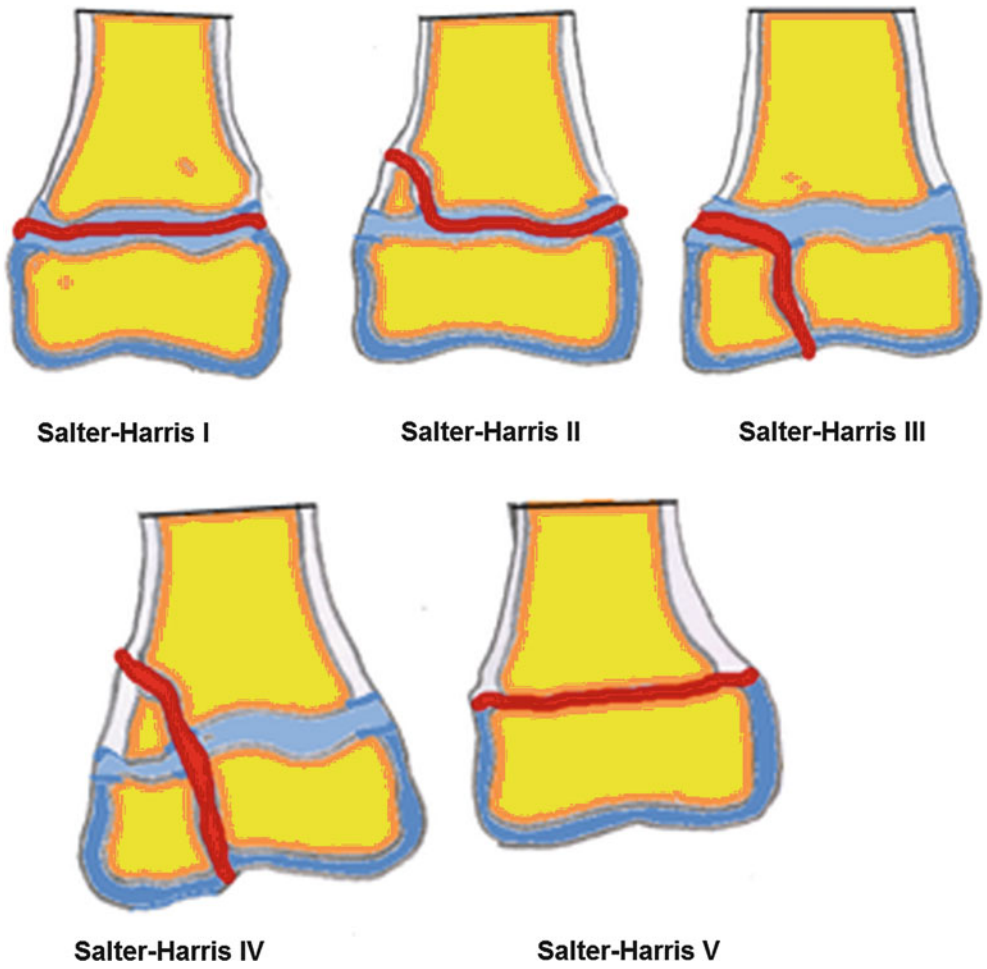

Fig. 22.2 Salter-Harris classification of physeal injury

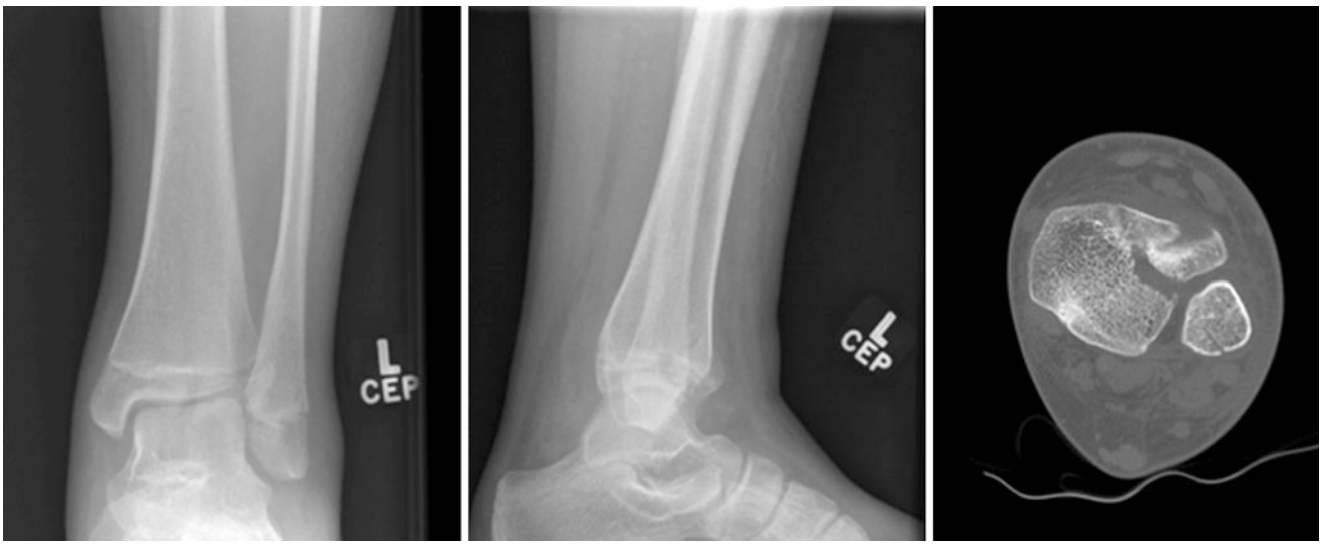

Fig. 22.3 Tillaux fracture

treatment. Fibular fractures of the malleolus without epiphyseal separation or dislocation (68 patients) were excluded. Of the 158 patients, 113 (70 %) were available for an average 6-year follow-up. They compared the degree of epiphyseal separation or dislocation, the Carothers-Crenshaw classification (a classification based on the mechanism of trauma) with the Salter-Harris classification (which is based on anatomical-radiographic criteria). They reported that fractures more likely to result in permanent damage to the physis are those caused by a traumatic adduction-supination mechanism that can produce SH-III, IV, and V fractures of the distal part of the tibia; they also reported that the combination of compression and

Table 22.3 The outcomes after various types of Salter-Harris fractures

Type of fractures	Good outcome	Poor outcome	Total
SH-I	180	1	181
SH-II	107	6	113
SH-III	58	8	66
SH-IV	15	1	16
Total	360	16	376

adduction may cause a SH- V injury with type III and IV fractures. However, type V lesions are often diagnosed late. In 11 of their 12 poor results, 6 were caused by adduction-supination injuries and 5 were compressive injuries.

In a large retrospective study of 376 children with distal tibial physeal injury, Schurz et al. [20] reported the outcomes after various types of Salter-Harris fractures (Table 22.3).

Vahvanen and Aalto [21] studied 310 children treated for ankle fractures. They were classified according to the classifications of Ashhurst-Bromer-Weber, Lauge-Hansen, and Salter-Harris. They found that grouping of the fractures according to Lauge-Hansen and Ashurst-Bromer-Weber classifications suited to adults was largely unsuccessful. Epiphyseal fractures were easily classified according to Salter-Harris. They proposed that ankle fractures in children can be roughly divided into avulsion and epiphyseal fractures. Adequately reduced avulsion fractures can be expected to heal well; epiphyseal fractures, however, may cause late complications.

What Is the Evidence Behind Ankle Fractures Investigation?

Ottawa Rule

Ankle injury is common and radiological tests are not always indicated. The Ottawa ankle rules [22, 23] have been shown to be accurate in predicting the need for radiography in the acute trauma situation in adults. They can be used by medical and nursing staff in a variety of settings, and can reduce unnecessary radiography ([24, 25]; Allerston and Justham [26–28]). Several studies showed their value in detecting ankle fractures in children [29–36]. In a review by Crocco [37] of 671 fractures, the sensitivities of the Ottawa ankle rules ranged from 83 % to 100 % and specificities from 7.9 % to 50 %. X-ray reduction rates ranged from 5 % to 44 % (pooled reduction rate 25 %, 95 % CI: 23–26 %).

Plain Radiograph

The standard plain radiographic views of the injured ankle are antero-posterior (AP), the mortise and lateral views. The mortise view is a modified AP with the ankle internally rotated so that the malleoli are in the same horizontal plane and the joint space is seen evenly on both sides of the ankle. This requires 10°–20° of internal rotation. The need for three views has been questioned. Brage [38] found that ankle fractures could be classified with two radiographic views as reliably as with three views. Four different observers independently evaluated 99 sets of ankle radiographs. The examiners classified the ankle fractures by using both the Lauge-Hansen and Weber systems. The interobserver and intraobserver variations were analyzed by kappa statistics. The study demonstrated that ankle fractures can be classified with two views, lateral or mortise, with a reliability as good as that achieved with three views. The best agreement was achieved with lateral and mortise views. Adding a true AP view did not add useful information.

In a study by Vangsness [39], 123 sets of emergency room ankle x-rays (AP, lateral and mortise) were retrospectively reviewed to determine whether all three views were necessary to diagnose the presence of an ankle fracture. Four physicians (two orthopaedic surgeons, one musculoskeletal radiologist, and one emergency room physician) reviewed all randomly ordered sets of films twice – once with all three views and once with only the lateral and mortise views. The overall accuracy of two views was within the 95 % expected threshold of accuracy using three views. The lateral and mortise views alone appear sufficient for ankle fracture diagnosis, and imply a substantial decrease in radiation and cost savings.

The Role of Medial Clear Space

The presence of an ankle fracture on plain x-ray is not an indication for surgery and stable fractures may do not even require cast protection. Signs of instability are more important than the presence of a fracture as such. Displacement is often used to indicate instability (Fig. 22.4). Murphy et al. [40] measured medial and superior clear space in 73 patients without ankle injuries. Seventeen percent of male x-rays and 1 % of female x-rays had a medial clear space >4 mm, while 2 % of males and no females had a medial clear space >5 mm. Thirteen percent of radiographs had a medial clear space greater than superior clear space. Measurements were symmetrical, so the authors suggest the use of contralateral comparison radiographs to evaluate apparent medial widening. Koval et al. [41] suggested that a 4 mm medial clear space indicates an intact medial deltoid ligament and a stable ankle.

Schuberth [42] showed that medial clear space was a poor predictor of arthroscopically-diagnosed deltoid ligament tears. They found the 88.5 % false positive rate for deltoid ligament rupture when medial clear space > or = 3 mm (P = .54, Fisher's exact test) and 53.6 % when medial clear space > or = 4 mm (P = .007).

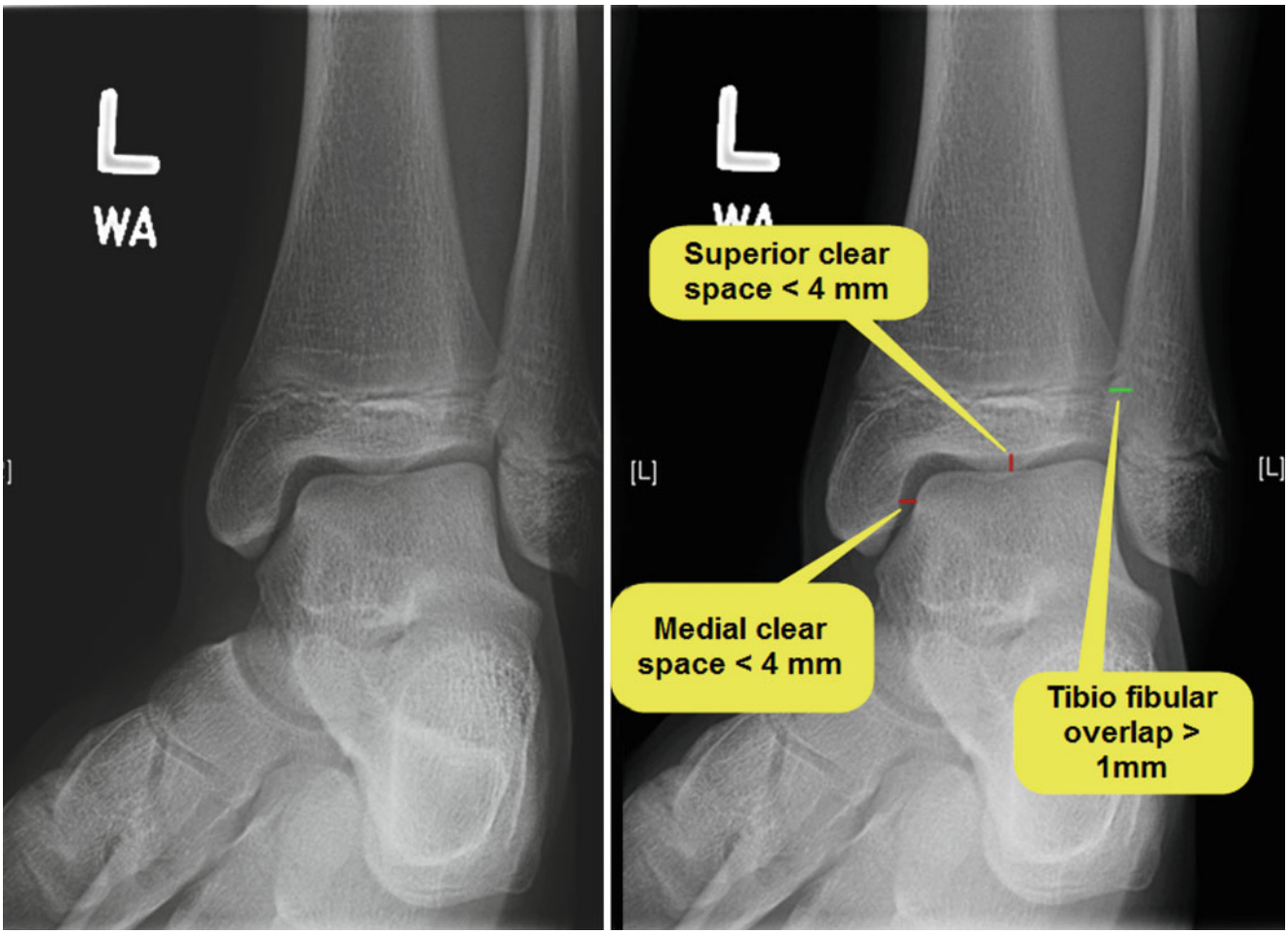

Fig. 22.4 Mortise view showing medial and superior clear space

Several studies [41, 43, 44] have suggested that external rotation stress (manual or gravitational) x-ray can differentiate stable from unstable undisplaced fractures when plain x-rays are inconclusive. These studies assumed ankle instability when the clear medial space increased more than 4 mm with stress. However, this has been contested. Koval [41] investigated the significance of positive stress radiographs (medial clear space >5 mm on stress testing) in 21 patients who. All had MRI scan; 19 patients had evidence of partially torn deep deltoid ligament and their ankles were not stabilised whereas two had a complete rupture of the deep deltoid ligament and underwent stabilisation. All fractures united without evidence of residual medial clear space widening or posttraumatic joint space narrowing.

Several other studies concluded that stress radiographs greatly over-estimate instability and that most undisplaced ankle fractures can be treated non-surgically [45–47]. Most of these studies include only a few children and adolescents so their findings may not be applicable to paediatric practice.

The Role of CT Scan

CT can be useful in assessing more complex ankle fractures for accurate diagnosis, preoperative planning, and assessment of reduction of intra-articular fractures. However, clear indications are still lacking and reasoning remains subjective. A recent study of 64 distal tibial fractures with intra-articular involvement demonstrated the importance of CT scan in assessment [48]. All patients had plain radiographs and CT scans. Findings based on radiographs and CT scan are summarised in Table 22.4. Authors concluded that CT scan led to changes in fracture classification and treatment decision.

Table 22.4 The difference in classification and decisions of surgical treatment when CT scan was used in 64 patients with ankle fracture

	Palin radiograph	Adding CT Scan
SH-III fracture	31	20
SH-IV fracture	8	12
Tillaux fracture	9	9
Triplane fracture	16	23
Decision for operative treatment	18	42
Decision for non operative	46	22

In a similar study of 25 triplane fractures by Eismann [49], there was poor inter-rater reliability (a kappa of 0.17) and intra-rater reliability (a kappa of 0.31) with radiographs alone but moderate inter-rater reliability (a kappa of 0.41) and intra-rater reliability (a kappa of 0.54) with the addition of computed tomography. More interestingly, the decision from non-operative to operative treatments was changed in 27 % and either the orientation or number of screws was changed in 41 % of the cases when raters reviewed CT scans.

Black et al. [50] performed a retrospective analysis on 100 consecutive patients treated for ankle fractures having both preoperative radiographs and CT scans. They found operative strategy was changed in 24 % of cases after CT review. Several authors suggested that CT scan should be routinely performed in clinical practice [51, 52].

The Role of Magnetic Resonance Imaging (MRI)

MRI provides fine detail of bones and soft-tissue structures and does not involve radiation. MRI scans have been used to diagnose occult fractures and, in cases of incomplete ossification of malleoli, to establish displacement [41, 53].

Lohman et al. [53] studied 60 children with acute ankle injuries with both conventional radiography and MR. Plain radiography produced 5 of 28 (18 %) false negative and 12 of 92 (13 %) false positive fracture diagnoses compared to MRI. However they found no complex ankle fracture was missed and the MRI did not change the treatment plan in any case.

Petit et al. [54] compared radiographs with MRIs for 29 children with physeal ankle injuries and found only 1 of 29 fractures (3 %) was misclassified by plain film radiography.

Contrary to these two studies, Carey et al. [55] reported on 14 patients with suspected physeal injuries and indicated that that the MRI scans led to a change in the Salter-Harris classification for two of the nine patients with fractures seen on radiographs, they identified two occult fractures, and changed the management in five patients.

What Is the Best Treatment for Non-displaced Ankle Fractures in Children?

There is a consensus on non operative treatment of stable and undisplaced ankle fractures. Many centres use a below knee full cast to treat undisplaced stable fractures although this may not be necessary in every case. In a randomised controlled study (level I) of 40 patients with Lauge-Hansen supination-eversion, stage II ankle fractures, the use of the air stirrup led to a significant improvement in early patient comfort, post-fracture swelling, range of ankle motion at union, and time to full rehabilitation in comparison to a below knee walking cast [56].Similar findings were found in 66 patients who were treated with Aircast Stirrup ankle braces or Don Joy R.O.M.-Walker braces [57]. Subjective satisfaction with comfort and ease of use was significantly higher with Aircast Stirrup while pain relief and an inflammatory score were significantly better in the R.O.M.-Walker group after 4 weeks. There was in difference in outcomes after 3 months.

The above may be not true for unstable fractures where there is a potential for fracture displacement. Defining stability when there is no displacement remains controversial and the cut-off between stable and non-stable fractures has not yet been fully defined [1]. Several clinical and radiological signs have been proposed to signify instability including medial tenderness, bruising or swelling, fibular fracture above the syndesmosis and high energy fractures but none is confirmatory. Stress views have been used to diagnose instability; however several studies showed that even in the presence of a positive stress views conservative treatment in a brace or cast usually produced satisfactory union in an undisplaced position [41, 44, 46, 58].

What Is the Treatment for Displaced Paediatric Ankle Fractures?

The decision for operative intervention is obvious when displacement is substantial. The aim is for gentle reduction to as anatomical position as possible is desirable and often achievable. However, when displacement is minor, the decision for intervention can be tricky. Correct identification of a child ankle fracture according to Dias-Tachjidian classification aids (Table 22.5) its close reduction. We describe below the current approach to treat displaced fractures based on Salter-Harris classification.

Table 22.5 Dias-Tachdjian classification of children ankle fracture

Dias and Tachdjian	Sequence of structures failure with increasing force caused by an injury
Supination – external rotation (SER) (foot is supinated and the force is external rotation)	Stage I: SH-II fracture of the distal tibial epiphysis with a posterior metaphyseal-epiphyseal fragment displaced posteriorly. The distal tibial fracture begins on the lateral distal aspect and spirals medially and proximally. The fibula remains intact. This fracture is similar to a supination–plantar flexion injury, especially when seen on the lateral radiograph; the distinction is that the distal tibial fracture line begins on the distal lateral aspect and spirals medially when viewed on the AP projection
	Stage II: spiral fracture of the fibula. The fracture begins medially and extends superiorly and posteriorly
Supination – inversion injury (foot is supinated and the force is adduction or inversion)	Stage I: traction by the lateral ligaments produces a SH-I or II fracture of the distal fibular physis. Lateral ligamentous injury can occur but is rare as the physis is usually weaker than the ligament
	Stage II: the talus impacts against the medial malleolus causing SH III or IV injury, occasionally a SH-II and rarely type I of the distal tibia
Pronation–eversion–external rotation fracture	SH-I or II fracture of the distal tibia with a transverse or short oblique fibular fracture located 4–7 cm proximal to the tip of the lateral malleolus When a SH-II fracture occurs, the metaphyseal fragment is located laterally or posterolaterally and the distal tibial fragment is displaced laterally and posteriorly
Supination–plantar flexion injury	SH-II physeal injury of the distal tibial physis with posterior displacement of the epiphyseal-metaphyseal fragment and no fracture of the fibula. The metaphyseal fragment of the tibia is posterior and best seen on a lateral radiograph

Table 22.6 Growth disturbance in distal tibial ankle fracture

Types of fractures	Rate of growth disturbance (%)
SH-III and IV (medial malleolar type) fractures	38
SH-I and II fractures	36
Triplane fracture	21
Tillaux fractures	0

Salter-Harris Type I and II Tibial

These two types of ankle fractures usually behave in the same way. SH-I is relatively rare (15 %) whereas SH-II is common (38 %) of all distal tibial fractures in children [18]. Treatment consists of gentle closed reduction by reversing the original mechanism of injury, followed by an above knee cast immobilisation. Follow-up is recommended in the first week to ensure no further displacement.

In a retrospective study of 92 distal tibial physeal fracture by Barmada [59], 25 fractures (27.2 %) were complicated by growth disturbance. These are summarised in Table 22.6.

They found that initial displacement, number of reduction attempts, or treatment method did not significantly affect the incidence of growth disturbance. However, the more anatomic reductions resulted in a statistically significant decrease in growth disturbance rates. The rate of growth disturbance was tripled when a residual gap was seen on the radiograph (60 % vs. 17 %).

In another study by Rohmiller et al. [60], around 40 % of 91 distal tibial SH-I or II developed growth disturbance. The mechanism of injury was significant in developing growth arrest: 35 % (17/48) in patients with a supination-external-rotation-type injury (SER) and 54 % (14/26) in patients with pronation-abduction-type injuries (ABD). Initial displacement was significantly greater in patients with ABD (11.7 + 8 mm) injuries than those with SER (4.9 + 3 mm) (P = 0.001). Non-operative treatment resulted in growth disturbance in 56 % of cases in comparison to 16 % with operative treatment, this was not statistically significant (P = 0.16). They also reported that the most important determinant of growth disturbance is the fracture displacement following reduction. It is of note that both above studies came from the same centre and almost the same time and likely to represent the same cohort of patients.

Salter-Harris Type III and IV Tibial

These fractures types are challenging because of the involvement of the articular surface as well as the physis. They occur in approximately 20 % and 1 % of all distal tibiofibular fractures in children. They encompass a variety of anatomically distinct fractures such as Tillaux fracture (SH-III of the anterolateral tibial plafond), McFarland fracture (SH-III or IV of the medial tibial plafond) and triplane fracture. Treatment of undisplaced fractures consists of protection with a cast and careful follow-up on a weekly basis to ensure maintenance of fracture. Fractures that are displaced more than 2 mm should be reduced by either closed or open reduction followed by screw fixation.

Spiegel [18] reported that 88 % (46/52) of these fractures occurred on the medial side of the tibial plafond. They are also called **McFarland fractures** (Fig 22.5) and they have been associated with a high rate of growth disturbance.

Petratos et al. [61] reported on 20 children with surgically treated McFarland fractures at a mean follow-up of 8.9 years. Seven children (35 %) developed growth disturbance and angular deformity. Initial displacement of more than 6 mm

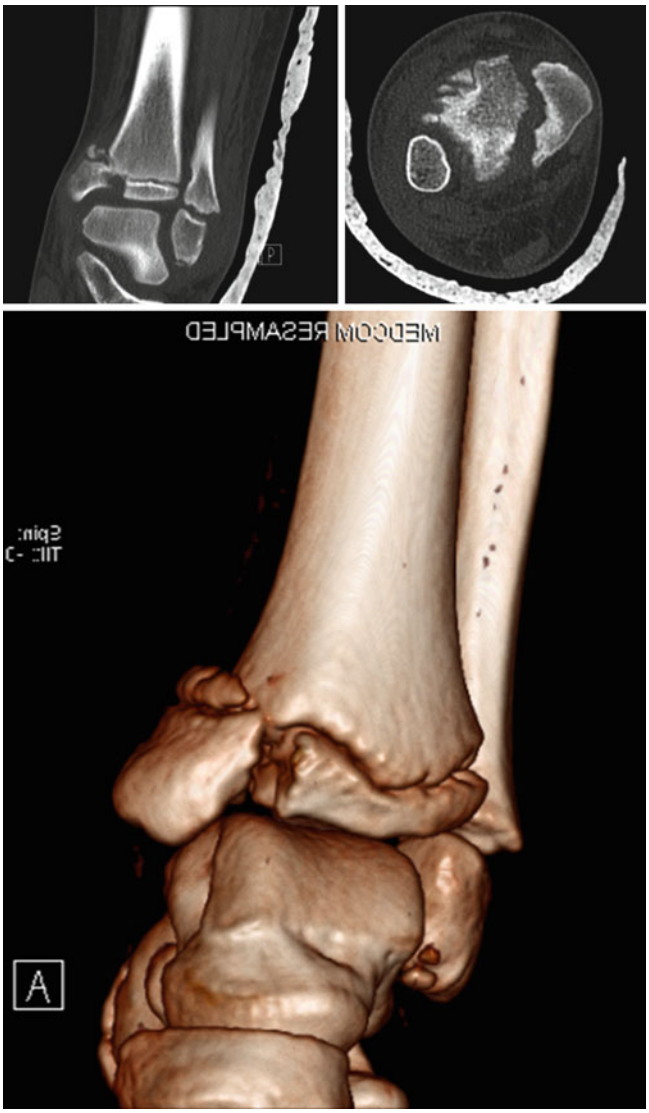

Fig. 22.5 McFarland fracture

(p = 0.004) and operative delay beyond 24 h (p = 0.007) were significant risk factors for growth disturbance.

Kling et al. [62] evaluated 37 children with established growth arrest (drawn from two different cohorts of ankle fractures) and found that 75 % had been treated by closed methods. Most of these were not anatomically reduced.

Cottalorda et al. [63] reviewed 48 patients with McFarland fractures (30 SH-III and 18 SH-IV). All fractures were displaced >1 mm, meeting their operative indications, and were treated with open arthrotomy and fracture reduction under direct visualisation with screw (46) or pin (2) fixation. They reported 45 good and 2 fair results, and only one patient with angular deformity (6° of varus) at a mean follow-up of 3.25 years.

Schurz et al. [20] reported on a large retrospective series of 376 patients. All non-displaced fractures were treated by plaster cast immobilisation (118 children). Displaced fractures, regardless the degree of displacement, were treated by open or closed reduction, with or without internal fixation, to achieve an absolutely anatomical reduction. They showed that 77 displaced physeal fractures of the distal tibia were reconstructed anatomically by open or closed reduction and produced 95 % excellent results. They recommended a perfect anatomical reduction, if necessary by open means, should be achieved to prevent a bone bridge with subsequent epiphysiodesis and post-traumatic deformities due to growth inhibition and/or retardation.

Tillaux fracture was first described by Cooper in 1822 then by Tillaux in 1848 [64–66] and it is the result of avulsion injuries of the lateral epiphyseal plate caused by external rotation of the ankle. The lateral part of physeal plate is usually the last to close. And the timeline for this fracture is during an 18 month period of late growth [67]. The current treatment vogue is non operative treatment when displacement is less than 2 mm and either closed or open reduction and stabilisation when the displacement is more than 2 mm [12]. Excellent outcomes have been widely reported with the above treatments options [68–74]. However there is a lack of long term studies to show similarly good outcome following non operative treatment in Tillaux fracture with displacement of more than 2 mm.

Triplane fractures of the distal tibia are relatively uncommon. They account for 6–8 % of all distal tibial physeal fracture [12, 18, 75]. The treatment options of triplane fractures are not different form that of Tillaux ones. The goal of the treatment is to achieve anatomic reduction of the distal tibial articular surface to prevent potential long-term degenerative changes. When fracture displacement is minimal (<2 mm), nonoperative treatment is recommended with an above knee case. However, if displacement > 2 mm, closed reduction can be performed with axial traction and internal rotation of the foot with the patient under general anaesthesia. Failure of closed reduction to achieve satisfactory position is an indication for open reduction and internal fixation.

Ertl et al. followed 23 children with triplane fractures, they found that favourable outcome was related to articular congruity of the weight bearing part of the distal tibia [76]. They found that residual displacement of 2 mm or more was associated with suboptimum result unless the epiphyseal fracture was outside the primary weight-bearing area of the ankle.

Another study of 35 children with triplane fractures who were followed up for more than 5 years reached the same conclusion; prognosis was good only when adequate reduction (< 2 mm displacement) had been achieved [77].

Weinberg et al. [78] in an evaluation of 50 children with triplane fractures, including 30 who had operative treatment and 20 who had non-operative treatment, reported that all

Table 22.7 Recommendations

Statement	Grade of recommendation
Stress views are not required to aid decision on treating undisplaced ankle fractures	B
CT scan should be requested in all physeal fractures	B/C
MRI scan is not routinely indicated in children ankle injury	C
Undisplaced ankle fracture in children can be safely treated non operatively	B
Displaced physeal and/ or intra-articular fracture should be reduced to anatomical position. Failure of closed reduction is an indication for open reduction	B
2 mm displacement is the threshold for reducing displaced fractures or accepting reduction as satisfactory	B/C
Above knee cast is better than below knee cast for physeal ankle injury	I

patients were doing well after a mean follow-up of 7.4 years. There were no significant differences between the group treated non-operatively and the group that underwent surgery. Only two of the initially undisplaced fractures later became displaced concluding that operative treatment is not indicated for all undisplaced fractures.

Recommendations for treatment are listed in Table 22.7.

References

1. Barrie J, Jackson G. 2013. Ankle fracture, Foot hyperbook. 2015, from http://www.foothyperbook.com/elective/neuroma/neuromaIntro.htm
2. Gardner MJ, Demetrakopoulos D, et al. The ability of the Lauge-Hansen classification to predict ligament injury and mechanism in ankle fractures: an MRI study. J Orthop Trauma. 2006;20(4): 267–72.
3. Michelson JD, Magid D, et al. Clinical utility of a stability-based ankle fracture classification system. J Orthop Trauma. 2007;21(5): 307–15.
4. Lauge-Hansen N. Fractures of the Ankle. Arch Surg. 1950; 60(5):957.
5. Haraguchi N. A new interpretation of the mechanism of ankle fracture. J Bone Joint Surg Am. 2009;91(4):821.
6. Michelson J, Solocoff D, et al. Ankle fractures. The Lauge-Hansen classification revisited. Clin Orthop Relat Res. 1997;(345): 198–205.
7. Hermans JJ, Wentink N, et al. Correlation between radiological assessment of acute ankle fractures and syndesmotic injury on MRI. Skeletal Radiol. 2011;41(7):787–801.
8. Alshryda, S. and Wright J.. Injuries involving the epiphyseal plate. Classic papers in Orthopaedics. Springer Science + Business Media; 2013. 555–7.
9. Salter R, Harris W. Injuries involving the epiphyseal plate. J Bone Joint Surg Am. 1963;1963(45A):587–622.
10. Leary JT, Handling M, et al. Physeal fractures of the distal tibia. J Pediatr Orthop. 2009;29(4):356–61.
11. Dias LS, Tachdjian MO. Physeal injuries of the ankle in children. Clin Orthop Relat Res. 1978;(136):230–33.
12. Tachdjians MO. Pediatric orthopaedics. Philadelphia: Saunders Elsevier; 2013.
13. Alshryda S, Jones S, et al. Postgraduate paediatric orthopaedics: the candidate's guide to the FRCS (Tr and Orth) examination. Cambridge University: Cambridge University Press; 2014.
14. Lynn MD. The triplane distal tibial epiphyseal fracture. Clin Orthop Relat Res. 1972;86:187–90.
15. van Laarhoven CJ, van der Werken C. 'Quadriplane' fracture of the distal tibia: a triplane fracture with a double metaphyseal fragment. Injury. 1992;23(7):497–9.
16. van Laarhoven CJHM, Severijnen RSVM, et al. Triplane fractures of the distal tibia. J Foot Ankle Surg. 1995;34(6):556–9.
17. Karrholm J, Hansson LI, et al. Prediction of growth pattern after ankle fractures in children. J Pediatr Orthop. 1983;3(3):319–25.
18. Spiegel PG, Cooperman DR, et al. Epiphyseal fractures of the distal ends of the tibia and fibula. A retrospective study of two hundred and thirty-seven cases in children. J Bone Joint Surg Am. 1978;60(8):1046–50.
19. de Sanctis N, Della Corte S, et al. Distal tibial and fibular epiphyseal fractures in children: prognostic criteria and long-term results in 158 patients. J Pediatr Orthop B. 2000;9(1):40–4.
20. Schurz M, Binder H, et al. Physeal injuries of the distal tibia: long-term results in 376 patients. Int Orthop. 2010;34(4):547–52.
21. Vahvanen V, Aalto K. Classification of ankle fractures in children. Arch Orthop Trauma Surg. 1980;97(1):1–5.
22. Jonckheer P, Willems T, et al. Evaluating fracture risk in acute ankle sprains: any news since the Ottawa ankle rules? A systematic review. Eur J Gen Pract. 2015:1–11.
23. Stiell I, Wells G, et al. Multicentre trial to introduce the Ottawa ankle rules for use of radiography in acute ankle injuries. BMJ. 1995;311(7005):594–7.
24. Aginaga Badiola JR, Ventura Huarte I, et al. Validation of the Ottawa ankle rules for the efficient utilization of radiographies in acute lesions of the ankle. Aten Primaria. 1999;24(4):203–8.
25. Curr S, Xyrichis A. Does nurse-led initiation of Ottawa ankle rules reduce ED length of stay? Int Emerg Nurs. 2015;23(4):317–22.
26. Allerston J, Justham D. A case-control study of the transit times through an accident and emergency department of ankle injured patients assessed using the Ottawa ankle rules. Accid Emerg Nurs. 2000a;8(3):148–54.
27. Allerston J, Justham D. Nurse practitioners and the Ottawa ankle rules: comparisons with medical staff in requesting X-rays for ankle injured patients. Accid Emerg Nurs. 2000b;8(2):110–5.
28. Derksen RJ, Bakker FC, et al. Diagnostic accuracy and reproducibility in the interpretation of Ottawa ankle and foot rules by specialized emergency nurses. Am J Emerg Med. 2005;23(6): 725–9.
29. Clark KD, Tanner S. Evaluation of the Ottawa ankle rules in children. Pediatr Emerg Care. 2003;19(2):73–8.
30. Cuello-Garcia C, Ruiz-Flores A, et al. The Ottawa ankle rules: evaluation in a pediatric emergency setting in Mexico. An Pediatr (Barc). 2004;60(5):454–8.
31. Dowling S, Spooner CH, et al. Accuracy of Ottawa ankle rules to exclude fractures of the ankle and midfoot in children: a meta-analysis. Acad Emerg Med. 2009;16(4):277–87.
32. Karpas A, Hennes H, et al. Utilization of the Ottawa ankle rules by nurses in a pediatric emergency department. Acad Emerg Med. 2002;9(2):130–3.
33. Libetta C, Burke D, et al. Validation of the Ottawa ankle rules in children. J Accid Emerg Med. 1999;16(5):342–4.
34. Plint AC, Bulloch B, et al. Validation of the Ottawa ankle rules in children with ankle injuries. Acad Emerg Med. 1999;6(10): 1005–9.

35. Runyon MS. Can we safely apply the Ottawa ankle rules to children? Acad Emerg Med. 2009;16(4):352–4.
36. Yuen MC, Saunders F. Towards evidence based emergency medicine: best BETs from the Manchester Royal Infirmary. The Ottawa ankle rules in children. Emerg Med J. 2001;18(6):466–7.
37. Crocco A. Review: the Ottawa Ankle Rules are accurate for excluding fractures in acute blunt ankle/midfoot injury in children. Evid Based Med. 2009;14(6):184.
38. Brage ME, Rockett M, et al. Ankle fracture classification: a comparison of reliability of three X-ray views versus two. Foot Ankle Int. 1998;19(8):555–62.
39. Vangsness CT, Carter V, et al. Radiographic diagnosis of ankle fractures: are three views necessary? Foot Ankle Int. 1994;15(4): 172–4.
40. Murphy JM, Kadakia AR, et al. Variability in radiographic medial clear space measurement of the normal weight-bearing ankle. Foot Ankle Int. 2012;33(11):956–63.
41. Koval KJ, Egol KA, et al. Does a positive ankle stress test indicate the need for operative treatment after lateral malleolus fracture? A preliminary report. J Orthop Trauma. 2007;21(7):449–55.
42. Schuberth JM, Collman DR, et al. Deltoid ligament integrity in lateral malleolar fractures: a comparative analysis of arthroscopic and radiographic assessments. J Foot Ankle Surg. 2004;43(1):20–9.
43. McConnell T, Creevy W, et al. Stress examination of supination external rotation-type fibular fractures. J Bone Joint Surg Am. 2004;86-A(10):2171–8.
44. Schock HJ, Pinzur M, et al. The use of gravity or manual-stress radiographs in the assessment of supination-external rotation fractures of the ankle. J Bone Joint Surg Br. 2007;89-B(8):1055–9.
45. Akhtar S, Fox A, et al. Pragmatic treatment of ankle fractures of uncertain stability: clinical features and risk of displacement. Injury Extra. 2009;41(12):185.
46. Hoshino CM, Nomoto EK, et al. Correlation of weightbearing radiographs and stability of stress positive ankle fractures. Foot Ankle Int. 2012;33(2):92–8.
47. Weber M, Burmeister H, et al. The use of weightbearing radiographs to assess the stability of supination-external rotation fractures of the ankle. Arch Orthop Trauma Surg. 2012;130(5):693–8.
48. Nenopoulos A, Beslikas T, et al. The role of CT in diagnosis and treatment of distal tibial fractures with intra-articular involvement in children. Injury. 2015;46(11):2177–80.
49. Eismann EA, Stephan ZA, et al. Pediatric triplane ankle fractures: impact of radiographs and computed tomography on fracture classification and treatment planning. J Bone Joint Surg Am. 2015;97(12):995–1002.
50. Black EM, Antoci V, et al. Role of preoperative computed tomography scans in operative planning for malleolar ankle fractures. Foot Ankle Int. 2013;34(5):697–704.
51. Horn BD, Crisci K, et al. Radiologic evaluation of juvenile tillaux fractures of the distal tibia. J Pediatr Orthop. 2001;21(2):162–4.
52. Hou Z, Zhang Q, et al. A occult and regular combination injury: the posterior malleolar fracture associated with spiral tibial shaft fracture. J Trauma. 2009;66(5):1385–90.
53. Lohman M, Kivisaari A, et al. Acute paediatric ankle trauma: MRI versus plain radiography. Skeletal Radiol. 2001;30(9):504–11.
54. Petit P, Panuel M, et al. Acute fracture of the distal tibial physis: role of gradient-echo MR imaging versus plain film examination. AJR Am J Roentgenol. 1996;166(5):1203–6.
55. Carey J, Spence L, et al. MRI of pediatric growth plate injury: correlation with plain film radiographs and clinical outcome. Skeletal Radiol. 1998;27(5):250–5.
56. Stuart PR, Brumby C, et al. Comparative study of functional bracing and plaster cast treatment of stable lateral malleolar fractures. Injury. 1989;20(6):323–6.
57. Brink O, Staunstrup H, et al. Stable lateral malleolar fractures treated with aircast ankle brace and DonJoy R.O.M.-Walker Brace: a prospective randomized study. Foot Ankle Int. 1996;17(11): 679–84.
58. Tornetta P, Axelrad TW, et al. Treatment of the stress positive ligamentous SE4 ankle fracture. J Orthop Trauma. 2012;26(11): 659–61.
59. Barmada A, Gaynor T, et al. Premature physeal closure following distal tibia physeal fractures. J Pediatr Orthop. 2003;23(6): 733–9.
60. Rohmiller MT, Gaynor TP, et al. Salter-Harris I and II fractures of the distal tibia. J Pediatr Orthop. 2006;26(3):322–8.
61. Petratos DV, Kokkinakis M, et al. Prognostic factors for premature growth plate arrest as a complication of the surgical treatment of fractures of the medial malleolus in children. Bone Joint J. 2013;95-B(3):419–23.
62. Kling Jr TF, Bright RW, et al. Distal tibial physeal fractures in children that may require open reduction. J Bone Joint Surg Am. 1984;66(5):647–57.
63. Cottalorda J, Beranger V, et al. Salter-Harris Type III and IV medial malleolar fractures: growth arrest: is it a fate? A retrospective study of 48 cases with open reduction. J Pediatr Orthop. 2008;28(6): 652–5.
64. Cooper A.. On dislocation of the ankle joints. A treatise on dislocations and on fractures of the joints. Longman, Hurst, Rees, Orme and Brown, E Cox and Son; 1822.
65. Sharma B, Reddy IS, et al.. The adult Tillaux fracture: one not to miss. BMJ Case Rep. 2013.
66. Tillaux P. Trait de Chirurgie clinique. Paris: Asselin and Houzeau; 1848.
67. Kleiger B, Mankin HJ. Fracture of the lateral portion of the distal tibial epiphysis. J Bone Joint Surg Am. 1964;46:25–32.
68. Cassas KJ, Jamison JP. Juvenile tillaux fracture in an adolescent basketball player. Phys Sportsmed. 2005;33(2):30–3.
69. Choudhry IK, Wall EJ, et al. Functional outcome analysis of triplane and tillaux fractures after closed reduction and percutaneous fixation. J Pediatr Orthop. 2014;34(2):139–43.
70. Crawford AH. Triplane and Tillaux fractures: is a 2 mm residual gap acceptable? J Pediatr Orthop. 2012;32(Suppl 1):S69–73.
71. Felman AH. Tillaux fractures of the tibia (in adolescents). Pediatr Radiol. 1989;20(1–2):87–9.
72. Kaya A, Altay T, et al. Open reduction and internal fixation in displaced juvenile Tillaux fractures. Injury. 2007;38(2):201–5.
73. Kim JR, Song KH, et al. Treatment outcomes of triplane and Tillaux fractures of the ankle in adolescence. Clin Orthop Surg. 2010;2(1): 34–8.
74. Leetun DT, Ireland ML. Arthroscopically assisted reduction and fixation of a juvenile Tillaux fracture. Arthroscopy. 2002;18(4): 427–9.
75. Dias LS, Giegerich CR. Fractures of the distal tibial epiphysis in adolescence. J Bone Joint Surg Am. 1983;65(4):438–44.
76. Ertl JP, Barrack RL, et al. Triplane fracture of the distal tibial epiphysis. Long-term follow-up. J Bone Joint Surg Am. 1988;70(7): 967–76.
77. Rapariz JM, Ocete G, et al. Distal tibial triplane fractures: long-term follow-up. J Pediatr Orthop. 1996;16(1):113–8.
78. Weinberg AM, Jablonski M, et al. Transitional fractures of the distal tibia. Injury. 2005;36(11):1371–8.

Part V

The Spine

Evidence-Based Treatment of Adolescent Idiopathic Scoliosis

23

David Lebel

Abstract

Adolescent Idiopathic scoliosis (AIS) is the most common deformity of the spinal column. Most of the diagnosed patients have mild or moderate deformity and therefore they need observation and some kind of conservative treatment but, those patients with significant deformities often require surgical treatment in order to correct and prevent further deterioration. The general guidelines for the treatment of AIS are based on the severity of the deformity, the age and development of the individual patient. The evidence for various treatments of AIS; whether conservative or surgical, is often suboptimal. In the following review we explored and summarised the best available evidence to support current practice.

Keywords

Scoliosis • Adolescent idiopathic scoliosis • Braces • Bracing • Pedicle screws • Hooks • Lenke classification

Introduction

Adolescent idiopathic scoliosis (AIS) is defined as side-to-side shift of the spine with a Cobb angle measured above 10° in patients older than 10 years. Although the definition is of a single plane deformity, scoliosis is a three dimensional deformity with sagittal hypo-kyphosis and axial rotation. Two classifications are in common use to characterize various types of curve: King-Moe and Lenke classifications. Lenke [1] claimed that his classification a more comprehensive, based on objective criteria from each curve type, emphasise consideration of sagittal alignment and help standardise treatment. According to Lenke, there are three spine regions:

1. Proximal Thoracic (Apical vertebra located at T3-5)
2. Main Thoracic (Apical vertebra located at T6-11 or T12 disc)
3. Thoracolumbar/Lumbar (Apical vertebra located at T12-L1 for thoracolumbar curves and between L1-2 disc and L4 for lumbar curves)

Each curve is further classified into a major curve (the curve segment with the largest Cobb angle) and a minor curve (the curve segment with the smaller Cobb angle). Major curves are always considered structural while minor curves can be considered structural only if minimal residual coronal curves of 25° on bending film and kyphosis of at least 20°. Therefore, there are six curve types corresponding to different combinations of structural and non-structural curve patterns (Table 23.1). Lenke further proposed two modifiers: lumbar modifier to emphasis the importance of deformities in the lumbar region and the thoracic spine sagittal modifier to introduce a three-dimensional analysis to the classification system.

Current treatment for AIS is tailored based on the severity of the deformity as measured by the Cobb method, patients' age and skeletal development. While severe deformities (Cobb angle >50°) are rather treated surgically, milder deformities are treated conservatively.

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Table 23.1 Lenke classification system

Curve type	Description	Proximal thoracic	Main thoracic	Thoraco-lumbar/lumbar
1	Main thoracic	Non-structural	Structural	Non-structural
2	Double thoracic	Structural	Structural (major)	Non-structural
3	Double major	Non-structural	Structural (major)	Structural
4	Triple major	Structural	Structural (major)	Structural
5	Thoraco-lumbar/lumbar	Non-structural	Non-structural	Structural
6	Thoraco-lumbar/ lumbar – main thoracic	Non-structural	Structural	Structural (major)

Key point for building a treatment rationale in a poor evidence environment is a thorough knowledge of the natural history of untreated AIS.

What Is the Natural History of Adolescent Idiopathic Scoliosis?

Mortality

Historical cohorts analyzed and published by Pehrsson et al. [2] revealed that differently from infantile and juvenile scoliosis where there is a strong correlation between curve magnitude and mortality rate, in AIS the mortality rate is as in the general population regardless the severity of the curve. Weinstein et al. [3] published their 51 years of follow up with similar conclusions regarding mortality rate in AIS.

Pulmonary Morbidity

While pulmonary involvement is evident among patients with severe AIS, respiratory compromise occurs only in severe scoliosis with Cobb angle greater that 80° [3].

Curve Progression

Long term follow up revealed slow deterioration of curve magnitude with the time. Rate of deterioration was calculated to be 0.79° per year in thoracic deformities with Cobb angle larger than 50° [4].

Untreated severe AIS might increase without treatment beyond skeletal growth and eventually may cause pulmonary impairment. This statement together with body image impairment is the rationale for treating AIS.

The question "What is the best treatment for AIS" will be answered in this chapter by looking for the best evidence to support it.

Non-surgical Treatment for AIS

Searching the literature for the best conservative approach to treat Scoliosis brought various options: Scoliosis specific exercise (SSE), manipulations, physiotherapy and Brace treatment.

Brace Treatment

Brace treatment for scoliosis exists for many years. Numerous studies regarding efficacy were published threw out the years although most of them were of low quality regarding level of evidence.

Weinstein et al. [5] published the BrAIST study; the only level 1 study that exists regarding scoliosis treatment. In which effectiveness of brace treatment was studied in a multicenter randomized control study. Inclusion criteria as for the scoliosis research society (SRS) guidelines were age 10–15 years. Riser sign 0–2 and deformity with severity of 20–40° measured by the Cobb technique. Failure was defined as curve progression to 50°. BrAIST was conducted in 25 centres in North America. Participants were treated either with a thoracolumbosacral orthosis or watchful waiting and followed every 6 months until they reached skeletal maturity or the surgical threshold of 50° Cobb angle.

The results were highly significant with success of 72 % among treatment group and 48 % among control group. Moreover, there was a strong correlation between number of treatment hours and the rate of treatment success.

Although several level 2 studies were published prior to this study, The BRAIST study is the benchmark for conservative treatment of [4].

Scoliosis Specific Exercise Treatment (SSE)

Literature search revealed few low level studies and several reviews regarding SSE. The latest Cochrane systemic review published in 2012 [6] concluded that there is not enough evidence to support SSE for the treatment of scoliosis.

Osteopathy, Yoga, Pilates

There is no good quality evidence to support these treatments for AIS.

Surgical Treatment for AIS

When non-surgical treatment fails and curve progress is noted, surgical treatment is the treatment of choice. Several questions regarding indication for surgery are a matter of ongoing debate with some weak evidence that helps the health care professionals to develop a treatment rationale. Again, knowledge of the natural history of untreated AIS helps with decision-making and setting treatment goals. The Indication for surgical intervention in AIS patient is deformity that is measured >50°. Although it is evident from natural history studies that severe scoliosis does not increase mortality rates [2, 3]. The impaired pulmonary function that is associated with severe scoliosis is probably the trigger for surgical intervention. The goals for surgical intervention, whether anterior, posterior or combined approach are spinal fusion aimed at stopping deformity progression, correction of the deformity, restoring coronal and sagittal balance and improvement of cosmetic appearance. Several questions are often raised when considering surgical treatment of AIS:

What Is the Best Surgical Approach to Treat Scoliosis?

Selecting the best surgical treatment for AIS is a matter of surgeons' expertise, location of the curve best described by classification, curves magnitude and preference of the patient. Essentially, surgical approach could be divided into posterior only, anterior only and combined approach. Anterior approach might be just release when combined with posterior fusion and fixation or, anterior fusion and fixation. With the development of modern surgical equipment based on pedicle screws for three-column fixation, posterior approach gained popularity and became the standard technique for most AIS surgeries.

Chen and Ron [7] published their recent meta-analysis in which they compared posterior only to anterior and posterior surgeries. They concluded that with posterior only approach, sagittal correction is better, surgeries are shorter, with less blood loss and less complications.

Schmidt et al. [8] studied retrospectively AIS patients that were treated with either anterior spinal fusion or posterior spinal fusion for Lenke type curve 1–3. Both groups showed similar coronal plane correction but the posterior group had significantly less sagittal correction, longer operating time and more blood loss.

Several studies compared surgical approach in Lenke 5 deformities (major thoracolumbar/lumbar curves). Geck et al. [9] compared retrospectively two groups of patients that were treated in two different institutions. The patients in the posterior only group had significantly better curve correction, less loss of correction over time and shorter hospital stay. There were no complications in both groups.

Li et al. [10] reviewed their group of Lenke 5 patients that were treated either with anterior spinal fusion or posterior spinal fusion. Two groups were comparable in their inclusion parameters and the authors concluded that both surgical methods are comparable in deformity correction but, posterior group had shorter operation time and hospital stay whereas the anterior group had less posterior fusion group had less fusion levels. Wang et al. [11] performed a prospective study that compared anterior versus posterior spinal fusions for AIS patients with Lenke 5 deformity. Although each group was very small, they concluded that the anterior group had less fusion levels and less blood loss. There were no complications in both groups. Their conclusion was that because of these results, anterior spinal fusion is preferable in AIS patients with Lenke 5 deformity.

The best surgical approach to treat AIS is probably the one that the surgeon is most comfortable with. The amount of correction achieved is comparable; the number of fused levels in Lenke 5 deformities is significantly smaller when performing anterior approach but, regarding blood loss, length of surgery and length of stay the data is diverse.

Interestingly, in none of the studies complications were reported and the conclusion was that both anterior and posterior surgeries are safe. The safety of different surgical approach was evaluated previously analyzing the morbidity mortality data of the scoliosis research society [12]. It was found that complication rate in anterior and posterior spinal surgeries is about 5 % but the complication rate of combined anterior- posterior surgeries was 10.2 % that is significantly higher.

What Is the Best Spinal Fixation Method (Pedicle Screws, Hooks or Hybrid)?

Surgical correction of AIS is based on the ability to get the best anchorage to the relevant spinal segment. The surgical apparatus and techniques were evolved from the era of the Harrington rod, which applied distraction forces on the spine, to the Cotrell Dubousset (CD) systems that allowed distraction, compression and translation of the spine to the pedicle screw systems that are widely used today and allow three dimensional correction by translation, distraction, compression and axial de-rotation of the vertebrae. Several studies compared these techniques.

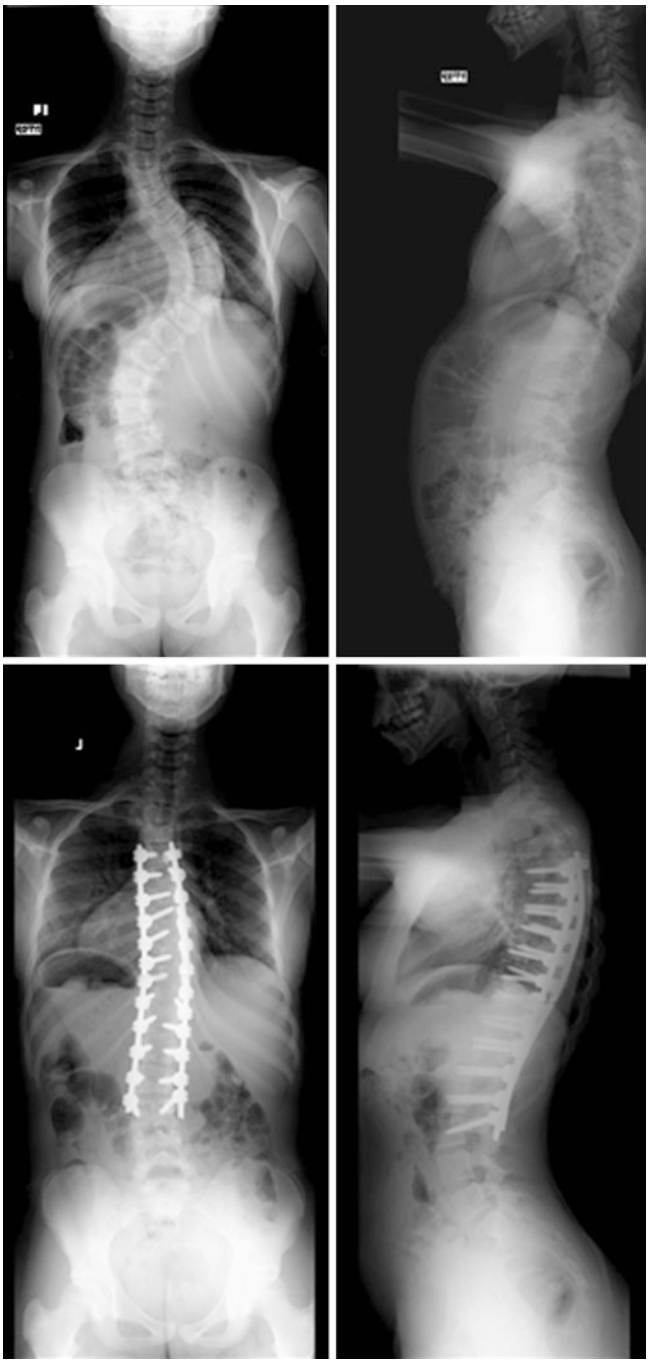

Fig. 23.1 A child with Lenke 1B scoliosis- treated with pedicle screw construction

Storer et al. [13] looked at the short-term results of patients that were treated with all pedicle screw construct (Fig. 23.1) versus patients that were treated with all hooks construct for thoracic AIS. Although very small number of patients in both groups, no difference was reported regarding deformity correction, apical vertebrae translation and coronal translation but it was noted that screw constructs are significantly more expensive. Dobbs et al. [14] investigated a larger group of patients with minimum 2 years of follow up. The pedicle screw group achieved significantly better correction immediately after surgery and less decompensating after minimum 2 years of follow up.

Several studies compared pedicle screw constructs to hybrid (distal screw combined with proximal hooks and/or wires) constructs [15–18]. Karatoprak et al. [15] compared pedicle screw treated AIS patients with Hybrid construct treated patients. The groups differed in the length of follow up with a significantly longer follow up time for the Hybrid treated patients. Nevertheless, no difference was found looking at the correction rate, coronal and sagittal balance, blood loss and duration of surgery. The pedicle screw group had a better correction durability and apical de-rotation when compared to the hybrid group. The author suggested that better correction durability is due to the three columns purchase of pedicle screws. However, significantly shorter follow up time (mean 60 months versus mean 29 months) might be another reason.

Kim et al. [16] in their retrospective comparative study looked at two matched cohorts with AIS treated with hybrid or pedicle screw constructs. They concluded that segmental pedicle screw instrumentation offers a significantly better major curve correction, less perioperative blood loss, and improved postoperative pulmonary function values without neurologic problems compared with hybrid constructs. Both instrumentation methods provide similar operative time, and postoperative SRS-24 outcome scores in the operative treatment of adolescent idiopathic scoliosis.

Looking at patients with AIS with severe deformity, curves with Cobb angel of 80° or greater, Di Silvestre et al. [17] compared hybrid to pedicle screw constructs retrospectively. The results were similar to results is previous studies. Better correction of coronal deformity with the screw construct with a better correction of the secondary non-instrumented curve. Nevertheless, quality of life and patient satisfaction didn't differ between the groups.

Recent study [18] emphasized the difference between hybrid and pedicle screws with regard to sagittal alignment. Independently of the surgical technique used, the cervical spine had a tendency to decompensation acquiring a kyphotic sagittal profile. The all pedicle screw construct found to have stronger thoracic hypo- kyphotic effect with a greater cervical decompensation compared to the hybrid construct. The coronal correction as in previous studies was significantly better with the pedicle screw constructs.

It is safe to conclude that the use of pedicle screws is probably as safe as hybrid constructs, the amount of coronal correction is comparable or maybe better with pedicle screws but, correction durability is probably better with screws. Sagittal correction might be better with hybrid constructs due to weaker hypo-kyphotic effect on the thoracic spine. Patient satisfaction as reported by SRS questionnaires is probably the same no matter what was the surgical technique.

Table 23.2 Recommendations

Statement	Grade of recommendation
Bracing can reduce the scoliosis progression and reduce the need for corrective surgery	C
Other conservative treatments (such as SSR and Osteopathy, Yoga, Pilates) are not effective in treating scoliosis	I
Surgery is indicated when curve magnitude is more than 50°	C
Clinical outcomes (correction, durability and complications) are not related to the surgical approach used	C
Clinical outcomes (correction, durability and complications) are not related to the types of fixation construct used	C

There are many other unsolved issues regarding treatment of AIS and there is a need for better evidence regarding the best treatment. Several new treatments are proposed during recent years, many of them are based on different strategies of growth modulation in order to avoid long spinal fusions. All this new techniques and treatment philosophy need to be supported by some evidence that does not exist at this time.

Current recommendations for treatment are outlined in Table 23.2**.**

References

1. Lenke LG, Betz RR, Harms J, et al. Adolescent idiopathic scoliosis: a new classification to determine the extent of spinal arthrodesis. J Bone Joint Surg Am. 2001;83:1160–81.
2. Pehrsson K, Larsson S, Oden A, et al. Long-term follow-up of patients with untreated scoliosis. A study of mortality, causes of death, and symptoms. Spine. 1992;17:1091–6.
3. Weinstein SL, Dolan LA, Spratt KF, et al. Health and function of patients with untreated idiopathic scoliosis: a 50-year natural history study. JAMA. 2003;289(5):559–67.
4. Weinstein SL, Zavala DC, Ponseti IV. Idiopathic scoliosis: long-term follow-up and prognosis in untreated patients. J Bone Joint Surg Am. 1981;63:702–12.
5. Weinstein SL, Dolan LA, Wright JG, Dobbs MB. Effects of bracing in adolescents with idiopathic scoliosis. N Engl J Med. 2013;369(16):1512–21.
6. Romano M, Minozzi S, Bettany-Saltikov J, Zaina F, Chockalingam N, Kotwicki T, Maier-Hennes A, Negrini S. Exercises for adolescent idiopathic scoliosis. Cochrane Database Syst Rev. 2012;8:CD007837.
7. Chen Z, Rong L. Comparison of combined anterior-posterior approach versus posterior-only approach in treating adolescent idiopathic scoliosis: a meta-analysis. Eur Spine J. 2015;22.
8. Schmidt C, Liljenqvist U, Lerner T, Schulte TL, Bullmann V. Sagittal balance of thoracic lordoscoliosis: anterior dual rod instrumentation versus posterior pedicle screw fixation. Eur Spine J. 2011;20(7):1118–26.
9. Geck MJ, Rinella A, Hawthorne D, Macagno A, Koester L, Sides B, Bridwell K, Lenke L, Shufflebarger H. Comparison of surgical treatment in Lenke 5C adolescent idiopathic scoliosis: anterior dual rod versus posterior pedicle fixation surgery: a comparison of two practices. Spine (Phila Pa 1976). 2009;34(18):1942–51.
10. Li M, Ni J, Fang X, Liu H, Zhu X, He S, Gu S, Wang X. Comparison of selective anterior versus posterior screw instrumentation in Lenke5C adolescent idiopathic scoliosis. Spine (Phila Pa 1976). 2009;34(11):1162–6.
11. Wang Y, Fei Q, Qiu G, Lee CI, Shen J, Zhang J, Zhao H, Zhao Y, Wang H, Yuan S. Anterior spinal fusion versus posterior spinal fusion for moderate lumbar/thoracolumbar adolescent idiopathic scoliosis: a prospective study. Spine (Phila Pa 1976). 2008;33(20):2166–72.
12. Coe JD, Arlet V, Donaldson W, Berven S, Hanson DS, Mudiyam R, Perra JH, Shaffrey CI. Complications in spinal fusion for adolescent idiopathic scoliosis in the new millennium. A report of the Scoliosis Research Society Morbidity and Mortality Committee. Spine (Phila Pa 1976). 2006;31(3):345–9.
13. Storer SK, Vitale MG, Hyman JE, Lee FY, Choe JC, Roye Jr DP. Correction of adolescent idiopathic scoliosis using thoracic pedicle screw fixation versus hook constructs. J Pediatr Orthop. 2005;25(4):415–9.
14. Dobbs MB, Lenke LG, Kim YJ, Kamath G, Peelle MW, Bridwell KH. Selective posterior thoracic fusions for adolescent idiopathic scoliosis: comparison of hooks versus pedicle screws. Spine (Phila Pa 1976). 2006;31(20):2400–4.
15. Karatoprak O, Unay K, Tezer M, Ozturk C, Aydogan M, Mirzanli C. Comparativeanalysis of pedicle screw versus hybrid instrumentation in adolescent idiopathic scoliosis surgery. Int Orthop. 2008;32(4):523–8 .discussion 529. Epub 2007 Apr 17
16. Kim YJ, Lenke LG, Kim J, Bridwell KH, Cho SK, Cheh G, Sides B. Comparative analysis of pedicle screw versus hybrid instrumentation in posterior spinal fusion of adolescent idiopathic scoliosis. Spine (Phila Pa 1976). 2006;31(3):291–8.
17. Di Silvestre M, Bakaloudis G, Lolli F, Vommaro F, Martikos K, Parisini P. Posterior fusion only for thoracic adolescent idiopathic scoliosis of more than 80 degrees: pedicle screws versus hybrid instrumentation. Eur Spine J. 2008;17(10):1336–49. doi:10.1007/s00586-008-0731-9.
18. Legarreta CA, Barrios C, Rositto GE, Reviriego JM, Maruenda JI, Escalada MN, Piza-Vallespir G, Burgos J, Hevia E. Cervical and thoracic sagittal misalignment after surgery for adolescent idiopathic scoliosis: a comparative study of all pedicle screws versus hybrid instrumentation. Spine (Phila Pa 1976). 2014;39(16):1330–7.

Evidence-Based Treatment of Neuromuscular Scoliosis

24

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Abstract

Scoliosis is a common clinical entity ubiquitous to most neurologic disorders encountered by the pediatric orthopaedic surgeon and associated care providers. Until recently, though widely performed, the evidence for scoliosis corrective surgery and its impact on patient-specific outcomes was sparse at best. In addition, the choice of surgical techniques applied during surgery for 'neuromuscular' scoliosis was based mainly on the results of historic case series with a paucity of higher-level evidence available. Over the last few years, however, the number and quality of relevant studies has increased substantially. The aim of this chapter is to provide a review of the best available evidence and provide recommendations as to most appropriate treatment(s) pertaining to the management of neuromuscular scoliosis. Given the results of this evidence-based review, it would seem that neuromuscular scoliosis correction is a worthwhile procedure with expected improvements in function, quality of life, and patient/caregiver satisfaction, albeit with high surgical risks. The ultimate decision as to whether or not scoliosis surgery is performed should be dependent on a disease-specific assessment of the risks and benefits with appropriate communication to the patient and/or caregiver. With respect to surgical fixation, the use of thoracic/lumbar segmental pedicle screws may offer improved outcomes over other methods but the best choice of pelvic fixation is still controversial. Regarding the use of anterior surgery for neuromuscular scoliosis, its role remains unclear except in the case of spina bifida where it is likely to reduce perioperative risks.

Keywords

Neuromuscular scoliosis • Cerebral palsy • Spinal bifida • Duchenne muscular dystrophy • Spinal fixation • Pelvic fixation • Anterior surgery • Quality of life

Introduction

Spinal deformities associated with neuromuscular diagnoses are common, typically progressive, and often require operative intervention. Despite a wealth of literature in this subject area, most of the published studies are retrospective, overly generalized by diagnosis, focused on radiographic rather than patient-centered outcomes, and are lacking a comparator group. This deficit in high quality studies makes it difficult to draw any hard conclusions regarding the most appropriate choice of operative strategy for scoliosis correction in these disorders. Recently however, there has been a shift towards comparative studies and the use of quality of

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life (QOL) related outcomes that serve to make the picture a little clearer.

The aim of this chapter is to provide a review of the best available evidence and provide recommendations as to most appropriate treatment(s) pertaining to the management of scoliosis associated with neuromuscular diagnoses [1].

Etiology and Morphology of Scoliosis in Neuromuscular Disorders

Spinal deformities secondary to neuromuscular disorders are common, with etiologies varied in pathophysiology and anatomic origin. These associated diagnoses can be categorized anatomically, stemming from upper motor neuron causes (e.g. cerebral palsy (CP), Friedrich's ataxia, syringomyelia), lower motor neuron causes (e.g. spinal muscular atrophy (SMA), poliomyelitis), or myopathic causes (e.g. Duchenne muscular dystrophy (DMD), myotonic dystrophy). From a pathophysiological point of view, scoliosis may be secondary to problems with weakness, coordination, and/or hypertonia of the truncal musculature coupled with a lack of effective compensatory mechanisms [1]. Neuromuscular curves typically present at a younger age and are known to exhibit rapid progression during growth that often continues, albeit more slowly, even after skeletal maturity [2].

Irrespective of diagnosis, the curve types associated with neuromuscular etiologies follow typical patterns. The most common curve type is long and C-shaped, often with associated pelvic obliquity, a collapsing kyphosis, and a loss of sitting balance (Fig. 24.1a) [2, 3]. The similarity in curve presentation across diagnoses has led to the convergence of surgical indications and operative strategies for the management of neuromuscular diagnoses, with little difference in approach between disorders except in specific cases (e.g. the exclusion of pelvic fixation in the absence of pelvic obliquity).

Goals of Scoliosis Correction: Patient Versus Surgical

The mainstay of treatment for neuromuscular curves involves posterior instrumentation and fusion from the upper thoracic spine (typically T2 or T3) to the pelvis. Many options for spinal fixation have been previously reported including the use of sublaminar wires or bands, segmental pedicle screw fixation, and hybrid methods involving more than one implant type (Fig. 24.1b–d) [4–6].

Indications for surgery have traditionally been met when the major curve reached a Cobb angle of 40–50° and/or there was a significant functional deficit, specifically with respect to sitting tolerance. The goals of the patient and/or caregiver, however, center on the expectation of improvements in activities of daily living (e.g. dressing, independent ambulation, personal hygiene), the absence of pain, ease of care-giving, social interaction, in addition to comfortable sitting [7]. In other words, the patient's major expectations revolve around issues pertaining to QOL.

Despite these goals, the published literature has, until recently, focused primarily on radiographic outcomes such as Cobb angle correction, implant type and density, pseudoarthrosis rates, and surgical approaches, in addition to reports of peri-operative complications [8–10]. This focus may be at least partially misplaced as several studies have reported a lack of correlation between the extent of Cobb angle correction and QOL improvements in children with neuromuscular disorders [7, 11, 12]. Arguably, the only radiographic goals that might have a substantial overall impact are the achievements of (1) a balanced spine over a level pelvis and (2) a solid spinal fusion.

Of course, the patient and/or caregiver have a substantial interest in the surgical risks associated with these procedures but this represents only one side of the risk-to-benefit ratio. A true evidence-based assessment of the available literature must give priority to those studies that report outcomes related to factors that matter to the patient and/or caregiver. At the present time, this may prove to be a difficult task given that, previous to 2011, there were very few published studies that reported QOL measures as outcomes of neuromuscular scoliosis correction [11]. However, in recent years, validated outcome measures have been developed which should prompt an increase in studies that prioritize outcomes on both sides of the risk-benefit ratio [13]. At this time, however, the number of studies that measure patient-specific outcomes remain scant as will be seen throughout the course of this chapter.

Is 'Neuromuscular Scoliosis' a Diagnosis to Be Analyzed?

The definition of what is considered a 'neuromuscular' diagnosis is variable amongst clinicians and is a source of some confusion, especially when reviewing the relevant literature on the subject. From the neurologists' view, the term typically refers to neurologic disorders that are progressive with respect to their primary etiology (e.g. DMD) versus those that that arise from a static neurologic lesion with associated progressive musculoskeletal manifestations (e.g. CP) [14]. The distinction is important given that these diagnoses have different natural histories, different levels of gross motor function (depending on disease severity), different surgical risk profiles (e.g. ventriculo-peritoneal shunt failure in myelomeningocele), and differing evidence regarding the utility of surgical interventions such as scoliosis correction [3, 7].

Despite these differences, for reasons likely related to improving sample sizes in studies investigating the role of scoliosis surgery for disorders with varying prevalence, disparate diagnoses such as cerebral palsy, spina bifida, Duchenne muscular dystrophy and others are often 'lumped together' and analyzed as if they were equivalent entities [9, 15]. Although the motivations are well understood, the

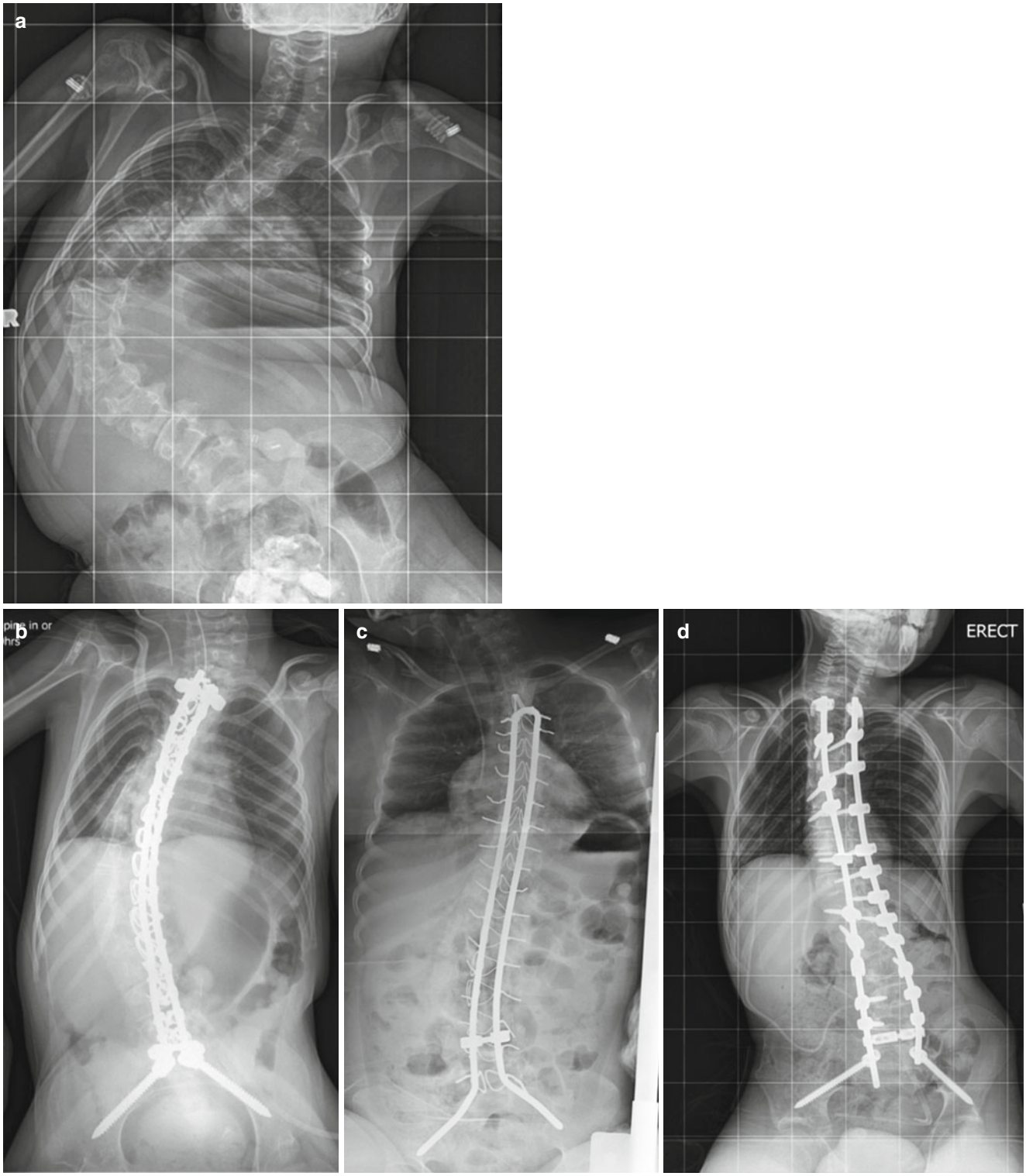

Fig. 24.1 Typical neuromuscular curve type and options for spinal fixation. (**a**) C-shaped thoracolumbar curve with associated pelvic obliquity. (**b**) Hybrid fixation including Luque sublaminar wires and sacral alar iliac screws. More commonly, lumbar pedicle screws are present in addition to sacropelvic screw fixation. (**c**) Unit rod including Luque sublaminar wires and Galveston pelvic fixation. (**d**) Segmental pedicle screw fixation with sacral alar iliac pelvic fixation

question remains as to the validity of this practice as it undoubtedly skews the interpretation of surgical results in favour of the diagnosis with the largest number of subjects within the analysis, most typically CP [10].

As with other musculoskeletal manifestations of the many diagnoses that fall under the 'neuromuscular' moniker, the prevalence of scoliosis in these populations is typically high and most often related to disease severity [16–18]. Like

incidence, surgical success seems to be related to neuromuscular disease severity. In CP, for example, the incidence of hip displacement has been reported to be significantly correlated to functional level (via the Gross Motor Function Classification System (GMFCS)) as have the success rates for hip adductor surgery [19, 20] Similar to the argument against merging results of different diagnoses, analyzing surgical outcomes without stratifying the analysis by functional level may also skew outcomes in favour of the higher functioning diagnoses and subjects [7].

Given the discussion above, coupled with the reality that most of the published literature regarding the treatment of neuromuscular scoliosis lacks a comparator group and are primarily retrospective in nature, one must be cautious when applying the conclusions of the available literature in clinical practice.

Search Strategy and Grade Recommendations

For the purposes of this evidence-based chapter, studies for consideration were identified via thorough searches of the PubMed, Cochrane and Web of Science databases, using a combination of keyword and controlled vocabulary searches. Search terms used included but were not limited to: scoliosis, neuromuscular scoliosis, neurogenic scoliosis, cerebral palsy, spina bifida, myelomeningocele and Duchenne muscular dystrophy. When assessing retrieved studies for inclusion, the Journal of Bone and Joint Surgery Level-of-Evidence ratings were utilized [21]. The search focused on systematic reviews, randomized control trials and studies that contained a comparator group (levels of evidence I, II and III). Lower level studies (level IV) were considered only when there were no higher-level studies available. Searches were limited to include only English-language studies. A sample PubMed search strategy is outlined in Table 24.1.

For each question asked we have provided an overview of the evidence and applied grades of recommendation according to Wright et al. [22]:

GRADE A – good evidence based on level I studies with consistent findings.
GRADE B – fair evidence based on consistent level II or Level III studies.
GRADE C – poor or conflicting evidence based on level IV/V evidence.
GRADE I – insufficient evidence to make a treatment recommendation.

What Is the Evidence for Scoliosis Correction?

Scoliosis Correction: Risks Versus Benefits

In the preceding section, it was established that the true benefits of scoliosis correction should be measured in terms of outcomes of interest to the patient and/or caregiver. In a general sense, taking neuromuscular diagnoses as a whole, there are several studies that met the inclusion criteria for this evidence-

Table 24.1 Sample PubMed search strategy

Search	Query	Items found
#20	Search ((((“Infant”[Mesh] OR “Child”[Mesh] OR “Adolescent”[Mesh] OR “Pediatrics”[Mesh] OR “Minors”[Mesh])) OR (infant[Title/Abstract] OR child[Title/Abstract] OR children[Title/Abstract] OR toddler *[Title/Abstract] OR kindergarten *[Title/Abstract] OR adolescent[Title/Abstract] OR adolescence[TIAB] OR minor[Title/Abstract] OR minors[Title/Abstract] OR boy[Title/Abstract] OR boys[Title/Abstract] OR girl[Title/Abstract] OR girls[Title/Abstract] OR pediatr *[Title/Abstract] OR juvenile[Title/Abstract] OR youth[Title/Abstract]))) AND ((((((“Scoliosis”[Mesh]) OR “Scoliosis/surgery”[Mesh])) OR (scoliosis OR “neuromuscular scoliosis” OR “neurogenic scoliosis”))) AND (((((“Cerebral Palsy/surgery”[Mesh]) OR “Muscular Dystrophy, Duchenne/surgery”[Mesh]) OR “Meningomyelocele/surgery”[Mesh])) OR (((“cerebral palsy” OR “spina bifida” OR myelomeningocele OR “Duchenne Muscular Dystrophy”)) AND (surgery OR surgical)))) Filters: English	507
#19	Search ((((“Infant”[Mesh] OR “Child”[Mesh] OR “Adolescent”[Mesh] OR “Pediatrics”[Mesh] OR “Minors”[Mesh])) OR (infant[Title/Abstract] OR child[Title/Abstract] OR children[Title/Abstract] OR toddler *[Title/Abstract] OR kindergarten *[Title/Abstract] OR adolescent[Title/Abstract] OR adolescence[TIAB] OR minor[Title/Abstract] OR minors[Title/Abstract] OR boy[Title/Abstract] OR boys[Title/Abstract] OR girl[Title/Abstract] OR girls[Title/Abstract] OR pediatr *[Title/Abstract] OR juvenile[Title/Abstract] OR youth[Title/Abstract]))) AND ((((((“Scoliosis”[Mesh]) OR “Scoliosis/surgery”[Mesh])) OR (scoliosis OR “neuromuscular scoliosis” OR “neurogenic scoliosis”))) AND (((((“Cerebral Palsy/surgery”[Mesh]) OR “Muscular Dystrophy, Duchenne/surgery”[Mesh]) OR “Meningomyelocele/surgery”[Mesh])) OR (((“cerebral palsy” OR “spina bifida” OR myelomeningocele OR “Duchenne Muscular Dystrophy”)) AND (surgery OR surgical))))	569
#15	Search (((((“Scoliosis”[Mesh]) OR “Scoliosis/surgery”[Mesh])) OR (scoliosis OR “neuromuscular scoliosis” OR “neurogenic scoliosis”))) AND (((((“Cerebral Palsy/surgery”[Mesh]) OR “Muscular Dystrophy, Duchenne/surgery”[Mesh]) OR “Meningomyelocele/surgery”[Mesh])) OR (((“cerebral palsy” OR “spina bifida” OR myelomeningocele OR “Duchenne Muscular Dystrophy”)) AND (surgery OR surgical)))	649

based review and used a QOL measure to assess patient-centered benefits.

In a prospective study utilizing the Swedish spine registry, Ersberg and colleagues used the EQ-5D (a validated instrument with sections that evaluate self-care, mobility, usual activities, pain, and anxiety) and the SRS-22 questionnaire to compare QOL between idiopathic, neuromuscular, and congenital scoliosis groups [15]. From a surgical risk perspective, patients with neuromuscular scoliosis experienced statistically significant increases in intra-operative blood loss, duration of surgery, and length of hospital stay as compared to the idiopathic group. With respect to QOL measures, there were significant increases in the post-operative EQ-5D total score and in particular the reduction of post-operative pain. When assessed via the SRS-22 instrument, neuromuscular patients experienced significantly improved function and better self-image. Interestingly, despite the significant number in complications in the neuromuscular group, when stratified into groups with and without complications, there were no significant differences in QOL scores. The conclusion of this study was that QOL was improved after scoliosis surgery even despite the high risk of complications.

In another prospective Swedish study, the impact of scoliosis surgery in neuromuscular patients was assessed in terms of outcomes pertaining to QOL, using a questionnaire that assessed sitting, care-giving, reaching, pain, rest time, seating supports, and activities of daily living (ADL), Cobb angle correction, and respiratory function as measured by vital capacity (VC) at a mean follow-up of 7 years [7]. This landmark study stratified patients who underwent scoliosis surgery into 4 subgroups including those that: (1) understood verbal instructions, (2) did not understand verbal instructions, (3) had progressive disease (e.g. DMD), and (4) had non-progressive disease (e.g. CP). Overall, surgical patients had significant QOL improvements in sitting balance, weight distribution, ADL, time used for resting, number of seating supports in addition to improvements in Cobb angle (~50 %) and respiratory function. When analyzed by subgroups, both the non-progressive and verbal instructions subgroups maintained improvements in outcomes as per the study results overall, but those that did not understand verbal instructions showed no improvement in ADL, ease of care giving, or respiratory function. Even more telling, scoliosis surgery had no impact on QOL-related outcomes or respiratory function in the progressive disease subgroup in which the only significant improvement was in Cobb angle correction. The sample size in this subgroup however, was quite small (only 14 patients) so this should be interpreted with caution.

In a general sense, the results of these two prospective studies support the surgical treatment of scoliosis in neuromuscular diagnoses, citing significant improvements in QOL-related outcomes, radiographic measures and, in some cases, respiratory function. That said, an objective assessment of the literature with respect to the risks involved with such procedures is required to balance the risk-benefit equation.

In a large retrospective study utilizing the Kids Inpatient Database (US based database of nationwide hospital discharges), 437 children with progressive neurodegenerative disorders (e.g. SMA, myopathies, etc.) who underwent scoliosis surgery were identified and their results were compared to non-progressive patients. In general, the progressive group had significant increases in length of stay (10.3 versus 7.7 days), pulmonary complications, in-hospital mortality (1.6 % versus 0.6 %), and hospital costs [14].

These results were similar to a study from the Scoliosis Research Society Morbidity and Mortality Committee, which analyzed complication rates from a database of 19,360 patients who underwent pediatric scoliosis surgery; 4657 of which had a neuromuscular diagnosis (including non-progressive and progressive diagnoses) [10]. Within this large sample, as compared to idiopathic scoliosis (IS), neuromuscular patients again had the highest risk of complications at 18 % versus 6 %. As with the previous study, mortality rates were significantly higher in the neuromuscular group (0.3 % versus 0.02 % for IS) with respiratory complications being the leading cause of death. Non-fatal complications such as blood loss (NM: 1.2 % versus IS: 0.2 %) and deep wound infection (NM: 3.8 % versus IS: 0.8 %) were also significantly higher in the neuromuscular group.

Continuing the trend, a systematic review analyzing the rate of complications in scoliosis surgery substantiated these results with significantly increased risks of death, infection, and pseudoarthrosis associated with a neuromuscular diagnosis when compared to IS [9].

The results of these studies confirm that the risks associated with scoliosis correction in neuromuscular diagnoses are substantial but, at the same time, the best evidence available also suggests that QOL is improved for these patients despite the risks. These points must be carefully considered and effectively communicated by the treating surgeon so the patient and/or caregiver can make an informed decision as to whether or not they proceed with surgery. Furthermore, any discussion regarding disease-specific risks and benefits needs to take into consideration the differences in outcomes that are beginning to emerge in the recent literature when analyzed by diagnosis.

Cerebral Palsy

Though typically associated with high patient/care-giver satisfaction, scoliosis surgery in children with CP is fraught with high complication rates likely related to the increased prevalence of co-morbidities inherent to this patient population including: poor nutritional status, epilepsy, infections of the urinary and respiratory tract, feeding disorders, and relative immunodeficiency [23]. Despite these risks, children with CP

seem to be highly tolerant of spinal surgery with a relatively long predicted life expectancy post-operatively [24]. The evidence regarding the risks and benefits of scoliosis correction in CP, as in other diagnoses, is scant but a few studies did meet our inclusion criteria.

In a recent systematic review with an aim to determine the risks, benefits of scoliosis correction in CP, in addition to the pre-operative factors affecting surgical outcome, only 1 prospective and 3 retrospective cohort studies were identified, with the rest being retrospective case series. Unfortunately, none of these studies analyzed included an observational group and the conclusions of the review were that the "overall strength of the evidence was insufficient" to make any firm recommendations for or against surgical intervention [25]. The authors also revealed that outcomes in these studies were "poorly delineated with limited or no use of validated outcome instruments". They suggested that future studies needed to employ validated outcomes relating to patient satisfaction and function.

Recently, the development of the Caregiver Priorities & Child Health Index of Life with Disabilities (CP CHILD) questionnaire by Narayanan and colleagues has provided a validated disease-specific outcome measure to apply to patients with CP [13]. In a prospective longitudinal cohort multi-center study investigating the utility of the CP CHILD questionnaire for children with severe CP who underwent scoliosis surgery, the authors found that by 12 months post-operatively, significant improvements in positioning/transfers, health, and overall QOL were achieved. The instrument was found to be sensitive to change and suggested as a meaningful outcome measure for evaluating this patient population [26].

These results were corroborated by a recent retrospective case-control study that also used the CP CHILD as its primary outcome measure [27]. In this study, children with severe CP (GMFCS IV and V) and scoliosis greater than 40 degrees were analyzed to determine the impact of scoliosis correction. The operative group demonstrated significant improvements in overall CP CHILD scores, personal care/activities of daily living, positioning/transferring/mobility, comfort/emotions, and communication/social interactions while the observational group deteriorated. In the surgical group, the complications included wound infections (22 %), pneumonia (17 %), reoperations due to post-surgical collections (12 %), pneumothorax (6 %), and recurrent hip dislocation (6 %).

Two other retrospective case-control studies also concurred with the findings of the previous studies, reporting a high level of patient and/or caregiver satisfaction but with a high rate of associated complications [28, 29].

In conclusion, given the best evidence available, it would seem that scoliosis correction improves quality of life in CP albeit with a high rate of complications.

Spina Bifida

Spina bifida represents a spectrum of disease severity and patho-anatomical variations (including deficiencies of the bony posterior elements in conjunction with congenital abnormalities of the spinal cord, brain stem, and peripheral nerves) that make it a difficult entity to assess from a disease-specific standpoint. To this point, many previous studies that refer to 'spina bifida' as the diagnosis in question typically focus on patients with myelomeningocele rather than typically higher functioning forms of spinal dysraphism including meningocele, lipomeningocele and others. As in other neuromuscular disorders, this distinction is relevant since the incidence of scoliosis and risks associated with curve correction vary according to these differing manifestations of the 'spina bifida' diagnosis. For example, disease severity in myelomeningocele is commonly related to neurosegmental level, with the risk of scoliosis progression being more prevalent at higher motor levels (e.g. T12 and above (high risk) versus L5 and below (low risk) [18, 30]. Furthermore, QOL and functional scores have also been linked to neurosegmental level, a potential confounder when assessing the impact of interventions without taking disease severity into consideration [31].

Scoliosis is one of the most common musculoskeletal manifestations in spina bifida with up to 50 % prevalence being reported [3]. As curve magnitude increases, a progressive loss in truncal balance and sitting stability can occur which may impact QOL [32]. The indications for scoliosis correction in spina bifida have mirrored that of other neuromuscular diagnoses including a progressive curve greater than 50° and functional concerns such as sitting balance and wheelchair tolerance. However, the operative risks associated with spinal fusion have been reported to be among the highest for scoliosis of any diagnosis which necessitates a comprehensive assessment of the benefits and risks of surgery, and their effective communication to patient and/or caregiver, before embarking on such a procedure.

Until recently, there were no validated outcome measures to evaluate the impact of scoliosis correction of QOL and patient satisfaction in spina bifida despite complication rates approaching 75 % [33]. To rectify this, Wai and colleagues developed the Spina Bifida Spine Questionnaire (SBSQ), a validated tool that evaluated self-perception and overall physical function for these children with associated spinal deformity [34]. Interestingly, in this cross-sectional study, the authors found no relationship between spinal deformity and self-perception or physical function.

In a follow-up retrospective case-control study involving the same institution, the SBSQ and 36-Item Short Form Health Survey (SF-36) were used to assess the impact of scoliosis correction on QOL in patients with spina bifida at a mean 14-year follow-up [35]. Like the previous study, they

found no difference in SBSQ and SF-36 scores between the operative and non-operative groups and no relation to curve magnitude. In addition, the study showed that, of the patients that could walk pre-operatively, only 50 % remained ambulatory after scoliosis correction. The authors suggest that this finding, coupled with an increased spinal stiffness after fusion and a lack of improvement in sitting balance between the groups, may have contributed to the lack of impact on QOL.

A prospective study from Poland also investigated the relationship between QOL/functional scores and the presence of spinal deformity in spina bifida in a non-operative cohort [32]. They used the Quality of Life in Spina Bifida Questionnaire (QLSBQ) to assess QOL and the SBSQ to assess physical function in addition to a self-perception outcome measure. They found that very large curve magnitudes did have a negative affect on QOL but no impact on physical function or self-perception was identified. Since there was no surgical arm in the study, no inference can be made as to whether surgery would improve these QOL limitations.

A recent evidence-based review on the subject identified 9 level III studies involving the spine but only two that evaluated physical function within both operative and non-operative groups [36]. The authors concluded that surgery had little effect on physical function and cautioned that the risks may outweigh the benefits when considering scoliosis correction for children with spina bifida. If surgery was to be done, a combined anterior and posterior fusion was reported to have a decreased complication rate over other approaches.

As previously mentioned, complication rates associated with scoliosis correction in spina bifida are very high and are an important consideration. In one retrospective comparative study investigating complication rates associated with different surgical approaches, an overall complication rate of 48–60 % was found with infection (19 %), shunt insufficiency (12 %), pseudoarthrosis (22 %), and hardware-related problems (30 %) being the most notable [33]. They also found that the addition of anterior instrumentation and fusion to a posterior fusion provided the lowest rate of hardware-related complications and loss of correction. Furthermore, one death from shunt insufficiency was identified and as such the authors stressed that shunts should be evaluated pre-operatively to help decrease the rates of peri-operative malfunction.

In summary, unlike CP, current evidence suggests that the risk-benefit ratio for scoliosis correction in spina bifida is not favourable given the lack of significant improvements in QOL or physical function coupled with very high complication rates. As such, unless a substantial functional problem (e.g. severe pain from costo-pelvic impingement; functional sitting imbalance) is identified which is unresponsive to conservative management (e.g. wheelchair modifications), scoliosis surgery should not be recommended for children with spina bifida.

Duchenne Muscular Dystrophy

Duchenne Muscular Dystrophy (DMD) is an inherited X-linked myopathic disorder secondary to mutations in the gene coding for dystrophin, a muscle cell membrane stabilizer. The absence, or reduced action, of dystrophin renders muscle cells susceptible to damage that is thought to result in an inflammatory response with eventual replacement of viable muscle with fibro fatty scar tissue [37]. As a result of this process, unlike CP and other non-progressive disorders, the natural history of DMD is typified by progressive deleterious changes in muscle strength, static contractures, and the development of scoliosis. Concomitant to these changes, progressive decreases in respiratory and cardiac function, along with a significantly reduced life span, increase the risks associated with the correction of scoliosis in this population.

Key questions regarding scoliosis correction in DMD center around whether it: (1) improves long-term survival, (2) improves respiratory function, (3) improves QOL and overall physical function, (4) has a positive benefit-to-risk ratio, and (5) is still required given the potential for scoliosis prevention or delay in onset by the use of newer corticosteroids with fewer side effects.

In an attempt to answer some for these questions, a recent Cochrane review was published which investigated the role of scoliosis correction in DMD [38]. Expectedly, no randomized controlled studies were identified and the authors' conclusions were that the available literature was insufficient to make any direct recommendations regarding the application of scoliosis correction in DMD. However, based on the available literature, some of which was at least level III, a few general statements were offered.

Most of the studies reviewed were in agreement that spinal surgery improved sitting position, patient satisfaction, and overall QOL, in addition to improvements in Cobb angle and pelvic obliquity for children with DMD. However, most failed to show significant improvements in respiratory function or long-term survival despite the many studies investigating this over the past 35 years [39–42]. In fact, one retrospective case-control study cited found no significant differences in respiratory function deterioration between operative and non-operative groups. Furthermore, the addition of surgery did not improve respiratory function despite significant Cobb angle correction (61.7 %) [43].

The published complication rates were significant as in other neuromuscular disorders. In a recent level III study comparing complication rates for scoliosis surgery in DMD and CP, it was suggested that DMD involves even higher perioperative risks with significantly higher complication rates (DMD: 38 % vs CP: 18 %) [44]. A systematic review from Mercado and colleagues supported these statements suggesting that cosmesis, QOL, and overall patient satisfaction are generally improved after scoliosis correction in

Table 24.2 GRADE recommendations for scoliosis correction

Evidence-based statement	GRADE recommendation
The risks of scoliosis correction for neuromuscular diagnoses are significantly more than for idiopathic diagnoses	B
Scoliosis correction in neuromuscular diagnoses, on the whole, improves QOL, radiographic measures and, in some cases, respiratory function	B
Scoliosis correction improves quality of life in CP	B
Scoliosis correction does not improve quality of life or physical function in spina bifida and has a very high risk profile	B
Scoliosis correction improves quality of life and patient satisfaction in DMD albeit with a high complication rate	B
Scoliosis correction does not improve or preserve respiratory function in DMD	B
The use of deflazacort significantly reduces the need for scoliosis correction in DMD	B

DMD despite the fact that most patients undergo surgery at a relatively early stage (e.g. 25° Cobb angle) [3]. Given the lack of controlled studies however, the authors stated that the relationship between spinal surgery and improvement in respiratory function is still unclear. They also cautioned that a consistent negative aspect was the adverse impact on self-feeding due to the combination of a stiff, straight spine and upper limb weakness post-operatively.

Recently, it has been suggested that perhaps scoliosis surgery in DMD might be completely avoided. In the absence of effective medical management, it has been reported that over 90 % of patients with DMD will develop scoliosis, most of which will require spinal stabilization [45, 46]. Previous studies have shown that corticosteroid administration can slow the decline in muscle strength, prolong walking in children, and delay the onset of scoliosis with DMD but often at the expense of unacceptable side effects [47]. However, several recent studies have demonstrated that deflazacort (an oxazolone derivative of prednisone) can achieve similar improvements in function with substantially better side effect profile.

In a prospective cohort study investigating the role of deflazacort in the development of scoliosis, the Kaplan-Meier survival rate of not developing scoliosis or needing spinal surgery was 78 % in the treatment group and 8.3 % in the non-treatment group at a mean follow-up of 15 years [48]. Scoliosis measuring 20° or more developed in 20 % of patients for the treatment group and in 92 % for the non-treatment group, all of which required surgery. Of further benefit, patients in the treatment group had significantly improved pulmonary function, prolongation of walking, and ability to climb stairs. Side effects were common with 70 % (21 patients) in the treatment group developing cataracts but only two patients required cataract surgery. Other side effects included decreased height (17 cm shorter), weight gain (55 kg vs 51 kg), and no difference in bone fractures (treatment group was given bisphosphonates). The authors concluded that glucocorticoids have a long-term protective effect against the development of scoliosis in DMD.

Two additional retrospective cohort studies investigating the role of deflazacort in DMD supported the findings above [46, 49]. In each of these studies, the treatment group showed a similar decrease in the need for spinal surgery with an acceptable side effect profile. In one of these cohorts with a mean 8-year follow-up, surgery was completely avoided in the treatment group while 43 % of the non-treatment group underwent surgery [46]. In addition to prolonged walking, this study also reported significant improvements in cardiac function.

Given the studies above, it would seem that the use of deflazacort prevents, or at least delays, the onset of scoliosis in DMD with secondary benefits of preserving respiratory and cardiac function with an acceptable side effect profile. When surgery is required, it can be expected to result in improvements in QOL and patient satisfaction albeit with high perioperative risks. At this time, the available evidence regarding scoliosis correction in DMD does not seem to support improvements in survival and/or respiratory function.

Evidence-based statements and GRADE recommendations for this section are provided in Table 24.2.

What Is the Best Choice of Spinal Fixation?

In previous sections, it was established that the goals of surgical management for neuromuscular scoliosis typically involved the achievement of a balanced spine over a level pelvis via a spinal fusion extending from the upper thoracic spine to the lower lumbar spine and/or the pelvis. Due to the high rate of vertebral osteopenia in neuromuscular patients, segmental fixation is typically the rule to help prevent implant-related complications. The choice of spinal fixation used to achieve these goals has evolved over the years from Harrington instrumentation to Luque sublaminar wires and, more recently, to segmental pedicle screw fixation [1, 2, 50].

Proponents of pedicle screw fixation cite improvements in Cobb correction, a decreased need for anterior release, and a favourable risk profile over earlier methods despite the associated increase in implant costs [51]. Proponents of sublaminar fixation cite the ability to achieve the desired goals, albeit with a smaller Cobb correction (~50 %), decreased costs, and comparable complication rates [52]. Although most of the literature in this area is comprised of uncontrolled case series, several level III studies were identified and available for review.

Hybrid Versus Pedicle Screw Fixation

Regarding the assessment of spinal fixation methods in neuromuscular scoliosis, our literature search revealed no prospective studies and a small number of retrospective comparative studies. One such study compared hybrid fixation (sublaminar wires/hooks in the thoracic spine, pedicle screws in the lumbosacral spine) to segmental pedicle screw fixation and found significant improvements in curve correction (75 % vs 59 %), operating time (6.0 vs 7.4 h), blood loss (1785 vs 3760 mL), and the need for anterior surgery (12 % vs 40 %) for the pedicle screw group as compared to the hybrid group [53]. As might be expected, the authors reported no change in QOL as measured by the SRS-24 questionnaire and no significant difference in overall complication rate between the two groups. They stressed that, although Cobb correction was greater in the pedicle screw group, the attainment of spinal balance was most important and was achieved by both groups.

In another retrospective comparative study with 44 of 68 patients having a neuromuscular diagnosis, four different types of apical spinal fixation were compared for large curves greater than 100°: sublaminar wires, hooks, anterior vertebral screws, and all pedicle screw constructs [5]. Like the previous study, all pedicle screw constructs demonstrated significant improvements in Cobb correction, a decreased rate of anterior release, and the lowest complication rate between the groups. The all-pedicle screw group was also better at maintaining curve correction by final follow-up as compared to other methods, which tended to be associated with a significant loss of curve correction over time. That said, there were no significant differences in the ability to achieve coronal/sagittal balance or in the neurologic risk profile between the wire and screw groups.

A third retrospective cohort comparing 'rigid' constructs (greater than 50 % pedicle screw fixation) against 'non-rigid' constructs (greater than 50 % sublaminar fixation) in neuromuscular scoliosis correction supported the claim that pedicle screw constructs achieved greater Cobb angle correction and had a decreased need for anterior release as compared to sublaminar wires [54]. The pseudoarthrosis rate was also significantly less in the rigid group (5 % vs 22 %). The authors concluded that, despite a fivefold increase in implant costs, the overall charges associated with the higher rates of pseudoarthrosis in the wire group would more than offset any implant-related differences in cost. They suggested that there was a need for a future study that incorporates a more comprehensive economic analysis to more fully elucidate the true value of using pedicle screws over cheaper implants such as sublaminar wires.

Lending further support to the evidence above, in a retrospective study investigating the use of sublaminar (SL) wires, hybrid (H) constructs, and segmental pedicle screw (PS) fixation in patients with DMD, significant increases in operative time, blood loss, and Cobb correction were again identified in the SL group [55]. The increased Cobb correction was likely related to the increased pre-operative Cobb angle in the wire group as compared to the other groups (50° (SL) vs 18° (H), 26° (PS)) rather than improved mechanical capabilities. The authors suggested that the increased blood loss in the SL group was likely due decreased vasoconstriction in DMD and epidural vessel injury during wire passage. In this study, based on radiographic and intra-operative outcomes, it would seem that pedicle screw constructs performed most favourably as compared to other implants.

Newer Implants

Regarding the Universal Clamp (Zimmer, Warsaw, USA), a relatively new titanium implant which incorporates a sublaminar polyester tape, only one comparative study was identified which compared its use to the standard sublaminar wire in scoliosis correction [6]. This cohort of 50 patients (25 in each group) showed no differences in Cobb correction and no change in operative time or blood loss between the two groups. The costs, though not stipulated in the study, are known to be quite high for the Universal Clamp as compared to the stainless steel sublaminar wire, leaving the sole reported benefit to be related to the MRI compatibility of the titanium implant. Additional studies comparing the Universal Clamp to other types of spinal fixation will be required to ascertain its place in the surgeon's armentarium.

Given the above discussion, it would seem that segmental pedicle screw fixation provides the best Cobb correction, decreased operative time, and a decreased need for anterior release, as compared to hooks or sublaminar wires. Prospective studies that incorporate comprehensive economic analyses would be beneficial to assess the outcomes-to-cost ratio as it pertains to the most appropriate choice of spinal fixation for these children.

Evidence-based statements and GRADE recommendations for this section are provided in Table 24.3.

Table 24.3 GRADE recommendations for choice of spinal fixation

Evidence-based statement	GRADE recommendation
The use of segmental pedicle screw fixation in neuromuscular scoliosis results in the largest Cobb angle correction as compared to other methods	B
The use of segmental pedicle screw fixation in neuromuscular scoliosis results in a reduced rate of anterior release as compared to other methods	B
The use of segmental pedicle screw fixation in neuromuscular scoliosis results in a reduced operative time over other methods	B
The use of segmental pedicle screw fixation in neuromuscular scoliosis results in a reduced rate of complications over other methods	I
Quality of life improvements are unrelated to the choice of spinal fixation in neuromuscular scoliosis	I

What Is the Best Choice of Pelvic Fixation?

In neuromuscular scoliosis, reducing pelvic obliquity is one of the major technical goals of spinal correction. Coupled with an unbalanced spine, its presence can adversely affect sitting balance, wheelchair tolerance, weight distribution, and QOL [56]. Pelvic obliquity correction is typically achieved by instrumentation and fusion to the sacropelvis through various implant types and configurations. There has been some controversy as to the indications for fusion to the pelvis but, when required, achieving stable fixation to the sacropelvic unit that resists post-operative loss of correction is essential to surgical success [56–59]. Although a myriad of implant configurations for the treatment of pelvic obliquity have been described, this section will focus on three of the most prevalent types of pelvic fixation: (1) Galveston, (2) iliac screws, and (3) sacral alar iliac (SAI) screws (Fig. 24.2).

Galveston Versus Iliac Fixation

Galveston bilateral iliac fixation is one of the best-known techniques for the treatment of pelvic obliquity in neuromuscular scoliosis although its use has been waning in favour of more modern techniques utilizing screw fixation. Typically, it has been utilized in conjunction with Luque segmental sublaminar wire fixation in the thoracic and lumbar spine and has evolved as a system incorporated into a single, pre-contoured 'unit rod' [60, 61]. The technique has been well described elsewhere but in essence, involves the insertion of stainless steel smooth tines into tracts made between the tables of the ilia bilaterally.

Iliac screw fixation takes a similar approach but substitutes long screws for the tines described for the Galveston technique. It has been suggested that the use of screws over smooth tines may diminish implant pullout and loss of pelvic obliquity correction [62]. A frequent need for lateral offset connectors, however, has been reported to increase pullout forces rather than decrease them and serves as an additional potential point of failure [63]. Clearly, comparative studies are needed to ascertain whether or not these 'more advanced' systems actually improve outcomes. Despite an exhaustive literature search, very few studies with comparator groups and/or adequate sample size were identified [64].

In a retrospective cohort of forty patients with neuromuscular diagnoses, radiographic outcomes and complication rates between Galveston and iliac screw pelvic fixation were compared to assess superiority of one technique over the other [62]. Each patient underwent the standard posterior instrumentation and fusion from the upper thoracic spine to the pelvis with no differences in curve correction or apical vertebral translation between the groups. In addition, an equal number of patients in each group underwent intra-operative halo femoral traction. The study showed no difference in initial pelvic obliquity correction between the two pelvic fixation groups. At a final follow-up of 3 years however, the percentage of patients with residual pelvic obliquity greater than 10° was significantly increased in the Galveston group; a fact the authors claimed was not clinically significant given that both groups achieved "an excellent overall correction". In addition, pelvic anchor motion ('wiper blading') was significantly increased in the Galveston group but with uncertain clinical significance. The complication rates were similar overall. The authors concluded that iliac screws are equivalent to the Galveston technique but the former allows for sacral and lumbar screw fixation that may offer better construct stability with the downside of having an increased implant profile.

Despite these claims, the Galveston technique may still prove to be more efficacious over more modern implants. In a multicenter retrospective study of 157 children with CP comparing the pre-contoured unit rod (including Galveston pelvic fixation) to custom-bent rods utilizing various methods of pelvic anchorage (including iliac screws or rods, S-rods, and sacral screws), pelvic obliquity was found to be significantly improved in the unit rod group (74 % vs 22 %) with no significant differences in operative time or intraoperative blood loss [65]. The authors offered that the likely reason for the improvement could be explained by the fixed 90° angle between the unit rod's pelvic limbs and spinal rods. Transfusion requirements, implant prominence, as well as inpatient and intensive care unit length of stays, were significantly increased in the unit rod group. Accordingly, results from the non-validated caregiver-reported outcome measure used in the study suggested a higher satisfaction rate in the custom-bent rod group.

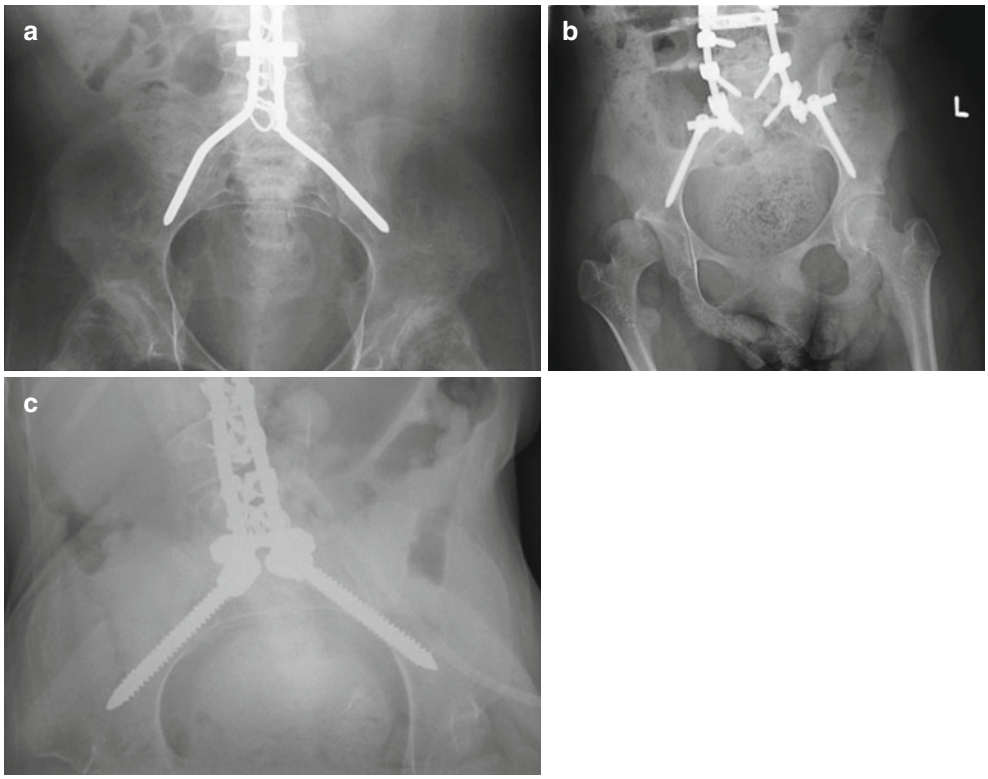

Fig. 24.2 Types of pelvic fixation. (**a**) Galveston, (**b**) Iliac screws with lateral offset connectors, and (**c**) Sacral alar iliac (SAI) screws

One potential advantage of the use of iliac screw fixation is that it allows for augmentation with additional implants to theoretically improve construct stability. In a retrospective study with 5-year follow-up, two pelvic fixation constructs utilizing 1 or 2 iliac screws were compared with respect to pelvic obliquity correction and associated complications [66]. The single screw group experienced 2.5 times greater rod dislodgement than the double screw group but this did not reach statistical significance. However, a sevenfold increase in proximal (i.e. thoracic and lumbar spine) implant failure was seen in the single screw group. The authors surmised that the extra screw provided a more secure base that dampened proximal motion and thus reduced this risk. The overall pelvic obliquity correction was reported to be 59 % (from 18.8° to 7.6°) with no mention of correction stratified by treatment group. Given the decreased complication rate, the authors recommended the adoption of the double screw technique over the single screw for neuromuscular pelvic fixation.

Sacral Alar Iliac Fixation

Recently, a new pelvic screw fixation technique was described that utilized a tract traversing the sacral ala, sacroiliac joint, and ilium [67]. The proposed advantages of this sacral alar iliac (SAI) screw fixation over iliac screws were reported to be due to its low profile and decreased need for both lateral offset connectors and the associated subparaspinal muscle dissection required. In a retrospective comparative study with 2-year follow-up, SAI screws demonstrated significant improvements in pelvic obliquity correction over an iliac screw group (70 % vs 50 %, respectively) with no difference in implant-related complications or pain between the groups. The authors suggested that the improvement in pelvic obliquity correction might have stemmed from a better mechanical advantage in manipulating the pelvis directly rather than working through the lateral offset connectors used in the iliac screw group. Additional studies investigating patient-specific outcomes would be beneficial to further elucidate the clinical superiority of SAI screws in pelvic fixation.

Choice of Surgical Approach

The choice of surgical approach may have an impact on the success of pelvic fixation. In a retrospective comparative study analyzing 54 patients with muscular dystrophy, one group underwent a posterior-only approach while the second underwent a combined anterior and posterior approach for the correction of pelvic obliquity [68]. The anterior approach chosen involved an extraperitoneal midline lumbosacral release and fusion with mesh cage and allograft. The anterior-posterior group achieved a significant improvement in pelvic obliquity correction over the posterior-only group (21° vs 13°, respectively) but at the expense of significant increases in intraoperative blood loss (2.4 L vs 345 mL, respectively) and operative time (611 vs 440 min, respectively). Since the study only evaluated radiographic parameters, rather than

sitting tolerance or other patient-specific outcomes, the clinical significance of this modest improvement in pelvic obliquity is not known and may not outweigh the additional risks imparted by the anterior approach.

Fusion to Lumbar Spine Versus Pelvis

The indications for pelvic fixation in neuromuscular scoliosis correction have been controversial with some authors calling for all non-ambulatory patients to be instrumented and others only in the face of substantial pelvic obliquity [2, 69]. Reasons to exclude the pelvis have included: minimal preoperative pelvic obliquity, increased operative time, potential loss of walking ability, and higher complication rates [56, 70].

In support of these reasons, the extent of caudal fixation for patients with and without pelvic obliquity was investigated in a retrospective cohort of 55 patients with neuromuscular scoliosis [69]. The cohort was stratified into 3 groups according to the distal fixation level and the severity of pelvic obliquity including: (1) pelvic obliquity greater than 15° with pelvic fixation, (2) pelvic obliquity greater than 15° without pelvic fixation, (3) pelvic obliquity less than 15 degrees with pelvic fixation. Pelvic obliquity correction was found to be significant for all groups but Group 2 displayed a significant loss in correction at final follow-up. The authors concluded that the presence of pelvic obliquity greater than 15° necessitates the addition of pelvic fixation while lesser amounts remain stable without fixation.

Lending further support to the exclusion of pelvic fixation for patients with minimal pelvic obliquity, a retrospective study of 36 patients with DMD compared two groups according to the magnitude of tilt [70]. In this cohort, pelvic obliquity was found to be (1) more than 15° in 10 patients and (2) less than 15° in 26 patients. In Group 1, pelvic tilt was improved by 62 % using iliac screw fixation. Group 2 also experienced stable pelvic tilt correction by 42 % even without pelvic fixation at a final follow-up of 37 months. As for complications, 24 % in Group 2 had postoperative coccygodynia compared with only 10 % in Group 1. The authors concluded that instrumentation of the pelvis is unwarranted for pelvic obliquity less than 15°.

Given the above discussion with respect to pelvic fixation, the evidence seems to support the use of the Galveston technique, a posterior-only approach, and the exclusion of pelvic fixation for patients with pelvic tilt less than 15°. However, the available evidence is based on a limited number of studies with small sample sizes and, as such, more studies are required to make definitive conclusions regarding pelvic fixation in neuromuscular scoliosis.

Evidence-based statements and GRADE recommendations for this section are provided in Table 24.4.

Table 24.4 GRADE recommendations for choice of pelvic fixation

Evidence-based statement	GRADE recommendation
For pelvic obliquity correction, iliac screw fixation is not superior to Galveston fixation	B
For pelvic obliquity correction, sacral alar iliac fixation may be superior to iliac screw fixation	I
Pelvic fixation is not be required for pelvic obliquity less than 15 degrees	B

What Is the Evidence for Anterior Fusion?

The indications for anterior release for neuromuscular scoliosis have traditionally been described for curves with residual magnitude greater than 50–70° on bending or traction radiographs and for the prevention of crankshaft in immature patients [71]. The rationale behind the anterior approach is twofold: (1) to improve the capacity for correction in large, stiff curves by releasing the anterior longitudinal ligament and performing discectomies at multiple levels about the apex of the major curve (Fig. 24.3), (2) to facilitate the development of an anterior fusion to reduce the risk of pseudoarthrosis [24]. It was believed that the addition of an anterior fusion allowed for a better curve correction than posterior fusion alone. With the advent of 'heavy constructs' utilizing segmental pedicle screw fixation, in addition to the use of peri-operative adjuncts such as skull-femoral traction, the routine use of the anterior approach has been challenged. Though few in number, several studies that focused in this area and met our inclusion criteria were identified for review.

Anterior Surgery: Risk to Benefit Balance

Like any surgical intervention, a comprehensive assessment of the risk-benefit balance is paramount, particularly for procedures that violate the thoracic cage in a patient population that is already predisposed to pulmonary complications and have significant co-morbidities. Indeed, the risks of anterior surgery are substantial. In a large retrospective comparative study comparing risks of anterior surgery as a function of diagnosis, children with CP were more likely to have a major complication, most of which were pulmonary, as compared to spina bifida (52 % vs 41 %, respectively). In addition, curve size greater than 100° was found to be a significant risk factor for a major complication [72].

Another retrospective study assessing the risks pertaining to surgical approach for children with neuromuscular scoliosis, reported significantly longer intensive care unit (ICU) stays and increased pulmonary complications with combined

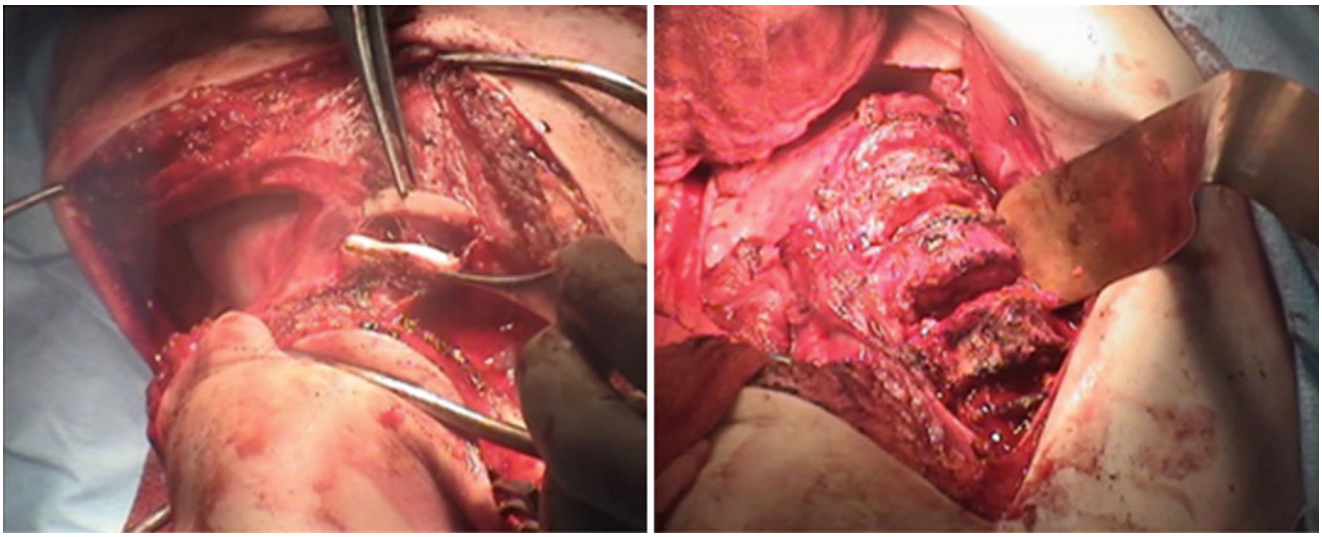

Fig. 24.3 Multilevel anterior discectomies via thoracoabdominal approach for severe scoliosis and pelvic obliquity in a 15 year-old with spastic quadriplegia

anterior-posterior fusions as compared to posterior instrumentation and fusion (PSF) alone [73].

Despite the findings presented above, there are instances where the benefits of an anterior fusion may very likely outweigh the potential risks. In a systematic review investigating the role of spine surgery in spina bifida, it was found that curve correction and fusion rates were significantly improved when an anterior fusion was added to the standard PSF [36]. Specifically, the risk of pseudoarthrosis was reported to be 46 % for PSF only compared to 14–23 % for combined anterior and posterior fusion. Wright assigned a B GRADE recommendation for the combined approach but stated there was insufficient evidence to recommend that the addition of anterior instrumentation improved outcomes further.

For children with neuromuscular scoliosis who, for whatever reason, require the addition of anterior fusion, the question of whether to do both procedures staged or same day is a topic of interest. In a retrospective comparative study involving children with spastic quadriplegic CP, significant increases in blood loss and operative time for single stage combined anterior-posterior fusion compared to performing the anterior and posterior procedures in two stages were demonstrated [24]. Based on this and other studies, it would seem that, although there are substantial risks associated with the combined approach, staging the procedures might mitigate the risk to some extent. Given the contradictory results of other comparative studies in this area, however, the evidence is likely insufficient to lend definitive guidance [74–76].

As outlined above, one of the most common reasons to perform an anterior release is to improve curve flexibility and accordingly, the potential for increased curve correction over PSF alone for large, stiff curves. A retrospective case-control study investigated the legitimacy of this practice by comparing matched groups of children with CP and scoliosis that underwent PSF with or without an anterior release [77]. The PSF-only group was augmented with intraoperative skull-femoral traction. There were no significant differences in curve correction, coronal/sagittal balance, or pelvic obliquity correction, between the groups. There were however, significantly increased complication rates demonstrated in the anterior release group including increased operative times, blood loss, postoperative intubation, and pneumonias. There were no traction-related complications reported. The authors concluded that the addition of an anterior release did not improve radiographic outcomes as compared to PSF (with skull-femoral traction) alone and demonstrated a significantly higher complication rate.

The Role of Intra-operative Skull-Femoral Traction

Although the preceding study suggested that curve correction was equivalent for both combined approaches and PSF alone, the addition of skull-femoral traction in the preceding study represented a significant confounding factor given that the technique has been shown to significantly improve major curve correction and pelvic obliquity when coupled to PSF versus PSF alone [78]. Therefore, one cannot conclude that scoliosis correction via a combined approach is equivalent to PSF alone without traction.

In recent years, intra-operative halo-femoral traction has become more commonplace, particularly for stiff curves, and

Table 24.5 GRADE recommendations for the role of anterior fusion

Evidence-based statement	GRADE recommendation
Overall, anterior release in addition to posterior fusion does not provide better outcomes over posterior fusion alone	I
The combined use of anterior and posterior fusion results in superior outcomes to posterior fusion alone in spina bifida	B
The risks associated with the combined use of anterior and posterior fusion are higher than for posterior fusion alone	I
Staging anterior and posterior fusion for combined approaches is associated with a decreased complication rate compared to single stage surgery	I
The use of skull-femoral traction as an adjunct to PSF may provide equivalent curve correction to the combined use of anterior and posterior fusion	C
The use of skull-femoral traction as an adjunct to PSF reduces the need for anterior release in neuromuscular scoliosis	I

is often combined with PSF following an anterior release. A recent systematic review on the subject suggested that the use of intraoperative skull-femoral traction may improve radiographic outcomes, decrease complication rates, and reduce the need for anterior procedures in neuromuscular scoliosis surgery [79]. The included studies in this review, however, were mixed in diagnosis and surgical approaches, and were marred by small sample sizes. As such, these recommendations should be interpreted with caution.

Overall, the availability of level III or better evidence studies concerning the role of anterior surgery in neuromuscular scoliosis is scarce and definitive conclusions in this area remain elusive. Given the available evidence, however, it is reasonably well established that the addition of anterior fusion to PSF in spina bifida leads to improved outcomes. Lesser evidence is available regarding the use of skull-femoral traction and its ability to obviate the need for anterior release in stiff curves.

Evidence-based statements and GRADE recommendations for this section are provided in Table 24.5.

Conclusions

Given the results of this evidence-based review of the best available literature, it would seem that for most cases, neuromuscular scoliosis correction is a worthwhile procedure with expected improvements in function, quality of life, and patient/caregiver satisfaction. These positive benefits likely outweigh the high surgical risks associated with these procedures but the ultimate decision as to whether or not scoliosis surgery is performed should be dependent on a disease-specific assessment of risks and benefits and appropriate communication with the patient and/or caregiver. The use of segmental pedicle screw fixation may offer improved outcomes over other methods of spinal fixation but the best choice of pelvic fixation is still controversial. Regarding the use of anterior surgery for neuromuscular scoliosis, its role is also controversial except in the case of spina bifida where it is likely to reduce perioperative risks.

References

1. Vialle R, Thevenin-Lemoine C, Mary P. Neuromuscular scoliosis. Orthop Traumatol Surg Res. 2013;99(1 Suppl):S124–39. doi:10.1016/j.otsr.2012.11.002.
2. McCarthy RE. Management of neuromuscular scoliosis. Orthop Clin North Am. 1999;30(3):435–49 .viii
3. Mercado E, Alman B, Wright JG. Does spinal fusion influence quality of life in neuromuscular scoliosis? Spine. 2007;32(19 Suppl):S120–5. doi:10.1097/BRS.0b013e318134eabe.
4. Modi HN, Suh SW, Yang JH, Cho JW, Hong JY, Singh SU, et al. Surgical complications in neuromuscular scoliosis operated with posterior – only approach using pedicle screw fixation. Scope. 2009;4:11. doi:10.1186/1748-7161-4-11.
5. Watanabe K, Lenke LG, Bridwell KH, Kim YJ, Watanabe K, Kim YW, et al. Comparison of radiographic outcomes for the treatment of scoliotic curves greater than 100 degrees: wires versus hooks versus screws. Spine. 2008;33(10):1084–92. doi:10.1097/BRS.0b013e31816f5f3a.
6. Caekebeke P, Moke L, Moens P. Sublaminar devices for the correction of scoliosis: metal wire versus polyester tape. Acta Orthop Belg. 2013;79(2):216–21.
7. Larsson EL, Aaro SI, Normelli HC, Oberg BE. Long-term follow-up of functioning after spinal surgery in patients with neuromuscular scoliosis. Spine. 2005;30(19):2145–52.
8. Sharma S, Wu C, Andersen T, Wang Y, Hansen ES, Bunger CE. Prevalence of complications in neuromuscular scoliosis surgery: a literature meta-analysis from the past 15 years. Eur Spine J. 2013;22(6):1230–49. doi:10.1007/s00586-012-2542-2.
9. Weiss HR, Goodall D. Rate of complications in scoliosis surgery – a systematic review of the Pub Med literature. Scope. 2008;3:9. doi:10.1186/1748-7161-3-9.
10. Reames DL, Smith JS, Fu KM, Polly Jr DW, Ames CP, Berven SH, et al. Complications in the surgical treatment of 19,360 cases of pediatric scoliosis: a review of the Scoliosis Research Society Morbidity and Mortality database. Spine. 2011;36(18):1484–91. doi:10.1097/BRS.0b013e3181f3a326.
11. Bohtz C, Meyer-Heim A, Min K. Changes in health-related quality of life after spinal fusion and scoliosis correction in patients with cerebral palsy. J Pediatr Orthop. 2011;31(6):668–73. doi:10.1097/BPO.0b013e318221093c.
12. Wilson PL, Newton PO, Wenger DR, Haher T, Merola A, Lenke L, et al. A multicenter study analyzing the relationship of a standardized radiographic scoring system of adolescent idiopathic scoliosis and the Scoliosis Research Society outcomes instrument. Spine (Phila Pa 1976). 2002;27(18):2036–40.
13. Narayanan UG, Fehlings D, Weir S, Knights S, Kiran S, Campbell K. Initial development and validation of the Caregiver Priorities and Child Health Index of Life with Disabilities (CPCHILD). Dev

Med Child Neurol. 2006;48(10):804–12. doi:10.1017/s0012162206001745.
14. Barsdorf AI, Sproule DM, Kaufmann P. Scoliosis surgery in children with neuromuscular disease: findings from the US National Inpatient Sample, 1997 to 2003. Arch Neurol. 2010;67(2):231–5. doi:10.1001/archneurol.2009.296.
15. Ersberg A, Gerdhem P. Pre – and postoperative quality of life in patients treated for scoliosis. Acta Orthop. 2013;84(6):537–43. doi :10.3109/17453674.2013.854667.
16. Persson-Bunke M, Hagglund G, Lauge-Pedersen H, Wagner P, Westbom L. Scoliosis in a total population of children with cerebral palsy. Spine. 2012;37(12):E708–13. doi:10.1097/BRS.0b013e318246a962.
17. Loeters MJ, Maathuis CG, Hadders-Algra M. Risk factors for emergence and progression of scoliosis in children with severe cerebral palsy: a systematic review. Dev Med Child Neurol. 2010;52(7):605–11. doi:10.1111/j.1469-8749.2010.03617.x.
18. Trivedi J, Thomson JD, Slakey JB, Banta JV, Jones PW. Clinical and radiographic predictors of scoliosis in patients with myelomeningocele. J Bone Joint Surg Am. 2002;84-a(8):1389–94.
19. Soo B, Howard JJ, Boyd RN, Reid SM, Lanigan A, Wolfe R, et al. Hip displacement in cerebral palsy. J Bone Joint Surg Am. 2006;88(1):121–9. doi:10.2106/jbjs.e.00071.
20. Shore BJ, Yu X, Desai S, Selber P, Wolfe R, Graham HK. Adductor surgery to prevent hip displacement in children with cerebral palsy: the predictive role of the Gross Motor Function Classification System. J Bone Joint Surg Am. 2012;94(4):326–34. doi:10.2106/jbjs.j.02003.
21. Wright JG, Swiontkowski MF, Heckman JD. Introducing levels of evidence to the journal. J Bone Joint Surg Am. 2003;85-a(1):1–3.
22. Wright JG, Einhorn TA, Heckman JD. Grades of recommendation. J Bone Joint Surg Am. 2005;87(9):1909–10. doi:10.2106/JBJS.8709.edit.
23. Comstock CP, Leach J, Wenger DR. Scoliosis in total-body-involvement cerebral palsy. Analysis of surgical treatment and patient and caregiver satisfaction. Spine (Phila Pa 1976). 1998;23(12):1412–24 .discussion 24-5
24. Tsirikos AI, Chang WN, Dabney KW, Miller F, Glutting J. Life expectancy in pediatric patients with cerebral palsy and neuromuscular scoliosis who underwent spinal fusion. Dev Med Child Neurol. 2003;45(10):677–82.
25. Legg J, Davies E, Raich AL, Dettori JR, Sherry N. Surgical correction of scoliosis in children with spastic quadriplegia: benefits, adverse effects, and patient selection. Evid Based Spine Care J. 2014;5(1):38–51. doi:10.1055/s-0034-1370898.
26. Narayanan UG, Sponseller P, Newton PO, Marks MC, editors. The CPCHILD questionnaire is sensitive to change following scoliosis surgery in children with cerebral palsy: PAPER# 62. Spine Journal Meeting Abstracts; LWW. 2011.
27. Sewell MD, Malagelada F, Wallace C, Gibson A, Noordeen H, Tucker S, et al. A preliminary study to assess whether spinal fusion for scoliosis improves carer-assessed quality of life for children with GMFCS level IV or V cerebral palsy. J Pediatr Orthop. 2015. doi:10.1097/bpo.0000000000000447.
28. Jones KB, Sponseller PD, Shindle MK, McCarthy ML. Longitudinal parental perceptions of spinal fusion for neuromuscular spine deformity in patients with totally involved cerebral palsy. J Pediatr Orthop. 2003;23(2):143–9.
29. Tsirikos AI, Chang WN, Dabney KW, Miller F. Comparison of parents' and caregivers' satisfaction after spinal fusion in children with cerebral palsy. J Pediatr Orthop. 2004;24(1):54–8.
30. Thomson JD, Segal LS. Orthopedic management of spina bifida. Dev Disabil Res Rev. 2010;16(1):96–103. doi:10.1002/ddrr.97.
31. Flanagan A, Gorzkowski M, Altiok H, Hassani S, Ahn KW. Activity level, functional health, and quality of life of children with myelomeningocele as perceived by parents. Clin Orthop Relat Res. 2011;469(5):1230–5. doi:10.1007/s11999-010-1651-7.
32. Sibinski M, Synder M, Higgs ZC, Kujawa J, Grzegorzewski A. Quality of life and functional disability in skeletally mature patients with myelomeningocele-related spinal deformity. J Pediatr Orthop B. 2013;22(2):106–9. doi:10.1097/BPB.0b013e32835c2a65.
33. Geiger F, Parsch D, Carstens C. Complications of scoliosis surgery in children with myelomeningocele. Eur Spine J. 1999;8(1):22–6.
34. Wai EK, Young NL, Feldman BM, Badley EM, Wright JG. The relationship between function, self-perception, and spinal deformity: implications for treatment of scoliosis in children with spina bifida. J Pediatr Orthop. 2005;25(1):64–9.
35. Khoshbin A, Vivas L, Law PW, Stephens D, Davis AM, Howard A, et al. The long-term outcome of patients treated operatively and non-operatively for scoliosis deformity secondary to spina bifida. Bone Joint J. 2014;96-b(9):1244–51. doi:10.1302/0301-620x.96b9.33857.
36. Wright JG. Hip and spine surgery is of questionable value in spina bifida: an evidence-based review. Clin Orthop Relat Res. 2011;469(5):1258–64. doi:10.1007/s11999-010-1595-y.
37. Sussman M. Duchenne muscular dystrophy. J Am Acad Orthop Surg. 2002;10(2):138–51.
38. Cheuk DK, Wong V, Wraige E, Baxter P, Cole A. Surgery for scoliosis in Duchenne muscular dystrophy. Cochrane Database Syst Rev. 2015;10:Cd005375. doi:10.1002/14651858.CD005375.pub4.
39. Eagle M, Bourke J, Bullock R, Gibson M, Mehta J, Giddings D, et al. Managing Duchenne muscular dystrophy–the additive effect of spinal surgery and home nocturnal ventilation in improving survival. Neuromuscul Disord. 2007;17(6):470–5. doi:10.1016/j.nmd.2007.03.002.
40. Galasko CS, Williamson JB, Delaney CM. Lung function in Duchenne muscular dystrophy. Eur Spine J. 1995;4(5):263–7.
41. Kennedy JD, Staples AJ, Brook PD, Parsons DW, Sutherland AD, Martin AJ, et al. Effect of spinal surgery on lung function in Duchenne muscular dystrophy. Thorax. 1995;50(11):1173–8.
42. Miller RG, Chalmers AC, Dao H, Filler-Katz A, Holman D, Bost F. The effect of spine fusion on respiratory function in Duchenne muscular dystrophy. Neurology. 1991;41(1):38–40.
43. Alexander WM, Smith M, Freeman BJ, Sutherland LM, Kennedy JD, Cundy PJ. The effect of posterior spinal fusion on respiratory function in Duchenne muscular dystrophy. Eur Spine J. 2013;22(2):411–6. doi:10.1007/s00586-012-2585-4.
44. Duckworth AD, Mitchell MJ, Tsirikos AI. Incidence and risk factors for post-operative complications after scoliosis surgery in patients with Duchenne muscular dystrophy: a comparison with other neuromuscular conditions. Bone Joint J. 2014;96-b(7):943–9. doi:10.1302/0301-620x.96b7.33423.
45. Galasko C, Delaney C, Morris P. Spinal stabilisation in Duchenne muscular dystrophy. J Bone Joint Surg Br. 1992;74(2):210–4.
46. Houde S, Filiatrault M, Fournier A, Dube J, D'Arcy S, Berube D, et al. Deflazacort use in Duchenne muscular dystrophy: an 8-year follow-up. Pediatr Neurol. 2008;38(3):200–6. doi:10.1016/j.pediatrneurol.2007.11.001.
47. King WM, Ruttencutter R, Nagaraja HN, Matkovic V, Landoll J, Hoyle C, et al. Orthopedic outcomes of long-term daily corticosteroid treatment in Duchenne muscular dystrophy. Neurology. 2007;68(19):1607–13. doi:10.1212/01.wnl.0000260974.41514.83.
48. Lebel DE, Corston JA, McAdam LC, Biggar WD, Alman BA. Glucocorticoid treatment for the prevention of scoliosis in children with Duchenne muscular dystrophy: long-term follow-up. J Bone Joint Surg Am. 2013;95(12):1057–61. doi:10.2106/jbjs.l.01577.
49. Dooley JM, Gordon KE, MacSween JM. Impact of steroids on surgical experiences of patients with duchenne muscular dystrophy. Pediatr Neurol. 2010;43(3):173–6. doi:10.1016/j.pediatrneurol.2010.04.013.

50. Luque ER. Segmental spinal instrumentation for correction of scoliosis. Clin Orthop Relat Res. 1982;163:192–8.
51. Modi HN, Suh SW, Song HR, Fernandez HM, Yang JH. Treatment of neuromuscular scoliosis with posterior-only pedicle screw fixation. J Orthop Surg Res. 2008;3:23. doi:10.1186/1749-799x-3-23.
52. Lonstein JE, Koop SE, Novachek TF, Perra JH. Results and complications after spinal fusion for neuromuscular scoliosis in cerebral palsy and static encephalopathy using luque galveston instrumentation: experience in 93 patients. Spine (Phila Pa 1976). 2012;37(7):583–91. doi:10.1097/BRS.0b013e318225ebd5.
53. Mattila M, Jalanko T, Puisto V, Pajulo O, Helenius IJ. Hybrid versus total pedicle screw instrumentation in patients undergoing surgery for neuromuscular scoliosis: a comparative study with matched cohorts. J Bone Joint Surg Br. 2012;94(10):1393–8. doi:10.1302/0301-620x.94b10.29383.
54. Funk S, Lovejoy S, Mencio G, Martus J. Rigid instrumentation for neuromuscular scoliosis improves deformity correction without increasing complications. Spine (Phila Pa 1976). 2015. doi:10.1097/brs.0000000000001170.
55. Arun R, Srinivas S, Mehdian SM. Scoliosis in Duchenne's muscular dystrophy: a changing trend in surgical management : a historical surgical outcome study comparing sublaminar, hybrid and pedicle screw instrumentation systems. Eur Spine J. 2010;19(3):376–83. doi:10.1007/s00586-009-1163-x.
56. Dayer R, Ouellet JA, Saran N. Pelvic fixation for neuromuscular scoliosis deformity correction. Curr Rev Musculoskelet Med. 2012;5(2):91–101. doi:10.1007/s12178-012-9122-2.
57. Alman BA, Kim HK. Pelvic obliquity after fusion of the spine in Duchenne muscular dystrophy. J Bone Joint Surg Br. 1999;81(5):821–4.
58. McCall RE, Hayes B. Long-term outcome in neuromuscular scoliosis fused only to lumbar 5. Spine. 2005;30(18):2056–60.
59. Whitaker C, Burton DC, Asher M. Treatment of selected neuromuscular patients with posterior instrumentation and arthrodesis ending with lumbar pedicle screw anchorage. Spine. 2000;25(18):2312–8.
60. Allen Jr BL, Ferguson RL. A 1988 perspective on the Galveston technique of pelvic fixation. Orthop Clin North Am. 1988;19(2):409–18.
61. Bell DF, Moseley CF, Koreska J. Unit rod segmental spinal instrumentation in the management of patients with progressive neuromuscular spinal deformity. Spine. 1989;14(12):1301–7.
62. Peelle MW, Lenke LG, Bridwell KH, Sides B. Comparison of pelvic fixation techniques in neuromuscular spinal deformity correction: Galveston rod versus iliac and lumbosacral screws. Spine 2006;31(20):2392–2398; discussion 9. doi:10.1097/01.brs.0000238973.13294.16.
63. Desrochers-Perrault F, Aubin CE, Wang X, Schwend RM. Biomechanical analysis of iliac screw fixation in spinal deformity instrumentation. Clin Biochem. 2014;29(6):614–21. doi:10.1016/j.clinbiomech.2014.04.016.
64. Carroll EA, Shilt JS, Jacks L. MW construct in fusion for neuromuscular scoliosis. Eur Spine J. 2007;16(3):373–7. doi:10.1007/s00586-006-0077-0.
65. Sponseller PD, Shah SA, Abel MF, Sucato D, Newton PO, Shufflebarger H, et al. Scoliosis surgery in cerebral palsy: differences between unit rod and custom rods. Spine. 2009;34(8):840–4. doi:10.1097/BRS.0b013e31819487b7.
66. Phillips JH, Gutheil JP, Knapp Jr DR. Iliac screw fixation in neuromuscular scoliosis. Spine. 2007;32(14):1566–70. doi:10.1097/BRS.0b013e318067dcff.
67. Chang TL, Sponseller PD, Kebaish KM, Fishman EK. Low profile pelvic fixation: anatomic parameters for sacral alar-iliac fixation versus traditional iliac fixation. Spine. 2009;34(5):436–40. doi:10.1097/BRS.0b013e318194128c.
68. Moon ES, Nanda A, Park JO, Moon SH, Lee HM, Kim JY, et al. Pelvic obliquity in neuromuscular scoliosis: radiologic comparative results of single-stage posterior versus two-stage anterior and posterior approach. Spine. 2011;36(2):146–52. doi:10.1097/BRS.0b013e3181cd2a55.
69. Modi HN, Suh SW, Song HR, Yang JH, Jajodia N. Evaluation of pelvic fixation in neuromuscular scoliosis: a retrospective study in 55 patients. Int Orthop. 2010;34(1):89–96. doi:10.1007/s00264-008-0703-z.
70. Mehta SS, Modi HN, Srinivasalu S, Suh SW, Yi JW, Cho JW, et al. Pedicle screw-only constructs with lumbar or pelvic fixation for spinal stabilization in patients with Duchenne muscular dystrophy. J Spinal Disord Tech. 2009;22(6):428–33. doi:10.1097/BSD.0b013e3181872d74.
71. Imrie MN, Yaszay B. Management of spinal deformity in cerebral palsy. Orthop Clin North Am. 2010;41(4):531–47. doi:10.1016/j.ocl.2010.06.008.
72. Sarwahi V, Sarwark JF, Schafer MF, Backer C, Lee M, King EC, et al. Standards in anterior spine surgery in pediatric patients with neuromuscular scoliosis. J Pediatr Orthop. 2001;21(6):756–60.
73. Hod-Feins R, Abu-Kishk I, Eshel G, Barr Y, Anekstein Y, Mirovsky Y. Risk factors affecting the immediate postoperative course in pediatric scoliosis surgery. Spine. 2007;32(21):2355–60. doi:10.1097/BRS.0b013e3181558393.
74. Ferguson RL, Hansen MM, Nicholas DA, Allen Jr BL. Same-day versus staged anterior-posterior spinal surgery in a neuromuscular scoliosis population: the evaluation of medical complications. J Pediatr Orthop. 1996;16(3):293–303.
75. Hopf CG, Eysel P. One-stage versus two-stage spinal fusion in neuromuscular scolioses. J Pediatr Orthop B. 2000;9(4):234–43.
76. O'Brien T, Akmakjian J, Ogin G, Eilert R. Comparison of one-stage versus two-stage anterior/posterior spinal fusion for neuromuscular scoliosis. J Pediatr Orthop. 1992;12(5):610–5.
77. Keeler KA, Lenke LG, Good CR, Bridwell KH, Sides B, Luhmann SJ. Spinal fusion for spastic neuromuscular scoliosis: is anterior releasing necessary when intraoperative halo-femoral traction is used? Spine. 2010;35(10):E427–33. doi:10.1097/BRS.0b013e3181d9527e.
78. Vialle R, Delecourt C, Morin C. Surgical treatment of scoliosis with pelvic obliquity in cerebral palsy: the influence of intraoperative traction. Spine (Phila Pa 1976). 2006;31(13):1461–6. doi:10.1097/01.brs.0000219874.46680.87.
79. LaMothe J, Al Sayegh S, Parsons D, Ferri-de-Barros F. The use of intraoperative traction in pediatric scoliosis surgery: a systematic review. Spine Deform. 2015;3(1):45–51.

Evidence-Based Treatment for Congenital Scoliosis

25

Firoz Miyanji

Abstract

Management of congenital scoliosis begins with the recognition of the patterns of congenital anomalies and the anticipation of the risk of deformity progression. The natural history and prognosis of a patient with congenital scoliosis can vary considerably as illustrated by many of the early descriptive reports. Despite a variety of different surgical strategies that have been described for congenital scoliosis to date, very little evidence exists supporting the most ideal management strategy from timing of intervention to most appropriate surgical technique, to efficacy of the available procedures in altering the natural history of congenital scoliosis. This chapter will outline the available literature, its limitations, and whether evidence-based recommendations can be made around treatment strategies for congenital scoliosis.

Keywords

Congenital scoliosis • Treatment • Hemivertebra resection • Hemiepiphysiodesis • VEPTR • Growing rods

Background

Congenital scoliosis is defined as a curvature of the spine due to one or more abnormalities of the vertebrae, which is generally due to a failure of formation or segmentation or a combination of these two. By virtue of its early gestational origin, congenital scoliosis is always an early-onset spinal deformity with the potential for progression as the child grows, which remains the fundamental guiding principle around which treatment decisions are made.

Although early reports estimated that congenital scoliosis had generally a benign course [1] subsequent natural history studies by Winter et al. [2] and McMaster et al. [3] refuted this, reporting that the rate of deterioration and the severity of the final deformity were predictable, according to the type of anomaly and curve location. In fact McMaster and Ohtsuka [3] narrowed the risk of progression to four key factors: the type of congenital anomaly, the curve location, the patient's age, and solitary or multiple curves. The authors found that nearly 75 % of patients in their series required treatment and 84 % of patients who were untreated developed curves greater than 40° at maturity. It is accepted now that the worst anomaly is a unilateral unsegmented bar combined with a single or multiple convex hemivertebrae, followed by a unilateral unsegmented bar, double convex hemivertebrae, and finally a single convex hemivertebra. A block vertebra has the best prognosis. Thoracolumbar curves have the poorest prognosis, and children with clinical deformities in the first year of life are also at risk of severe progression (Fig. 25.1) [4].

Management of congenital scoliosis begins with the recognition of those curves with a bad prognosis at an early stage. Treatment should be aimed at altering the natural

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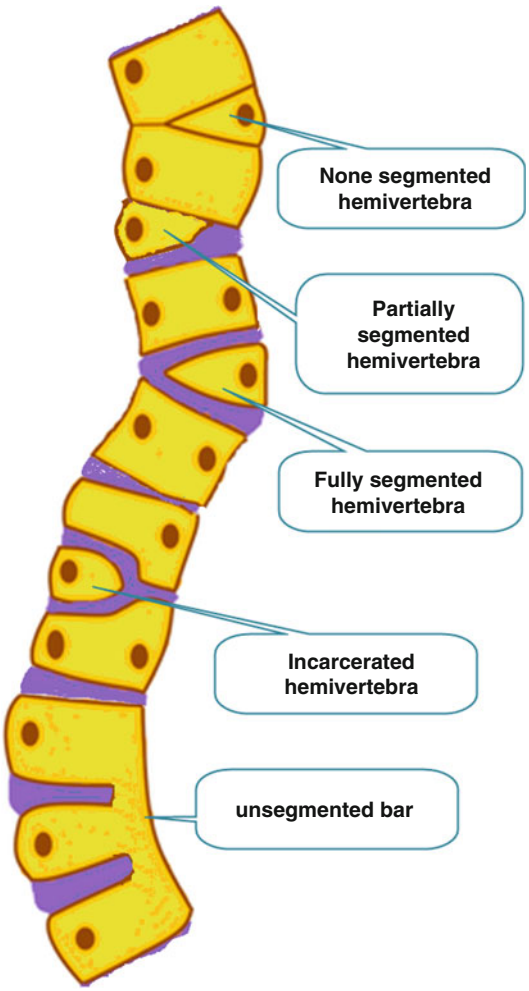

Fig. 25.1 Diagram showing various abnormal vertebra that lead to congenital scoliosis

history as described by Winter et al. [2] and McMaster et al. [3], specifically in preventing deformity progression, maintaining chest wall and lung development, achieving maximum overall spinal growth, and minimizing potential complications. Observation is appropriate when the natural history suggests that the curve will not progress, and bracing has been considered by some to control secondary noncongenital curves in the presence of a primary congenital curve. The evidence around brace treatment in congenital scoliosis is however limited to scattered case series and case reports.

A number of surgical strategies have been described for the management of congenital scoliosis either to prevent further curve progression (in situ fusion; growth-friendly technologies) or in addition to obtain deformity correction in either a gradual (hemiepiphysiodesis) or acute fashion (hemivertebrae excision). Effectiveness of non-operative therapies of bracing and casting, timing of surgical intervention, the most appropriate surgical strategy, and the efficacy of the available surgical options remain at the center of continued controversy.

Four Key Questions

1. Is brace/cast treatment effective in congenital scoliosis?
2. When ideally should surgery be considered?
3. What is the ideal surgical strategy for congenital scoliosis secondary to a hemivertebra?
4. Is there any role for growth-friendly technologies in congenital scoliosis?

Is Brace/Cast Treatment Effective in Congenital Scoliosis?

Winter's retrospective multicenter review of patients with congenital scoliosis treated in a Milwaukee brace reported on 63 patients with a mean age of 8 years and mean duration of brace wear of 4 years [5]. The authors classified their outcome of brace treatment as "very good", "good", "fair", or "poor" determined a priori as whether curve progression occurred ultimately requiring surgical intervention or not. They found 2 patients had results classified as "very good" (surgery was avoided), 33 were "good" (surgery either prevented or delayed), 8 were "fair" (curve stabilized), and 20 were "poor" (continued deterioration of the curve and surgery was neither delayed nor prevented). They also analyzed curve characteristics that responded to brace treatment and suggested that longer, flexible curves, with a mixture of anomalous and nonanomalous vertebrae responded best whereas short, sharp, and rigid curves did not do well with brace treatment. The authors did not report on the percentage of total patients that went on to surgery. Their conclusions are to consider the use of Milwaukee brace as a trial in some cases of congenital scoliosis. This study is limited to Level IV evidence, providing little evidence as to the true effect of brace treatment on congenital scoliosis.

More recently Demirkiran et al. [6] analyzed their experience in 11 patients with congenital scoliosis who were treated with serial cast application. The average age of the patients was 3.3 years and follow-up was unclear, though all patients had a minimum of 1 year follow-up. The mean precasting Cobb angle was 70.7°, with a mean correction in cast at final follow-up of 55.1°. The mean compensatory curve angle was 55.8°, which corrected in cast to a mean 39.8° at final follow-up. The authors concluded that surgery is delayed in such patients by an average of 26.3 months. The small sample size, retrospective design, significant selection bias, and the exclusion of a priori defined indications for surgery limit this study in providing evidence for serial casting.

When Ideally Should Surgery Be Considered?

In Situ Fusion

Although generally accepted that the younger the child, the more potential for significant progression of the deformity, the variety of combinations of congenital deformities seems

limitless and hence the prediction about what will happen with growth remains difficult. Nonetheless, expert opinion has recommended early fusion with the assumption that "it is safe to say that it is easier to prevent a deformity than to correct a deformity" [7].

To date there remains a paucity of strong evidence on which recommendations for early in situ fusion for congenital scoliosis can be made. Of the available studies, the majority of the literature remains retrospective in nature with limited follow-up. McMaster's review of a single type of congenital scoliosis (unilateral bar and contralateral hemivertebrae) described significant variability in the natural history [4]. Nine patients who had an upper thoracic scoliosis had variable rates of progression: two reached curves of 70° and 80° at skeletal maturity while 5 others were ≤50°. One 4 year-old was managed by immediate arthrodesis because of a curve of 48°, yet a 5 year old with a curve of 45° was observed and had a curve of 51° by age 10. Similarly, substantial variability in timing of arthrodesis, surgical approach, number of segments fused, as well as magnitude of preoperative curve makes it difficult to support the conclusions that early surgery, preferably in the first year of life, is warranted.

Given that, at best, studies reporting on *in situ* early fusion for congenital scoliosis remain retrospective case series, it is important to review those studies with longer-term follow-up.

Goldberg et al. [8] reported their experience of in situ fusion in the setting of congenital scoliosis and included a retrospective review of 43 patients who were at least 15 years of age at final follow-up. They noted that although the localized fusion was effective in preventing progression of the Cobb angle of the congenitally malformed area, it did not control the overall deformity that developed and progressed with growth. Their case series noted a 25.6 % reoperation rate due to continued progression of the deformity. The type of congenital scoliosis was heterogeneous in this study.

Subsequently Vitale and colleagues [9] reported clinical and radiographic outcomes, also in a retrospective manner on 21 patients. In this study, 5 of the 21 patients had multiple procedures and the authors also reported pulmonary function data as well as Child Health Quality of Life Outcome (QOL) questionnaires. The group compared the results with healthy children and concluded that the cohort of congenital scoliosis patients with early fusions had worse pulmonary function tests (PFTs) and QOL scores compared to healthy peers, cautioning against early fusion in this patient population. The comparative group, however, was that of healthy peers which limits the interpretation of their data, as patients with congenital scoliosis often have other comorbidities. The authors did not report on pre-operative PFTs or QOL data for comparison.

A more recent retrospective case-control study reported on outcomes of instrumented versus non-instrumented spinal fusion in a total of 51 patients [10]. Although the instrumented group had better post-operative curve correction, continued and marked curve progression was noted in both groups with a 25 % reoperation rate (both groups). The authors questioned whether early fusion for congenital scoliosis was meeting the goal of progression prevention.

Karol et al.'s review of in situ fusion in early onset scoliosis although having mixed etiologies, 20 of the reported on 28 patients had congenital scoliosis [11]. They had a 39.3 % revision rate. The authors cautioned against early fusion as it put patients at risk of restrictive pulmonary disease, especially those with proximal thoracic deformity who were fused over more than four segments. Again, pre-operative pulmonary data on these patients were lacking, making it difficult to support their conclusion that early fusions have a significant direct negative effect on pulmonary function in the long-term.

Both earlier and the more recent studies remain retrospective cross-sectional case series with no longitudinal or prospective component and so remain as weak evidence only supporting/refuting early fusion as a treatment for congenital scoliosis.

Hemiepiphysiodesis

Convex growth arrest as a treatment for congenital scoliosis has been previously described as effective by a number of authors [12–14]. It has been described for patients with hemivertebrae but others have also reported on its outcomes for failure of segmentation defects and mixed anomalies. Demirkiran et al. [15] most recently described their experience with this technique in a series of 13 patients (mean operative age, 5.4 years; mean follow-up, 4.7 years). The authors' indication for surgery was a long sweeping curve, including >4–5 segments, and not suitable for single hemivertebrectomy. Although they report promising results with a mean curve correction of 33.5°± 12.4° at final follow-up, from an average preoperative Cobb angle of 49°± 10.9°, none of these patients were evaluated to skeletal maturity. In addition, the authors note that in nine patients the curve improved, in three patients there was no change, and in one patient there was curve progression, illustrating that the procedure may have either an "epiphysiodesis effect" (what it is intended for) or a "fusion effect" where the curve may not improve but also will not progress. They admit that, at present, with the absence of any reliable technique to evaluate the growth potential of the vertebral apophysis, it is impossible to predict the outcome of this procedure. Therefore which patients would be the most appropriate candidates for an instrumented convex hemiepiphysiodesis remains unclear.

In contrast, Thompson et al. [16] reviewed retrospectively their case series of 30 patients (mean age, 6.3 years), 63 % of whom reached skeletal maturity. They noted an improvement in Cobb angle in 23 patients, progression arrested or slowed in 5 patients, and progression in 2 patients. Their technique involved a combined anterior/posterior approach.

Marks et al. [17] examined a further 53 patients with a mean follow-up of 8.8 years. Thirty-four patients were skeletally mature. This series reported on the effects of convex hemiepiphysiodesis on a variety of types of congenital anomaly. Those with failure of segmentation defects, as well as complex anomalies, continued to progress despite surgery, with a final Cobb angle increase from a mean of 61°–70°. In contrast, 97 % of hemivertebrae patients had reversal in their curves or slowed progression with an average preoperative Cobb angle of 41° improving to 35° at final follow-up. The authors felt that a younger age at surgery and a hemivertebrae located in the lumbar spine yielded the best results.

Keller et al. [18] reported their experience in 16 patients (mean operative age, 4.8 years; mean follow-up, 4.8 years). Their series had a variety of formation and segmentation anomalies and they found 37 % of curves improved, 42 % were unchanged or progressed less than 7°, 16 % progressed 10°–15°, and 5 % progressed greater than 15°. The best results were in patients with a hemivertebrae, whereas progression despite surgery was seen in patients with an unsegmented bar and a contralateral hemivertebrae. The authors felt that their results were most predictable at producing an arrest of the deformity and the epiphysiodesis effect to be less predictable. The authors conclude that it remains unknown what the final outcome will be, as these patients were not followed to skeletal maturity.

The remaining retrospective reviews pooled data shows improvement in curves in 48 % of cases (range 20–77 %), no change (fusion effect) in 40 % (range 17–70%), and curve progression in 12 % (range 0–21 %) with conclusions that earlier intervention provides the best results [12–14, 19, 20]. Given that all these studies are retrospective single center case series without the benefit of comparative controls, the effect of age as well as type of congenital anomaly on the results of surgical intervention cannot truly be determined and the small number of patients in each study limit broad conclusions to be made. Therefore, there is weak evidence at best to suggest that early hemiepiphysiodesis (before the age of 5 years) is most appropriate for congenital scoliosis.

Hemivertebra Excision

Hemivertebra excision, initially described as via a combination of anterior and posterior approaches, has recently been refined to an all-posterior approach. Most studies report on the technique and feasibility of hemivertebra excision with expert opinion on the advantages and disadvantages of either a combination anterior/posterior approach or an all-posterior approach [21–24]. Studies reporting on hemivertebra excision in young children have attempted to report on children younger than 10 years of age with a substantial variation in timing of surgery. Crostelli et al.'s 15 cases had a mean age of 5.5 years, with a range from 2 to 9.5 years [25]. Although they reported a percentage curve correction of 72.5 % (mean preoperative curve of 44°) and no major complications, the average follow-up was extremely limited (3.3 years). Chang and colleagues [26] performed a retrospective review of 18 patients less than 10 years of age (mean 6.6 years, range 2.6–9.8 years); they noted an average preoperative curve of 34.4° correcting to 8.4° (75 % correction) immediately postoperatively. This was an average of 12.9° at final follow-up (62.5 % correction). Their series had a mean follow-up of 11.4 years and also reported no complications from surgery.

Chang et al. [27] also reported their results of hemivertebra resection and the effect of age on outcomes. They arbitrarily assigned patients into two groups (nine in each group) based on age at time of surgery: those under 6 years of age (mean age 4.2 years) and those between 6 and 10 years of age (mean age 9 years). The authors hypothesized that those that had surgery before the age of six had better deformity correction with no impact on vertebral or spinal growth. The younger patients had, on average, a preoperative curve magnitude of 32.4° improving to 9.1° at final follow-up, compared to the older age group with a preoperative curve of 36.5° improving to 14.5°. As a retrospective review of patients treated at a single center, without the benefit of true controls, the effect of age on the results of surgical intervention cannot truly be determined. It seems more interesting that although the average difference in age between the groups was nearly 5 years, the mean difference in preoperative Cobb angle was only 4.1°, suggesting that there was not a dramatic risk of progression during this time; therefore a delay in treatment until the child is older could be equally considered.

Bollini and colleagues reported homogeneous cohorts of thoracolumbar and lumbar hemivertebrae resections, in two separate studies [22, 23]. The studies had equivalent mean age at time of surgery (3.5 years for the report on thoracolumbar hemivertebrae resections; 3.3 years for the lumbar resections). The average follow-up was 6 and 8.6 years, respectively. In the thoracolumbar resections, there was a mean improvement of 69.3 % in the segmental curve, from an average 34.8° before surgery to 10.7° at final follow-up. Seven of the thirty-four patients had immediate postoperative complications and late complications occurred in 12 patients (34 %), which included pseudarthrosis in 5 patients and progression of curve in 6 patients. Of the 34 patients, 18 required additional surgery during the follow-up period.

In their report of lumbar hemivertebrae resections, Bollini et al. [23] noted a more significant improvement in the segmental scoliosis curve of 71.4 % with a mean curve of 32.9° preoperatively, improving to 9.4° at final follow-up. Of the 21 patients in this series 3 experienced postoperative complications, which included radiculopathy, wound infection, and acute renal insufficiency. The authors concluded in both studies that hemivertebra excision should be performed as early as possible.

Similarly, retrospective case series from Wang et al. [28] and Ruf et al. [29] conclude that early hemivertebra resection surgery should be considered to prevent the development of severe local deformities and secondary structural curves. The mean preoperative curve in Wang et al.'s study was 36.6°, which improved to 5.1° [28]. Ruf et al. [29] had a preoperative Cobb angle of 36° in patients without a bar formation that improved to 7° following surgery; those with a bar formation had a preoperative curve of 69° improving on average to 23°. The average age was 4.9 and 3.4 years, respectively.

Nakamura et al. [30] reported on five patients over an average 12.8-year follow-up period for hemivertebra resection. Their illustrative case report on one patient described the age at operation to be 13 years 7 months while another case had surgery at the age of 3 years. Despite satisfactory long-term results reported by the authors, definitive conclusions with a sample size of 5 cannot be made. Zang et al. [31] retrospectively reviewed 58 hemivertebra resections in 56 patients with a significant range in age at the time of surgery (mean age 9.9 years with range 1.5–17 years). There was a mean improvement of 72.9 % in this study in the Cobb angle at final follow-up. The mean follow-up was 37.9 months (range 24–58 months). The authors did not analyze the results as a function of age yet conclude that hemivertebra resection is ideal for very young children.

Interestingly, earlier reports from Deviren et al. [24] demonstrated that hemivertebra excision in larger curves (mean preoperative Cobb angle78.2°) and in older patients (mean age 13.4 years) yielded equally favourable results without major complications and therefore the literature remains inconclusive as regards to timing of intervention on outcomes of congenital scoliosis after hemivertebra excision. All studies to date are retrospective in nature providing Level IV evidence with inconsistent surgical indications, variable approaches (anterior/posterior, all-posterior, number of fusion segments), heterogeneity of congenital anomalies, and age of intervention such that specific guidelines for surgical timing cannot be made.

What Is the Ideal Surgical Strategy for Congenital Scoliosis Secondary to a Hemivertebra?

A very limited number of comparative studies exist in which recommendations can be made regarding the optimum surgical strategy for congenital scoliosis. As the previous section outlined, the majority of the existing literature is limited to retrospective single center reviews of case series with a small sample size. This precludes definitive conclusions to be made around timing of surgery as well as the ideal surgical strategy (Fig. 25.2).

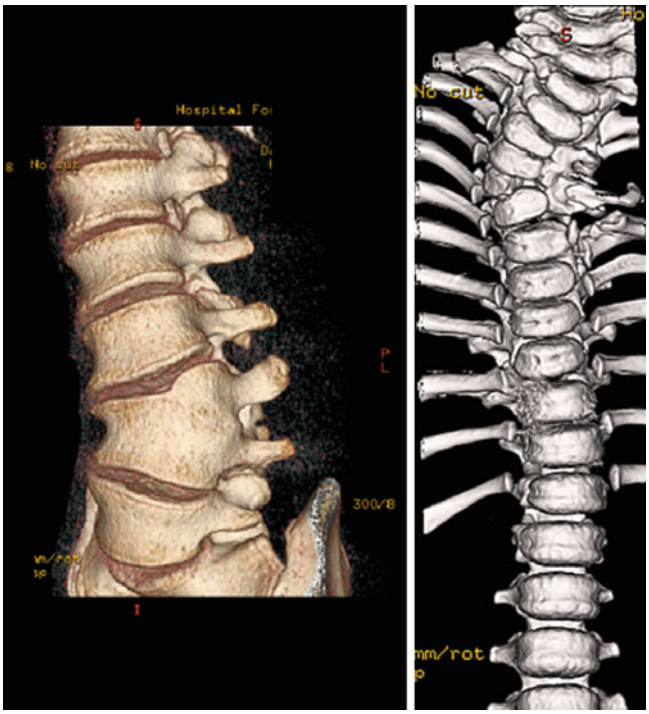

Fig. 25.2 Reconstructive CT images showing a partially segmented and fully segmented hemivertebrae

Yaszay et al. [32] reported the only Level III multi-center study comparing three different surgical techniques. This was a retrospective review of 76 patients with a minimum 2 year follow-up. They compared in situ fusion or hemiepiphysiodesis (Group 1) to instrumented fusion without hemivertebra excision (Group 2), and instrumented fusions with hemivertebra excision (Group 3). They had 14 patients in group 1, 20 in group 2, and 42 in group 3, with mean ages of 9.8, 10.3, and 5 years, respectively. Groups 1 and 3 had smaller preoperative curves than group 2. At 2 year follow-up, group 3 had the best overall curve correction at 73 %, compared to 42 % for group 2 and 27 % for group 1. Group 3 also had shorter fusions and less blood loss than group 2, and a trend towards shorter operative times. Group 1 had the lowest blood loss and operative time. The complication rate also differed: group 3 had the highest rate at 44 %, compared to 23 % in group 1 and 17 % in group 2. Groups 1 and 3 each had a patient with a pseudarthrosis requiring revision surgery, as well as progression of deformity in two patients requiring repeat surgery. A total of six instrumentation failures occurred, one in group 2 and five in group 3. A total of six neurologic complications occurred; one was in group 2, and five were in group 3. The authors highlight that group 1 had the smallest curve correction but also the lowest complication rate and blood loss. Group 2 had the longest operative time and blood loss likely secondary to the longer fusions. Group 3, having the most aggressive deformity correction, had the highest complication rate including a higher risk of

neurological injury. The retrospective nature of the study lends itself to significant limitations. The treatment selection criteria were not standardized and the heterogeneity of the patient population, especially with curve magnitude, patient age, location of the hemivertebra, and other confounders influencing treatment selection (ultimately made by individual surgeons) should be taken into account when interpreting these results.

Lee and colleagues [33] attempted to compare two different treatments in patients with congenital scoliosis: instrumented fusions and hemivertebrectomy. Their report would suggest that the instrumentation only group had comparable and satisfactory curve correction (60.3 % compared to 73.7 % in the hemivertebrectomy group), with shorter operative times and less blood loss. However, this study had significant limitations from a methodological perspective in that the decision to consider a hemivertebrectomy was made intraoperatively, if the temporary intraoperative deformity correction was not satisfactory. If, however, the deformity correction was satisfactory intra-operatively, then the authors simply performed an instrumented fusion without hemivertebrectomy. As such the study is more in keeping with an observational case series than a true comparative study (with the results representing Level IV evidence).

Evidence supporting a particular surgical strategy for congenital scoliosis presently relies strongly on expert opinion. A strong recommendation of a single surgical strategy over another in the setting of a hemivertebra cannot be made on the basis of the available literature.

Is There Any Role for Growth-Friendly Technologies in Congenital Scoliosis?

By nature of its gestational origin, congenital scoliosis is always an early-onset condition. For such children, early literature strongly encouraged spinal arthrodesis at a young age. Some recommended surgery within the first year of life [4] and others [17, 22, 23, 29, 32] report promising results if surgery was completed before the age of 5 years. These principles rely primarily on expert opinion and the evidence surrounding this remains Level IV. In more recent years, longer term follow-up studies, albeit retrospective in nature, have highlighted potential concerns of early fusions and its negative impact on pulmonary function, lung development, and spinal growth [8, 9, 11]. Goldberg's comparative case-control study showed substantial decline in vital capacity at skeletal maturity in children who had spinal fusions (FVC 40.8 %) before the age of 10 compared to those without fusion (FVC 96.6 %) providing fair evidence that the prognosis following early spinal fusion should be guarded [34]. Campbell also highlighted that early spinal fusion not only results in short stature but also influences the thorax and may result in severe extrinsic restrictive lung disease by volume restriction of the growing lungs and motion restriction of the ribs [35]. He termed the inability of the thorax to support normal lung growth and respiration *thoracic insufficiency syndrome* [36].

Hell et al. [37] reviewed their experience with VEPTR in congenital scoliosis. The authors simply reported on the Cobb angle at final follow-up and the subjective patient and parent impressions. Most patients were younger than 10 and the follow-up period is unclear. The authors conclude that the VEPTR addressed the thoracic insufficiency syndrome with probable benefit to the remaining lung growth. However, they failed to provide any measure of preoperative and postoperative lung function data. They also concluded that this method seems superior to other concepts in treating congenital scoliosis, again with no comparative group data. The study provides poor-quality evidence to make recommendations regarding VEPTR in congenital scoliosis.

More recently, Flynn et al. [38] reviewed VEPTR treatment for congenital scoliosis, reporting a multicenter retrospective analysis on 24 children (mean age at implantation, 3.3 years – with a broad range of 8 months to 12.5 years; average follow-up, 3.3 years). Indications for surgery and types of congenital anomalies treated are not clear. Their primary outcome was Cobb angle correction and thoracic height change. They had no objective measures of pulmonary assessment but report that 92 % of patients had no change in ventilation status following surgery. They found 83 % of patients have an improvement of the Cobb angle, an average of 8.9°, and 17 % had progression of their deformity on average by 18.8°. They found *all* patients had an improvement in thoracic height (average change, 3.41 cm) – those that had improvement in their curves as well as patients in whom curve *progression* occurred. They also noted 16 adverse events in 8 patients, of which 94 % required surgical intervention. The authors conclude that VEPTR is a "successful" treatment for congenital scoliosis; however, the small sample size, limited follow-up, and retrospective design do not support such a broad conclusion.

Growing rods have also been reported on as potential effective treatments for congenital scoliosis. Elsebai et al. [39] report on the most homogeneous population of patients with congenital scoliosis; other studies concerning the growing rod included patients with congenital scoliosis among other aetiologies – hence interpretation of their results is challenging. Elsebai and colleagues' retrospective review of 19 patients (mean age of index surgery, 6.9 years; mean fol-

low-up period, 4 years) had a slightly older group of patients and narrower age-range (3.2–10.7 years) than Flynn et al.'s [38] VEPTR study. However, 10 of the 19 patients had had previous surgery, including hemiepiphysiodesis and apical fusions. The authors reported a 29 % improvement in the major curve Cobb angle (mean preoperative 66° improved to mean 47° at final follow-up). Analysis of individual cases showed deformity improvement in only six patients, no change in two, and worsening of deformity in nine. It is unclear whether these nine patients were the ones that had only growing rod placement and no prior surgery. Patients with failure of formation, failure of segmentation, mixed anomalies, and unclassified types of congenital deformity were all included in the study. There was considerable variability of implant configuration with differences in proximal and distal anchors as well as single (12 patients) versus dual rod (7 patients) implants. The authors also demonstrate an improvement of T1–S1 spinal length as well as space available for lung (SAL) following repeated distractions. However, correlation of these parameters to pulmonary function is not presented. From this study, it may be suggested that deformity correction, spinal growth, and SAL is improved following growing rod placement. However, the effect of this treatment on the natural history of congenital scoliosis cannot be determined from this Level IV study.

More recently Wang et al. [40] reported their case series of growing rod for 30 congenital scoliosis patients (mean operative age, 7.3 years; range, 2–13 years), with a mean follow-up of 2 years. The authors reported that they chose this method for severe congenital scoliosis patients, wherein the curve could not be controlled by other methods. In some of their patients, a 1-stage posterior apical osteotomy or hemivertebra resection with a short segmental fusion was also performed in conjunction with the growing rod implantation. Similarly to Elsebai's study [39], outcomes included curve correction, T1–S1 length, and SAL which all showed improvements postoperatively. There were 13 complications in 7 patients. The limited follow-up, sample size, surgical technique variability, and bias inherent in retrospective reviews makes it difficult to support of the author's conclusions that dual growing rod technique is safe and effective for treating congenital scoliosis.

Therefore, the evidence concerning the true treatment effect of growth-friendly technologies on congenital scoliosis is insufficient. Studies, to date, are of poor quality, unable to provide any recommendations for the routine use of growth-friendly technologies in congenital scoliosis.

Conclusion

Natural history studies have provided valuable knowledge regarding congenital scoliosis through descriptive retrospective large case series. They have alerted clinicians to definite curve progression for *some* patients with congenital scoliosis, but there is not evidence that *all* patients with congenital scoliosis will progress.

Treatment principles initially guided by preventing deformity progression and obtaining correction have evolved through expert opinion into an emphasis on chest wall and lung development, and maintenance of spinal length. Initial reports emphasizing early arthrodesis for congenital scoliosis were based primarily on retrospective observational case series. Recently, these have been challenged by Level IV studies which caution against this approach. The substantial variation of age at time of surgery, surgical indications, as well as follow-up between all available Level IV studies reporting on outcomes of early fusion, hemiepiphysiodesis, and hemivertebra excision for congenital scoliosis provide poor-quality evidence for the ideal time for surgical intervention. Therefore, recommendations in support of early intervention versus a delayed approach cannot be made.

Congenital scoliosis secondary to a hemivertebra is the most common anomaly encountered. Prospective comparative studies on treatment outcomes following different surgical strategies are lacking. The single study to date is a retrospective comparison among a heterogeneous cohort of patients providing fair evidence in support of early fusion, hemiepiphysiodesis, or hemivertebra excision in this setting. A strong recommendation of one strategy over another cannot be made.

Although principles of growth-friendly technologies are promising and may be extrapolated to congenital scoliosis, currently evidence to support their use as an effective treatment in this setting is insufficient.

Table 25.1 lists current recommendations for treatment of congenital scoliosis.

Table 25.1 Recommendations

Recommendation	Level of evidence	Grade
Bracing/cast treatment for congenital scoliosis may be effective	IV	I
Surgery for congenital scoliosis should be considered at an early age (<5 year)	IV	I, C
The optimum surgical strategy for a congenital hemivertebra is established	III, IV	I
Growth-friendly technologies are safe and effective in treating congenital scoliosis	IV, V	I

References

1. Kuhns JG, Hormell RS. Management of congenital scoliosis. Review of one hundred seventy cases. Arch Surg. 1952;65: 250–63.
2. Winter RB, Moe JH, Eilers VE. Congenital scoliosis: a study of 234 patients treated and untreated. J Bone Joint Surg A. 1968;50: 15–47.
3. McMaster MJ, Ohtsuka K. The natural history of congenital scoliosis. A study of two hundred and fifty-one patients. J Bone Joint Surg A. 1982;64:1128–47.
4. McMaster MJ. Congenital scoliosis caused by a unilateral failure of vertebral segmentation with contralateral hemivertebrae. Spine. 1998;23(9):998–1005.
5. Winter RB, Moe JH, MacEwen D, et al. The Milwaukee brace in the nonoperative treatment of congenital scoliosis. Spine. 1976;1:85–96.
6. Demirkiran HG, Bekmez S, Celilov R, et al. Serial derotational casting in congenital scoliosis as a time-buying strategy. J Pediatr Orthop. 2015;35(1):43–9.
7. Hedden D. Management themes in congenital scoliosis. J Bone Joint Surg A. 2007;89(suppl 1):72–8.
8. Goldberg CJ, Moore DP, Fogarty EE, et al. Long-term results from in situ fusion for congenital vertebral deformity. Spine. 2002;27(6):619–28.
9. Vitale MG, Matsumoto H, Bye MR, et al. A retrospective cohort study of pulmonary function, radiographic measures, and quality of life in children with congenital scoliosis: an evaluation of patient outcomes after early spinal fusion. Spine. 2008;33(11):1242–9.
10. Farley FA, Li Y, Jong N, Powell CC, et al. Congenital scoliosis SRS-22 outcomes in children treated with observation, surgery, and VEPTR. Spine. 2014;39(22):1868–74.
11. Karol LA, Johnston C, Mladenov K, et al. Pulmonary function following early thoracic fusion in non-neuromuscular scoliosis. J Bone Joint Surg A. 2008;90:1272–81.
12. Andrew T, Piggot H. Growth arrest for progressive scoliosis: combined anterior and posterior fusion of the convexity. J Bone Joint Surg Br. 1985;67:193–7.
13. Uzumcugil A, Cil A, Yazici M, et al. Convex growth arrest in the treatment of congenital spinal deformities: revisited. J Pediatr Orthop. 2004;24:658–66.
14. Winter RB, Lonstein JE, Dennis F. Convex growth arerest for progressive congenital scoliosis due to hemivertebra. J Pediatr Orthop. 1988;8:633–8.
15. Demirkiran G, Yilmaz G, Kaymaz B, et al. Safety and efficacy of instrumented convex growth arrest in treatment of congenital scoliosis. J Pediatr Orthop. 2014;34(3):275–81.
16. Thompson AG, Marks DS, Sayampanathan SR, et al. Long-term results of combined anterior and posterior convex epiphysiodesis for congenital scoliosis due to hemivertebrae. Spine. 1995;20(12): 1380–5.
17. Marks DS, Sayampanathan SR, Thompson AG, et al. Long-term results of convex epiphysiodesis for congenital scoliosis. Eur Spine J. 1995;4(5):296–301.
18. Keller PM, Lindseth RE, DeRosa GP. Progressive congenital scoliosis treatment using a transpedicular anterior and posterior convex hemiepiphysiodesis and hemiarthrodesis: a preliminary report. Spine. 1994;19(17):1933–9.
19. Kieffer J, Dubousset J. Combined anterior and posterior convex epiphysiodesis for progressive congenital scoliosis in children aged < or = 5 years. Eur Spine J. 1994;3:120–5.
20. Walhout RJ, van Rhijn LW, Pruijs JE. Hemi-epiphysiodesis for unclassified congenital scoliosis: immediate results and mid-term follow-up. Eur Spine J. 2002;11:543–9.
21. Jalanko T, Rintala R, Puisto V, et al. Hemivertebra resection for congenital scoliosis in young children: comparison of clinical, radiographic, and health-related quality of life outcomes between the anteroposterior and posterolateral approaches. Spine. 2011;36(1):41–9.
22. Bollini G, Docquier PL, Viehweger E, Launay F, Jouve JL. Thoracolumbar hemivertebrae resection by double approach in a single procedure: long-term follow-up. Spine. 2006;31(15): 1745–57.
23. Bollini G, Docquier PL, Viehweger E, Launay F, Jouve JL. Lumbar hemivertebra resection. J Bone Joint Surg A. 2006;88(5):1043–52.
24. Deviren V, Berven S, Smith A, et al. Excision of hemivertebrae in the management of congenital scoliosis involving the thoracic and thoracolumbar spine. J Bone Joint Surg Br. 2001;83(4):496–500.
25. Crostelli M, Mazza O, Mariani M. Posterior approach lumbar and thoracolumbar hemivertebra resection in congenital scoliosis in children under 10 years of age: results with 3 years mean follow up. Eur Spine J. 2014;23(1):209–15.
26. Chang DG, Kim JH, Ha KY, et al. Posterior hemivertebra resection and short segment fusion with pedicle screw fixation for congenital scoliosis in children younger than 10 years: greater than 7-year follow-up. Spine. 2015;40(8):E484–91.
27. Chang DG, Suk SI, Kim JH, et al. Surgical outcomes by the age at the time of surgery in the treatment of congenital scoliosis in children under age 10 years. Spine J. 2015;15:1783–95.
28. Wang S, Zhang J, Qiu G, et al. Posterior hemivertebra resection with bisegmental fusion for congenital scoliosis: more than 3 year outcomes and analysis of unanticipated surgeries. Eur Spine J. 2013;22(2):387–93.
29. Ruf M, Jensen R, Letko L, et al. Hemivertebra resection and osteotomies in congenital spine deformity. Spine. 2009;34(17): 1791–9.
30. Nakamura H, Matsuda H, Konishi S, et al. Single-stage excision of hemivertebrae via the posterior approach alone for congenital scoliosis. Spine. 2002;27(1):110–5.
31. Zhang J, Shengru W, Qui G, et al. The efficacy and complications of posterior hemivertebra resection. Eur Spine J. 2011;20:1692–702.
32. Yaszay B, O'Brien M, Shufflebarger HL, et al. Efficacy of hemivertebra resection for congenital scoliosis: a multicenter retrospective comparison of three surgical techniques. Spine. 2011;36(24):2052–60.
33. Lee CS, Hwang CJ, Kim DJ, et al. Feasibility of correction with instrumentation only in congenital scoliosis. Neurosurgery. 2014;74(1):35–41.
34. Goldberg CJ, Gillic I, Connaughton O, et al. Respiratory function and cosmesis at maturity in infantile-onset scoliosis. Spine. 2003;28:2397–406.
35. Kose N, Campbell Jr RM. Congenital scoliosis. Med Sci Monit. 2004;10(5):RA104–10.
36. Campbell RM, Smith MD, Mayes TE, et al. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. J Bone Joint Surg A. 2003;85:399–408.
37. Hell AK, Campbell RM, Hefti F. The vertical expandable prosthetic titanium rib implant for the treatment of thoracic insufficiency syndrome associated with congenital and neuromuscular scoliosis in young children. J Pediatr Orthop B. 2005;14(4):287–93.
38. Flynn JM, Emans JB, Smith JT, et al. VEPTR to treat nonsyndromic congenital scoliosis: a multicenter, mid-term follow-up study. J Pediatr Orthop. 2013;33(7):679–84.
39. Elsebai HB, Yazici M, Thompson GH, et al. Safety and efficacy of growing rod technique for pediatric congenital spinal deformities. J Pediatr Orthop. 2011;31(1):1–5.
40. Wang S, Zhang J, Qiu G, et al. Dual growing rods technique for congenital scoliosis: more than 2 years outcomes: preliminary results of a single center. Spine. 2012;37(26):E1639–44.

Evidence-Based Treatment of Spondylolysis and Spondylolisthesis

26

Athanasios I. Tsirikos and George Mataliotakis

Abstract

Spondylolysis and spondylolisthesis are common pathological conditions affecting the lumbosacral junction and resulting in a forward displacement of lumbar vertebra number 5 (L5) in relation to the sacrum. It is a frequent cause of back pain in children and adolescents and may also produce neurological deficits or physical disability in severe forms.

Many treatment modalities have been proposed over the years but there is still no consensus on the most effective. This chapter explores the evidence that underpin the current practice for treating the various degree of severity of the condition.

In isolated spondylolysis (pars defect <3 mm) affecting L5 the treatment of choice is in situ posterolateral fusion (PLF), whereas if the isthmic lesion involves levels proximal to L5 pars repair is recommended. In Grade I and II lumbosacral spondylolisthesis the preferred treatment is in situ non-instrumented PLF supplemented by brace support for 4 months. In Grade IV lumbosacral spondylolisthesis which does not produce global sagittal and spino-pelvic imbalance the recommended treatment is in situ un-instrumented PLF or instrumented posterior spinal fusion. In the presence of Grade IV or reducible Grade V lumbosacral spondylolisthesis, the preferred treatment options include either reduction of L5 and instrumented circumferential fusion or instrumented posterior spinal fusion using the transfixation lumbosacral technique. Finally, in fixed lumbosacral spondyloptosis (Grade V spondylolisthesis) the recommended treatment is an L5 vertebrectomy and L4/S1 anterior/posterior fusion (Table 26.1).

Keywords

Pars defect • Spondylolysis • Low grade spondylolisthesis • High grade spondylolisthesis • Spondyloptosis • Pars repair • Lumbosacral fusion • Sagittal imbalance • Children • Adolescents

Introduction

Spondylolysis refers to a defect of the pars interarticularis (Fig. 26.1). Spondylolisthesis represents the forward translation of one vertebral segment over the one beneath it (Fig. 26.2). It is the most common cause of back pain in children and adolescents. In severe forms it may be progressive and produce neurological deficits, global sagittal and spino-pelvic imbalance and severe physical disability. In the presence of persistently symptomatic or progressive deformities surgical treatment is often recommended. Since the 1960s, when the first surgical techniques were described, there is controversy on the best indicated operative management. The aim of the present chapter is to include the most relevant studies, in order to present with an evidence based approach the preferred treatment for each sub-group of this condition (Table 26.1).

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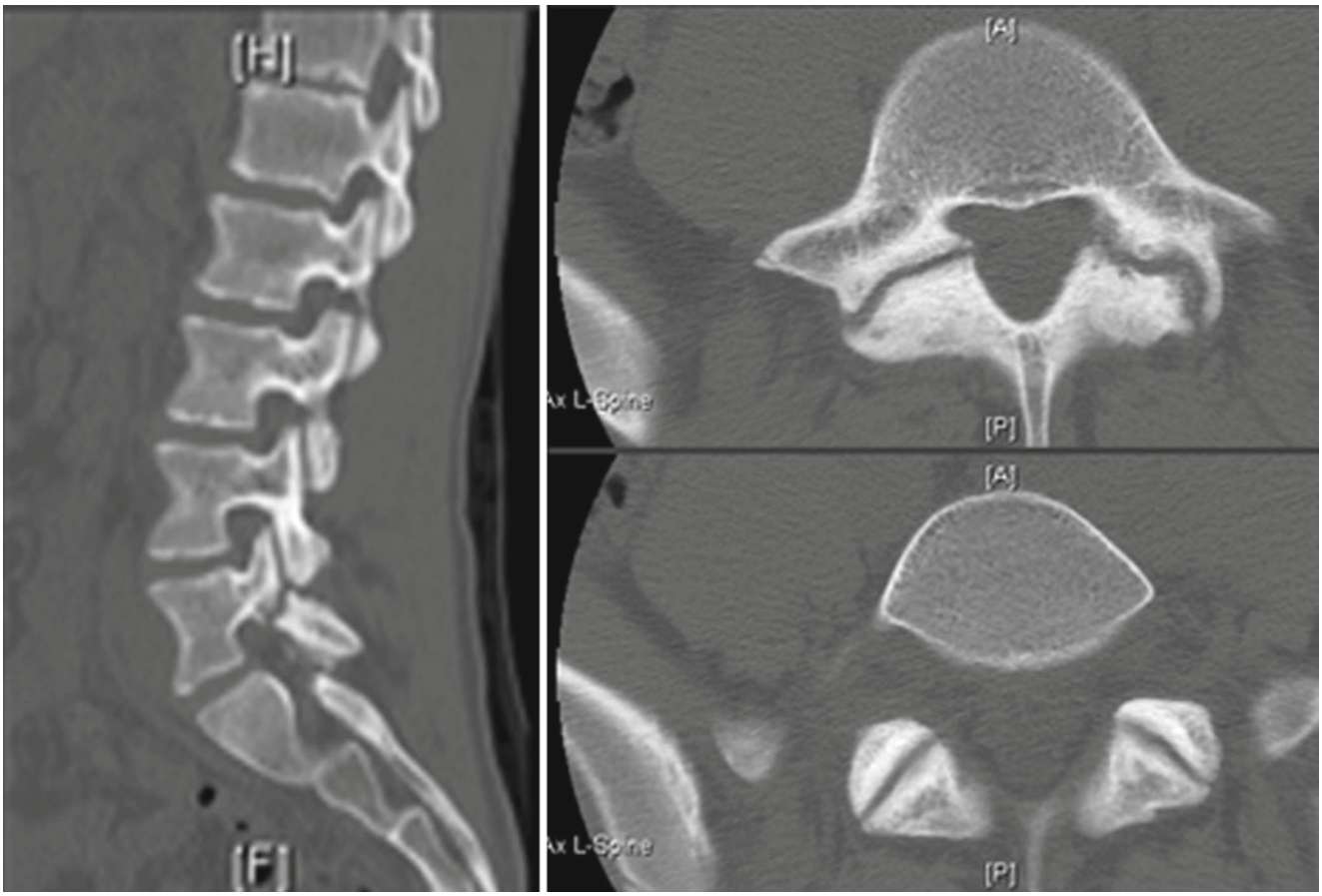

Fig. 26.1 CT Scan cuts showing a bilateral defect of the pars interarticularis. Notice the difference between the defect *top right* image and the facet joint (*bottom right* image)

Spondylolysis is considered a stress fracture of the pars interarticularis due to increased mechanical forces developing in that area. The most commonly affected level is L5 due to the thin structure of the pars and the unique anatomy of the L5/S1 articulation [1]. Predisposing factors to increased stresses across the L5 pars are: (a) small distance between the superior S1 and inferior L4 facets, which creates pinching of the L5 lamina in trunk hyperextension; (b) repetitive flexion-extension movements; (c) hyperlordosis of the lumbar spine [1].

Following a bilateral spondylolysis, the elongation of the disco-ligamentous restraints leads to subsequent spondylolisthesis. The long-term natural course of the condition in the presence of a spondylolysis with or without low grade spondylolisthesis is associated with disc degeneration and often spontaneous stabilization of the displaced segment which occurs between 13 and 17 years, with no further slip after skeletal maturity [1–3]. The tendency for segmental displacement progression is greatest around the pubertal growth spurt, with the girls showing more significant clinical and radiological deterioration than boys [4, 5].

The degree of slip is graded by dividing the superior endplate of S1 into quarters and observing how far the postero-inferior corner of the L5 vertebral body slips forward on S1 [6] such that grade I = 0–25 % slip, grade II = 25–50 %, grade III = 50–75 %, grade IV = 75–100 % and grade 5 > 100 % slip. The latter is often called spondyloptosis.

Spondylolysis manifests with lumbosacral mechanical pain induced by lumbar extension. 90 % of the segmental displacement has already occurred when the patient first presents in clinic [7]. Grade I and II isthmic spondylolisthesis may be symptomatic but the global sagittal balance and spino-pelvic parameters remain within normal limits. Apart from the isthmic lytic defect, the pain may originate from the paraspinal muscles and ligamentous structures, as well as the intervening disc due to stretching, degeneration and instability. There is no correlation between the subjective low back pain and the degree of disc degeneration or the number of affected levels [3].

High grade spondylolisthesis produces a global sagittal deformity affecting the spine and pelvis. There is always a level of dysplasia of the lumbosacral junction. At the time of diagnosis there may also be secondary changes of a long-standing underlying spondylolisthesis including trapezoidal deformation of the L5 vertebral body, rounding of the sacral

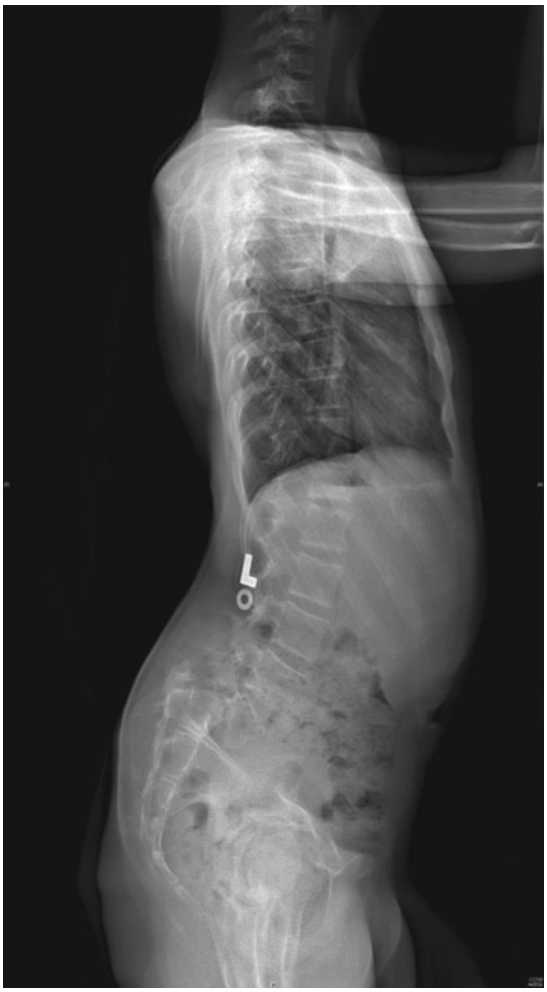

Fig. 26.2 A plain radiograph showing a severe spondylolisthesis of L5 over S1

Table 26.1 Summary of recommendation for the spondylolisthesis treatment

Type of deformity	Treatment recommendation	Level of evidence
Spondylolysis-early spondylolisthesis (<15 % vertebral displacement)	Conservative Surgical (if symptoms persist >6 months of conservative management L5/S1: In situ postero-lateral lumbosacral fusion Isolated and proximal to L5: isthmic repair	IV
Low grade spondylolisthesis (grades I & II)	Non-instrumented in situ postero-lateral lumbosacral fusion	III, IV
High grade spondylolisthesis/ spondyloptosis	Grade III: in situ instrumented posterior fusion Grade IV and reducible Grade V: reduction and instrumented circumferential fusion or Transfixation posterior fusion	III, IV
Fixed spondyloptosis (Grade V)	Staged reduction, decompression and instrumented circumferential fusion or L5 vertebrectomy and L4/S1 fusion	IV, V

dome (dome-shaped upper surface of the sacrum), facet insufficiency, and disc space narrowing [8]. The higher the degree of anterior vertebral displacement the greater the risk of neurological injury to the cauda equina or the exiting nerve roots at the affected levels.

Beyond grade III lumbosacral spondylolisthesis, in order to maintain global sagittal balance compensatory mechanisms create an increased lumbar lordosis and pelvic tilt. At grade V spondylolisthesis (spondyloptosis) the sacrum is markedly retroverted to a vertical position and the patient is compensating by flexing the hips and knees to maintain an upright posture. The resulting global sagittal malalignment leads to higher energy expenditure in both standing and walking [8, 9].

The clinical symptoms of spondylolisthesis include lumbosacral back pain, neurological deficits and development of a crouched gait. The back and leg pain is due to: (a) the pars defect; (b) degenerative changes in the discs and lumbosacral facets; (c) segmental instability producing increased strain and fatigue of the ligaments and paraspinal muscles; (d) global spino-pelvic imbalance causing further muscle stresses aggravated by hamstring tightness; (e) L5 radiculopathy or sacral root compression due to foraminal or central canal stenosis.

The descriptive classification systems currently used are based on the aetiology by Wiltse et al. [10] and on the degree of spondylolisthesis by Meyerding [6, 11]. The only radiological parameter with predictive value of progression is the percentage of primary vertebral displacement [5, 12]. The spondylolysis healing potential is classified by the Tokushima defect grading classification [13] into: (a) early: focal bony absorption or a hair-line defect; (b) progressive: wide defect and small fragments present; (c) terminal: sclerotic changes. The gold standard for diagnosis of spondylolysis is computed tomography (CT) scan. T2 weighted magnetic resonance (MRI) scans and/or SPECT give information regarding active lesions with healing potential or chronic lesions with higher rates of non-union if managed conservatively [14, 15].

Apart from the Meyerding classification, the severity of high grade spondylolisthesis is also defined by measuring the slip angle and lumbo-sacral kyphosis (Newman classification) [16]. The Newman classification is helpful to assess the position of L5 in relation to the sacral dome; it divides

the superior and anterior surfaces of the sacrum into 10 equal areas respectively. Depending on the position of the postero-inferior corner of L5 in relation to the sacral dome (x) and the position of the antero-inferior corner of L5 in relation to the anterior surface of the sacrum (y), the Newman score is expressed as (x) + (y) with the highest possible score being 10 + 10.

Spondylolysis-Early Spondylolisthesis (<15 % Vertebral Displacement)

The mainstay of treatment is conservative with the use of non-steroidal anti-inflammatory medication (NSAIDS), activity modification, physiotherapy and bracing. Physiotherapy focuses on relieving the extension stresses from the lumbosacral junction (hamstring and hip flexor stretching exercises), as well as working on core strengthening (deep abdominal muscles and lumbar multifidus strengthening exercises) [1, 13, 14, 17]. These modalities can achieve control of pain symptoms with an overall success rate of 86 % without and 89 % with the addition of an underarm brace [18].

Early active isthmic lesions may be treated as stress fractures by immobilization in a brace for 3–6 months [1]. The non-operative overall radiographic pars healing rate is 28 % (71 % for unilateral and 18.1 % for bilateral defects; 68.1 % for acute, 28.3 % for progressive and 0 % for terminal lesions) [18]. The healing rate is significantly greater in isthmic defects with less than 5 % displacement on the associated spondylolisthesis. The patients return to their normal activities once the symptoms resolve but they remain under 6 or 12 monthly follow up until skeletal maturity [1].

Persistent pain despite a 6-month conservative management (due to instability, adjacent disc disease or poor pars healing potential), progression to symptomatic spondylolisthesis and development of neurological deficits are the main indications to consider operative treatment [1, 14, 19–22]. Pars injection contributes to diagnosis confirmation of a symptomatic spondylolysis and to initial pain management but mainly provides prognostic information regarding a successful outcome following a surgical pars repair [23].

The surgical treatment options are following two main concepts; either repairing the pars defect or fusing the spondylolisthetic segment in situ.

Isthmic Repair

Indications for pars repair are: (a) patient age below 20 years; (b) isthmic defect less than 3 mm, (c) absence of significant disc degeneration, as evident on preoperative MRI scan; (d) no instability in lateral spinal flexion-extension view [19–26]. Presence of spina bifida occulta and a thin lamina are contra-indications for pars repair [19, 27]. The lamina should be at least 3 mm thick to provide adequate instrumentation purchase [20].

The theoretical advantages of pars repair are: (a) preservation of a motion segment (especially if the isthmic lesion affects levels proximal to L5), therefore minimizing the risk of adjacent segment disc degeneration; (b) no effect on spinal growth [20, 28–31]. Mild disc degeneration is not necessarily a contra-indication for pars repair as long as it is preoperatively confirmed by a local anaesthetic injection that the painful symptomatic area is the spondylolysis and not the adjacent disc [23, 30, 31]. Technically demanding positioning of the implants and need for instrumentation removal at a later stage according to the patient's symptoms or needs (for example, if recurrent back pain develops often in athletes who have high physical demands) are the potential disadvantages of pars repair [1].

The isthmic repair represents a true reconstructive procedure of the lumbar spine. This is accomplished by debridement of the non-union, refreshing of the pars defect bony edges and autologous bone grafting. Local stabilization is achieved either though application of a brace [32, 33] or most commonly by internal fixation of the free floating posterior fragment using various techniques. These may include: (a) trans-defect screws [20, 23, 24]; (b) a figure of 8 wire fixation [34]; (c) a hook-screw construct [27, 35]; (d) a pedicle screw U-shaped rod construct [28]. The repair techniques are not recommended for gaps greater than 3 mm and the exposure of the transverse processes may lead to bleeding and a higher risk of nerve root damage. The hook-screw technique overcomes the difficulty of fixation in the presence of dysplasia of the posterior arch and avoids crossing the isthmic defect with a screw. It also provides a larger surface area for bone healing and produces a more stable construct. Various modifications of the technique have been developed since the initial report [19, 20, 30, 35, 36]. More recently, the U-shaped pedicle screw/rod technique has promoted union of the defect by applying compression forces through the spinous process of the affected segment. Advantages of this technique include stable fixation and wide surface for bone grafting which allows early patient mobilization and no need for instrumentation removal as this is not interfering with the integrity of the facets.

In Situ Fusion

This is the mainstay of surgical treatment in bilateral L5 spondylolysis and associated low grade lumbosacral spondylolisthesis [1, 8, 37]. It consists of either a posterior or posterolateral fusion with the use of autologous bone graft

commonly harvested from the iliac crest. A postoperative lumbar support is recommended for a period of 4–6 months.

The advantages of the in situ fusion are: (a) there is a large area for bone grafting; (b) it can address a dysplastic spondylolisthesis; (c) it can be used to treat more severe spondylolisthesis; (d) the complete fusion addresses other potentially symptomatic areas of the affected level (L5/S1 disc and lumbosacral facets); (e) it can be performed with the use of autologous bone graft alone with no need for placement or removal of spinal instrumentation; (f) it involves a lesser risk of neurological injury compared to an instrumented fusion. The restriction of movement in the fused segment is the only disadvantage of this procedure.

The most commonly used technique to achieve an in situ lumbosacral fusion is that described by Wiltse and Spencer [38]. The fusion is performed using a bilateral paraspinal sacrospinalis muscle splitting approach. The sacral ala, posterior surface of the L5 transverse processes and L5/S1 facet joints are decorticated and the area is packed with iliac crest bone autograft (ICBG). The patients are mobilized on the 2nd or 3rd postoperative day with a soft lumbar brace applied for approximately 4 months. Previous reports have combined the technique of direct pars repair with interposition and in situ fusion of the L5/S1 facet joints using autologous bone in patients who already have degenerative changes affecting the L5/S1 disc [32, 33].

Discussion-Levels of Evidence

There is no evidence on superiority of conservative versus operative treatment for spondylolysis and low grade spondylolisthesis in regards to clinical outcomes (Table 26.2). The long term retrospective level IV studies of the Finnish group [2, 3, 5] regarding conservative versus in-situ fusion suggest: (a) that none of the patients for whom conservative treatment was decided required operative treatment at a later stage; (b) that there is significantly higher progression of slip in the first review but not in the final follow-up in the operated group; (c) the operated group had better clinical results and less pain at final follow up; (d) that fusion operations do not significantly increase the rate of disc degeneration in the adjacent disc level above after a mean postoperative follow-up of 13.8 years.

There is only level IV evidence assessing the clinical and radiological outcomes of the various pars repair techniques (Table 26.3). It seems that the rod-pedicle screw construct achieves the highest mean fusion rate (93.08 %). According to the same analysis the best clinical outcome has been achieved by the hook-screw Morsher-type repair (90.87 %); however this is a subjective result and the various authors used different criteria for evaluating their outcomes. The rod-pedicle screw technique produces 85 % good to excellent clinical results. No clear recommendation can be suggested on the basis of this level of evidence. These are complex and technically challenging procedures which require surgical expertise. It seems that every group of surgeons evolve their preferred method of pars repair with eventually improved clinical results [46] (Table 26.4).

Lumbosacral fusion is considered the surgical treatment of choice and is applied for the entire spectrum of grades of symptomatic spondylolisthesis. The studies which are mostly relevant to spondylolysis and mild spondylolisthesis are by Lenke et al. [48] and Helenius et al. [37] (Table 26.5). The fusion rate achieved after surgery varies from 50 % to 81.48 %; despite the relatively low fusion rates both studies report a high percent of good to excellent clinical outcomes.

Two author groups [32, 49, 50] compared the effectiveness of isthmic repair versus in situ fusion in the treatment of spondylolysis and low grade spondylolisthesis (Table 26.6). The conclusions of these studies regarding spondylolisthesis affecting L5/S1 are that: (a) both pars repair and fusion showed no significantly different radiological or clinical outcomes in the short-term follow-up; (b) the in situ fusion achieved superior results in the long term; (c) the theoretical advantage of motion-segment preservation in pars repair

Table 26.2 Level IV retrospective case series comparing surgical versus conservative treatment for spondylolysis

Authors	Patient number	Technique	F/U	Fusion rate (%)	Slips at initial F/U (%)	Slips at final F/U (%)	Good or excellent clinical outcome (%)	Criteria used for clinical outcome
Seitsalo et al. [2]	77	PF:49, PLF:28	159.6	88.30	19.00	19.00	94.80	descriptive
	72	conservative (bracing only in 7 pts)		NR	4	16.70	87.50	
Seitsalo et al. [4]	32	PF:20, PLF:10, AF:1, laminectomy: 1	NR	NR	NR	NR	NR	NR
	24	conservative		NR	NR	NR	NR	
Seitsalo et al. [5]	190	PF and PLF	14.8	NR	10 in the first year	NR	NR	NR
	82	conservative		NR	NR	NR	NR	

F/U follow-up in months, PF posterior fusion, PLF posterolateral fusion, AF anterior fusion, NR not reported

Table 26.3 Level IV retrospective case series assessing spondylolysis repair techniques. The studies were grouped on the basis of the technique used. Four groups were identified and their results were processed to demonstrate average values for each technique. Three techniques could not be fitted into those 4 groups and are presented separately at the end of the table

Authors	Patient number	Technique	F/U (months)	Fusion rate after first operation (%)	Good or Excellent clinical outcome (%)	Criteria used for clinical outcome
Buck [24]	16	Trans-defect, laminar compression screws (Bucks' technique)	NR	93.75	87.50	Descriptive
Pedersen and Hagen [39]	18		41	NR	83	Descriptive
Suh et al. [23]	10		NR	100	90	Descriptive
Ohmori et al. [21]	31		32.5	64.51	90.32	Henderson's
Hardcastle [26]	10		NR	90	90	Descriptive
Kim [22]	25		71	72	88	Kirkaldy-Willis
Menga et al. [40]	31		60	93.50	90	VAS
Snyder et al. [17]	16		13.2	89.60	94	Descriptive
Total	157		43.54	86.19	89.1	
Morsher et al. [27]	12	Hook-screw construct (Morsher type)	NR	NR	83.33	Descriptive
Hefti et al. [35]	33		41	73	79	Descriptive
Tokuhashi and Matsuzaki [36]	6		29.8	91.66	100	MacNab
Kakiuchi [30]	16		25	100	100	Descriptive
Ivanic et al. [19]	113		132	86.70	92	Descriptive
Total	180		56.95	87.84	90.87	
Bradford and Iza [34]	22	Figure-of-8 wire fixation (Nicol & Scott)	NR	90	80	Descriptive
Johnson and Thompson [41]	22		48	90.91	90.91	Descriptive
Total	44		48	90.46	85.46	
Gillet and Petit [28]	10	Rod-pedicle screws construct	35	100	70	Prolo score
Roca et al. [31][a]	19		30	92.31	79	Prolo score
Ulibarri et al. [29]	5		55.2	100	100	ODI, VAS, SRS-22
Altaf et al. [42][a]	20		48	80	90	ODI, VAS
Total	54		42.05	93	85	
Louis [43]	65	Butterfly-plate fixation	52.8	93.50	86	Descriptive
Hambly et al. [44]	13	Tension and wiring: intra-segmental (spondylolysis) & inter-segmental & one level fusion (spondylolisthesis)	20	100	92	Descriptive
Songer and Rovin [45]	7	Pedicle screws-cable	25.5	100	100	Prolo score

F/U follow-up in months, *VAS* visual analogue scale, *ODI* oswestry disability index, *SRS* scoliosis research society, *NR* not reported
[a]Level IV-prospective case series

cannot be proven in the clinical setting; (d) the isthmic repair does not prevent disc degeneration; (e) fusion should be performed in patients with preoperative MRI proven disc degeneration; (f) the repair has better results in younger patients; (g) the clinical and functional scores (VAS: Visual Analogue Scale, ODI: Oswestry Disability Index, SRS: Scoliosis Research Society) are significantly better in the fusion when compared to the direct repair group.

Many authors, therefore, suggest that the repair of spondylolisthesis at L5 produces less favourable results than lumbosacral in situ fusion [1]. In contrast, repair of the pars defect is indicated for L1 through to L4 spondylolytic defects, spondylolytic defects of multiple vertebral levels, and low-grade but reducible spondylolisthesis at levels cephalad to L5 with an intact intervertebral disc at the level of the displacement [8]. This is possibly due to: (a) anatomical reasons; (b) the fact that many lytic defects at L5 are the end stage of a developmentally weakened and elongated pars; (c) fusion for an isthmic defect cephalad to L5 would create a much stiffer lumbar spine.

Table 26.4 Level IV retrospective case series comparing different techniques for pars repair in spondylolysis treatment

Authors	Patient Number	Technique	F/U	Fusion rate after first operation (%)	Good or excellent clinical outcome (%)	Criteria used for clinical outcome
Guidici et al. [46]	7	Trans-defect, laminar compression screw (Bucks')	108	NR	28.50	Odom
	8	Figure-of-8 TP wiring (Nicol & Scott)		NR	62.50	
	37	Wiring with pedicle screws (modified Nicol & Scott)		NR	83.80	
Karatas et al. [47]	9	Laminar compression screw (Bucks')	21	100 after 6.5 months	88.89	McNab
	7	Hook-screw construct (Morsher type)	24	100 after 6.2 months	85.70	

F/U follow-up in months, *TP* transverse process, *NR* not reported

Table 26.5 Level IV retrospective case series assessing in situ lumbosacral fusion for spondylolysis and early grade spondylolisthesis

Authors	Patient number	Technique	Average follow up (months)	Fusion rate (%)	Good or excellent clinical outcome (%)	Criteria used for clinical outcome
Lenke et al. [48]	56	In situ PLF	NR	50	80	Descriptive
Helenius et al. [37]	108	In situ PF and PLF	240	81.48	86–92	Descriptive

F/U follow-up in months , PF posterior fusion, PLF posterolateral fusion, NR not reported

Table 26.6 Level III retrospective matched cohort studies comparing pars repair versus posterolateral fusion for spondylolysis/early spondylolisthesis

Authors	Patient number	Technique	F/U	Fusion rate (%)[a], pars repair/fusion	Good or excellent clinical outcome (%), pars repair/fusion	Criteria used for clinical outcome
Schlenzka et al. [50]	48	Figure-of-8 wiring (Nicol & Scott) versus PLF	54	61/88	87/97	ODI
Schlenzka et al. [49]	56	Figure-of-8 wiring (Nicol & Scott) versus PLF	180	43/89	64/87	Descriptive
Dai et al. [32]	46 (20 pars repair & 26 fusion)	Un-instrumented pars repair (Kimura) versus repair with facet fusion	50	95/92.3	95/92.3	Descriptive

F/U follow-up in months, *PLF* posterolateral fusion, *ODI* oswestry disability index
[a]Either uni – or bi-lateral

Recommendations

In the current literature, there is no solid evidence to lead to strong recommendation for the treatment of spondylolysis and/or mild spondylolisthesis (<15 % vertebral displacement). The existing studies are an amalgamation of different concepts, treatment of different levels of slips, inclusion of different age groups and application of same techniques regardless of the affected level.

The mainstay of surgical treatment in spondylolysis/mild spondylolisthesis affecting L5 is in situ lumbosacral fusion. However, adolescent patients with increased healing potential (as evident on SPECT), symptomatic (injection proven) spondylolysis (<3 mm) or early-reducible spondylolisthesis without degenerative changes affecting the discs/facets, dysplastic bony changes or spina bifida may be considered for direct pars repair. The threshold towards isthmic repair if the pathology involves the L1-L4 levels is lower.

Surgical treatment should be offered after at least 6 months of unsuccessful conservative management and should be accompanied: a) by brace immobilization of the lumbosacral junction for 4–6 months based on the surgical method and postoperative progress and b) by activity modification and limitations.

Low Grade Spondylolisthesis (Grade I & II)

Low-grade spondylolisthesis (Meyerding grades I–II) generally has a benign course and favourable prognosis and can often be managed non-operatively if the degree of vertebral displacement remains stable [51]. As with spondylolysis, surgical treatment is indicated for persistently symptomatic patients despite a structured conservative management or in the presence of a progressive spondylolisthesis. There is no ground for pars repair as this is not recommended for isthmic defects greater than 3 mm [1, 24]. Reduction of the spondylolisthesis carries a risk of neurological injury [51]. Therefore in highly symptomatic low grade lumbosacral spondylolisthesis surgical treatment is recommended and consists of in situ fusion between the displaced segment and the level below in order to prevent further anterior displacement and maintain global spino-pelvic balance.

Surgical treatment of spondylolisthesis should be considered in growing children with: (a) slip >30 % because of the high risk of further progression; (b) radiological evidence of displacement deterioration; (c) persistent back pain not relieved by conservative measures; (d) development of neurological deficits; (e) development of symptomatic hamstring tightness [52, 53].

Non-instrumented posterior and posterolateral fusion techniques have been previously used [1, 7, 21, 22, 24, 54]. The use of instrumentation is not required in young patients and postoperative support can be provided with a lumbar brace to enhance a bony fusion [1]. Instrumented fusion has been used in: (a) the presence of dysplasia and congenital deformities of the lumbosacral junction; (b) older/young adult patients as the fusion rates in this group are lower than in children; (c) in revision cases following initial uninstrumented in situ fusion which failed to achieve adequate stabilisation of the lumbosacral junction [37, 55, 56].

A reduction in the slip of about 1 % is noted between early and final follow-up and this is due to on-going bony remodelling [2]. In situ fusion can be associated with: (a) development of degenerative changes at the level of the spondylolisthesis and the level above the fusion; (b) neural foraminal stenosis which is relative to the severity of the slip; (c) mild muscular atrophy. With successful fusion of the lumbosacral junction in situ, tightness of the hamstrings resolves in most patients within 12–18 months from surgery [57].

Discussion-Levels of Evidence

Among the available level IV studies the mean fusion rate achieved when a posterolateral fusion is performed for spondylolisthesis is 86.93 % [37, 49, 58]. The fusion rate increases above 92 % in long term follow-up series with large experience on this technique [49]. A posterior fusion is less effective reaching mean fusion rates of 82.67 % (Table 26.7). In

Table 26.7 Level IV retrospective case series for grade I & II spondylolisthesis treatment

Authors	Patient number	Technique	F/U	Fusion rate after first operation (%)	Average slip (%) (preop/final follow up)	Good or excellent clinical outcome at final follow up (%)	Criteria used for clinical outcome
Helenius et al. [37]	108	PF: 29, PLF: 79	249.6	PF 10 (64), PLF 10 (87)	25.2/24.2	SRS-24: 94, ODI: 8.2	ODI, SRS-24
Schlenska et al. [49]	28	PLF	180	92.80	13.1/5.6	SRS-24: 96.4, ODI: 4.3, VAS: 15.5	ODI, SRS-24, VAS
de Loubresse et al. [58]	48	PLF	32	81	NR	88.00	Descriptive
Seitsalo [2]	77	PF: 49, PLF: 28	162	88.3	16.6/19.4	94.80	Descriptive
Remes et al. [52]	102	PF: 29, PLF: 73	252	89.2	27/26	86, ODI: 7.7 (PF:11.3/ PLF:6.3)	Descriptive, ODI
Jalanko et al. [59][a]	44	pars repair: 4, PF: 11, PLF: 29	204	77[b]	27.5/25.1	SRS-24: 94, ODI: 4.35, VAS: 18	ODI, SRS-24, VAS
Lenke LG et al. [48]	56	PLF	NR	50	NR	80.00	Descriptive

F/U follow-up in months, *PF* posterior fusion, *PLF* posterolateral fusion, *ODI* oswestry disability index, *SRS* scoliosis research society

[a] Level III-retrospective matched cohort study

[b] Direct pars repair excluded

the same line, the studies of the Finnish group (level III & IV) show that the SRS-24, ODI and VAS scores were better when the fusion was achieved through the posterolateral compared to the posterior approach [37, 49, 59]. However, the functional/clinical results are difficult to interpret, as different methods of assessment have been used among the available studies. There is no correlation between patient outcome (ODI) and abnormal lumbar MRI findings [52]. Also the development of a non-union following index surgery does not affect the final clinical outcome [59]. The presence of abnormal preoperative neurology may justify additional root decompression. Even though the difference has not reached significance, there is level III evidence that the pain in exertion and at rest is lesser with fusion alone than with fusion and decompression [58].

Conclusion-Recommendation

Posterolateral fusion in situ remains the mainstay of surgical treatment for children and adolescents with a grade I–II L5/S1 spondylolisthesis and produces satisfactory, long-lasting results [1, 4, 48, 51, 60]. The recommended technique involves a bilateral muscle-splitting approach through a midline skin incision, enabling fusion with the use of autologous bone harvested from the iliac crest across the transverse processes of L5 and the sacral ala [60]. The presence of neurological deficits associated with radiological evidence of neural compromise requires nerve root decompression which can be performed either through a posterolateral or a posterior approach to the spine.

High Grade Spondylolisthesis/ Spondyloptosis

The treatment of high grade spondylolisthesis (grade III–V) is surgical and the goals are to: (a) stop progression of segmental vertebral displacement; (b) improve/correct global sagittal balance and spino-pelvic parameters; (c) address nerve root compression or prevent neurological deterioration and permanent neural deficits.

The global sagittal balance is measured by the position of the C7-plumb line (C7PL); however, its restoration is unpredictable and only few studies have used it as a measure of assessing results postoperatively [61]. The improvement in the slip angle has been used instead as a parameter to demonstrate maintenance of the reduction achieved intra-operatively or the possible progression of the anterior displacement due to bending of the fusion mass in non-instrumented fusions.

If the spine remains in adequate global sagittal balance and spondylolisthesis reduction can be achieved only by patient positioning, a posterolateral fusion from L4 to S1 with the use of instrumentation without attempt to further correct the segmental displacement of L5 on the sacrum remains the preferred treatment of high-grade spondylolisthesis in children and adolescents [8]. Instrumentation needs to be used due to the high shear forces exerted across the lumbosacral junction, which may lead to non-union, bending of the fusion mass and delayed foraminal or canal stenosis [57, 62–66]. The fusion may be achieved through a posterolateral Wiltse approach [60] or through a posterior approach if central neural decompression is necessary [8].

Despite the fact that achieving a solid lumbosacral fusion remains the primary surgical goal, in the presence of severe global sagittal imbalance associated with retroversion of the pelvis, a vertical sacrum and lumbosacral kyphosis a degree of spondylolisthesis reduction is required to improve spino-pelvic alignment [66, 67]. The reduction also improves the load distribution across the instrumented segments leading to enhanced fusion rates [1]. However, the greater the degree of attempted spondylolisthesis reduction, the higher the risk of iatrogenic neurological injury affecting the L5 or sacral nerve roots [8]. The commonest injury is to the L5 root, as it gets stretched due to elongation and forward tilt of the sacrum [8, 68]. Partial reduction and instrumented fusion of spondylolisthesis may lead to increased pelvic incidence due to sacroiliac joint motion or sacral remodelling [67]. Reported complications following reduction are: (a) sacral insufficiency fractures; (b) bending of the sacrum; (c) spinal decompensation occurring above the fused levels [1]. Overall, the degree of slip reduction is less important in regards to clinical outcomes as long as stability of the lumbosacral junction and global spino-pelvic sagittal balance can be achieved [8, 69]. For those reasons, a wide decompression followed by an instrumented partial reduction and posterolateral fusion has been advocated for high-grade spondylolisthesis [8, 69].

In Situ Fusion Versus Spondylolisthesis Reduction

In situ un-instrumented lumbosacral fusion has been reported for spondylolisthesis of mean 67.02 % L5/S1 displacement (range: 60.7–80 %) [65, 70, 71] (Tables 26.8 and 26.9). There is level III evidence that after in situ fusion the best clinical results and the less slip progression at final follow up can be achieved with circumferential fusion (CF). The fusion rate with CF reaches approximately 96 % with a solid anterior fusion documented in 100 % of patients in previous series [65, 70]. Postoperative spica support for approximately 3–4 months is recommended until fusion becomes radiologically evident.

Spondylolisthesis reduction has been performed in the presence of anterior displacement of mean 76.03 % (range: 64–93 %) and there is level III evidence that the reduction and instrumented fusion achieves better radiological results

Table 26.8 Level III retrospective matched cohort studies assessing the treatment of high grade spondylolisthesis

Authors	Patient number	F/U	Fusion type	D	Graft	In situ/reduction	Fusion rate after first operation (%)	Average slip (%) (preop/final F/U)	Slip angle° (preop/postop)	Good or excellent clinical outcome at final F/U	ND	RS
Helenius et al. [70]	21	206.4	PLF	No	ICBG	In situ	86	61/67	NR	ODI: 9.7, VAS: 22.6	NR	1
	20	206.4	AF	No	ICBG	In situ	100	63/59	NR	ODI: 8.9, VAS: 24.5	NR	1
	25	206.4	CF	No	ICBG	In situ	96	71/70	NR	ODI: 3.0, VAS: 5.2	NR	0
Remes et al. [65]	21	207.6	PLF	No	ICBG	In situ	90.47	60.7/66.8	NR	ODI: 9.5	NR	NR
	22	207.6	AF	No	ICBG	In situ	100	62.6/59.9	NR	ODI: 8.0	NR	NR
	24	207.6	CF	No	ICBG	In situ	95.83	70.9/69.4	NR	ODI: 2.3	NR	NR
Poussa et al. [71]	11	177.6	CF	Yes	NR	In situ	100	80/78	27/23	ODI: 1.6, SRS-24: 103.9	no	2
	11	177.6	CF	Yes	NR	Reduction	100	93/57	34/20	ODI: 7.2, SRS-24: 90	no	2
Muschik et al. [72]	29	125	AF	No	ICBG	In situ	75.8	66/59	28/26	68.96 % symptom free	1	2
	30	67	CF	No	ICBG	Reduction	93.33	75/36	36/14	86.67 % symptom free	1	1
Shufflebarger et al. [73][a]	18	39.6	CF (PLIF)	Yes	ICBG	Reduction	100	77/13	35/4.3	NR	0	1
Poussa et al. [74]	11	56.5	CF	NR	NR	Reduction	NR	36.1[b]	NR	NR	0	NR
	11	59.8	CF, PLF, AF	NR	NR	In situ	NR	7.7[b]	NR	NR	0	0

F/U follow-up in months, *D* decompression, *ND* neurology at final follow up (permanent deficit), *RS* total revision surgery, *CF* circumferential fusion, *PLF* posterolateral fusion, *PF* posterior fusion, *ICBG* iliac crest bone graft, *AF* anterior fusion, *ODI* oswestry disability index, *VAS* visual analogue scale, *SRS-24* scoliosis research society, *NR* not reported

[a]Prospective cohort study

[b]Improvement at follow up

Table 26.9 Comparison of the results of different techniques in the treatment of high grade spondylolisthesis [65, 69–86]

Technique	Patient number	F/U	Fusion rate (%)	Slip improvement (%) ([a])	Slip angle improvement (°)[a]	ND (number, %)	RS (number, %)
Non-instrumented in situ fusion	423	154	86.58	0.20	–3[b]	2 (0.47 %)	24 (7.77)
Reduction and instrumented fusion	327	51.83	94.77	41.32	23.09	9 (2.75 %)	22 (6.96)
Instrumented PLF following reduction	52	37.4	90.21	35.18	25.57	3 (5.77 %)	5 (9.62)
Instrumented CF following reduction	242	54.08	97.57	44.74	23.4	6 (2.48 %)	16 (6.93)
Instrumented CF: fibula graft-TSIF (in situ or following reduction)	65	42.73	94.96	–	–	0	1 (1.54)
Instrumented CF: ALIF (in situ or following reduction)	174	127.79	96.12	–	–	3 (3.03 %)	7 (5.47)

F/U follow-up in months, *ND* Permanent neurological deficit at final follow up, *RS* Total revision surgery for symptomatic complications, *CF* circumferential fusion, *PLF* posterolateral fusion, *TSIF* trans-sacral inter-body fusion-fibula graft, *ALIF* anterior lumbar interbody fusion
[a]Of the patients reported
[b]Minus prefix represents worsening

compared to the in situ fusion [71–74]. In the available level III studies the average fusion rate following reduction and instrumented fusion is 97.78 %, the average slip reduction is 44.5 % (36–64 %) and the average slip angle improvement is 23.16° (14–30.7) [71–74]. It is unclear, whether the higher fusion rate recorded following an instrumented spondylolisthesis reduction compared to in situ fusion is due to correction of either L5 translation or angulation on top of the sacrum [75]. The clinical outcomes are difficult to assess among the previously reported level III studies due to different outcome measures used [65, 70–74] (Table 26.8).

Amongst all studies the average slip angle worsens by 3° at final follow up, whereas the slip improves by 0.18 % on average [65, 69–86]; (Table 26.9). The latter may be attributed either to casting postoperatively or to lumbosacral remodelling occurring during remaining skeletal growth [76]. Postoperative permanent neurological deficits have been reported in 2.75 % (0–14 %) of patients following spondylolisthesis reduction and fusion compared to approximately 0.47 % (0–4 %) after in situ fusion [67, 72, 77–82, 87]. The revision rate due to symptomatic complications is comparable among the 2 groups; 7.77 % and 6.96 % following in situ fusion and instrumented reduction with fusion respectively (Table 26.10).

CF Versus PLF Following Spondylolisthesis Reduction

There is no level III study comparing antero-posterior versus posterolateral instrumented fusion after spondylolisthesis reduction. Most of the authors recommend CF following reduction of a high grade spondylolisthesis [71–74, 77–80, 82–85, 87–89] (Table 26.10). This recommendation is supported by the fact that the average postoperative slip improvement following CF is better by 9.56 % as compared to instrumented PLF; 44.74 % (8–81 %) for CF versus 35.18 % (9–60 %) for PLF. The average rate of permanent postoperative neurological deficits is 2.48 % (0–10 %) following a CF compared to 5.77 % (0–14 %) after a PLF. The average rate of revision surgery due to symptomatic complications has been reported at 6.93 % (0–19 %) in CF versus 9.62 % (0–43 %) in PLF.

Fibula Trans-sacral Interbody Fusion Versus Anterior Lumbar Interbody Fusion

The bridging of the S1 and L5 vertebral bodies with the use of a fibular strut graft (Fibula trans-sacral interbody fusion (TSIF)) is a technique which can achieve fusion across the lumbosacral disc through a posterior approach to the spine [56, 83, 84, 86]. It can result in similar fusion rates to the anterior lumbar interbody fusion (ALIF) with a lower complication rate. There are no postoperative neurological deficits reported in the TSIF technique with a revision rate of 1.54 % in comparison to 3.03 % for neural damage and 5.47 % for need of revision surgery in ALIF procedures respectively (Table 26.10). There is no difference between the use of autologous or allograft fibula grafts in the TSIF technique [83]. Small numbers of patients and variations of the technique have been reported in the literature and this does not allow demonstrating a superiority of the fibula-TSIF when compared to the isolated ALIF technique in regards to biomechanical stability and fusion rates.

Table 26.10 Level IV retrospective studies assessing treatment in high grade spondylolisthesis

Authors	Patient number	F/U	Technique	D	Graft	In situ/ reduction	Fusion rate after first operation (%)	Slip (%) (preop/ final F/U)	Slip angle (°) (preop/postop)	Good or excellent clinical outcome at F/U	ND	RS
Mehdian et al. [85]	8	72	CF	Yes	ICBG	Reduction	100	86/5	48/7	ODI:6, VAS:1	0	0
Sasso et al. [56]	25	39	PLF & fibula graft-TSIF	Yes	Fibula	In situ	100	74/70	37/27	96 % satisfaction in SRS, VAS: 3.4	0	0
Helenius et al. [81]	21	206.4	PLF	No	ICBG	In situ	86	60.7/66.8	4.6/17.4	SRS: 89.7, ODI: 9.7, VAS: 22.6	0	1
	23	206.4	AF	No	ICBG	In situ	100	62.9/59.6	9.1/14.4	SRS: 93.2, ODI: 8.9, VAS: 24.1	1	3
	26	206.4	CF	No	ICBG	In situ	96	70.8/69.6	22.0/22.5	SRS: 100, ODI: 3, VAS: 5.5	0	1
Fabris et al. [88]	12	16	CF (posterior)	Yes	ICBG	Reduction	100	70.4/11.8	NR	91.67 %	0	1
Seitsalo et al. [66]	87	13.8	PLF (30), PF (54), AF(3)	No	ICBG/tibia	In situ	87	76/78	78/84	81.61 %	0	13
Hu et al. [78]	16	45.6	PLF (4 pts.: fibula strut graft also)	Yes	ICBG	Reduction	94	89/29	50/24	93.75 %	2	1
Roca et al. [86]	14	30	CF (posteriorly with fibula graft)	Yes	Fibula / ICBG	In situ	86	77/–	36/–	92.86 %	0	0
Molinari et al. [80]	11	41.4	PLF	No	ICBG	In situ	55	66.2/66.2	13.6/17.96	Function: 84 %, pain: 30 %, satisfaction: 83 %	0	2
	7	27.7	PLF	Yes	ICBG	Reduction	71	68.57/22.85	21/4.3	Function: 91 % , pain: 36 %, satisfaction: 93 %	1	3
	19	39.7	CF (16 ALIF, 3 PLIF)	Yes	ICBG	Reduction	0	70/24.21	31/4.7	Function:92 % , pain: 25 %, satisfaction:97.3 %	0	1
Hanson et al. [83]	17	55.2	CF (posterior + anterior approach)	No	Fibula	Reduction	94	77.6/46.47	32/18	ODI: 11.4, SRS: 37.3 + 14.1 (max: 45 + 15)	0	1
Bartolozzi et al. [77]	15	31.4	CF (titanium cage transfixation)	Yes	ICBG	Reduction	100	69.3/55.8	31.2/21.4	94.11 % satisfaction, SRS: 85.57	1	0
Smith et al. [84]	9	43	Fibula-TSIF	Yes	Fibula/ICBG	Reduction	100	78/70	41.2/21	100 % satisfaction, SRS: 73.94	0	0

Molinari et al. [71]	11	44.72	Posterior fusion + cast	No	NR	In situ	55	66.36/66.36	14.9	Function: 12.57/25, pain: 30 %, satisfaction: 84.3 %	0	0
	7	27.71	Posterior fusion	Yes	NR	Reduction	71	68.57/22.85	20.85/4.29	Function: 90.6 %, pain: 36 %, satisfaction: 94 %	0	1
	19	39.78	CF (ALIF or PLIF)	Yes	NR	Reduction	100	70/24.12	31.47/11.84	Function: 37.3 %, pain: 24.5 %, satisfaction: 97.3 %	1	1
Boachie-Adjei et al. [69]	6	42.6	Transfixation	Yes	ICBG	Reduction	100	89/80	62/28	SRS: 4.3	0	0
	25	36	CF (ALIF or PLIF)	Yes	ICBG	Reduction	100	73.2/13.6	34.2/−2.2	VAS: 2.1, low back outcome score: 43.7, SF-12: 41.3	0	1
	26	90	PLF or CF (ALIF in 5)	Yes[a]	ICBG	Reduction	100	17 ([b])	19 ([b])	NR	1	0
Saihan et al. [54]	23	24	PLF	No	ICBG	Reduction	96	Total: 64/38	64/48.6 ([c])	LBP: preop 19 pts./ postop 6 pts. , leg pain: preop 11 pts./ postop 1 pt	0	1
	21	24	CF (ALIF)	No	ICBG	Reduction	81			LBP: preop 22 pts./ postop 6 pts., leg pain: preop 8 pts./ postop 1 pt	2	4
Ruf et al. [82]	27	45	CF (ALIF or PLIF)	Yes	ICB	Reduction	100	74/11	LSA: 19.9/−8.5	SRS 30: 122.5/150	1	3
Grzegorzewski et al. [76]	21	153.6	PLF	No	ICBG	In situ	100	70.7/65.5	16.7/4.8	81 % had no pain postop	0	0

F/U follow-up in months, *ND* Permanent neurological deficit at final follow up, *RS* Total revision surgery for symptomatic complications, *CF* circumferential fusion, *PLF* posterolateral fusion, *PF* posterior fusion, *TSIF* trans sacral inter-body fusion, *ICBG* iliac crest bone graft, *NR* not reported, *VAS* Visual Analogue Scale, *LSA* lumbosacral segmental angle, *LBP* low back pain, *SRS* Scoliosis Research Society, *ODI* Oswestry Disability Index, *LBOS* low back outcome score, pts. patients

[a]Where required

[b]Improvement (preop-postop)

[c]L5 incidence

A variation of the fibula trans-sacral technique uses transfixation screws (placed through S1 into the antero-inferior region of the anteriorly displaced L5 vertebral body) either in isolation or as part of a longer construct extending proximally to L4, occasionally supplemented by iliac fixation in patients with grade IV or partly reducible grade V spondylolisthesis [69, 77]. The fixation is biomechanically more rigid compared to the fibula trans-sacral technique or the instrumented PLF [1]; however the presence of the transfixation screw along S1 into L5 doesn't produce direct interbody fusion across the lumbosacral disc into the body of L5.

Fusion Levels

Stress fractures of S1 (usually across the inferior aspect) or along the S1/S2 junction may occur in 13.69 % of the lumbosacral instrumented fusions ending to S1 due to increased shear forces and result in bending and anterior angulation of the S1 vertebral body [71, 73]. In the presence of inherent bone weakness across the sacrum which is primarily cancellous, caudal stability of the construct can be achieved with the use of supplementary iliac screws in order to widely distribute the increased stresses of the distal fixation [78, 79]. The addition of anterior column support provides a better biomechanical environment which can lead to increased fusion rates and maintenance of the surgically achieved correction [80].

In grade IV and V spondylolisthesis the fixation usually extends to L4 in order to: (a) achieve more stable proximal bony fixation; (b) prevent proximal junctional decompensation; (c) better control the low lumbar lordosis in view of restoring and maintaining spino-pelvic and global sagittal balance correction. Lamartina et al. [89] introduced the concept of the "stable zone rectangle", according to which all vertebrae within the rectangle should be included in the fusion. The stable zone rectangle consists of a square, whose area is identified by a segment of the horizontal line that passes through the centre of S2, which is intersected by the vertical line passing through the midpoint of the inferior end plate of L5 representing the gravity line and by the vertical line passing through the centre of the femoral head representing the ground reaction force.

Complications

In the early postoperative period, transient neurological deficits affecting the L5 nerve roots and producing radiculitis or neurapraxia have been reported to occur in 9.1–15 % of patients with high grade spondylolisthesis [56, 78, 80, 87]. If the symptoms appear immediate after surgery, these are often improved or resolved by partially releasing the spondylolisthesis reduction [73, 80]. Transient neurology may appear between 2 and 10 days postoperatively and often requires up to 6 months to resolve [73, 80, 87]. The number and rates of permanent neurological deficits among the different techniques are presented in Table 26.10. Permanent neurological damage has been reported to affect the L5 nerve root producing a foot drop which can be bilateral [72, 77, 79, 81, 87], the L4 nerve root [67] or cause an isolated sensory deficit [82].

The instrumented fusion following reduction shows higher average fusion rate than the non-instrumented in situ fusion (Table 26.10). CF presents an average fusion rate of 97.6 % (81–100 %) [72–74, 77, 79, 80, 82–85, 87–89], whereas PLF has achieved a fusion of 90.21 % (71.43–100 %) with greater risk of pseudarthrosis [69, 78, 80, 87]. Indication for reoperation, in an otherwise asymptomatic non-union, is progression of more than 30 % of L5/S1 displacement [72]. Apart from occasional wound infections, the complication which leads to revision surgery in the immediate postoperative period is transient nerve palsy due to nerve root distraction [71, 81]; partial reduction of the spondylolisthesis correction usually restores neurological function. Late symptomatic complications requiring revision surgery include non-union [66, 70–72, 81, 83], nerve root compression [66, 82, 89] and further slip progression [70, 73, 81] needing decompression and revision instrumented fusion accordingly.

Recurrence of pain symptoms long term is often associated with disc degeneration occurring at the segment proximal to the fusion regardless of the technique and approach used [65, 66, 70]. Muscle degeneration following fusion (involving the psoas and erector spinae) is noted during postoperative follow-up, especially following spondylolisthesis reduction with the use of instrumentation [56, 65].

Spondyloptosis

A lumbosacral spondyloptosis which still maintains some flexibility can be reduced using closed techniques under general anaesthesia to a grade IV spondylolisthesis by positioning the patient on the operating table with extension of the hips in order to produce anteversion of the pelvis and a more oblique position of the sacrum with correction of lumbosacral kyphosis. In this case, many of the techniques described for high grade spondylolisthesis may be also used in the treatment of spondyloptosis including the transfixation and instrumented reduction techniques [61]. For fixed spondyloptosis a staged reduction or a vertebral column shortening procedure (L5 vertebrectomy) may be required in order to achieve deformity correction and enhance fusion [61] (Table 26.11). There is only level IV and V evidence on these techniques and apart from the series presented by Gaines et al. [54, 61, 90] the numbers of patients reported are small. The technique

Table 26.11 Level IV retrospective studies for the treatment of spondyloptosis

Authors	Patient number	F/U	Technique	Fusion rate after first operation (%)	Slip correction (%)	Slip angle correction (%)	Good or excellent clinical outcome at final follow up	Criteria used for clinical outcome	ND	RS
Lehmer et al. (Gaines) [90]	16	46.8	Gaines procedure (L5 vertebrectomy and L4/S1 posterior instrumented fusion)	NR	NR	NR	High patient satisfaction (pain/function/appearance)	Descriptive	7/12/1	NR
Gaines et al. [9]	30	180	Gaines procedure (L5 vertebrectomy and L4/S1 posterior instrumented fusion)	93.33	NR	NR	High satisfaction rate	Descriptive	25/10/7	9
Karla et al. [91] ([a])	1	24	Modified Gaines (excision of inferior half of L5 body and L3-S1 posterior instrumented fusion)	100	100	80	ODI: 6	ODI	0	0
Wild et al. [94]	11	36	Staged reduction with external fixator and instrumented CF	100	NR	NR	Improvement in pain/ function/cosmesis	Descriptive	NR	NR
Suslu et al. [92] [a]	1	25	Posterior decompression, sacral dome resection, CF (PLIF) and instrumented L3-S1 posterior fixation	100	NR	60	No pain/no activity restriction	Descriptive	NR	NR
Ferris et al. [16]	3	126	Closed reduction-plaster cast-posterior fusion- anterior fusion after 3–6 months	100	52	59	Satisfactory clinical results	Descriptive	NR	NR

F/U follow-up in months, *ND* Neurological deficit preop/early postop/final follow up, *RS* Total revision surgery, *CF* circumferential fusion, *PLIF* posterior lumbar interbody fusion, *ODI* Oswestry Disability Index, *NR* not reported

[a]Level V case report

described by Gaines and Nichols in 1979 consists of an L5 complete vertebrectomy followed by circumferential instrumented fusion of L4 on the sacrum. Variations of this technique have been subsequently reported [85, 91, 92]. A staged reduction of L5 on S1 carries the advantage of lower complication rates when compared to the L5 vertebrectomy and it may be achieved either through closed techniques by casting or with the use of an external fixator [93] (Table 26.11).

Discussion-Levels of Evidence

There is no level I or II evidence study assessing the best treatment for high grade spondylolisthesis and spondyloptosis. There are few available studies with level III evidence (Tables 26.7 and 26.8). Even though the authors present matched cohorts of patients, they offer different surgical treatment for greater slips. A further combined analysis with all retrospective case series (level IV) is shown in Table 26.10.

Conclusion-Recommendation

In grade III spondylolisthesis the treatment of choice is in situ non-instrumented PLF followed by lumbar brace support for 4 months or in situ instrumented posterior fusion. In the presence of severe global sagittal and spino-pelvic imbalance, usually encountered in grade IV or reducible grade V spondylolisthesis, the treatment is reduction and instrumented CF/or transfixation PLF. Finally, in fixed spondyloptosis the recommended treatment is instrumented reduction, decompression and CF fusion or L5 vertebrectomy and L4/S1 fusion.

References

1. Tsirikos AI, Garrido EG. Spondylolysis and spondylolisthesis in children and adolescents. J Bone Joint Surg Br. 2010;92:751–9.
2. Seitsalo S. Operative and conservative treatment of moderate spondylolisthesis in young patients. J Bone Joint Surg Br. 1990;72:908–13.
3. Seitsalo S, Schlenzka D, Poussa M, et al. Disc degeneration in young patients with isthmic spondylolisthesis treated operatively or conservatively: a long-term follow-up. Eur Spine J. 1997;6:393–7.
4. Seitsalo S, Osterman K, Poussa M, et al. Spondylolisthesis in children under 12 years of age: long-term results of 56 patients treated conservatively or operatively. J Pediatr Orthop. 1988;8:516–21.
5. Seitsalo S, Osterman K, Hyvärinen H, et al. Progression of spondylolisthesis in children and adolescents. A long-term follow-up of 272 patients. Spine (Phila Pa 1976). 1991;16:417–21.
6. Meyerding HW. Spondylolisthesis. Surg Gynecol Obstet. 1932;54:371–7.
7. Osterman K, Schlenzka D, Poussa M, et al. Isthmic spondylolisthesis in symptomatic and asymptomatic subjects, epidemiology, and natural history with special reference to disk abnormality and mode of treatment. Clin Orthop Relat Res. 1993;297:65–70.
8. Cheung EV, Herman MJ, Cavalier R, et al. Spondylolysis and spondylolisthesis in children and adolescents: II. Surg Manag J Am Acad Orthop Surg. 2006;14:488–98.
9. Gaines RW (2005). L5 vertebrectomy for the surgical treatment of spondyloptosis: thirty cases in 25 years. Spine (Phila Pa 1976) ;30(6 Suppl):66–70
10. Wiltse LL, Newman PH, Macnab I. Classification of spondylolysis and spondylolisthesis. Clin Orthop Relat Res. 1976;117:23–9.
11. Nazarian S. Spondylolysis and spondylolytic spondylolisthesis. A review of current concepts on pathogenesis, natural history, clinical symptoms, imaging, and therapeutic management. Eur Spine J. 1992;1:62–83.
12. Huang RP, Bohlman HH, Thompson GH, et al. Predictive value of pelvic incidence in progression of spondylolisthesis. Spine (Phila Pa 1976). 2003;28:2381–5.
13. Morita T, Ikata T, Katoh S, et al. Lumbar spondylolysis in children and adolescents. J Bone Joint Surg Br. 1995;77:620–5.
14. Altaf F, Heran MK, Wilson LF. Back pain in children and adolescents. Bone Joint J. 2014;96B:717–23.
15. Payne 3rd WK, Ogilvie JW. Back pain in children and adolescents. Pediatr Clin North Am. 1996;43:899–917.
16. Newman PH. A clinical syndrome associated with severe lumbosacral subluxation. J Bone Joint Surg Br. 1976;47:472–81.
17. Snyder LA, Shufflebarger H, O'Brien MF, et al. Spondylolysis outcomes in adolescents after direct screw repair of the pars interarticularis. J Neurosurg Spine. 2014;21:329–33.
18. Klein G, Mehlman CT, McCarty M. Nonoperative treatment of spondylolysis and grade I spondylolisthesis in children and young adults: a meta-analysis of observational studies. J Pediatr Orthop. 2009;29:146–56.
19. Ivanic GM, Pink TP, Achatz W, et al. Direct stabilization of lumbar spondylolysis with a hook screw: mean 11-year follow-up period for 113 patients. Spine. 2003;28:255–9.
20. Jeanneret B. Direct repair of spondylolysis. Acta Orthop Scand Suppl. 1993;251:111–5.
21. Ohmori K, Suzuki K, Ishida Y. Translamino-pedicular screw fixation with bone grafting for symptomatic isthmic lumbar spondylolysis. Neurosurgery. 1992;30:379–84.
22. Kim YT, Lee H, Lee CS, et al. Direct repair of the pars interarticularis defect in spondylolysis. J Spinal Disord Tech. 2012. doi:10.1097/bsd.0b013e31827069e4.
23. Suh PB, Esses SI, Kostuik JP. Repair of pars interarticularis defect: the prognostic value of pars infiltration. Spine. 1991;16(Suppl):445–8.
24. Buck JE. Direct repair of the defect in spondylolisthesis: preliminary report. J Bone Joint Surg Br. 1970;52B:432–7.
25. Albassir A, Samson I, Hendrickx L. Treatment of painful spondylolysis using Morscher's hook. Acta Orthop Belg. 1990;56:489–95.
26. Hardcastle PH. Repair of spondylolysis in young fast bowlers. J Bone Joint Surg Br. 1993;75:398–402.
27. Morscher E, Gerber B, Fasel J. Surgical treatment of spondylolisthesis by bone grafting and direct stabilization of spondylolysis by means of a hook screw. Arch Orthop Trauma Surg. 1984;103:175–8.
28. Gillet P, Petit M. Direct repair of spondylolysis without spondylolisthesis, using a rodscrew construct and bone grafting of the pars defect. Spine. 1999;24:1252–6.
29. Ulibarri JA, Anderson PA, Escarcega T, et al. Biomechanical and clinical evaluation of a novel technique for surgical repair of spondylolysis in adolescents. Spine. 2006;31:2067–72.
30. Kakiuchi M. Repair of the defect in spondylolysis: durable fixation with pedicle screws and laminar hooks. J Bone Joint Surg Am. 1997;79A:818–25.
31. Roca J, Iborra M, Cavanilles-Walker JM, et al. Direct repair of spondylolysis using a new pedicle screw hook fixation: clinical and

CT-assessed study: an analysis of 19 patients. J Spinal Disord Tech. 2005;18(Suppl):82–9.
32. Dai LY, Jia LS, Yuan W, et al. Direct repair of defect in lumbar spondylolysis and mild isthmic spondylolisthesis by bone grafting, with or without facet joint fusion. Eur Spine J. 2001;10:78–83.
33. Kimura M. My method of filing the lesion with spongy bone in spondylolysis and spondylolistesis. Seikei Geka. 1968;19:285–96.
34. Bradford DS, Iza J. Repair of the defect in spondylolysis or minimal degrees of spondylolisthesis by segmental wire fixation and bone grafting. Spine. 1985;10:673–9.
35. Hefti F, Seelig W, Morscher E. Repair of lumbar spondylolysis with a hook-screw. Int Orthop. 1992;16:81–5.
36. Tokuhashi Y, Matsuzaki H. Repair of defects in spondylolysis by segmental pedicular screw hook fixation. A preliminary report. Spine. 1996;21:2041–5.
37. Helenius I, Lamberg T, Osterman K, et al. Scoliosis research society outcome instrument in evaluation of long-term surgical results in spondylolysis and low-grade isthmic spondylolisthesis in young patients. Spine. 2005;30:336–41.
38. Wiltse LL, Spencer CW. New uses and refinements of the paraspinal approach to the lumbar spine. Spine. 1988;13:696–706.
39. Pedersen AK, Hagen R. Spondylolysis and spondylolisthesis. Treatment by internal fixation and bone-grafting of the defect. J Bone Joint Surg Am. 1988;70:15–24.
40. Menga EN, Kebaish KM, Jain A. Clinical results and functional outcomes after direct intralaminar screw repair of spondylolysis. Spine. 2014;39:104–10.
41. Johnson GV, Thompson AG. The Scott wiring technique for direct repair of lumbar spondylolysis. J Bone Joint Surg Br. 1992;74:426–30.
42. Altaf F, Osei NA, Garrido E, et al. Repair of spondylolysis using compression with a modular link and screws. J Bone Joint Surg Br. 2011;93:73–7.
43. Louis R. Pars interarticularis reconstruction of spondylolysis using plates and screws with grafting without arthrodesis. A propos of 78 cases. Rev Chir Orthop Reparatrice Appar Mot. 1988;74:549–57.
44. Hambly M, Lee CK, Gutteling E, et al. Tension band wiring-bone grafting for spondylolysis and spondylolisthesis. A clinical and biomechanical study. Spine. 1989;14:455–60.
45. Songer MN, Rovin R. Repair of the pars interarticularis defect with a cable-screw construct: a preliminary report. Spine. 1998;23:263–9.
46. Giudici F, Minoia L, Archetti M, et al. Long-term results of the direct repair of spondylolisthesis. Eur Spine J. 2011;20(Suppl 1):115–20.
47. Karatas AF, Dede O, Atanda AA, et al. Comparison of direct pars repair techniques of spondylolysis in pediatric and adolescent Patients: Pars Compression Screw versus Pedicle Screw-Rod-Hook. J Spinal Disord Tech. 2012. doi:10.1097/bsd.0b013e318277cb7d.
48. Lenke LG, Bridwell KH, Bullis D, et al. Results of in situ fusion for isthmic spondylolisthesis. J Spinal Disord. 1992;5:433–42.
49. Schlenzka D, Remes V, Helenius I, et al. Direct repair for treatment of symptomatic spondylolysis and low-grade isthmic spondylolisthesis in young patients: no benefit in comparison to segmental fusion after a mean follow-up of 14.8 years. Eur Spine J. 2006;15:1437–47.
50. Schlenzka D, Seitsalo S, Poussa M. Operative treatment of symptomatic lumbar spondylolysis and mild isthmic spondylolisthesis in young patients: direct repair of the defect or segmental spinal fusion? Eur Spine J. 1993;2:104–12.
51. Smith JA, Hu SS. Management of spondylolysis and spondylolisthesis in the pediatric and adolescent population. Orthop Clin North Am. 1999;30:487–99.
52. Remes VM, Lamberg TS, Tervahartiala PO, et al. No correlation between patient outcome and abnormal lumbar MRI findings 21 years after posterior or posterolateral fusion for isthmic spondylolisthesis in children and adolescents. Eur Spine J. 2005;14:833–42.
53. Laurent LE, Osterman K. Operative treatment of spondylolisthesis in young patients. Clin Orthop Relat Res. 1976;117:85–91.
54. Gaines RW, Nichols WK. Treatment of spondyloptosis by two stage L5 vertebrectomy and reduction of L4 onto S1. Spine. 1985;10:680–6.
55. El Masry MA, El Assuity WI, El Hawary YK, et al. Instrumented in situ posterolateral fusion for low-grade lytic spondylolisthesis in adults. Acta Orthop Belg. 2005;71:83–7.
56. Sasso RC, Shively KD, Reilly TM. Transvertebral Transsacral strut grafting for high-grade isthmic spondylolisthesis L5-S1 with fibular allograft. J Spinal Disord Tech. 2008;21:328–33.
57. Newton PO, Johnston 2nd CE. Analysis and treatment of poor outcomes following in situ arthrodesis in adolescent spondylolisthesis. J Pediatr Orthop. 1997;17:754–61.
58. de Loubresse CG, Bon T, Deburge A, et al. Posterolateral fusion for radicular pain in isthmic spondylolisthesis. Clin Orthop Relat Res. 1996;323:194–201.
59. Jalanko T, Helenius I, Remes V, et al. Operative treatment of isthmic spondylolisthesis in children: a long-term, retrospective comparative study with matched cohorts. Eur Spine J. 2011;20:766–75.
60. Wiltse LL, Jackson DW. Treatment of spondylolisthesis and spondylolysis in children. Clin Orthop Relat Res. 1976;117:92–100.
61. Gaines RW. L5 vertebrectomy for the surgical treatment of spondyloptosis: thirty cases in 25 years. Spine. 2005;30(6 Suppl):66–70.
62. Boxall D, Bradford DS, Winter RB, et al. Management of severe spondylolisthesis in children and adolescents. J Bone Joint Surg Am. 1979;61A:479–95.
63. Schoenecker PL, Cole HO, Herring JA, et al. Cauda equinus syndrome after in situ arthrodesis for severe spondylolisthesis at the lumbosacral junction. J Bone Joint Surg Am. 1990;72A:369–77.
64. Harris IE, Weinstein SL. Long-term follow-up of patients with grade-III and IV spondylolisthesis: treatment with and without posterior fusion. J Bone Joint Surg Am. 1987;69A:960–9.
65. Remes V, Lamberg T, Tervahartiala P, et al. Long-term outcome after posterolateral, anterior, and circumferential fusion for high-grade isthmic spondylolisthesis in children and adolescents: magnetic resonance imaging findings after average of 17-year follow-up. Spine. 2006;31:2491–9.
66. Seitsalo S, Osterman K, Hyvärinen H, et al. Severe spondylolisthesis in children and adolescents. A long-term review of fusion in situ. J Bone Joint Surg Br. 1990;72:259–65.
67. Hresko MT, Hirschfeld R, Buerk AA, et al. The effect of reduction and instrumentation of spondylolisthesis on spinopelvic sagittal alignment. J Pediatr Orthop. 2009;29:157–62.
68. Petraco DM, Spivak JM, Cappadona JG, et al. An anatomic evaluation of L5 nerve stretch in spondylolisthesis reduction. Spine. 1996;21:1133–8.
69. Boachie-Adjei O, Do T, Rawlins BA. Partial lumbosacral kyphosis reduction, decompression, and posterior lumbosacral transfixation in high-grade isthmic spondylolisthesis: clinical and radiographic results in six patients. Spine. 2002;27:E161–8.
70. Lamberg T, Remes V, Helenius I, et al. Uninstrumented in situ fusion for high-grade childhood and adolescent isthmic spondylolisthesis: long-term outcome. J Bone Joint Surg Am. 2007;89:512–8.
71. Poussa M, Remes V, Lamberg T, et al. Treatment of severe spondylolisthesis in adolescence with reduction or fusion in situ: long-term clinical, radiologic, and functional outcome. Spine. 2006;31:583–90.
72. Muschik M, Zippel H, Perka C. Surgical management of severe spondylolisthesis in children and adolescents. Anterior fusion in situ versus anterior spondylodesis with posterior transpedicular instrumentation and reduction. Spine. 1997;22:2036–42.

73. Shufflebarger HL, Geck MJ. High-grade isthmic dysplastic spondylolisthesis: monosegmental surgical treatment. Spine. 2005;30(6 Suppl):42–8.
74. Poussa M, Schlenzka D, Seitsalo S, et al. Surgical treatment of severe isthmic spondylolisthesis in adolescents. Reduction or fusion in situ. Spine. 1993;18:894–901.
75. Transfeldt EE, Mehbod AA. Evidence-based medicine analysis of isthmic spondylolisthesis treatment including reduction versus fusion in situ for high-grade slips. Spine. 2007;32(19 Suppl): 126–9.
76. Grzegorzewski A, Kumar SJ. In situ posterolateral spine arthrodesis for grades III, IV, and V spondylolisthesis in children and adolescents. J Pediatr Orthop. 2000;20:506–11.
77. Bartolozzi P, Sandri A, Cassini M, et al. One-stage posterior decompression-stabilization and trans-sacral interbody fusion after partial reduction for severe L5-S1 spondylolisthesis. Spine. 2003;28:1135–41.
78. Hu SS, Bradford DS, Transfeldt EE. Reduction of high-grade spondylolisthesis using Edwards instrumentation. Spine. 1996;21:367–71.
79. Molinari RW, Bridwell KH, Lenke LG, et al. Complications in the surgical treatment of pediatric high-grade, isthmic dysplastic spondylolisthesis. A comparison of three surgical approaches. Spine. 1999;24:1701–11.
80. Molinari RW, Bridwell KH, Lenke LG, et al. Anterior column support in surgery for high-grade, isthmic spondylolisthesis. Clin Orthop Relat Res. 2002;394:109–20.
81. Helenius I, Lamberg T, Osterman K, et al. Posterolateral, anterior, or circumferential fusion in situ for high-grade spondylolisthesis in young patients: a long-term evaluation using the Scoliosis Research Society questionnaire. Spine. 2006;31:190–6.
82. Ruf M, Koch H, Melcher RP, et al. Anatomic reduction and monosegmental fusion in high-grade developmental spondylolisthesis. Spine. 2006;31:269–74.
83. Hanson DS, Bridwell KH, Rhee JM, et al. Dowel fibular strut grafts for high-grade dysplastic isthmic spondylolisthesis. Spine. 2002;27: 1982–8.
84. Smith JA, Deviren V, Berven S, et al. Clinical outcome of transsacral interbody fusion after partial reduction for high-grade l5-s1 spondylolisthesis. Spine. 2001;26:2227–34.
85. Mehdian SH, Arun R. A new three-stage spinal shortening procedure for reduction of severe adolescent isthmic spondylolisthesis: a case series with medium- to long-term follow-up. Spine. 2011;36:E705–11.
86. Roca J, Ubierna MT, Cáceres E, et al. One-stage decompression and posterolateral and interbody fusion for severe spondylolisthesis. An analysis of 14 patients. Spine. 1999;24:709–14.
87. Sailhan F, Gollogly S, Roussouly P. The radiographic results and neurologic complications of instrumented reduction and fusion of high-grade spondylolisthesis without decompression of the neural elements: a retrospective review of 44 patients. Spine. 2006;31:161–9.
88. Fabris DA, Costantini S, Nena U. Surgical treatment of severe L5–S1 spondylolisthesis in children and adolescents. Results of intraoperative reduction, posterior interbody fusion, and segmental pedicle fixation. Spine. 1996;21:728–33.
89. Lamartina C, Zavatsky JM, Petruzzi M. Novel concepts in the evaluation and treatment of high-dysplastic spondylolisthesis. Eur Spine J. 2009;18(Suppl 1):133–42.
90. Lehmer SM, Steffee AD, Gaines Jr RW. Treatment of L5-S1 spondyloptosis by staged L5 resection with reduction and fusion of L4 onto S1 (Gaines procedure). Spine. 1994;19:1916–25.
91. Kalra K, Kohli S, Dhar S. A modified Gaines procedure for spondyloptosis. J Bone Joint Surg Br. 2010;92:1589–91.
92. Suslu HT, Celikoglu E, Borekcı A, et al. One-stage surgery through posterior approach-for L5-S1 spondyloptosis. J Craniovertebr Junction Spine. 2011;2:89–92.
93. Wild A, Jäger M, Webb JK. Staged reposition and fusion with external fixator in spondyloptosis. Z Orthop Ihre Grenzgeb. 2001;139:152–6.

Evidence-Based Treatment of Spina Bifida

27

Emmanouil Morakis and James Wright

Abstract

Children with spina bifida manifest several congenital or developmental spine and limb deformities. Hip dislocation and scoliosis are among the most common musculoskeletal deformities in these patients. Surgical correction of these deformities has been advocated to improve function and quality of life. Current evidence shows that surgical reduction of hip dislocation does not improve their function or ambulatory capacity. Correction of scoliosis may improve sitting balance, but does not change the function or quality of life for these patients. If spine deformity correction is performed, combined anterior-posterior spinal fusion with instrumentation provides the best correction with the lowest rates of pseudarthrosis. Current evidence should be used to counsel patients and their families. Any surgical intervention should aim to better physical function that may also translate into improved quality of life.

Keywords

Spina bifida • Myelomeningocele • Hip dislocation • Scoliosis • Spine deformity • Evidence-based medicine • Surgical treatment • Complications • Quality of life • Treatment outcome

Introduction

Spina bifida is a neural tube malformation resulting from abnormal closure of the caudal part of the neural tube during embryonic development. Spina bifida is of variable severity, but the most common presentation, myelomeningocele, is characterized by a failure of formation of the dorsal vertebral elements, a defect of the overlying skin with exposure of the meninges and spinal cord. Dysplasia of the spinal cord and nerve roots leads to partial or complete paralysis of bladder, bowel, motor, and sensory function below the level of the lesion. Other lesions may co-exist that affect the neurologic function such as a Chiari malformation, cerebellar hypoplasia, hydrocephalus, syringomyelia or diastematomyelia.

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Children with spina bifida present with variable musculoskeletal deformities that can affect their ability to ambulate and their general physical function. The musculoskeletal deformities can be congenital (e.g. kyphosis, congenital scoliosis, teratologic hip dislocation, clubfoot, vertical talus) or acquired during development of the child as a result of muscle abnormalities, paralysis, joint contracture, bone deformity and decreased sensation of the lower extremities.

Orthopaedic management of these deformities must be part of a multidisciplinary approach. The presence of multiple medical co-morbidities makes orthopaedic treatment of these deformities challenging. The main goal of orthopaedic care is to maximize function and independence. While many factors affect physical function, the most important factor affecting the potential for ambulation is the level of neurologic involvement [1].

Correction of limb and spine deformities is frequently part of the orthopaedic management of patients with spina bifida. The correction of these deformities has a high rate of complications, such as wound infections, skin breakdown, allergic reactions to latex, pathologic fractures and nonunion after orthopedic surgery [2–4].

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Consistent with other aspects of paediatric orhopaedic, the results of orthopaedic interventions in patients with neuromuscular conditions, especially spina bifida, are assessed with instrumented gait analysis, measures of quality of life and functional benefits. Patient-oriented measures outcomes allow us to evaluate the function and satisfaction of patients with spina bifida undergoing orthopaedic surgery [5–7].

As discussed below, even satisfactory correction of structural deformities such as scoliosis or hip dislocation did not always improve, as expected, physical function and independence [2]. Since surgical treatment in patients with spina bifida can be a major task with high complication rates, it is important that patients and their families are provided with the best evidence to make the appropriate treatment choice.

Is Surgical Reduction of a Hip Dislocation Beneficial in Patients with Spina Bifida?

Muscle and sensory abnormalities in patients with spina bifida are associated with hip problems including hip contractures, subluxation or dislocation (Fig. 27.1). Hip deformities are not usually painful in patients with spina bifida. Thus, an important aim of correcting these deformities would be to maintain or improve function [8].

In the past, muscle balancing and hip relocation procedures were performed to address these deformities [9]. The functional outcome following these procedures was not always as expected. Postoperative complications, such as redislocation, hip joint stiffness or pathologic fractures, compromised the functional result [2, 10].

Studies suggested that these surgical interventions probably did not improve the ability to ambulate, reduce pain or the need for bracing [5, 11]. A systematic review of the literature by J. Wright identified only level 3 and 4 studies [12]. The recommendations of this review was that a dislocated hip, in above L4 level patient, should not be reduced surgically since it does not improve function and surgical treatment has a high risk of complications (30–45 %). The role of hip surgery in ambulatory patients with unilateral dislocation is uncertain.

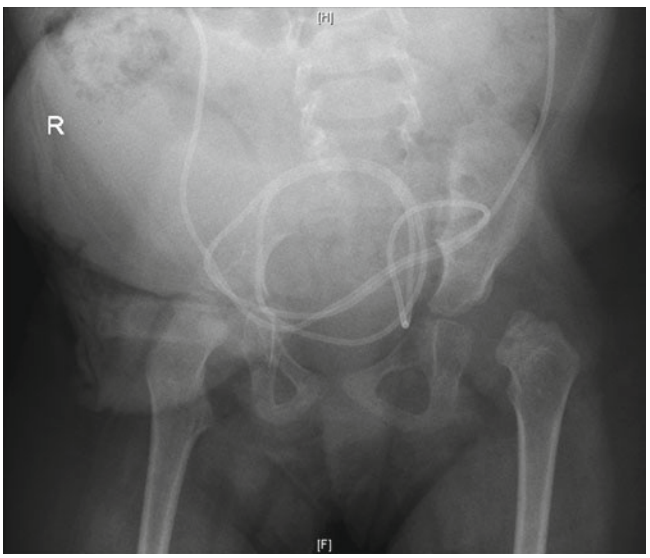

Fig. 27.1 A child with spina bifida developed bilateral dislocated hips

In a retrospective study of 55 patients with L3–L4 spina bifida, Fraser at al. evaluated the influence of several factors into their ability to walk [13]. Among these factors the neurologic level of the lesion was the most important determinant of their walking ability, while hip stability had no effect. They recommended that surgical procedures in an unstable hip are not beneficial.

Feiwell et al. compared 41 patients with spina bifida that had no operative treatment for the hip with 35 that received surgical treatment to reduce a dislocated hip [2]. They reported no relation between hip status, walking ability, requirement for bracing, hip pain or presence of hip contractures. They concluded that surgical relocation procedures are not indicated, the only surgical treatment should be release of joint contractures.

Bazih and Gross compared the ambulatory status between groups of patients that had surgery to relocate a dysplastic hip with a group of non-operative managed patients [14]. They reported 45 % failure rate to maintain hip reduction. There was no difference in the ambulatory status between the patients that had a reduced hip and the ones that had hip dysplasia (subluxation or dislocation). Fifteen out of twenty-eight (55 %) of the ambulatory patients with a lumbar level of function had one or both hips subluxed or dislocated. Hip reduction surgery did not provide any functional benefits in this group of patients.

Crandall et al. evaluated the ambulatory ability, patient-reported hip pain and presence of skin ulcers in 100 patients with spina bifida [15]. They found no correlation between the hip status (reduced vs. dislocated) and any of these factors. Seven out of thirteen patients that reported hip pain had reduced hips. Only one patient had relief from hip pain following hip reduction surgery, while several patients developed pain following hip surgery. The authors concluded that surgery to reduce a dislocated hip should not be expected to increase function.

In a retrospective comparative study, Sherk et al. compared the ambulatory function, bracing requirements and difficulty in sitting between 30 ambulatory patients with displaced hips and 11 ambulatory patients that had surgical reduction of a displaced hip [16]. The authors reported no difference between these two groups. The ability to walk depended on neurological level and the presence of contractures. They concluded that surgical hip reduction surgery provided little benefit, was costly and had high complication risk.

Fifty-two children with spina bifida affecting L3 and L4 were evaluated by Alman et al. [17]. They compared the function, range of motion, leg-length discrepancy and the metabolic energy consumption in two group of ambulatory patient that developed hip dislocation. One group of patients was treated non-operatively, while the other surgically. They found no significant difference in function between the operated and non-operative groups. Their redislocation rate was 31 % and in these patients function was particularly poor. In a small subset of surgically treated group of patients, who had gait analysis, gait was 30 % more efficient. This was though weakly related to better function. Thus the conclusion of this study was that successful surgical treatment had at best a marginal benefit.

Gabrieli et al. used gait analysis to assess the influence of hip dislocation or subluxation on gait asymmetry, since an asymmetrical gait would be less energy efficient [5]. They evaluated 20 patients with low lumbar spina bifida and unilateral hip subluxation or dislocation. They found no relation between gait symmetry and hip displacement. The walking speed of these patients was similar to the walking speed in low-lumbar patients without hip dislocation observed in a previous study from the same institution. The authors concluded that surgical reduction of the unilaterally unstable hip is not indicated.

The best evidence (Level III studies) that we have indicates that surgical reduction of a dislocated hip does not improve the walking ability, function, requirement for bracing, sitting ability or pain. On the contrary, surgical interventions to reduce a dislocated hip in patients with spina bifida have a significant complication risk and can deteriorate the functional status of a patient if they are not successful.

As already pointed out by Swaroop et al. a small group of patients with spina bifida, level L4 and below, that walk without assistive devices and have a unilateral hip subluxation or dislocation could benefit from a surgical intervention to reduce the hip [18]. These patients are typically home or community ambulators and may have gluteus medius function. A hip dislocation in these patients would cause significant gait asymmetry and decreased gait efficiency. Unfortunately there is no evidence yet in the literature that such an intervention would be translated into improved function.

Is Surgical Treatment of Scoliosis Beneficial in Patients with Spina Bifida?

Spine deformities are common in patients with spina bifida (Fig. 27.2) [19]. These deformities, scoliosis or kyphosis, can be either congenital or developmental. More than 80 %

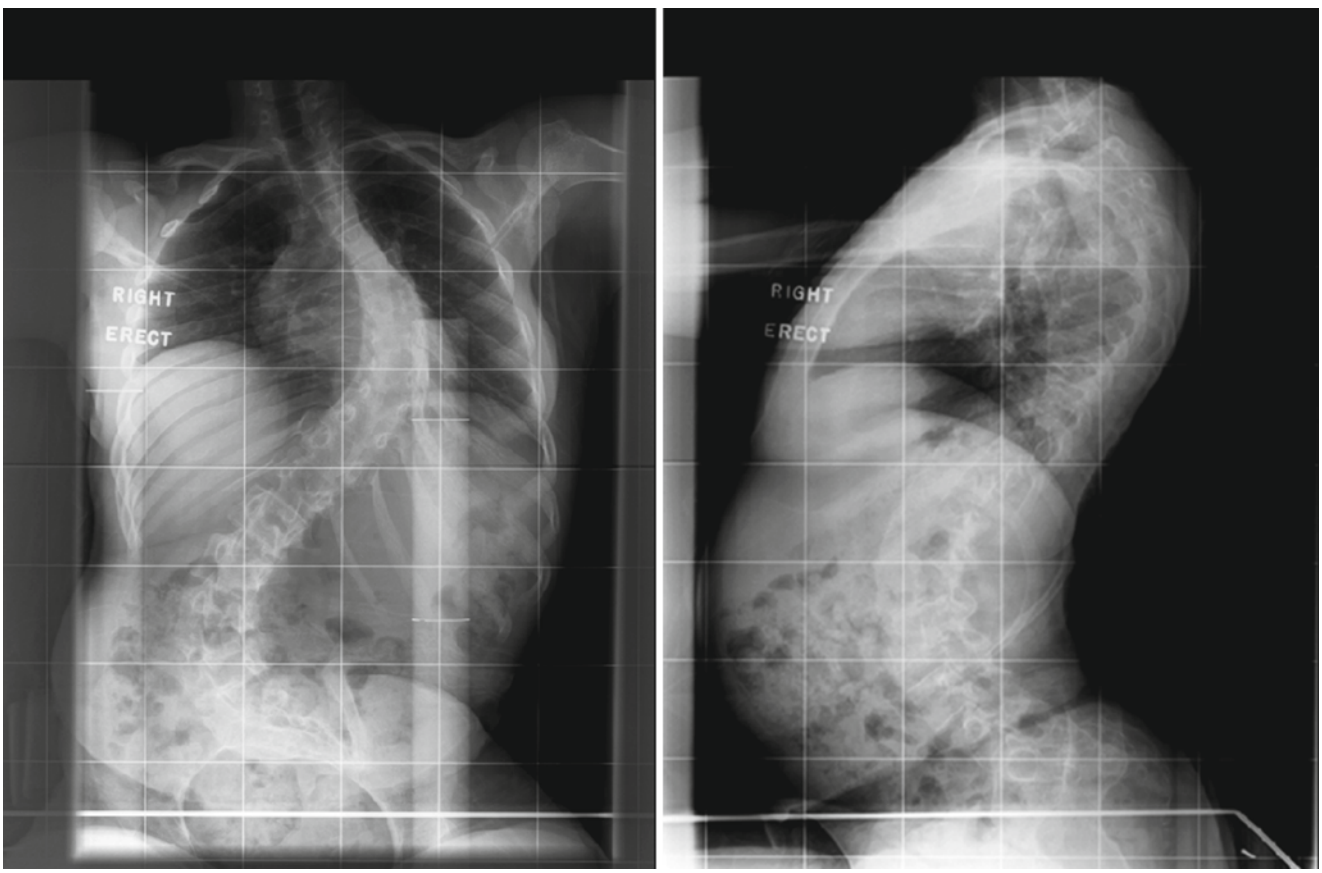

Fig. 27.2 Scoliosis in a patient with spina bifida

of the patients older than 10 years develop scoliosis [20]. The prevalence of scoliosis has been found to correlate with the level of the lesion and the level of the last intact laminar arch [19, 21]. Larger curves in non-ambulatory patients tend to progress faster during the early teenage years [22, 23].

Significant spinal deformity is associated with trunk imbalance and pelvic obliquity. Patients may have to use their hands to support their unbalanced trunk and loose function. Skin ulcers in the sacral area can sometimes be a problem for patients that use wheelchairs and have significant pelvic obliquity.

Surgical treatment to halt progression and correct deformities has been advocated with the hope that deformity correction will improve sitting imbalance; prevent pressure sores; improve mobility, transfer ability, physical function, activities of daily living and quality of life [24–26]. As with hip surgery, surgical treatment of spine deformities in patients with spina bifida has been associated with high complication rates. Post-operative wound infections, pseudarthrosis, fractures of the extremities and neurological deterioration have been reported [3, 4].

Several retrospective case series reported on the surgical outcomes of scoliosis correction on patients with spina bifida. Correction of spinal deformities has been achieved using different surgical techniques. How do these radiological and anatomical corrections correlate with the function and quality of life of these patients?

Unfortunately, the natural history of untreated scoliosis in patients with spina bifida is unknown. Most of the evidence available includes retrospective case series (Level IV). The only Level III study compared the health-related quality of life between two groups of patient with spina bifida and scoliosis [27]. In one group the patients were treated conservatively, while on the other they had instrumented fusion of the spine. The mean follow up of these patients was 14 years. No statistically significant difference was found between these groups in health-related quality of life, despite the operative group had improved coronal deformity with smaller average Cobb angle. Five out of eleven patients that had surgical treatment had lost their ability to walk. On the contrary, none of the walking patients that had non-operative management lost their ability to walk. The authors concluded that spine surgery to correct scoliosis "has no clear effect on health-related quality of life".

Similar findings were reported by other retrospective studies. Mazur et al. reviewed 49 patients with spina bifida and scoliosis, which were treated surgically [26]. There was no significant change post-operatively in overall function and activities of daily living. Sitting ability improved in these patients, but mobility (ambulation or mobility in a wheelchair) deteriorated in a significant number of patients (53 %). Despite the pelvic obliquity correction, most patients had persistent or developed problems with pressure sores.

In a retrospective review of 80 patients with spina bifida and scoliosis, Wai et al. evaluated the relationship of spinal deformity with overall physical function and self-perception using multivariable analysis [28]. Twenty-four patients had surgical treatment (anterior release, discectomy and posterior instrumentation to the pelvis with segmental unit rod fixation). Surgical treatment was correlated with improved Cobb angle and pelvic obliquity. Coronal imbalance of the spine was significantly correlated with sitting imbalance. Ambulation, hand function, self-perception and overall physical function were not found to be correlated with any aspect of spinal deformity. The only potential benefit of surgical correction of scoliosis according to this study would be improvement in sitting balance. As an alternative to surgery, chair modifications could be explored as an alternative to correct coronal imbalance and improve sitting function.

Schoenmakers et al. followed prospectively, for 18 months, ten patients with spina bifida that had surgical correction of their spine deformity. They evaluated their functional skills, the amount of caregiver assistance and their mobility, pre – and post – surgery. The authors reported complications in 8/10 patients (infection, loss of correction, increase of deformity above-below fusion mass). Following surgery the patients required less caregiver assistance on self-care. Their functional skills showed a trend of improvement at 18 months post-surgery, following an initial decline. Three out of four patients that were walking before surgery, had a deterioration of their ambulatory ability.

Muller et al. compared the ambulatory and functional status of 14 patients with spine bifida before and after surgical correction of scoliosis [29]. The mean follow-up was 3.4 years after surgery. Eight out fourteen patients had deterioration of their ability to ambulate. They reported no change overall in their function with activities of daily life, but 13/14 had loss in motor skills.

Thirty-nine skeletally mature patients with spina bifida and scoliosis were assessed by Kahanovitz and Duncan [25]. Their average follow up was 19.9 years. They evaluated the correlation between scoliosis, ambulatory ability and sitting balance. Fifteen patients had surgical stabilization of their spine deformity before the age of 16. The surgical complication rate was high. None of the surgically treated patients had better function after surgery and 7/15 had deterioration of their ambulatory status.

Two retrospective studies demonstrated that surgical correction of scoliosis in patients with spina bifida was associated with improved pulmonary mechanics and function, especially in severe deformities [30, 31]. The clinical significance of that improvement is not known.

The current evidence shows that surgical intervention can improve the coronal alignment of the spine in patients with spina bifida and scoliosis. Unfortunately a straighter and stiffer spine following surgery, in these patients does not

appear to improve their quality of life. On the contrary it may affect and compromise their ambulatory capacity and mobility. Scoliosis correction surgery may improve sitting balance and patients may require less caregiver assistance following surgery.

The decision to proceed with a surgical intervention to correct a scoliosis deformity in a patient with spina bifida is difficult. Surgery may be beneficial for a non-ambulatory patient with sitting imbalance. Since spine surgery in this group of patients has a high complication risk, non-operative means to improve sitting position and trunk support, such as chair modification, should be investigated. Patient that walk should be counseled that scoliosis correction surgery could compromise their ambulatory capacity.

Which Is the Best Surgical Technique to Correct Scoliosis in Patients with Spina Bifida?

Surgical correction of scoliosis in patients with spina bifida is challenging and has been associated with high complication rate [3, 4]. The rate of pseudarthrosis has been reported up to 76 % [32]. At the same time the spinal curves can be severe and rigid. The dysraphic spine makes instrumentation placement and purchase difficult.

Different surgical strategies and techniques have been described over the years. The main strategies include posterior only, anterior only or staged anterior and posterior spinal procedures. Which strategy and technique provides the best surgical outcomes and lowest complication rate?

Strategies and techniques have been changing over the years as new implants and technologies have been developing. Combined anterior and posterior instrumentation and fusion has provided the best surgical outcomes and lowest failure rates [12].

Osebold et al. reported on the positive outcomes of combined anterior and posterior fusion and instrumentation [33]. They reviewed the surgical outcomes and complications of patients with spina bifida treated for scoliosis correction in their institution. They compared the different surgical strategies they used over the years, including posterior spinal fusion alone, posterior fusion with Harrington instrumentation, combined posterior fusion with Harrington instrumentation with anterior fusion, anterior fusion with Dwyer instrumentation and finally combined posterior fusion with Harrington instrumentation with anterior fusion and Dwyer or Zielke instrumentation. Among these strategies, combined fusion with instrumentation gave the best correction with the lowest complications rates (pseudarthrosis, infection).

Three groups of patients with spina bifida and scoliosis that had surgical correction of their spine deformity with different techniques were reviewed by Mazur et al. [26]. The first group had posterior fusion and Harrington instrumentation, the second had anterior fusion and Dwyer instrumentation and the third group combined fusion and instrumentation. The patients that had combined fusion and instrumentation had better correction of their spine deformity with lowest rate of pseudarthrosis and hardware failure. The authors also reported better sitting balance in this group, since they achieved better correction of the pelvic obliquity and torso decompensation. There was no difference in the functional status and mobility between the three groups following surgery.

Five different surgical strategies to correct the scoliosis deformity in 41 patients with spina bifida were compared by Ward et al. [3]. The first group of patients had isolated anterior or posterior fusion with or without instrumentation. The other four groups had combined approach with anterior and posterior fusion, with or without anterior instrumentation, and posterior Harrington or Luque instrumentation. The combined approaches gave the best correction of the spine deformity comparing to isolated anterior or posterior. The authors did not find a significant difference amongst the combined approach strategies.

Parsch et al. compared three surgical techniques to correct scoliosis in 54 patients with spina bifida [34]. Thirty-three patients had posterior fusion and Cotrel-Dubousset or Spine-Fix instrumentation, twelve patients had anterior osteodiscectomy with posterior fusion and instrumentation and twenty-two patients had combined anterior-posterior fusion and instrumentation. Again these authors reported lower rates of pseudarthrosis and loss of correction with the combined approach. The amount of deformity correction was not significantly different amongst the groups, but the posterior only group of patients had smaller curves pre-operatively. The authors recommended combined approach for these patients, especially the ones with a thoracic level of paralysis. These patients had a higher risk of pseudarthrosis, hardware failure and loss of correction in mid-term follow-up.

In another multi-center retrospective study, Stella et al., reported the surgical outcomes of 29 patients with spina bifida and scoliosis [35]. Seven patients had only posterior fusion and instrumentation, three only anterior and nineteen had combined anterior –posterior approach using different types of instrumentation. Better correction was achieved with the combined approach, while overall rate of pseudarthrosis was 14 %.

Sponseller et al. and Basobas et al. advocated the use of an anterior only approach on selected patients with spina bifida and scoliosis [36, 37]. The anterior only approach has the advantages of lower risk of post-operative infection and fusion of fewer levels, which may allow more flexibility to the spine. Their indications include patients with a single major curve less than 80°, no syrinx or tethered cord, correction less than 66 % and kyphosis less than 30°.

Advances in spine instrumentation have improved fixation options and strength. The use of segmental instrumentation with pedicle screws allows secure fixation to the dysraphic and deficient posterior elements of the lumbar spine. Better fixation allows dramatic correction of severe and rigid curves in patients with neuromuscular scoliosis without the need of an anterior approach [38, 39]. All pedicle constructs have been shown to provide better correction with less bleeding comparing with hybrid constructs [40].

The use of pedicle screws in the dysraphic spine can be challenging. Pedicle orientation and landmarks can be altered [41]. The use of segmental fixation with pedicle screws may decrease the number of levels fused and stop fusion short of the sacrum. Preserving the lumbosacral junction and sacro-iliac joint allows more flexibility to the spine, which would be especially important in ambulatory patients. The use of segmental fixation with pedicle screws has allowed corrections comparable to the ones following combined anterior and posterior fusion with instrumentation.

Rodgers et al. reported the outcomes of 24 patients with spina bifida and scoliosis, which had posterior segmental instrumentation with pedicle screws [41]. Fourteen of these patients had posterior only surgery. One of these patients developed pseudarthrosis and required revision to anterior-posterior fusion and instrumentation. No cases with infection were reported. Most importantly, none of the ambulatory patients' function was affected. The overall correction of the spine deformity in the 24 patients was similar to other reports, where all patients had combined anterior-posterior fusion with instrumentation.

The effectiveness of posterior-only spinal fusion with segmental pedicle instrumentation was evaluated by Parisini et al. [42]. They compared the outcomes of 30 patients with spina bifida, which were operated for scoliosis. Ten patients had posterior spinal fusion and Harrington or Luque instrumentation; ten patients had combined anterior-posterior fusion and instrumentation; ten patients had posterior fusion with lumbar pedicle instrumentation. At 3 years of follow up, the last group of patients had significant improvement of their spine deformity both in the coronal and sagittal plane. The correction achieved was comparable to the group of patients that had combined anterior-posterior approach. The rate of pseudarthrosis was the lowest reported (10 %).

Currently, evidence shows that a combined anterior and posterior approach of fusion and instrumentation can provide the most reliable and lasting spine deformity correction with the lowest rates of pseudarthrosis. A selected group of patients may be candidate of an all-anterior approach. There is emerging evidence that an all-posterior approach with pedicle segmental instrumentation could provide equal effectiveness with the combined approach. Recommendations for treatment are listed in Table 27.1.

Table 27.1 Recommendations for treatment of spina bifida

Statements	Grade of recommendation
Hip dislocation should not be reduced surgically in patients with spina bifida	B
Unilateral hip subluxation or dislocation in patients with spina bifida L4 or below may be reduced surgically	I
Surgical correction of scoliosis may improve sitting balance	C
Patients should not have surgical correction of scoliosis to improve their quality of life	B
Patients should not have surgical correction of scoliosis to improve their ambulatory ability, mobility, activities of daily living and self-perception	C
Patients that have surgical correction for scoliosis should be treated with combined anterior-posterior spinal fusion and instrumentation	B
Patients that have surgical correction for scoliosis may be treated with posterior spinal fusion and segmental pedicle instrumentation	I

References

1. Asher M, Olson J. Factors affecting the ambulatory status of patients with spina bifida cystica. J Bone Joint Surg Am. 1983; 65(3):350–6.
2. Feiwell E, Sakai D, Blatt T. The effect of hip reduction on function in patients with myelomeningocele. Potential gains and hazards of surgical treatment. J Bone Joint Surg Am. 1978;60(2):169–73.
3. Ward WT, Wenger DR, Roach JW. Surgical correction of myelomeningocele scoliosis: a critical appraisal of various spinal instrumentation systems. J Pediatr Orthop. 1989;9(3):262–8.
4. Geiger F, Parsch D, Carstens C. Complications of scoliosis surgery in children with myelomeningocele. Eur Spine J. 1999;8(1): 22–6.
5. Gabrieli AP, Vankoski SJ, Dias LS, Milani C, Lourenco A, Filho JL, et al. Gait analysis in low lumbar myelomeningocele patients with unilateral hip dislocation or subluxation. J Pediatr Orthop. 2003;23(3):330–4.
6. Smith PL, Owen JL, Fehlings D, Wright JG. Measuring physical function in children with spina bifida and dislocated hips: the spina bifida hips questionnaire. J Pediatr Orthop. 2005;25(3):273–9.
7. Schoenmakers MA, Uiterwaal CS, Gulmans VA, Gooskens RH, Helders PJ. Determinants of functional independence and quality of life in children with spina bifida. Clin Rehabil. 2005;19(6): 677–85.
8. Beaty JH, Canale ST. Orthopaedic aspects of myelomeningocele. J Bone Joint Surg Am. 1990;72(4):626–30.
9. Heeg M, Broughton NS, Menelaus MB. Bilateral dislocation of the hip in spina bifida: a long-term follow-up study. J Pediatr Orthop. 1998;18(4):434–6.
10. Sherk HH, Ames MD. Functional results of iliopsoas transfer in myelomeningocele hip dislocations. Clin Orthop Relat Res. 1978;137:181–6.
11. Guille JT, Sarwark JF, Sherk HH, Kumar SJ. Congenital and developmental deformities of the spine in children with myelomeningocele. J Am Acad Orthop Surg. 2006;14(5):294–302.

12. Wright JG. Hip and spine surgery is of questionable value in spina bifida: an evidence-based review. Clin Orthop Relat Res. 2011; 469(5):1258–64.
13. Fraser RK, Hoffman EB, Sparks LT, Buccimazza SS. The unstable hip and mid-lumbar myelomeningocele. J Bone Joint Surg Br. 1992;74(1):143–6.
14. Bazih J, Gross RH. Hip surgery in the lumbar level myelomeningocele patient. J Pediatr Orthop. 1981;1(4):405–11.
15. Crandall RC, Birkebak RC, Winter RB. The role of hip location and dislocation in the functional status of the myelodysplastic patient. A review of 100 patients. Orthopedics. 1989;12(5):675–84.
16. Sherk HH, Uppal GS, Lane G, Melchionni J. Treatment versus nontreatment of hip dislocations in ambulatory patients with myelomeningocele. Dev Med Child Neurol. 1991;33(6):491–4.
17. Alman BA, Bhandari M, Wright JG. Function of dislocated hips in children with lower level spina bifida. J Bone Joint Surg Br. 1996;78(2):294–8.
18. Swaroop VT, Dias L. Orthopedic management of spina bifida. Part I: hip, knee, and rotational deformities. J Child Orthop. 2009; 3(6):441–9.
19. Piggott H. The natural history of scoliosis in myelodysplasia. J Bone Joint Surg Br. 1980;62-B(1):54–8.
20. Samuelsson L, Eklof O. Scoliosis in myelomeningocele. Acta Orthop Scand. 1988;59(2):122–7.
21. Trivedi J, Thomson JD, Slakey JB, Banta JV, Jones PW. Clinical and radiographic predictors of scoliosis in patients with myelomeningocele. J Bone Joint Surg Am. 2002;84-A(8):1389–94.
22. Muller EB, Nordwall A, Oden A. Progression of scoliosis in children with myelomeningocele. Spine (Phila Pa 1976). 1994;19(2): 147–50.
23. Marchesi D, Rudeberg A, Aebi M. Development in conservatively treated scoliosis in patients with myelomeningocele (patients of the years 1964–1977). Acta Orthop Belg. 1991;57(4):390–8.
24. McMaster MJ. Anterior and posterior instrumentation and fusion of thoracolumbar scoliosis due to myelomeningocele. J Bone Joint Surg Br. 1987;69(1):20–5.
25. Kahanovitz N, Duncan JW. The role of scoliosis and pelvic obliquity on functional disability in myelomeningocele. Spine (Phila Pa 1976). 1981;6(5):494–7.
26. Mazur J, Menelaus MB, Dickens DR, Doig WG. Efficacy of surgical management for scoliosis in myelomeningocele: correction of deformity and alteration of functional status. J Pediatr Orthop. 1986;6(5):568–75.
27. Khoshbin A, Vivas L, Law PW, Stephens D, Davis AM, Howard A, et al. The long-term outcome of patients treated operatively and non-operatively for scoliosis deformity secondary to spina bifida. Bone Joint J. 2014;96-B(9):1244–51.
28. Wai EK, Young NL, Feldman BM, Badley EM, Wright JG. The relationship between function, self-perception, and spinal deformity: implications for treatment of scoliosis in children with spina bifida. J Pediatr Orthop. 2005;25(1):64–9.
29. Muller EB, Nordwall A, von Wendt L. Influence of surgical treatment of scoliosis in children with spina bifida on ambulation and motoric skills. Acta Paediatr. 1992;81(2):173–6.
30. Banta JV, Park SM. Improvement in pulmonary function in patients having combined anterior and posterior spine fusion for myelomeningocele scoliosis. Spine (Phila Pa 1976). 1983;8(7):765–70.
31. Carstens C, Paul K, Niethard FU, Pfeil J. Effect of scoliosis surgery on pulmonary function in patients with myelomeningocele. J Pediatr Orthop. 1991;11(4):459–64.
32. Hull WJ, Moe, J.H., Winter, R.B. Spine deformity in myelomeningocele: natural history, evaluation, and treatment. J Bone Joint Surg Am. 1974;56:1767.
33. Osebold WR, Mayfield JK, Winter RB, Moe JH. Surgical treatment of paralytic scoliosis associated with myelomeningocele. J Bone Joint Surg Am. 1982;64(6):841–56.
34. Parsch D, Geiger F, Brocai DR, Lang RD, Carstens C. Surgical management of paralytic scoliosis in myelomeningocele. J Pediatr Orthop B. 2001;10(1):10–7.
35. Stella G, Ascani E, Cervellati S, Bettini N, Scarsi M, Vicini M, et al. Surgical treatment of scoliosis associated with myelomeningocele. Eur J Pediatr Surg. 1998;8(Suppl 1):22–5.
36. Sponseller PD, Young AT, Sarwark JF, Lim R. Anterior only fusion for scoliosis in patients with myelomeningocele. Clin Orthop Relat Res. 1999;364:117–24.
37. Basobas L, Mardjetko S, Hammerberg K, Lubicky J. Selective anterior fusion and instrumentation for the treatment of neuromuscular scoliosis. Spine (Phila Pa 1976). 2003;28(20):S245–8.
38. Modi HN, Suh SW, Song HR, Lee SH, Yang JH. Correction of apical axial rotation with pedicular screws in neuromuscular scoliosis. J Spinal Disord Tech. 2008;21(8):606–13.
39. Funk S, Lovejoy S, Mencio G, Martus J. Rigid instrumentation for neuromuscular scoliosis improves deformity correction without increasing complications. Spine (Phila Pa 1976). 2016;41(1):46–52.
40. Mattila M, Jalanko T, Puisto V, Pajulo O, Helenius IJ. Hybrid versus total pedicle screw instrumentation in patients undergoing surgery for neuromuscular scoliosis: a comparative study with matched cohorts. J Bone Joint Surg Br. 2012;94(10):1393–8.
41. Rodgers WB, Williams MS, Schwend RM, Emans JB. Spinal deformity in myelodysplasia. Correction with posterior pedicle screw instrumentation. Spine (Phila Pa 1976). 1997;22(20):2435–43.
42. Parisini P, Greggi T, Di Silvestre M, Giardina F, Bakaloudis G. Surgical treatment of scoliosis in myelomeningocele. Stud Health Technol Inform. 2002;91:442–7.

Evidence-Based Treatment of Spinal Trauma

28

Morag Harris, Peter Millner, and Odhrán Murray

Abstract

Numerous types and combinations of spinal column injury exist, with some rarely encountered. Consequently, clinicians, including subspecialists regularly involved in paediatric trauma must rely on evidence to supplement experience and guide practice. Conversely, there is a sparsity of high quality evidence and validated classification systems to help guide diagnosis, prognosis and management.

Children less than 8 years-of-age have significant biomechanical and anatomical spinal differences compared to their adult counterparts. Examples include, (1) a relatively large head size leading to an increased prevalence of proximal cervical injuries, and (2) the presence and subsequent closure of physeal growth plates can make the interpretation of radiographs more challenging. The three leading causes of spinal column injury in children are road traffic accidents, falls from height and non-accidental trauma. Spinal column injury is often accompanied by other and often significant trauma to the head, thorax and/or abdominal cavity.

The absolute requirement for radiation-based investigations such as CT scans must be considered carefully as the developing child's soft tissues are very sensitive to radiation. In this chapter, we explore the quality of evidence surrounding childhood spinal trauma from the occipital-cervical junction to the thoracolumbar spine, and outline the grade of recommendation to aid decision-making.

Keywords

Spinal column injury • Spinal cord injury • SCIWORA • Cervical • Thoracic • Lumbar • Anatomy • Biomechanics • Paediatric • Children

Introduction

Paediatric spinal trauma is thankfully rare, accounting for only 5 % of all paediatric traumas. Due to a child's unique anatomical development and the exposure to differing mechanisms of injury, the level and type of spinal injuries are distinctive when compared to adults. Children have a characteristically elastic spine, a large head size compared to their body, and distinct biomechanics, all of which contribute to a predominance for cervical spine injuries and higher rates of spinal cord injury without radiological abnormality (SCIWORA).

Special considerations need to be made when managing children with spinal trauma: pre-hospital immobilisation and imaging are two examples.

Unfortunately, there is a general lack of evidence regarding the management of paediatric spinal trauma. This chapter aims to identify the common causes of paediatric spinal injury along with the appropriate imaging techniques and management options based on the most up to date information available.

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Anatomy and Development

The developing spine is unique and, in general, children have immature bones that are more pliable and prone to injuries around the physis. Children under 8 years of age are more susceptible to upper cervical spine injuries (C1–C3) than older children because of unique aspects of their anatomical development. The relatively large head and weak nuchal muscles cause a shift in the fulcrum of movement in the upper cervical spine with maximum movement at C2–C3. As the child grows, this point of flexion shifts inferiorly and eventually ends at C5–C6 in adults [1–3]. This explains the increased number of C0–C2 injuries in very young children compared to their older peers who sustain more subaxial cervical and thoracolumbar injuries as their spine becomes more adult-like. Table 28.1 provides a summary of the anatomical differences found in children and their consequences in relation to injury.

Table 28.1 Anatomical and biomechanical differences between the adult and paediatric spine and the clinical significance in the traumatic setting

Anatomy and biomechanics	
Anatomical difference to adult spine	Consequence
Greater head to body size ratio Relative paraspinal muscle immaturity Shallow, horizontally oriented facet joints Greater ligamentous elasticity Incomplete vertebral ossification Paediatric spine reaches adult like maturity at around 8–9 years-of-age	• Higher rates of upper cervical spine injuries • Neutralisation of spinal alignment on spinal board required with torso boosting pad or occipital recess to prevent neck hyperflexion, airway closure and/or neurological deterioration • SCIWORA much more common • Higher proportion of TL junction injuries where rigid thorax meets elastic lumbar spine • Greater risk of intra-abdominal/ intra-thoracic injuries • Injury patters more adult like ≥8 years • Normal variants found on imaging
Nucleus pulposus has increased water and decreased collagen content	Greater elasticity and ability to dissipate force with less osseous fractures
Spinal cords ends at L3 at birth migrating caudally to end at L1 or L2 by adolescence	Spinal cord injury possible more distally secondary to trauma or surgical fixation
Vertebral physis/apophysis persists	Salter Harris I fractures possible with traction injuries (excellent prognosis with conservative management) Apophyseal fracture and herniation possible
Wedge shaped small vertebral bodies	Compression fractures more common Multilevel fractures more often
Developing body more sensitive to radiation	Avoid excess radiation (CT's) where possible; increased overall risk of cancer

Ossification

Ossification centers and synchondroses (Fig. 28.1) can be troublesome when interpreting imaging and are therefore important to take into consideration. The atlas has three ossification centers, one anterior arch and two posterior arches. The anterior and posterior ossification centers are separated by the right and left neuro-central synchondroses which usually fuse by 7 years of age [2]. The posterior synchondrosis separates the posterior ossification centers and usually closes by 3 years of age. Beware; the posterior synchondrosis can remain unfused giving the appearance of a fracture [4, 5].

The axis has a total of five primary ossification centers, one for each neural arch, one for the body, and two for the odontoid process. The odontoid is separated from the body of the axis by a cartilaginous physis that fuses between 3 and 6 years of age. It is situated below the level of C1–C2 facet joints and can therefore be mistaken for a fracture up to 11 years of age [2, 4]. The posterior arches fuse in the midline by 2–3 years and fuse to the body by 3–6 years of age.

Subaxial cervical, thoracic and lumbar vertebrae are all typical vertebrae and have a similar ossification pattern: one ossification center for the body and one for each neural arch. These fuse in the midline between 2 and 4 years and the neuro-central synchondroses close at 3–6 years. With aging, these ossification centers enlarge and the ratio of bone to cartilage increases. Apophyseal rings on the upper and lower surfaces of the vertebral bodies begin to ossify late in childhood and complete their fusion to the vertebral body by 25 years of age. Fractures of the apophyseal ring (Salter Harris type I) have been reported and are more common in the inferior endplates.

Cause of Injury

Spinal injury should be suspected in all children who are severely injured or involved in a high-energy accident. Children less than 8 years old are more likely to have spinal injuries resulting from automobile versus pedestrian accidents, motor vehicle collision (MVC), a fall from a height or non-accidental trauma (NAT) [2, 3, 6]. NAT is a common cause of injury in patients less than 2 years old and should not be overlooked in this age group [7]. Motor Vehicle Collisions (MVCs) are the major cause of injury

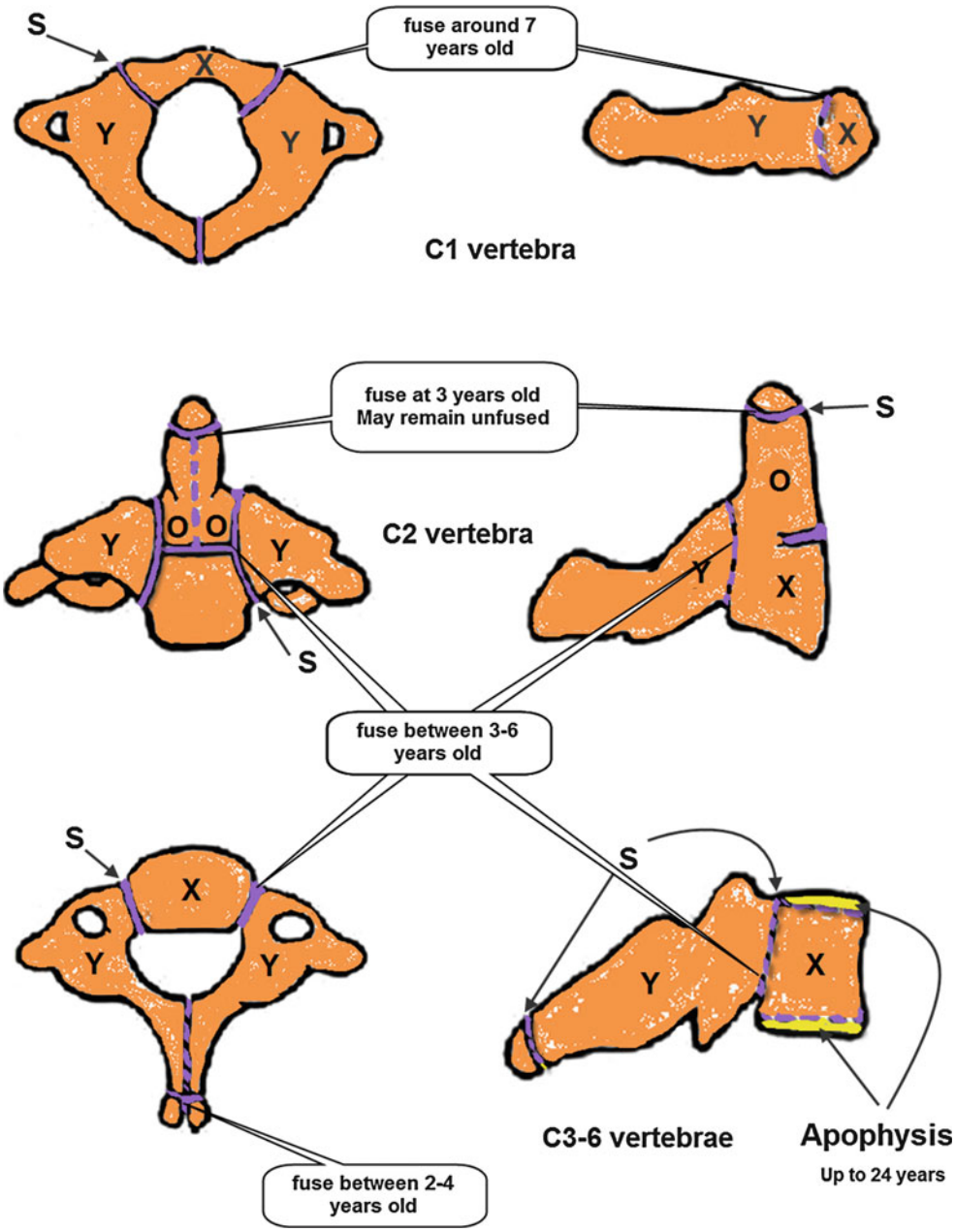

Fig. 28.1 Diagram of the ossification centres and synchondroses of the cervical vertebrae. Top images: Lateral and axial views of C1 and its three ossification centres. Middle images: Lateral and coronal views of the five ossification centres of C2. Bottom images: Axial and lateral views of a typical subaxial cervical vertebrae (thoracic and lumbar vertebrae have similar ossification patterns) X anterior ossification centre, Y posterior ossification centre, O odontoid ossification centre, *Purple dashed* line/gaps between ossification centres = synchondroses, *yellow line* = apophysis

across all age groups causing a high number of cervical and thoracolumbar fractures from inappropriate seat belt use. In older children and adolescents, one would expect to see more sporting injuries and motorcycle accidents. In general, serious head-injury and multi-injury trauma are commonly associated with spinal injuries. Patients with cervical spine injuries commonly have other associated traumatic injuries to the head, face, chest, abdomen or limbs [1, 8].

Important features to elicit from the history include conditions that predispose to spinal instability: e.g. Down's syndrome (15 % have atlanto-axial instability), previous cervical spine injuries, arthritis or previous spinal surgery.

Birth trauma resulting in spinal injury or cervical instability has a high mortality rate and is more often than not, fatal. The hallmark for upper cervical spinal cord injury at birth is apnoea with flaccid quadriplegia following either breech or prolonged forceps manipulation. This should be treated with immobilisation for presumed C-spine injury: the length of immobilisation remains arbitrary however [2, 7].

Pre Hospital Immobilisation

The aim of immobilisation is to prevent further damage or injury whilst the child is transported to an appropriate care

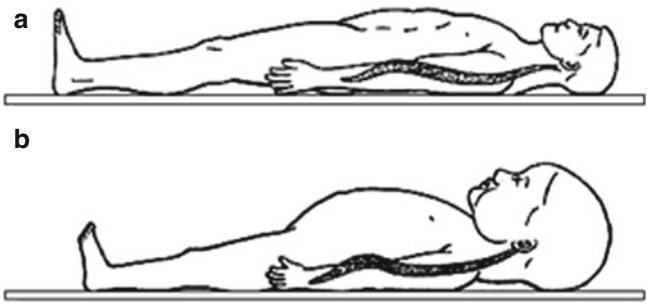

Fig. 28.2 The effect of relatively large size of a child's head. (**a**) The relatively large size of a child's head compared to their body results in cervical flexion when placed on a rigid spinal board (compared with adults (**b**)) (Reproduced by kind permission of Elsevier from Nypaver and Treloar [10])

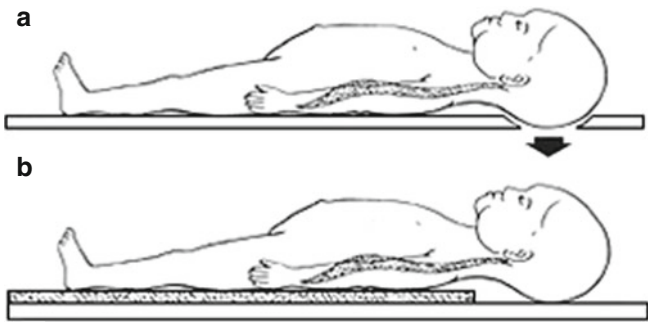

Fig. 28.3 Occipital recess and thoracic elevation allow for neutral alignment of the cervical spine. Use of **(a)** an occipital recess or **(b)** thoracic elevation (25 mm) allows for neutral alignment of the cervical spine (Reproduced by kind permission of Elsevier from Nypaver and Treloar [10])

facility. As with adults, all children with suspected spinal injuries should be immobilised at the scene as soon as possible. However, unlike adults, immobilisation of a child on a spinal board requires specific paediatric modifications.

Due to the relatively large size of a child's head compared to their body, evidence suggests that children less than 8 years of age should have thoracic elevation or an occipital recess to achieve better neutral alignment of their spine (25 mm on average) [9], (Figs. 28.2 and 28.3).

In general, the type of immobilisation should take into account the child's age and physical maturity. Ideally there should be a combination of a spinal board, rigid collar and torso tape. One must remember however that these may negatively influence the child's respiratory function, which should be monitored vigilantly [11, 12].

Safe removal of every patient from a spinal board must be an early priority after the primary survey and resuscitation phase. This prevents the complications of prolonged immobilisation as well as avoiding the agitation of a strapped down child.

Imaging

Figure 28.4 displays the criteria employed by the 2014 NICE guideline (CG176) for determining whether cervical spine imaging is required [13]. Although this is taken from a guideline that primarily manages children with a head injury, the algorithm is derived from the Canadian C-Spine Rule [14] which has successfully been used to reduce the amount of inappropriate radiation in children. There is some evidence that great caution is advised when applying these to children less than 11 years of age [15, 16].

Whilst is it important to consider the different anatomical and physiological differences in a growing child, health professionals must also be aware of the significant risks involved in exposing children to high doses of radiation. CT confers 90–200 times more thyroid radiation than standard plain films with an increased risk in children less than 4 [17]. Children have developing tissues which are significantly more sensitive to radiation and a longer life expectancy than adults, resulting in a greater time period to express such radiation damage.

As discussed previously, the spinal injury patterns that occur in young children differ from those in adults. The diagnostic tests and imaging required to exclude spinal injury also differ to those required in adults. Interpretation of paediatric radiographs requires a sound knowledge of age related ossification and anatomical variations and, more often than not, will require discussion with a musculoskeletal radiologist with specialised paediatric experience. (See Tables 28.2 and 28.3 for common normal variants found on cervical spine radiographs and their satisfactory measurements [2, 5, 18].)

Cervical spine imaging is not recommended in all children who have experienced trauma. It is vital, however, to image the whole spine in the presence of a fracture as there is a significant (8–24 %) risk of a non-contiguous fracture [5, 9, 19]. Unexplained hypotension should always raise suspicion of spinal cord injury.

Table 28.4 demonstrates the most up to date evidence available for imaging of the cervical spine in children [12].

When plain radiographs are indicated, an adequate cervical spine series must include:

1. Lateral cervical spine x-ray to include the base of skull and the junction of C7 and T1.
2. Antero-posterior cervical spine x-ray to include C2 to T1.
3. Adequate peg view if attainable*.

*Due to a child's age and difficulty with cooperation, current evidence suggests that open-mouth views only need to be obtained if the child is ≥9 years of age [4, 5, 12].

Unfortunately, there are no well-established, evidence-based recommendations for suspected thoracolumbar spine injuries following trauma. However, AP and lateral plain radio-

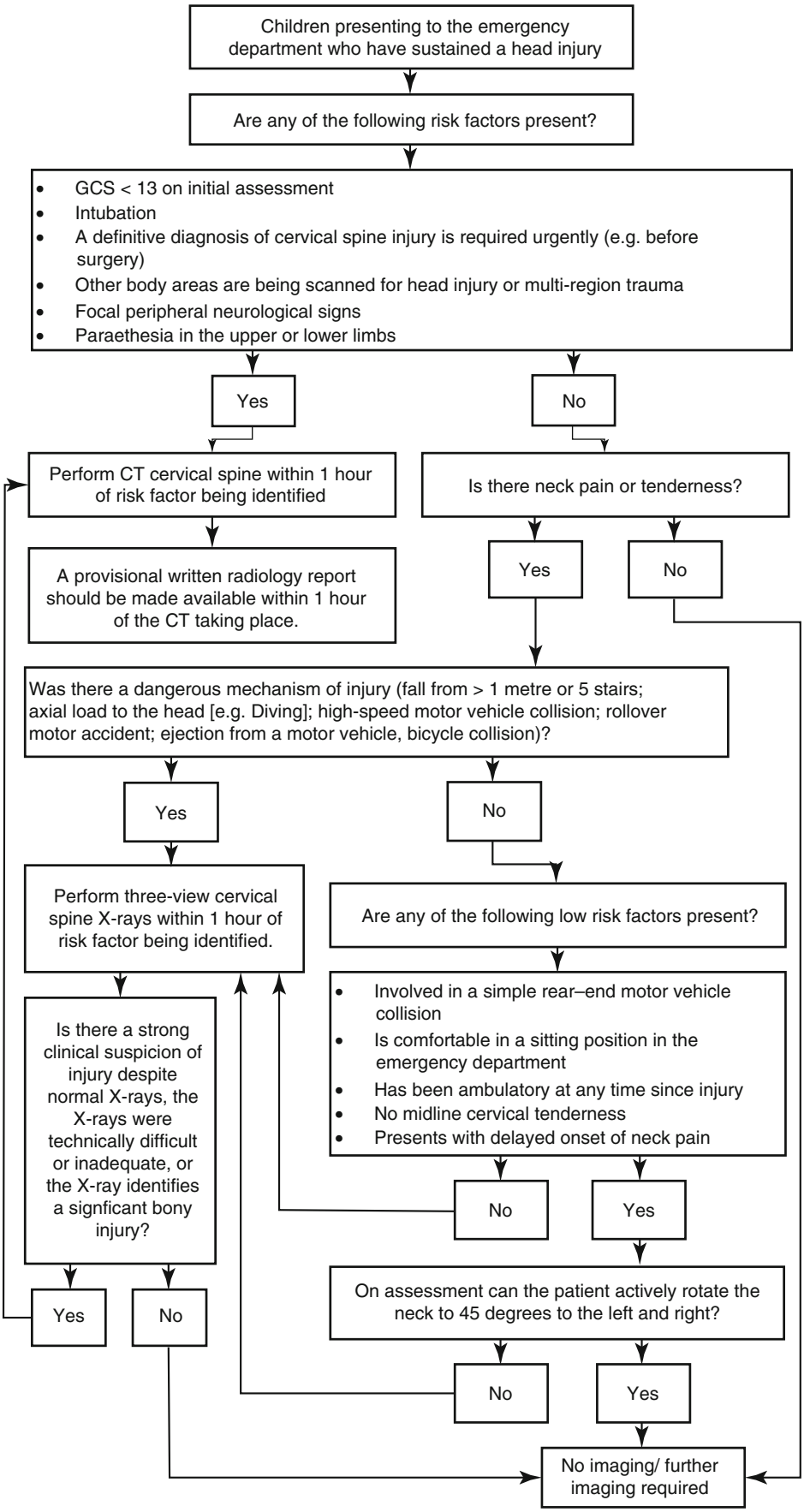

Fig. 28.4 NICE Clinical Guideline 176: Selection of children for imaging of the cervical spine (algorithm 4)

Table 28.2 Common normal variants found on cervical spine radiographs

Common normal variants	Comments
Absence of cervical lordosis	Seen in children up to 16 years of age
Prevertebral soft-tissue thickening	Can result from expiration, especially if the child is crying
'Pseudospread' of atlas	Results from a discrepancy between the "neural" growth pattern of the atlas and the "somatic" pattern of the axis
Increased atlanto-dental interval (ADI)	Potentially reflects incomplete ossification of the dens and laxity of the transverse ligament
Overriding arch of C1 above the odontoid	Can be mistaken for atlantoaxial instability, normal in 20 % of children <8 years
Incomplete ossification of posterior arch of C1	
Anterior wedging of vertebrae (up to 3 mm)	Can be present until endplates fuse.
Pseudosubluxation (anterior displacement of C2 on C3)	Seen as a normal variant in up to 40 % of young children
Odontoid and body of C2 synchondrosis mistaken for fracture	

Table 28.3 Acceptable measurements for cervical spine radiographs

Parameter	Normal value (mm)
C1 facet-occipital condyle distance	≤5
Atlanto-dens interval (ADI)	≤4 (3 mm in adults)
Space available for the cord (SAC)	≥14
Pseudosubluxation of C2 on C3 (most common misinterpretation)	≤4
Pseudosubluxation of C3 on C4	≤3
Retropharyngeal space	≤8 (at C2)
Retrotracheal space	≤14 (at C6, under age 15 years)

graphs are useful initial imaging techniques along with MRI for those children presenting with neurological deficit. Children who have sustained significant poly-trauma or high-energy accidents will commonly have CT imaging. A high clinical suspicion of spinal injury exists in the presence of: neurological deficit, high energy mechanism of injury, spinal step-off or crepitus, concurrent head or facial injuries, abnormal GCS and children who are non-compliant with examination [20].

Types of Injury

Cervical Spine Injuries

Occipital Condyle Fractures

These are rare fractures that require a high degree of suspicion. They can be associated with high-speed blunt traumas and may result in cranial nerve palsies and cervical spine instability. Use CT scan of the cranio-vertebral junction to promptly confirm the diagnosis.

A number of different classification systems have been developed over the years to help guide treatment options: reported evidence to date suggests that these classifications contribute little to management [2].

Below is the classification identified by Anderson and Montesano in 1988:

Type I – Impaction fracture.

Type II – Basilar skull fracture with extension into the condyle.

Type III – Avulsion of the alar ligament off the infero-medial portion of the condyle.

Appropriate choice of treatment is therefore based on the CT assessment of the fracture with or without MRI, rather than relying on fracture type alone [21, 22].

Management should involve occipito-cervical arthrodesis or halo fixation for those cases with occipito-cervical malalignment. If malalignment is not present, treat with immobilisation in a cervical orthosis with follow up imaging in a clinic setting [18, 23, 24]. C1–C2 transarticular screws, or C2 pedicle screws with rigid loops and plate or rods, have been successful in pediatric patients as young as 11 months old [2].

Atlanto-Occiptal Dislocation (AOD)

This is a very unstable injury and usually fatal. It results from sudden deceleration leading to hyperflexion and cranio-vertebral separation (Fig. 28.5). This causes disruption of the alar ligaments, articular capsules and tectorial membrane (ligamentous injury). Level I evidence recommends CT imaging to determine the condyle-C1 interval (CCI) [12] but requires a high index of suspicion: there is frequent association with head and face injuries following high-energy traumas [2]. Surgical stabilisation with occiput to C2 fusion or halo immobilisation is required [25].

Atlas Fractures (Jefferson Fractures)

These injuries result from rupture or avulsion of the transverse ligament caused by axial compression. The ring of C1 is commonly broken at more than one site. Greater than 6.9 mm of combined lateral overhang of the lateral masses on AP radiograph is diagnostic. As mentioned previously, up to 6 mm of pseudospread may be seen in children under 7-years (secondary to faster growth of atlas compared with axis). These require immobilisation in a cervical orthosis or halo device for up to 6 months. Halo traction followed by halo immobilisation maybe necessary if there is >6.9 mm or widening of lateral masses.

Table 28.4 Recommendations of radiological investigations

Level I evidence	• Use CT to determine the condyle-C1 interval (CCI) for children with suspected atlanto-occipital dislocation (AOD)
Level II evidence	• Cervical spine imaging is not recommended in children who are >3 years of age, who have experienced trauma and who: – Are alert – Have no neurological deficit – Have no midline cervical tenderness – Have no painful distracting injury – Do not have unexplained hypotension – Are not intoxicated • Cervical spine imaging is not recommended in children who are <3 years of age, who have experienced trauma and who: – Have a Glasgow Coma Scale (GCS) >13 – Have no neurological deficit – Have no midline cervical tenderness – Have no painful distracting injury – Are not intoxicated – Do not have unexplained hypotension – Do not have motor vehicle collision (MVC), a fall from a height > 10 ft, or non-accidental trauma (NAT) as a known or suspected MOI • Cervical spine radiographs or CT are recommended for children who have experienced trauma and who do not meet either set of criteria above • Use three-position CT with C1–C2 motion analysis to confirm and classify the diagnosis in atlantoaxial rotatory fixation (AARF)
Level III evidence	• Use AP, lateral and open mouth cervical spine radiographs or CT to assess the cervical spine • Use CT scan with attention to the suspected level of injury to exclude occult fractures or to evaluate regions not adequately visualized on plain radiographs • Flexion and extension cervical radiographs or fluoroscopy are recommended to exclude gross ligamentous instability when cervical instability remains a possibility • Use MRI to exclude spinal cord or nerve root compression, assess ligamentous integrity, or provide information regarding neurological prognosis

Odontoid Epiphysiolysis

The neuro-central synchondrosis of C2 may not fuse completely until age 7 years and can be a possible site of injury in young children (odontoid synchondrolysis). Lateral C-spine radiographs will reveal the odontoid process to be angulated anteriorly, and rarely, posteriorly [12]. Treatment is closed reduction and external immobilisation with a Minerva or halo for approximately 10 weeks (80 % fusion rate). Confirm healing on removal of the immobilisation device with flexion/extension radiographs [5]. Primary surgical stabilisation has a limited literature base and plays more of a role when closed external immobilisation is unable to maintain alignment of the odontoid atop the body of C2 [2].

Atlanto-Axial Injuries

The atlanto-axial joint is primarily stabilised by the transverse ligament and secondarily by the alar and apical ligaments. In children, it is more common to find rupture of the ligament as opposed to avulsion of the ligamentous attachments in adults. Current evidence supports early posterior atlanto-axial arthrodesis (successful fusion with C1–C2 transarticular screw fixation has been reported) with instrumentation for ligamentous disruptions and immobilisation in cervical orthosis or halo device for bony avulsions [18]. Surgery should be reserved for patients with non-union and persistent instability after 3–4 months of immobilisation. Imaging should be used pre-operatively to determine appropriate screw size, entry points and trajectories, before consideration of a Goel-Harms Procedure [2, 18].

Atlanto-Axial Rotatory Fixation (AARF)

AARF is not unique to children but more common during childhood. It presents with a 'Cock-robin' appearance, leaving the child unable to turn his/her head past the midline often causing pain. There is almost always a normal neurological examination although altered sensation radiating down the arms is sometimes encountered [26–28]. It may present following minor trauma, or non-traumatically e.g. following pharyngeal surgery or upper respiratory tract infection. Non-traumatic subluxation of the atlanto-axial joint caused by inflammation of the adjacent tissues is known as Grisel syndrome (Fig. 28.6).

Four types have been described by Fielding and Hawkins [2]:

- Type I (most common)
  - Intact transverse ligament.
  - Unilateral anterior rotation of atlas pivoting around the odontoid (no anterior shift).
  - ADI is normal, fixation within normal ROM.
- Type II
  - Disruption of the transverse ligament.
  - Unilateral anterior subluxation of the atlas with the pivot at the contralateral C1–C2 facet.

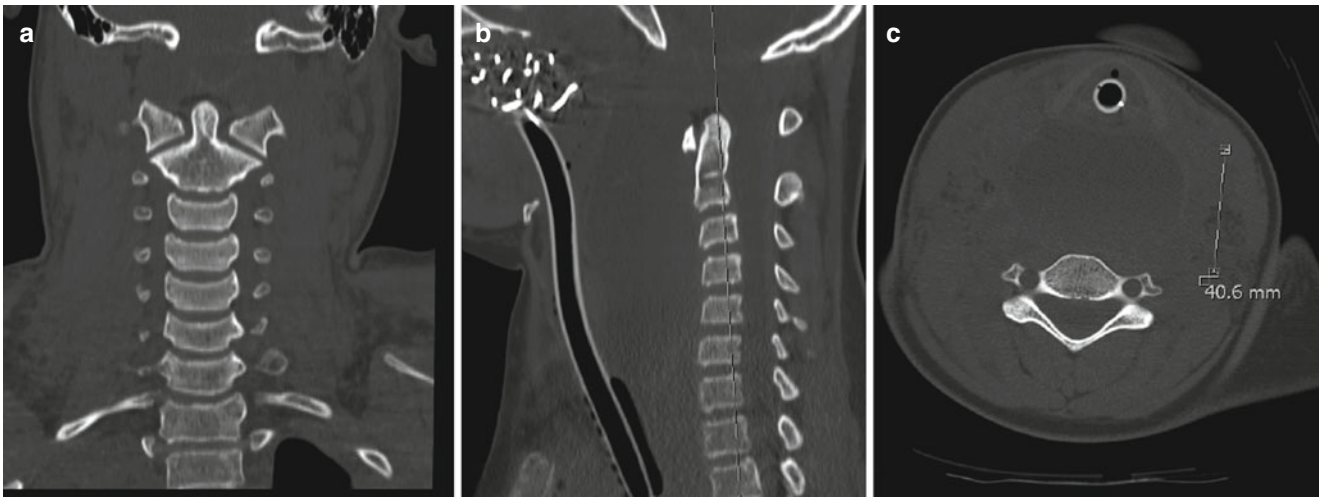

Fig. 28.5 Atlanto-occiptal dislocation. **(a)** Coronal, **(b)** sagittal and **(c)** axial (at C4 level) MRI images demonstrating significant atlanto-occiptal dislocation with associate soft tissue swelling

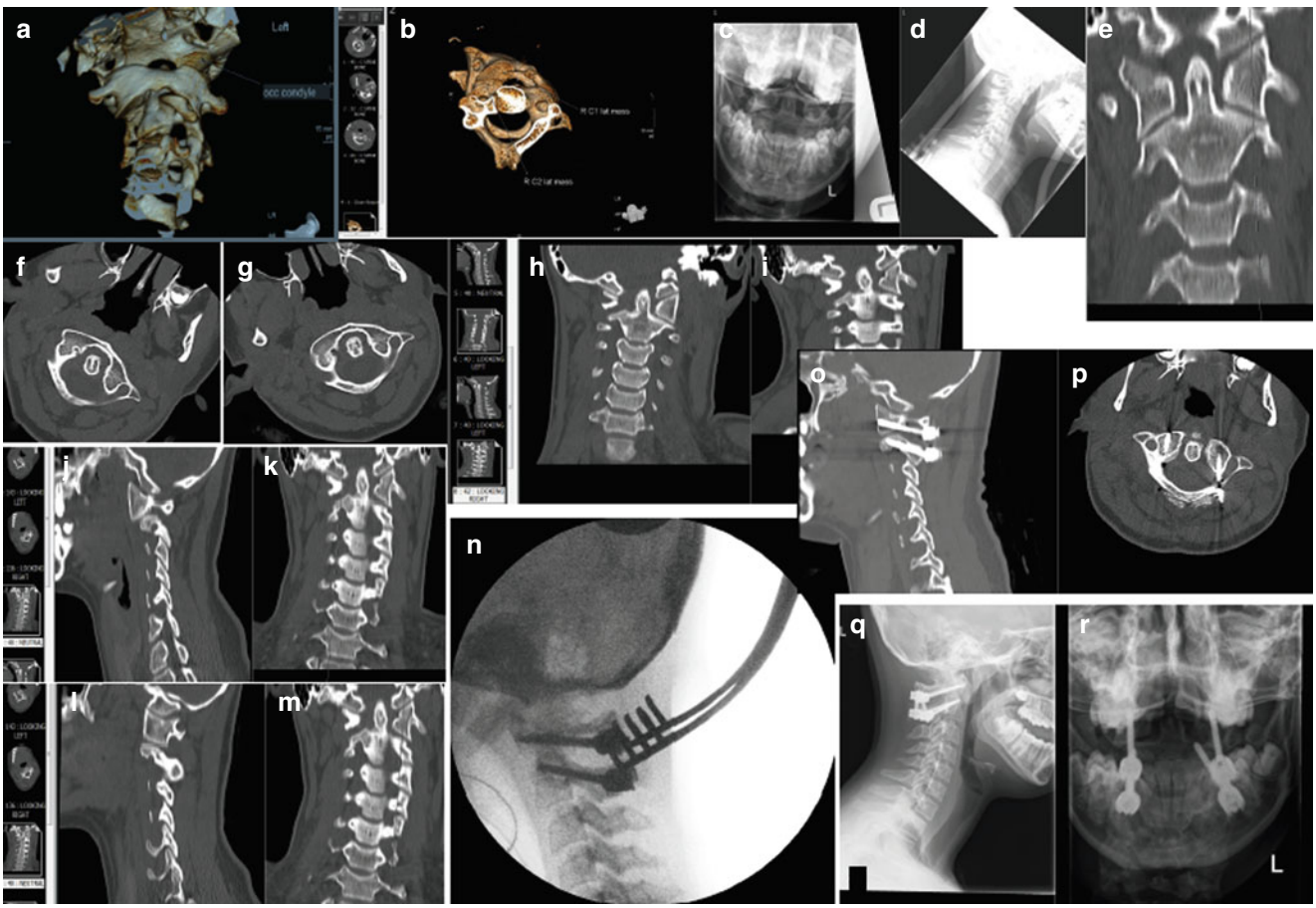

Fig. 28.6 Atlanto-axial rotatory fixation. CT reconstructions demonstrating rotatory subluxation of C1 on **(a)** C0 and **(b)** C2 at initial presentation following 6-weeks of torticollis. After 6-weeks of no improvement in a cervical collar, reduction was achieved by traction and placement in a halo. **(c)** AP open mouth and **(d)** lateral C-spine radiographs with **(e)** coronal CT and **(f)** axial CT to the left then **(g)** right demonstrated marked improvement after 10 days traction. Check dynamic CT at 3-months demonstrates fixed atlanto-axial subluxation; CT **(h)** looking left, **(i)** then right. Neutral CT **(j)** right para-sagittal and **(k)** corresponding coronal compared to **(l)** left para-sagittal and **(m)** corresponding coronal images. Failure of conservative management followed by Goel-Harms fusion; **(n)** intraoperative fluoroscopy, **(o)** postoperative sagittal and **(p)** axial CT. Postoperative 6-week follow up **(q)** lateral and **(r)** AP c-spine radiographs

- Type III
  - Disruption of both transverse and alar ligaments.
  - Anterior subluxation of both C1 facets.
- Type IV (rare)
  - Posterior displacement of C1 relative to C2.
  - Can only occur if absent or hypoplastic odontoid process.

Use three-position CT (first with the head in the injured position, then turned maximally to the right and then to the left) with C1–C2 motion analysis (fluoroscopy) to confirm and classify the diagnosis for children suspected of having AARF [12, 29].

Management depends upon the severity and duration of the abnormality: in general types II, III and IV usually require surgical stabilisation of the atlanto-axial complex [2]. Minor and acute cases (less than 1 week duration) can be treated with soft collar, rest and analgesia. Subsequent immobilisation should be proportional to the length of time that the subluxation was present before treatment. Surgical arthrodesis should be considered for those with irreducible subluxations, recurrent subluxations, or subluxations present for >3 months duration [2]. Consider botulin toxin injections as an adjunct to conservative treatment [30]. Hangman's fracture.

Traumatic spondylolisthesis of C2 usually occurs in children less than 2 years of age, due to their unique biomechanics and large head size. The mechanism of injury consists of hyperextension and axial loading that may be secondary to NAT (which must be excluded). Treatment consists of gentle closed reduction in extension and immobilisation in Minerva cast or halo device. If there is non-union or continual instability, posterior C1–3 arthrodesis or anterior C2–3 arthrodesis show good results [18].

Lower Cervical Spine Injuries

These injuries are more common in older children and adolescents as their spine develops a more adult structure. Injuries include posterior ligamentous disruptions, compression fractures, burst fractures, facet dislocations, spondylolysis and spondylolisthesis. C2–C3 subluxation can cause difficulties in diagnosis due to the similar findings in pseudosubluxation at this level.

Compression fractures are the most common in this region: these are stable fractures which heal with immobilisation in a cervical orthosis for 3–6 weeks with flexion-extension radiographs at 2–4 weeks post injury to help ascertain fracture stability. The second most common injury is facet dislocations, the management of which requires reduction with traction or open reduction and arthrodesis.

Burst fractures require CT to detect spinal canal compromise from retropulsed fragments and occult laminar fractures and MRI to delineate the state of the spinal cord and presence of associated ligamentous injury: halo immobilisation will be sufficient if the spinal canal is not compromised. Arthrodesis is required, with or without spinal cord decompression where there is canal compromise. If the fracture occurs through the endplate (more commonly inferior endplate), the injury is unstable with a high risk of neurological damage. This requires closed reduction and halo immobilisation as the primary treatment.

Teardrop fractures can be divided into two categories: flexion teardrop fractures and extension teardrop fractures. Flexion teardrop fractures are the most severe fracture of the cervical spine. They typically occur as a result of extreme flexion and compression (e.g. diving into a shallow pool) and commonly occur at the C5–6 level. They can be associated with an anterior cervical cord syndrome and quadriplegia. These fractures have been shown in adults to respond well to anterior plate stabilisation [31]. Additional posterior fixation may be required in the presence of posterior oesteoligamentous disruption. In the absence of specific paediatric evidence, one should apply sound biomechanical principles when deciding on the method of fixation.

Extension type teardrop fractures typically occur following forced extension of the neck resulting in avulsion of the antero-inferior corner of the vertebral body. They are not as severe as the flexion type and can often be treated conservatively.

SCIWORA

Spinal cord injury without radiological abnormality (SCIWORA) is more common in younger children (0–8 years old). There is a gradual decline in prevalence of SCIWORA as children become older and adopt an adult spinal alignment around 8–10 years of age [1, 8, 32]. The young spinal column may stretch up to 5 cm prior to rupture. The spinal cord however is tethered superiorly and inferiorly and can therefore rupture after only 5–6 mm of traction.

This entity was originally described when radiographs were the only imaging modality available. Now, due to advances in radiology, SCIWORA includes patients with neurological signs or symptoms and no abnormalities on plain film or CT, but with pathological findings on MRI. These can include ligament rupture, disc herniation, muscle oedema, complete cord disruption, cord haemorrhage and cord oedema [1, 8].

SCIWORA can have a delayed onset of up to 4 days: neurologically normal children with a history of transient neurological symptoms at the time of injury should be taken seriously and monitored closely [12]. MRI helps to exclude lesions that may require emergency decompression, such as haematomas and disc herniation, and it serves as a prognosticator for neurological recovery [33, 34]. The upper cervical spine is the most commonly affected region in those under 8 years of age.

SCIWORA is typically managed with external immobilisation for up to 12 weeks followed by activity modification to allow for healing of presumed ligamentous injury and prevent further injury to the spinal cord [35, 36]. Disagreement exists in the literature however and some authors state that immobilisation with bracing can be discontinued once certain criteria have been met. These include resolution of spinal tenderness, stabilisation of neurological examination and resolution of instability confirmed on imaging (including MRI) [35].

Non-Surgical Management of Cervical Spine

Several unique concerns regarding traction in children must always be considered before commencing treatment. Children have relatively thinner skulls, a lighter body weight, more elastic ligaments and less developed musculature: all of these increase the potential for over-distraction. CT is recommended prior to halo placement to assess skull thickness and safe areas for pin placement. The most common complications in children include: pin-site infections requiring antibiotics and on going pin-site care; loosening of pins; and issues with foreign bodies getting lodged in vests [1, 2, 6].

Immobilisation with thermoplastic Minerva orthoses rather than halos is favoured in young children as it has been demonstrated to provide superior stabilisation at all levels of the c-spine with the exception of C1–C2 [21–24]. Children as young as 1 year old can be successfully managed with a halo device [18].

Thoraco-Lumbar (TL) Spine Injuries

Thoracolumbar spinal injuries are associated with high-energy trauma and are less common in children <8 years of age due to the unique aspects of development mentioned earlier in this chapter. MVCs are the most common cause of injury of the thoracolumbar spine in all age groups. Inappropriate seat belt placement can lead to intra-abdominal and spinal injuries: up to 42 % of thoraco-lumbar fractures are associated with intra-abdominal injury [20].

The nucleus pulposus in children has a greater water content and smaller amount of collagen cross-linking than in adults. This provides more elasticity and a superior ability to dissipate force [20]. Therefore, when the immature spine is compressed, the vertebral body breaks through the endplate apophysis (which is formed of generally weaker cartilage) before the normal disc gives way, resulting in a Salter Harris type fracture.

Depending on the mechanism of injury and the skeletal maturity of the child, these fractures have a greater propensity to heal and remodel [20]. True fracture lines are seldom seen in the young child prior to puberty [5]. Table 28.1 summarises the epidemiological features of TL spine injuries.

In adults, health professionals use the Thoracolumbar Injury Classification and Severity Score (TLICS) to help provide better predictions of surgical intervention and outcomes [20].

The TLICS is based on:

1. Morphology of the injury.
2. Integrity of the posterior ligamentous complex.
3. Neurological status.

If the score is >4, surgical intervention should be considered. Conservative management is advocated at <4. Unfortunately, at the time of writing, there is no published paediatric equivalent and there is minimal data on its use on a paediatric population.

Compression Fractures

These are the most common type of fracture in the TL region and are caused by flexion and axial loading, usually as the result of low-energy mechanisms including falls and sporting injuries [20]. Use lateral radiographs to view the anterior vertebral body and to identify non-contiguous/contiguous fractures, as these are common. They are typically stable fractures with no neurological injury. If, however, >50 % of the anterior height is lost consider MRI as there may be disruption of the posterior elements. If the fracture is isolated, it may require TLSO brace immobilisation and activity modification for 6–8 weeks. In the scenario of multiple fractures with kyphosis, posterior spinal fusion should be considered.

Burst Fractures

These occur from axial loading which forces the nucleus pulopsus into the vertebral body. Retropulsion of bone fragments, and lamina fracture fragments, into the spinal canal may produce neurological deficits and biomechanical instability [20]. CT imaging is used to assess the posterior elements and the retropulsed fragments whilst MRI allows assessment of the posterior ligamentous complex to identify unstable injuries. If the injury is deemed stable and there is no progressive neurological deficit, treat with hyperextension casting or TLSO bracing for 8–12 weeks. If surgical stabilisation is required, the options include anterior corpectomy and strut grafting with anterior or posterior spinal instrumentation and fusion, or posterior spinal instrumentation and fusion with indirect or direct spinal canal decompression.

Current evidence suggests no difference in functional outcome for both operative and non-operative management in

burst fractures and the risk of SCI does not actually correlate with the degree of canal compromise. It is believed that neurological damage actually occurs at the time of injury and not to on going compression caused by retropulsed fragments: 'surgical clearance' of the canal does not necessarily affect the neurological outcome [37, 38]. It is important however to consider the development of post-traumatic kyphosis in the paediatric population: this has been shown to have a better outcome with a surgical management [39].

Flexion-Distraction Injuries (Chance Fractures)

These injuries are classically caused by a lap belt and up to 40 % are associated with visceral or head injuries [20]. The presence of the "seat belt sign" should always raise suspicion of a Chance fracture and associated intra-abdominal injury [24]. They most commonly occur between L1 and L3 but can occur at L4. They result in bony or ligamentous disruption of the posterior elements with anterior compression and distraction. They can be purely osseous, purely ligamentous or a combination of both involving just one or multiple levels [5].

If there is primarily bone involvement rather than ligamentous damage and no intra-abdominal injury, treat with a hyperextension cast or TLSO brace for 8 weeks. Standing radiographs whilst wearing the brace or cast must be obtained prior to discharge to ensure stable alignment. If however ligamentous injury predominates or acceptable alignment for purely osseous fractures cannot be obtained, manage with posterior spinal instrumented fusion [20, 24, 40].

Fracture Dislocations

Fracture dislocations are very unstable. They result from shearing and/or rotational displacement of the spinal column, often caused by blunt trauma. They are commonly associated with spinal cord injury and require posterior instrumentation and fusion to help maintain cord function, aid rehabilitation and allow sitting upright. Children display better recovery than adults. Unfortunately, however, children under 10-years-of-age with complete spinal cord injury almost always develop a paralytic scoliosis resistant to bracing. Vigilant follow-up of this cohort is therefore mandatory.

Apophyseal Injuries

These are unique to the paediatric population, most typically presenting in the adolescent male spine (10–14 years) suggesting the vertebral endplate is more susceptible during this phase of rapid growth. This injury is comparable to slipped upper femoral epiphysis (SUFE) [5]. They result from separation of the vertebral apophysis from the spongiosa layer of the vertebral body [20] commonly following lumbar flexion injury and cause the posterior portion of the ring apophysis to fracture (through the hypertrophic zone) and displace into the spinal canal. Presentation is similar to disc herniation with low back and radicular leg pain. In the absence of neurological deficit or cauda equina syndrome, these injuries are treated with anti-inflammatories, physiotherapy and TLSO bracing for 8 weeks. Surgical decompression is required in the presence of refractory pain or deteriorating neurological signs.

Spondylolysis and Spondylolisthesis

Isthmic spondylolysis occurs in approximately 4–6 % of Caucasian children. It can result from an acute fracture but more often is secondary to fatigue fractures through the pars interarticularis caused by repetitive microtrauma. It is more common in those who partake in hyperextension activities such as gymnastics or cricket. Most commonly it occurs at L5 (85–95 %), and less often at L4. Spondylolysis is usually asymptomatic but may present with focal lower back pain, with or without radiculopathy.

Plain radiographs are useful but often will not identify a pars defect. CT best defines the lesion but MRI is the preferred modality as there is no radiation exposure. Oblique radiographs can reveal the typical "scotty dog" of Lachapelle and can be a helpful visual aid: however, these emit high doses of radiation and are therefore being phased out. Spondylolisthesis occurs in a significant proportion of those with spondylolysis presenting with activity related lower back pain, radicular symptoms and neurological deficit. Examination findings include reduced/painful lumbar extension, hamstring tightness and a step off deformity with high-grade slips.

The lack of large-scale controlled trials makes it difficult to define optimal treatment. General principles for the management of proven pain due to spondylolysis include activity modification, physiotherapy and bracing; surgical in-situ fusion is reserved for patients who fail such treatment. Partial reduction with decompression and interbody fusion has been advocated for progressive and high-grade slips, but the patient must be warned about potential intraoperative neurological deterioration. All children or adolescents with symptoms due to spondylolysis, especially those under 10 years of age, should be followed closely for progression to spondylolisthesis.

Thoracolumbar Injury Management

As a rule of thumb, many stable fractures can be managed conservatively whereas unstable fractures require surgical

stabilisation. Different braces exist for thoraco-lumbar immobilisation: thoracolumbosacral orthoses (TLSO) or Jewett braces (if hyperextension is required) and sterno-occipito-mandibular immobilisers (SOMI) can be used in conjunction with TLSO for upper thoracic spine fractures.

Early surgical treatment, instrumentation and fusion are mandatory for unstable fractures and injuries associated with spinal cord injury. Surgical stabilisation for unstable lumbar fractures can often be managed via a posterior approach [41]. Adolescents can be stabilised with adult-type instrumentation. As mentioned previously, the paediatric spine develops an adult-like form between 8 and 10 years. Younger children have smaller pedicles therefore pedicle screw placement may be challenging.

Late Complications of Paediatric Spinal Injuries

Unlike the adult spine, children have a propensity to heal and remodel and therefore have more favourable long-term outcomes following spinal trauma [41]. However, it is important to note that the majority of children who experience spinal cord injury before their growth spurt, develop spinal deformities: girls under 12 years and boys under 14 years of age are at high risk of developing paralytic scoliosis and therefore must followed up closely [5, 20]. Children with more proximal injuries are more likely to have a progressive deformity then those injured at or below the level of the conus medullaris [5].

Evidence for the role for prophylactic bracing is unclear. It has been suggested that bracing coronal curves <10° may prevent the need for surgical correction and delay surgery in curves between 10° and 20° [30]. Unfortunately, bracing curves >20° is unlikely to be successful in controlling curve progression in patients with SCI [30, 36].

Deformity correction in SCI is often complicated by poor wound healing secondary to the lack of protective sensation and wound contamination from urinary/faecal incontinence [20]. Laminectomy is associated with significant post-operative kyphosis and, if absolutely necessary, should be accompanied by a short segment spinal fusion [5].

Conclusion

Spinal trauma in paediatric patients is uncommon but may lead to substantial morbidity and mortality.

Children require special attention due to unique features of their anatomical development. Children acquire a more adult-like spine beyond 8–10 years of age.

Never overlook NAT as a possible mechanism of spinal injury in children.

A large head size in comparison to the body, elastic spine and differing biomechanics means cervical spine injuries predominate in younger children.

Initial immobilisation should include a spinal board with an occipital recess or thoracic elevation in those children less than 8 years of age.

SCIWORA is more common in children and should always be suspected in those who have transient neurological signs and symptoms. SCIWORA has been reported up to 4 days post-injury.

Careful consideration of radiological modalities is fundamental. Although adult protocols exist, there is a general lack of evidence for their use in young children. The need for paediatric imaging protocols is urgent and should be the mainstay of future developments.

There is a lack of a validated thoraco-lumbar classification to guide management and aid prognosis. The application of thoracolumbar classification systems to children deserves the attention of future research.

In general, children pose a challenge to spine surgeons because of their immature bone quality, extensive anatomical variability, and smaller osseous structures.

Whilst children have a greater propensity to heal and remodel, they have specific long-term sequelae that must be closely monitored and if possible corrected.

Currently, management should be individualised on a case-by-case basis with the local resources and skills of the surgeon taken into account.

A summary of recommendations is provided in Table 28.5.

Table 28.5 Grade of recommendation

Statement	Level of evidence	Grade of recommendation
Children less than 8 years of age should be immobilised on a spinal board with an occipital recess or thoracic elevation	III	B
Taping a child to a spinal board causes forced vital capacity (FVC) to drop	III	B
Adult imaging guidelines should not be applied to children less than 11 years old	II	B
If AOD is clinically suspected use CT to diagnose	I	A
Radiographs may not be necessary to clear the cervical spine in children fulfilling low-risk criteria (See Table 28.5)	II	B
Do not attempt open mouth peg views if the child is less than 9 years old	III	B
Initially treat odontoid injuries with external immobilisation	III	B
Prolonged delay in treatment of AARF may adversely affect outcome	III	B
With regards to AARF; consider surgical arthrodesis for those with irreducible subluxations, recurrent subluxations, or subluxations present for >3 months duration	III	B
Use early posterior atlanto-axial arthrodesis for ligamentous disruptions	III	B
Consider primary surgical stabilisation for unstable cervical spine fractures to prevent late deformity	III	B
Treat stable cervical spine fractures conservatively	III	B
SCIWORA can have a delayed onset of up to 4 days	IV	C
Treat SCIWORA with external immobilisation for 12 weeks with further activity modification	III	B
The Minerva vest provides superior stabilisation all levels except for C1–C2 when compared to the halo device	III	B
Pin-site infection is the most common complication of the halo device	III	B
Post-traumatic kyphosis has a better outcome with surgical management compared to conservative management	III	B
The risk of SCI in TL burst fractures does not correlate to the degree of canal compromise	III	B

References

1. Basu S. Spinal injuries in children. Front Neurol. 2012;3:96.
2. Mortazavi M, Gore P, Chang S. Pediatric cervical spine injuries: a comprehensive review. Childs Nerv Syst. 2011;27:705–17.
3. Martin BW, Dykes E, Lecky FE. Patterns and risks in spinal trauma. Arch Dis Child. 2004;89(9):860–5.
4. Reddy SP, Junewick JJ, Backstrom JW. Distribution of spinal fractures in children: does age, mechanism of injury, or gender play a significant role? Pediatr Radiol. 2003;33(11):776–81.
5. Benson M. Children's orthopedics and fractures. 3rd ed. London: Springer; 2010.
6. Brown RL, Brunn MA, Garcia VF. Cervical spine injuries in children: a review of 103 patients treated consecutively at a level 1 pediatric trauma center. J Pediatr Surg. 2001;36(8):1107–14.
7. Knox J, Schneider J, Wimberly RL, Riccio AI. Characteristics of spinal injuries secondary to nonaccidental trauma. J Pediatr Orthop. 2014;34(4):376–81. doi: 10.1097/BPO.0000000000000111.
8. Hutchings L. Clearing the cervical spine in children. Trauma. 2011;13(4):340–52.
9. Nypaver M, Treloar D. Neutral cervical spine positioning in children. Ann Emerg Med. 1994;23(2):208–11.
10. Nypaver M, Treloar D. Neutral cervical spine positioning in children. Ann Emerg Med. 1994;17 23(2):208e211.
11. Schafermeyer RW, Ribbeck BM, Gaskins J, Thomason S, Harlan M, Attkisson A. Respiratory effects of spinal immobilization in children. Ann Emerg Med. 1991;20(9):1017–9.
12. Rozzelle CJ, Aarabi B, Dhall SS, Gelb DE, Hurlbert RJ, Ryken TC, et al. Management of pediatric cervical spine and spinal cord injuries. Neurosurgery. 2013;72(Suppl 2):205–26.
13. NICE guidelines. https://www.nice.org.uk/guidance/indevelopment/gid-cgwave0645
14. Burns E, Yanchar N. Using cervical spine clearance guidelines in a pediatric population: a survey of physician practices and opinions. CJEM. 2011;13(1):1–6.
15. Ehrlich PF, Wee C, Drongowski R, Rana AR. Canadian C-spine rule and the national emergency X-radiography utilization low-risk criteria for C-spine radiography in young trauma patients. J Pediatr Surg. 2009;44(5):987–91.
16. The Royal College of Radiologists. Paediatric trauma protocols. 2014; Available at: https://www.rcr.ac.uk/docs/radiology/pdf/BFCR(14)8_paeds_trauma.pdf, 2015.
17. Jimenez RR, Deguzman MA, Shiran S, Karrellas A, Lorenzo RL. CT versus plain radiographs for evaluation of c-spine injury in young children: do benefits outweigh risks? Pediatr Radiol. 2008;38(6):635–44.
18. Li Y, Glotzbecker MP, Hedequist D, ST M. Pediatric spinal trauma. Trauma. 2011;14:82–96.
19. Laham JL, Cotcamp DH, Gibbons PA, Kahana MD, Crone KR. Isolated head injuries versus multiple trauma in pediatric patients: do the same indications for cervical spine evaluation apply? Pediatr Neurosurg. 1994;21(4):221–6 .discussion 226
20. Daniels AH, Sobel AD, Eberson CP. Pediatric thoracolumbar spine trauma. J Am Acad Orthop Surg. 2013;21(12):707–16.
21. Tomaszewski R, Wiktor L. Occipital Condyle Fractures in Adolescents. Ortop Traumatol Rehabil. 2015;17(3):219–27.

22. Strehle E-M, Tolinov V. Occipital condylar fractures in children: rare or underdiagnosed? Dentomaxillofacial Radiology. 2012;41(2): 175–6.
23. Maserati MB, Stephens B, Zohny Z, Lee JY, Kanter AS, Spiro RM, et al. Occipital condyle fractures: clinical decision rule and surgical management. J Neurosurg Spine. 2009;11(4):388–95.
24. Dormans JP, Drummond DS, Sutton LN, Ecker ML, Kopacz KJ. Occipitocervical arthrodesis in children. A new technique and analysis of results. J Bone Joint Surg Am. 1995;77(8):1234–40.
25. Tauchi R, Imagama S, Ito Z. Complications and outcomes of posterior fusion in children with atlantoaxial instability. Eur Spine J. 2012;21:1346–52.
26. Joaquim AF, Ghizoni E, Tedeschi H, et al. Upper cervical injuries – A rational approach to guide surgical management. J Spinal Cord Med. 2014;37(2):139–51.
27. Pang D, Li V. Atlantoaxial rotatory fixation: part 2–new diagnostic paradigm and a new classification based on motion analysis using computed tomographic imaging. Neurosurgery. 2005;57(5):941–53 .discussion 941–53
28. Grogaard B, Dullerud R, Magnaes B. Acute torticollis in children due to atlanto-axial rotary fixation. Arch Orthop Trauma Surg. 1993;112(4):185–8.
29. Phillips WA, Hensinger RN. The management of rotatory atlantoaxial subluxation in children. J Bone Joint Surg Am. 1989;71(5): 664–8.
30. Janselewitz S1, Wehby M, Hume J. Pediatric atlantoaxial rotary subluxation relapse treated with botulinum toxin and repeated manual reduction. PM R. 2011;3(11):1072–4. doi: 10.1016/j.pmrj.2011.08.532.
31. Kim HJ, Lee KY, Kim WC. Treatment outcome of cervical tear drop fracture. Asian Spine J. 2009;3(2):73–9.
32. Liao CC, Lui TN, Chen LR, Chuang CC, Huang YC. Spinal cord injury without radiological abnormality in preschool-aged children: correlation of magnetic resonance imaging findings with neurological outcomes. J Neurosurg. 2005;103(1 Suppl):17–23.
33. Leonard M, Sproule J, McCormack D. Paediatric spinal trauma and associated injuries. Injury. 2007;38(2):188–93.
34. Pang D. Spinal cord injury without radiographic abnormality in children, 2 decades later. Neurosurgery. 2004;55(6):1325–42 .discussion 1342–3
35. Jones TM, Anderson PA, Noonan KJ. Pediatric cervical spine trauma. J Am Acad Orthop Surg. 2011;19(10):600–11.
36. Kim SH, Yoon SH, Cho KH, Kim SH. Spinal cord injury without radiological abnormality in an infant with delayed presentation of symptoms after a minor injury. Spine (Phila Pa 1976). 2008; 33(21):E792–4.
37. Limb D, Shaw DL, Dickson RA. Neurological injury in thoracolumbar burst fractures. J Bone Joint Surg Br. 1995;77(5):774–7.
38. Boerger TO, Limb D, Dickson RA. Does 'canal clearance' affect neurological outcome after thoracolumbar burst fractures? J Bone Joint Surg. 2000;82(5):629–35.
39. Vander Have KL, Caird MS, Gross S, Farley FA, Graziano GA, Stuaff M, Segal LS. Burst fractures of the thoracic and lumbar spine in children and adolescents. J Paediatric Orthop. 2009;29(7):713–9.
40. Arkader A, Warner W, Tolo T. Pediatric chance fractures: a multicenter perspective. J Pediatr Orthop. 2011;31:741–4.
41. Sayama C, Chen T, Trost G, Jea A. A review of pediatric lumbar spine trauma. Neurosurg Focus. 2014;37(1):E6.

Part VI

The Shoulder

Evidence-Based Treatment of Congenital Clavicular Pseudarthrosis

29

James S. Huntley

Abstract

Clavicular pseudarthrosis is a rare condition. Its natural history is not well established, as to the risks of progression in size, deformity or pain. It is known that it does not heal spontaneously. Operative indications, best age for surgery and principles/type of surgery are not agreed; there is a diversity of opinion on all these aspects. An assessment of the literature defines the paucity of evidence. There are only 3 level III studies (all of which have small numbers and possible confounders, limitations sufficient to warrant downgrading their quality of evidence). Furthermore, the conclusions of two of the retrospective comparative studies, in terms of superior results from type of fixation (plate or intramedullary wire), are contradictory. There are 16 case series of note (level IV). Although there is little by way of concrete recommendation, it is hoped that this analysis and summary can aid pragmatic and considered decision-making when encountering a patient with this diagnosis.

Keywords

Congenital • Clavicular pseudarthrosis • Cleidocranial dysostosis

Introduction

Congenital clavicular pseudarthrosis is a rare condition, first described in 1910 [1], with, as of 2011, less than 200 cases reported [2]. The most common presentation is of a painless, non-tender mobile lump over the right clavicle [3]. The differential diagnosis includes neonatal clavicle fracture, brachial plexus palsy, osteomyelitis (congenital syphilis), cleidocranial dysostosis and neurofibromatosis [2]. The diagnosis can usually be made reliably on the basis of the history, examination and plain films. Occasionally a pseudarthrosis may be associated with a thoracic outlet syndrome [4]. If the diagnosis is in doubt, CT scanning can be a useful aid [5].

The vast majority of clavicular pseudarthroses are on the right side, though it can occur bilaterally. Occasionally there is a positive family history. This has prompted the assertion that a genetic link is likely in some cases [6, 7]. The aetiology is thought to be related to abnormal embryogenesis [6], possibly involving the position/effect of the right subclavian artery [8].

In 1963, Alldred [3] was able to collect a case series of 9 patients, and to comment on a further two from the literature. In his opening paragraph he surmised: '*The case histories of these children indicate that the condition is not well-known, that its natural history has never been adequately recorded and that advice given to parents has often been based on speculation*'. Little has changed over the intervening half-century.

Questions

Three key questions were identified:

1. What is the natural history of congenital clavicular pseudarthrosis?

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2. What is the best treatment for congenital clavicular pseudarthrosis?
3. What is the best timing for operative treatment of congenital pseudarthrosis?

Search Strategy

The Pubmed database was searched using the following strategy (accessed 20/09/2015): ((“clavicle”[MeSH Terms] OR “clavicle”[All Fields]) AND (“pseudarthrosis”[MeSH Terms] OR “pseudarthrosis”[All Fields] OR “pseudoarthrosis”[All Fields])) OR ((“clavicle”[MeSH Terms] OR “clavicle”[All Fields]) AND (“pseudarthrosis”[MeSH Terms] OR “pseudarthrosis”[All Fields])) OR ((“clavicle”[MeSH Terms] OR “clavicle”[All Fields] OR “clavicular”[All Fields]) AND (“pseudarthrosis”[MeSH Terms] OR “pseudarthrosis”[All Fields])) OR ((“clavicle”[MeSH Terms] OR “clavicle”[All Fields] OR “clavicular”[All Fields]) AND (“pseudarthrosis”[MeSH Terms] OR “pseudarthrosis”[All Fields] OR “pseudoarthrosis”[All Fields]))

This yielded 270 references. Titles, abstracts and – where relevant – papers, were reviewed, allowing elimination of articles referring to diagnoses other than congenital clavicular pseudarthrosis. Single case reports were also excluded from the main analysis. This yielded 19 papers: 3 level III retrospective comparative studies, and 16 level IV case series. Searches of the Cochrane controlled trials register and Cochrane database (accessed 28/09/2015) did not provide additional relevant material. The 19 studies are summarized in Table 29.1.

What Is the Natural History of Congenital Clavicular Pseudarthrosis?

The natural history of this condition remains unclear in absolute terms, though some lesions enlarge with time and/or become painful. Others remain painless throughout life with no functional deficit [25]. Once identified, there is no record of spontaneous fusion/resolution [14, 16]. Pooling the numbers from the 19 studies above (which excludes single case reports) yields a total of 190 patients, of whom 61 were treated conservatively. Given that this data set largely represents ‘surgical’ series, and even when the studies identify patients treated non-operatively there is the potential for selection bias, it seems likely that the proportion identified as being treated non-operatively (61/190) is an underestimate. Moreover, given the limitations on follow-up, there is little information as to the ‘success’, or otherwise, of non-operative treatment. Therefore there is little evidence *per se* to guide the surgeon or parents of a child with an *asymptomatic* congenital clavicular pseudarthrosis.

What Is the Best Treatment for Congenital Clavicular Pseudarthrosis?

As outlined above, it is difficult to advise children and parents as to the natural history of such a pseudarthrosis. On this basis, for asymptomatic lesions, it is unclear if surgery should be advised. There are substantial operative risks including failure (infection and non-union), anticosmesis and other complications – including post-operative brachial plexopathy [26]. A balanced opinion is that of Quinlan et al. [20] who suggested that surgery is indicated only if there is increasing deformity or pain. However, earlier surgery at infant age has been advocated by some (see next section).

Types of surgery advocated include subperiosteal excision of pseudoarthrosis ± bone graft (itself structural/non-structural) ± fixation, usually by either plate or wire (K-wire/threaded pin).

The retrospective comparative study of Chandran et al. [9] concerned 10 patients treated by surgery involving pseudarthrosis resection, iliac bone grafting and fixation, either by plate or intramedullary wire (fixation mode defining the two groups considered). The median age (5 years) was similar between the two groups. Although they observed that the median time to healing was shorter in the reconstruction plate group, there were insufficient numbers for statistical analysis of difference. It is of note that one patient in in the plate group achieved union only at 22 months post procedure, whereas a non-union was defined at 8 months in the wire group. Furthermore, no indication was given as to reasons for selecting one form of fixation (plate vs. wire) over the other – it is possible, for instance, that the size of segmental defect (and equivalent bone graft size) post excision might prejudice the surgeon in favour of a particular mode of fixation. Their conclusion that ‘*Plate fixation achieved more reliable union quicker and with fewer complications compared to pin*’ is therefore subject to major reservations, and is in contradistinction to the series of Persianni et al. 2008 (below).

Persianni et al. (Persianni et al. [11]) treated 17 patients by surgery involving pseudarthrosis excision, with iliac bone graft in 12, and fixation (9 by plate and 8 by K-wire). There were five complications requiring revisional surgery: with (3/12) or without (2/5) bone graft, with plate (4/9) or wire (1/8) at index surgery. They found a lower reoperation rate and better results when the initial mode of fixation was a wire (compared to a plate). Similar to the study by Chandran et al. [9], the numbers were so small as to preclude any analysis of significance, and no indication was given as to reasons for initially selecting one form of fixation over the other.

Table 29.1 Summary of case series and retrospective comparative studies concerning congenital clavicular pseudarthrosis management

Paper	Design	Number patients	Details/results	LoE
Chandran et al. [9]	Retrospective comparative study	12	2 no treatment 10 patients underwent operative treatment (median age in both groups was 5 years): Group A (five patients): excision + iliac BG + fully threaded pin Group B (five patients – one with bilateral pseudarthroses): excision + iliac BG + reconstruction plate 2 failures of surgery – both in group A: one with non-union, one with wound breakdown and infection	III
Elliot and Richards [10]	Case series	2	Both patients (both aged 5 years) were treated surgically with resection, Tutobone (bovine cancellous xenograft) and IF (with 3.5 mm reconstruction plate) Both failed with '*significant osteolysis and failure of incorporation of the graft*'	IV
Persiani et al. [11]	Retrospective comparative study	17	All 17 were treated surgically; 12 with BG (iliac crest); 9 with plate fixation, 8 with Kirschner wire fixation Operative age {mean (range)} was 5.8 (4–7.5) years and 6.4 (5.25–7.6) in the plate and K-wire groups respectively Five patients (4 with plate, 1 with K-wire) required a secondary procedure in their grading system, 11 patients achieved a good result (4 with plate, 7 with K-wires), 3 patients achieved a fair result (2 with plates, 1 with K-wire), 3 patients had a poor result (all with plate)	III
Ullot Font et al. [12]	Case series	9	4 no treatment 5 treated with resection + iliac BG + IF (4 with plate and one with threaded pin); mean operative age, 8.4 years; range 5–14 years One patient (the one with threaded pin) required early removal metalwork because of protrusion	IV
Ettl et al. [13]	Case series	3	All 3 were treated surgically (operative ages: 4, 6, 8 years) by resection + BG + reconstruction plate; mean follow-up 44 months No failures of surgery	IV
Lorente Molto et al. [14]	Case series	6	1 no treatment 5 treated surgically (1 bilateral; ages 18 month to 4 years) with BG + IF with K-wire All healed by 6–8 weeks	IV

(continued)

Table 29.1 (continued)

Paper	Design	Number patients	Details/results	LoE
Cadhilac et al. [15]	Retrospective comparative study	25	8 treated non-operatively 17 treated surgically (at mean age 6 years 4 months; mean age at end follow-up 11.5 years) with resection and internal fixation with wire or plate: BG [6], no BG [1] Non-union: 0/9 in BG group, 3/8 in no BG group	III
Koster et al. [16]	Case series	2	2 treated operatively by resection + BG + IF with K-wire Both successful, with union at 8 weeks	IV
Price and Price [7]	Case series	2 (father and daughter)	Father previously operated on aged 10 years – unsuccessfully	IV
Schoenecker et al. [17]	Case series	5	All 5 treated surgically: resection, BG and IF with plate for all (BG: 4 from iliac crest, 1 local) Consolidation at average 3 months for all; MW removed at average 16 months post-op; average FU 4 years; all full function	IV
Grogan et al. [18]	Case series	8	All 8 were treated operatively [of note, at operation, 6 of the 8 were <2.5 years age; one was 5 years; one was 6 years]: Resection with maintenance of periosteal sleeve [there was no BG (though resected bone was placed around the approximated bon ends); no IF was used except for a loop of absorbable suture, threaded through bicortical drill holes on either side of the defect, to '*loosely bring the bone ends togther*']. All were fully healed by 14 weeks	IV
Schnall et al. [19]	Case series	6	All 6 were treated surgically by resection, BG and IF [5 with plate (mean operative age 10 years, range 4–15.5 years), 1 with wire (4.5 years)] All healed; no surgical failures	IV
Quinlan et al. [20]	Case series	4	1 no treatment 3 treated surgically (ages 4, 4.5, 6): excision [3], BG [21], wiring [22] No surgical failures	IV
Toledo and MacEwen [26]	Case series	10	6 no treatment 4 treated surgically (ages not ascertainable from manuscript): excision + BG + Steinmann pin 1 acute neurological compromise (termed '*massive neuropraxia of the brachial plexus*') necessitating immediate removal of internal fixation	IV
Ahmadi and Steel [22]	Case series	5	3 no treatment 2 treated surgically (operative ages 5 and 6 years) with excision, BG + IF (IM pin) No surgical failures	IV

(continued)

Table 29.1 (continued)

Paper	Design	Number patients	Details/results	LoE
Gibson and Carroll [6]	Case series	27	13 no treatment 14 treated surgically (mean operative age, 10 years.; range, 2–19 years): 11 BG, 8 with K-wires and 1 with compression plate (one unspecified type of surgery) 3/14 failure of surgery	IV
Wall [23]	Case series	5	Conservative treatment recommended	IV
Owen [24]	Case series	33	13 no treatment 20 treated surgically: (16 BG, 4 excision; 10 with wire or Rush pin) 1/20 non-union	IV
Alldred [3]	Case series	9	4 no treatment 5 treated surgically (age range 2–16 years): resection [9] + IF (IM pin (3), encircling wire [22]) + BG (4; iliac (1), tibial [22], rib [22], local chips [22]) 1 treatment failure in 2 year-old; and 1 further non-union (presumably intentional, in 16 year-old managed by resection alone)	IV

IF internal fixation, *BG* bone graft, *IM* intramedullary

These reservations downgrade the quailty of evidence [27] and the confidence in any conclusions.

Cadhilac et al. [15] reported 17 patients treated surgically (mean age, 6 years 4 months; mean age at end follow-up, 11.5 years) with resection and internal fixation with wire or plate, with [6] or without bone graft [1]. They reported a non-union rate of 0/9 and 3/8, with and without bone graft, respectively. This suggests that bone graft is a useful component of the surgery in this age group.

Conversely, Grogan et al. [18] reported full union in 8 patients after resection and cerclage suturing with no additional bone grafting. Of note, 6 of their patients were <2.5 years age. They commented that '*children undergoing surgery before age 3 years probably do not require temporary internal fixation.*'

In addition to the above studies, there is an array of case series documenting impressive results for pseudarthrosis resection with bone-grafting and internal fixation with either plate [12, 13, 17, 19] or wire [6, 14, 16, 22, 24, 26]; although the case series here have been accorded to plate or wire by the predominant fixation mode employed, those series of Schnall et al. [19] and Ullot Font et al. [12] contained 1 patient apiece in whom a wire was used, and that of Gibson and Carrol [6] contained 1 patient in whom a plate was used (see Table 29.1).

As regards plating, and strictly outwith the scope of this review (case report of a single patient), Sloan and Paton [5] advocated use of a reconstruction locking plate, which allowed for appropriate contouring, and the use of unicortical screws to minimise the risk of local neurovascular complications.

The experience of Elliot and Richards [10], attempting to obviate the need of iliac crest autograft, involved failure in 2/2 patients in whom clavicular reconstruction was attempted using Tutobone (bovine cancellous xenograft) rather than autogenous bone graft, with failure and substantial osteolysis. They therefore caution against the use of such material.

What Is the Best Timing for Operative Treatment of Congenital Pseudarthrosis?

As to timing of surgery, Gibson and Carroll (1970) [6] advocated deferring operation until pre-school (4–5 years) age, on grounds of technical ease at this stage. On the basis of progressive changes, Persiani et al. [11] recommended 'early' operation in all cases (but failed to specify a preferred age range). As above, Grogan et al. [18] reported full union in 8 patients after resection and cerclage suturing with no additional bone grafting, and 6 of their patients were <2.5 years age. They commented that '*children undergoing surgery before age 3 years probably do not require temporary internal fixation.*'

Lorente Molto et al. [14] presented a case series of 6 patients, treated surgically by resection, bone graft and internal fixation with K-wire in patients between 18 months and 4 years (healing of all by 6–8 weeks). In this age-group, they held that only minimal bone graft was required because of

Table 29.2 Summary of grades of recommendation

Recommendation	Level of evidence	Grade
If asymptomatic, it is unclear if surgery confers benefits (justifying the risks) in the long term	–	I
If symptoms or deformity are progressive, surgery can be justified	IV	C
In the children >4 years, bone graft is a useful component to the procedure	III, IV	C
In children >4 years a plate is preferred to an intramedullary wire for internal fixation	III (contradictory results)	I
The optimum time for surgery (infant vs. child) is not established	IV	I

the remodeling capacity. They advocated '*early surgical treatment… … during infancy or early childhood*', the rationale being that, in their young group, they found union to be achieved in a short time, with only K-wire fixation.

The small case series of Grogan et al. [18] and Lorente Molto et al. [14] above could be held to support operation in infancy. The difficulty is in establishing at an early stage which patients would progress to become symptomatic in the future, and offset numbers needed to treat against the risks of the procedure.

Conclusion

Given the rarity of this condition, it is unsurprising there are only small retrospective comparative studies, case series and single case reports (these latter being excluded). Only three studies could be termed comparative, all with numbers below threshold for statistical analysis and with multiple possible confounders, mandating downgrading of the quality of evidence [27]. Two of these studies [9, 11] had contradictory conclusions over the superior mode of fixation (wire vs. plate) in children of around 5 years of age. In the future, it remains to be seen whether other techniques applied generally to segmental bone defects, such as that of Masquelet (which has been applied successfully to paediatric clavicle pseudarthrosis on a case report basis [28]) give any advantage over the more established techniques. Recommendations and their grades are given in Table 29.2.

Acknowledgement JSH acknowledges the translation/interpretation services of Miss Itza Tellez for the manuscript by Ullot Font et al. [12].

References

1. Fitzwilliams D. Hereditary cranio-cleido-dysostosis. Lancet. 1910;2:1466.
2. Hudson BJ, Eastwood DM. Congenital upper limb deformities. Chapter 13.11. In: Bulstrode C, editor. Oxford textbook of trauma and orthopaedics. Oxford: Oxford University Press; 2011. p. 1504–11.
3. Alldred AJ. Congenital pseudarthrosis of the clavicle. J Bone Joint Surg Br. 1963;45B:312–9.
4. Hahn K, Shah R, Shalev Y, Schmidt DH, Bajwa T. Congenital clavicular pseudarthrosis associated with vascular thoracic outlet syndrome: case prersentation and review of the literature. Cathet Cardiovasc Diagn. 1995;35:321–7.
5. Sloan A, Paton R. Congenital pseudarthrosis of the clavicle: the role of CT scanning. Acta Orthop Belg. 2006;72:356–8.
6. Gibson DA, Carroll N. Congenital pseudarthrosis of the clavicle. J Bone Joint Surg. 1970;52B:629–43.
7. Price BD, Price CT. Familial congenital pseudarthrosis of the clavicle: case repport and literature review. Iowa Orthop J. 1996;16:153–6.
8. Lloyd-Roberts GC, Apley AG, Owen R. Reflections upon the aetiology of congenital pseudarthrosis of the clavicle. With a note on cranio-cleido dysostis. J Bone Joint Surg Br. 1975;57B:24–9.
9. Chandran P, George H, James LA. Congenital clavicular pseudarthrosis: comparison of two treatment methods. J Child Orthop. 2011;5:1–4.
10. Elliot RR, Richards RH. Failed operative treatment in 2 cases of pseudarthrosis of the clavicle using internal fixation and bovine cancellous xenograft (Tutobone). J Pediatr Orthop B. 2011;20:349–53.
11. Persiani P, Molayem I, Villani C, Cadhilac C, Glorion C. Surgical treatment of congenital pseudarthrosis of the clavicle: a report on 17 cases. Acta Orthop Belg. 2008;74:161–6.
12. Ullot Font R, Cepero Campa S, Gargantilla Vazquez A, Hernandez BS. Congenital pseudarthrosis of the clavicle: a review of nine cases. An Pediatr. 2006;64:273–6.
13. Ettl V, Wild A, Krauspe R, Raab P. Surgical treatment of congenital pseudarthrosis of the clavicle: a report of three cases and review of the literature. Eur J Pediatr Surg. 2005;15:56–60.
14. Lorente Molto FJ, Bonete Lluch DJ, Garrido IM. Congenital pseudarthrosis of the clavicle: a proposal for early surgical treatment. J Pediatr Orthop. 2001;21:689–93.
15. Cadhilac C, Fenoll B, Peretti A, Padovani JP, Pouliquen JC, Rigault P. Congenital pseudarthrosis of the clavicle: 25 childhood cases. Rev Chir Orthop. 2000;86:575–80.
16. Koster G, Kunze E, von Knoch M. Surgical treatment of congenital clavicular pseudarthrosis. Z Orthop Ihre Grenzgeb. 1999;137:414–8.
17. Schoenecker PL, Johnson GE, Howard B, Capelli AM. Congenital pseudarthosis. Orthop Rev. 1992;21:855–60.
18. Grogan DP, Love SM, Guidera KJ, Ogden JA. Operative treatment of congenital pseudarthrosis of the clavicle. J Pediatr Orthop. 1991;11:176–80.
19. Schnall SB, King JD, Marrero G. Congenital pseudarthrosis of the clavicle: a review of the literature and surgical results of six cases. J Pediatr Orthop. 1988;8:316–21.
20. Quinlan WR, Brady PG, Regan BF. Congenital pseudarthrosis of the clavicle. Acta Orthop Scand. 1980;51:489–92.
21. Beals RK, Sauser DD. Nontraumatic disorders of the clavicle. J Am Acad Orthop Surg. 2006;14:205–14.

22. Ahmadi B, Steel HH. Congenital pseudarthrosis of the clavicle. Clin Orthop Relat Res. 1977;126:129–34.
23. Wall JJ. Congenital pseudarthrosis of the clavicle. J Bone Joint Surg. 1970;52A:1003–9.
24. Owen R. Congenital pseudarthrosis of the clavicle. J Bone Joint Surg Br. 1970;52B:644–52.
25. Shalom A, Khermosh, Wientroub S. The natural history of congenital pseudarthrosis of the clavicle. J Bone Joint Surg Br. 1994;76:846–7.
26. Toledo LC, MacEwen GD. Severe complication of surgical treatment of congenital pseudarthrosis of the clavicle. Clin Orthop Relat Res. 1979;139:64–7.
27. Guyatt G, Oxman AD, Akl EA, Kunz R, Vist G, Brozek J, Norris S, Falck-Ytter Y, Glasziou P, deBeer H, Jaeschke R, Rind D, Meerpohl J, Dahm P, Schunemann HJ. GRADE guidelines: 1. Introduction – GRADE evidence profiles and summary of findings tables. J Clin Epidemiol. 2011;64:383–94.
28. Gouron R, Deroussen F, Juvet-Segerra M, Plancq MC, Collet LM. Reconstruction of congenital pseudarthrosis of the clavicle with use of the Masquelet technique. J Bone Joint Surg Am Case Connect. 2012;2:e77.

Evidence-Based Treatment of Sprengel Deformity

30

Talal Ibrahim and Abdelsalam Hegazy

Abstract

Various surgical procedures have been proposed for Sprengel deformity with the goal of improving shoulder function and cosmesis. They fall into two main categories: i) release of any omovertebral connection with resection of part of the scapula without an attempt to relocate the scapula, and/or ii) surgical procedures to relocate the scapula by muscle detachment at their origin or insertion with re-implantation. The indication for surgery, time of intervention, type of surgery are often based on surgeon's preference and expertise rather than evidence. A literature search revealed a small number of case series (level IV studies) with short to medium follow-up which preclude clear recommendation to aid decision making. In this chapter, we have summarised the available evidence and reported outcomes of the various surgical interventions for Sprengel deformity.

Keywords

Sprengel deformity • Congenital elevation of the scapula • Undescended scapula • Congenital anomaly of the shoulder • Woodward procedure • Green procedure • Mears Technique • Scapula osteotomy

Introduction

Sprengel deformity is defined as the congenital elevation of the scapula and is the most common congenital anomaly of the shoulder girdle [1, 2]. Although Eulenberg was the first to describe the deformity in 1863 [3], the name of the deformity has been attributed to Sprengel after the report of four cases in 1891 [4]. The scapula fails to descend between the 9th and 12th weeks of intrauterine life to its normal position leaving the scapula hypoplastic associated with abnormal development of the soft tissue in the shoulder girdle [5, 6]. The periscapular muscles are hypoplastic and develop contractures with time [7, 8]. In addition, the scapula is rotated medially with the inferior angle medialised and the glenoid facing inferiorly. Another anomaly associated with Sprengel deformity is the omovertebral connection extending from the superomedial border of the scapula to the cervical spine [6, 7]. The omovertebral connection has been reported to be present in approximately one-third of the patients with Sprengel deformity [6, 9, 10] and can be fibrous, cartilaginous or osseous. Furthermore, other associated abnormalities such as scoliosis, rib anomalies, torticollis and renal anomalies may be present [10, 11]. The aetiology and pathogenesis of this congenital deformity including the presence of the omovertebral connection remain unknown.

The main clinical features of Sprengel deformity include cosmetic deformity and functional shoulder limitation particularly abduction. The cosmetic deformity has been classified by Cavendish [10] and does not consider shoulder function as part of the grading system (Table 30.1; Figs. 30.1 and 30.2).

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Table 30.1 Cavendish classification

Grade	Description
Grade I (very mild)	Shoulder joints are level and the deformity is invisible when the patient is dressed
Grade II (mild)	Shoulder joints are level or almost level but the deformity is visible when the patient is dressed as a lump in the web of the neck
Grade III (moderate)	Shoulder joint is elevated 2–5 cm and the deformity is easily visible
Grade IV (severe)	Shoulder is much elevated with the superior angle of the scapula near the occiput, with or without neck webbing

Treatment

The decision of treatment of Sprengel deformity depends on several factors such as the cosmetic appearance (Cavendish grade – Table 30.1), functional shoulder impairment, presence of other anomalies, unilateral or bilateral deformities and the age of the patient.

Nonsurgical Treatment

Non-surgical treatment has been recommended for patients with mild Sprengel deformity. Non-surgical treatment includes modalities such as physiotherapy to improve the shoulder range of motion and prevent any torticollis. In the classic Cavendish study [10], different grades of Sprengel deformity were treated non-surgically. The study concluded that very mild (grade I) Sprengel deformity patients do not confer a worthwhile benefit with surgery. However, 34 of the 66 (52 %) patients treated non-surgically in the study were Cavendish grade II, III and IV Sprengel deformities [10].

Farsetti et al. [12] reported the long-term follow-up of 15 patients with Sprengel deformity treated non-surgically. At a mean follow-up of 26 years (range: 10–55 years), the final mean shoulder abduction was 125° (range: 90°–160°) and did not change over time. In addition, all patients were Cavendish grade I and II at final follow-up and remained in their same grade over time and were satisfied with the cosmetic outcome.

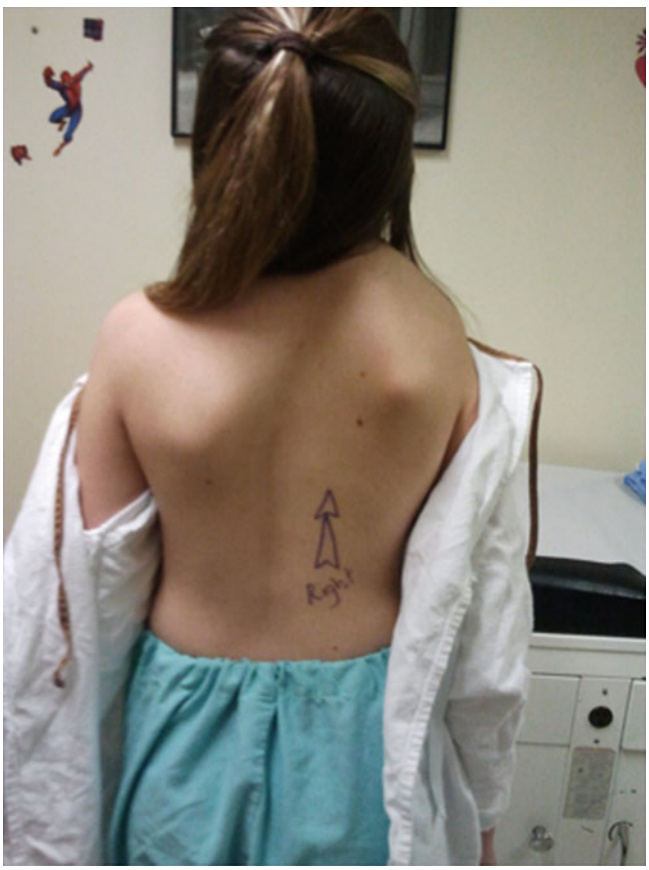

Fig. 30.1 Clinical photograph of a child with Sprengel shoulder

Surgical Treatment

Multiple surgical procedures have been described for the treatment of Sprengel deformity. The aim of surgical procedures is to lower the position of the scapula with respect to the chest wall with resection of the omovertebral connection if present. Soft tissue surgical procedures described in the literature include the release of either the muscular insertions at the medial border of the scapula or the corresponding origins with a relocation of the scapula inferiorly. Ross and Cruess [11] reported better results in patients whom undergo scapula relocation surgery compared to scapula excision in terms of both shoulder function and cosmesis.

The main goals of surgery for Sprengel deformity are to improve cosmetic appearance and shoulder function. Some authors believe that the cosmetic outcome is more important than the functional outcome for the majority of surgical procedures. The results of surgical treatment can be complicated by malformations and contractures of the soft tissue. Lowering the scapula to the level of the normal side is not advocated because of the increased risk of brachial plexus palsy. The optimal age for surgery remains controversial but has been suggested to be between 4 and 6 years old [1, 6, 13].

The most common surgical treatment for Sprengel deformity are the Woodward and Green procedures. A third category includes scapula osteotomies.

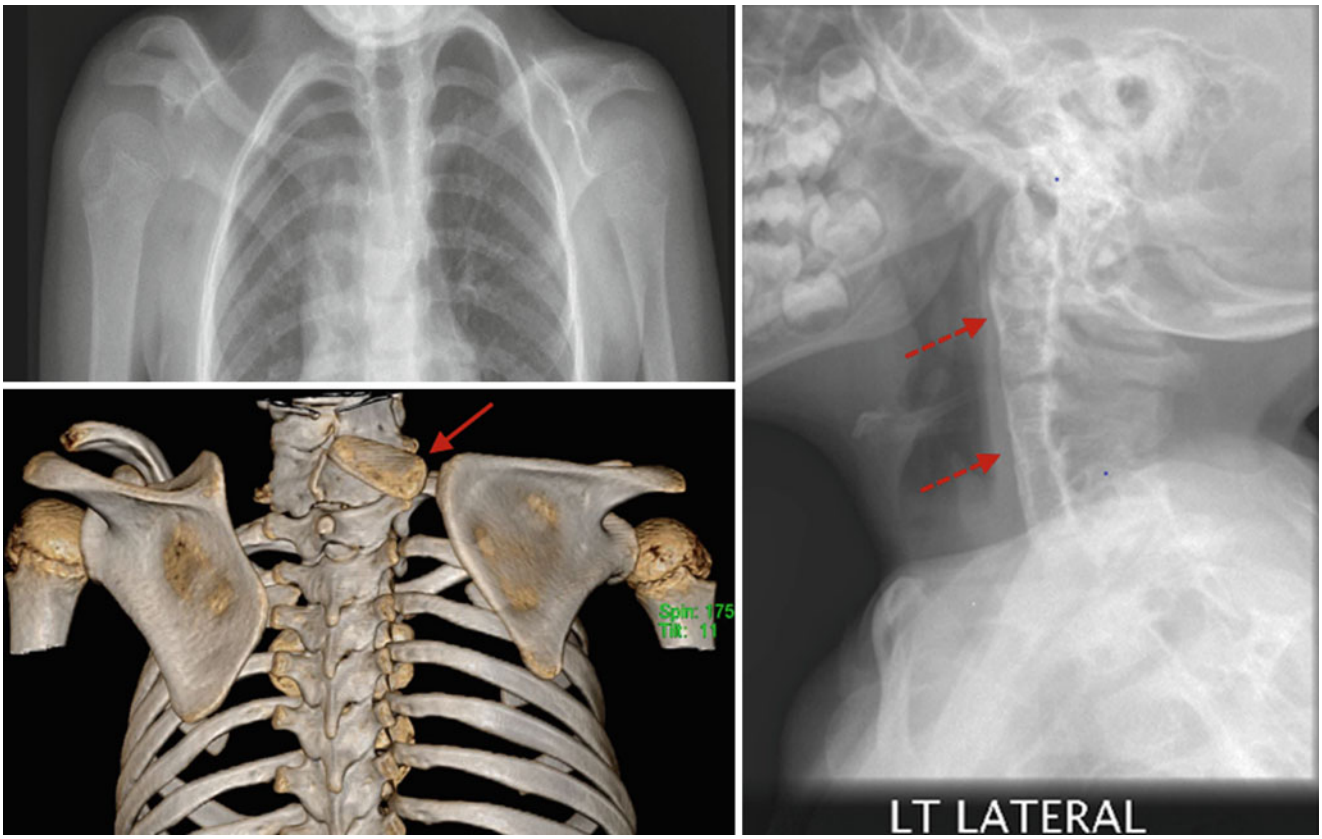

Fig. 30.2 Radiological findings in Sprengel shoulder. Plain radiographs and 3D CT-scan of a child with a right Sprengel shoulder. The right scapula is high riding. Cervical vertebrae fusion (*dashed arrows*) and omovertberal bony connection (*solid arrow*)

Woodward Procedure

The Woodward procedure was first described in 1961 and is stated more frequently in the literature than any other surgical procedure for Sprengel deformity [14]. In the Woodward procedure, the trapezius, rhomboid and levator scapulae muscles are detached from their origin along the spinous processes. The scapula is lowered and derotated with reattachment of the muscle to the spinous processes in a more inferior position. The muscles attached on the superior and medial borders of the scapula are reflected extraperiosteally. The procedure is accompanied with excision of any omovertebral connection present and a clavicle osteotomy if required.

Walstra et al. [15] reviewed seven patients (eight shoulders) who underwent the Woodward procedure and reported the long-term results at three different time intervals. The mean age at the time of surgery was 8.75 years (range: 3.4–15.1 years). At a mean follow-up of 13.5 years (range: 8–26 years), the mean shoulder abduction improved by 56° (range: 95°–175°). Cosmesis improved by at least one grade on the Cavendish scale in all patients. Other outcome measures such as the Constant, the DASH (Disabilities of the Arm, Shoulder and Hand) scores and simple shoulder test were also recorded. No long-term complications occurred in this case series. The authors concluded that the Woodward procedure is effective in improving shoulder function and cosmetic appearance in patients with Sprengel deformity in the long-term.

Similarly, Siu et al. [16] reviewed eight patients (nine shoulders) who underwent the Woodward procedure. At a mean follow-up of 113 months (range: 78–140 months), the mean shoulder abduction was 157° (range: 125°–180°). Cosmesis improved significantly on the Cavendish scale and all patients were satisfied with the results. The Constant score was also recorded for the patients. No complications were noted in this case series.

Other case series in the literature report the results of the Woodward procedure with satisfactory improvement in the mean shoulder abduction and cosmetic appearance according to the Cavendish grade [1, 6].

Modifications of the Woodward Procedure

Borges et al.

Borges et al. [13] modified the original Woodward procedure by adding an excision of the medial border of the scapula and resection of the supraspinatous portion of the scapula. Borges et al. [13] reviewed the long-term results of 15 patients (16

shoulders) in their original study who underwent the modified Woodward procedure. The mean pre-operative shoulder abduction was 115° (range: 90°–160°). At a mean follow-up of 8 years (range: 3.2–15 years), the mean shoulder abduction was 150° (range: 100°–180°). Cosmesis improved by at least one grade on the Cavendish scale in all patients. One patient developed a temporary brachial plexus palsy that resolved 3 months after a clavicular osteotomy. Borges et al. [13] concluded that the modified Woodward procedure correction did not change with growth of the patient and was maintained beyond skeletal maturity.

Ahmad

Ahmad [17] modified the Woodward procedure by combining the lowering of the scapula with correction of the varus position of the glenoid by placement of an absorbable suture through the superomedial portion of the scapula. This modification was associated with immediate post-operative range of motion of the shoulder in 11 patients (15 shoulders). At a mean follow-up of 36.5 months, the mean shoulder abduction was 139° (range: 90°–170°) translating into a gain of 49° of shoulder abduction. Cosmetic appearance improved in all patients. Three patients developed winging of the scapula post-operatively. The author concluded that this double correction results in an improvement of shoulder function and cosmesis.

Green Procedure

The Green procedure is one of the classic surgical procedures for the treatment of Sprengel deformity [18]. In the Green procedure, the muscles on the medial border of the scapula are detached from their insertions. The trapezius muscle is elevated extraperiosteally and reflected medially. The underlying medial and superior scapular musculature (latissimus dorsi, serratus anterior, levator scapulae, supraspinatus and rhomboids) are resected extraperiosteally. The supraspinatus fossa is excised to avoid damage to the neurovascular bundle. The omovertebral connection when present is also excised. The scapula is freed and mobilized distally to the appropriate level. Once the scapula is in the desired position, the muscle insertions are re-attached to the scapula lengthening each muscle as required. In the Green procedure, the scapula is both lowered and rotated. Muscle insertion is modified distally rather than proximally which provides better biomechanical outcome [19].

Modifications of the Green Procedure

Leibovic et al.

Leibovic et al. [20] modified the Green procedure by repositioning the scapula into a pocket that was developed in the latissimus dorsi. Leibovic et al. [20] reviewed 14 patients (16 shoulders) in their original study. The mean pre-operative shoulder abduction was 91° (range: 60°–120°). At a mean follow-up of 6.5 years (range: 3–14 years), the mean shoulder abduction was 148° (range: 100°–180°). Leibovic et al. [20] concluded that repositioning of the scapula in a pocket in latissimus dorsi did result in rotation of the scapula in the short-term but improvement was not maintained.

In another case series, Gonen et al. [21] reviewed 24 patients (28 shoulders) who underwent the Leibovic modified Green procedure and reported the long-term results. The mean age at the time of surgery was 4.5 years (range: 1.5–17 years). Seventeen shoulders were Cavendish grade III and 11 were grade IV. The mean pre-operative shoulder abduction was 101.3° (range: 70°–130°). At a mean follow-up of 11.3 years (range: 4.3–17 years), the mean shoulder abduction was 145.9° (range: 95°–175°). Cosmesis improved by at least one grade on the Cavendish scale in 89 % of patients. Two shoulders developed post-operative winging and hypertrophic scars in six shoulders. The authors concluded that the Leibovic modified Green procedure is relatively safe and reliable in the treatment of severe Sprengel deformity.

Bellemans and Lamoureux

Bellemans and Lamourex [22] modified the Green procedure by omitting the serratus anterior dissection and encouraging immediate shoulder range of motion exercises in seven children (7 shoulders). At a mean follow-up of 7.5 years (range: 5–13 years), the mean shoulder abduction was 170° (range: 145°–180°) translating into a gain of 77° of shoulder abduction. The authors concluded that preservation of the integrity of the serratus anterior muscle with early range of motion of the shoulder provided an improvement over the original technique.

Scapular Osteotomy

Vertical Scapular Osteotomy

Vertical scapular osteotomy was first described by Konig in 1914 [23] (Fig. 30.3). The procedure involves a medial vertical osteotomy approximately 1 cm from the vertebral border of the scapula allowing a downward displacement of the scapula. An osteotome is used to complete the osteotomy starting at the inferior angle moving upwards. All muscle attachments and fibrous bands are freed extraperiosteally from the superomedial angle of the scapula, which is then excised along with any omovertebral connection. The osteotomy is secured with silk sutures passed though drill holes. The patient remains in a sling for 6 weeks then starts full shoulder range of motion exercises.

Two case series have reported the results of vertical scapular osteotomy for Sprengel deformity. McMurtry et al. [24]

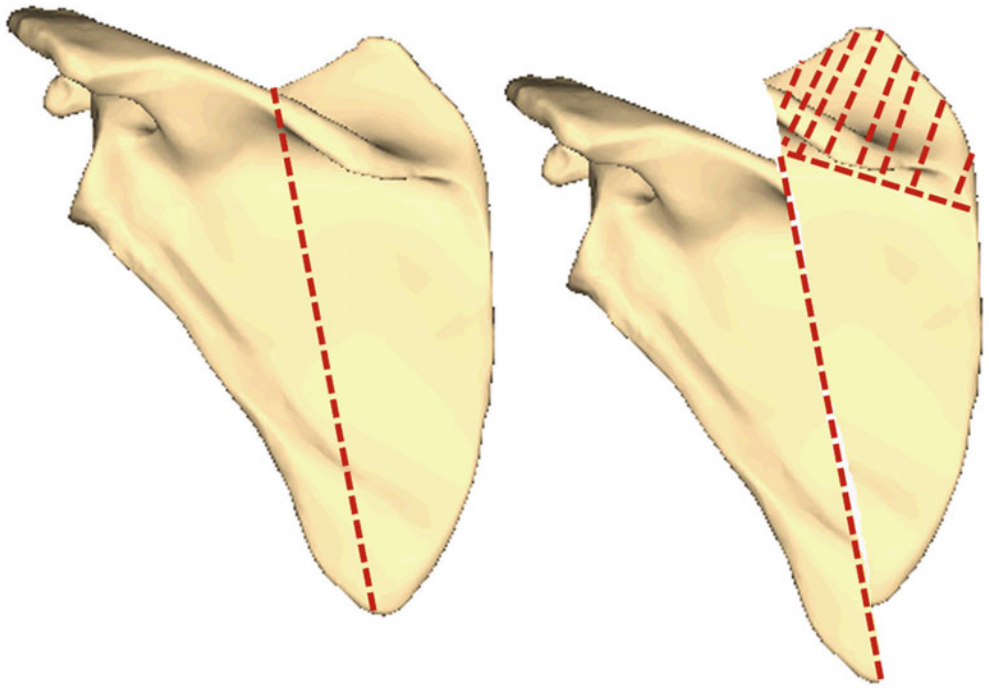

Fig. 30.3 A diagram showing Konig osteotomy for Sprengel deformity

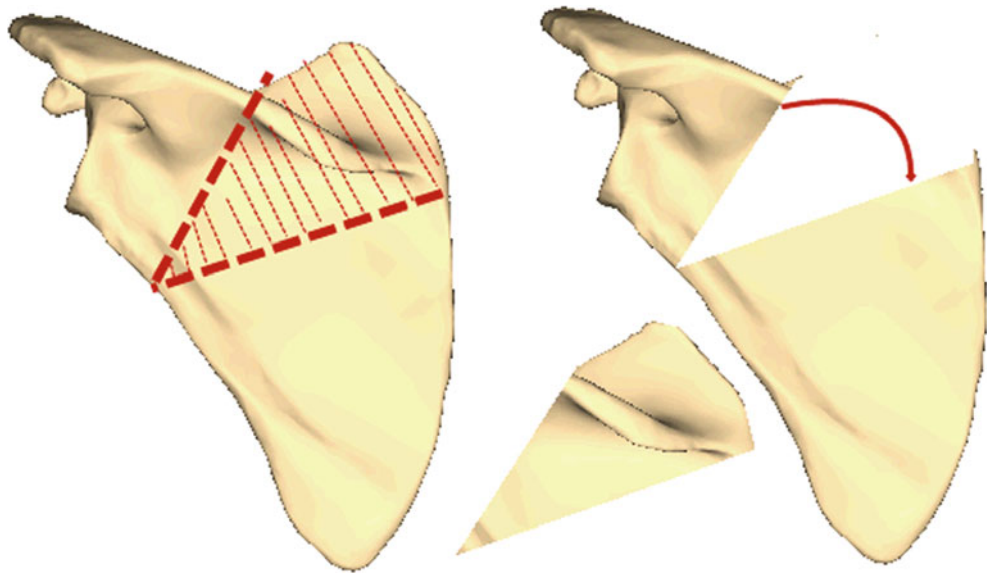

Fig. 30.4 A diagram showing Mears osteotomy for Sprengel deformity

reported the results of 12 patients (12 shoulders) over a 16-year period. Ten were Cavendish grade III, one grade IV and one grade II. The mean pre-operative shoulder abduction was 88° (range: 70°–180°). At a mean follow-up of 10.4 years (range: 1–17 years), the mean shoulder abduction was 132° (range: 50°–180°). Cosmesis also improved by a mean of 1.5 grades on the Cavendish scale. Neither function nor cosmesis deteriorated with time. One patient with obstetric brachial plexus palsy developed a recurrence of the upper plexus palsy that did not recover. Wilkinson and Campbell reported similar results in 12 patients (12 shoulders) over a 10-year period describing the procedure as safe and reliable [25].

Partial Resection of Scapula and Release of Long Head of Triceps (Mears Technique)

In 2001, a novel surgical technique was devised by Mears [26] (Fig. 30.4) that included a partial superomedial scapular resection, removal of any omovertebral connection and release of the long head of triceps from the scapula followed by early active and active-assisted post-operative shoulder

Table 30.2 Level of evidence

Level of evidence	References
Level III studies	[9, 11, 27]
Level IV studies	[1–8, 10, 12–26, 28]

Table 30.3 Summary of recommendations

Clinical questions	Recommendation
1. Surgical correction is recommended for Cavendish grade II and III Sprengel deformity?	C
2. Is there an optimal age to perform corrective surgery?	C
3. Are there any studies evaluating the superiority of one surgical procedure compared to others?	I

range of motion exercises. The procedure also includes an oblique osteotomy through the body of the scapula with resection to avoid bony impingement during full abduction. Mears [26] reported the results of eight patients (eight shoulders) with Sprengel deformity who were managed with this procedure. The mean age at the time of surgery was 5.8 years (range: 1.6–9 years). The mean pre-operative shoulder flexion and abduction were 100° (range: 90°–120°) and 90° (range: 85°–110°) respectively. At a mean follow-up of 5.5 years, the mean shoulder flexion and abduction were 175° (range: 170°–180°) and 150° (range: 100°–180°) respectively. Both shoulder function and cosmetic appearance improved significantly in all patients and no cases of brachial plexus palsy were recorded. Only one patient required surgery to excise residual exostosis.

In a prospective cohort study, Masquijo et al. [27] evaluated 14 patients (14 shoulders) who underwent scapular osteotomy with partial resection and release of the long head of triceps. The mean age at the time of surgery was 6.7 years (range: 4.7–10 years). Ten were Cavendish grade III and four were grade IV. The mean pre-operative shoulder flexion and abduction were 83.9° (range: 50°–120°) and 81° (range: 50°–120°) respectively. At a mean follow-up of 45 months (range: 12–74 months), the mean shoulder flexion and abduction were 152.1° (range: 110°–180°) and 145° (range: 100°–180°) respectively. Cosmesis also improved by a mean of two grades on the Cavendish scale. Two patients developed keloid scars and two other patients required further surgery to excise residual exostosis.

Partial Scapulectomy

In 1972, Cavendish treated 18 patients surgically with a partial excision of the superomedial portion of the scapula and excision of the omovertebral connection if present [10]. Most of the patients that benefited from this procedure were those with Cavendish grade II and III deformities or functional shoulder impairment. More recently, Zhang et al. [28] modified this procedure to release contracted tissue adjacent to the medial edge of the scapula and reviewed 26 patients (28 shoulders) at a mean follow-up of 3.9 years (range: 10 months to 7 years). The mean pre-operative shoulder abduction improved from 110° to 150° post-operatively. The majority of shoulders (23 shoulders) improved cosmetically following surgery. Furthermore, there were no neurological or scar complications following surgery. Hence, Zhang et al. [28] concluded that excision of the superomedial portion of the scapula and the omovertebral connection when present is a safe and effective procedure to treat Sprengel deformity.

Conclusion

Sprengel deformity is a rare condition and hence it is difficult to conduct prospective studies to clarify if one surgical procedure is superior to the other. Surgical treatment appears to be best indicated in patients with Cavendish grade II and III Sprengel deformity. Most studies in the literature are based on small number of patients and retrospective in nature. Some authors believe that patients who are older than 6 years of age are not suitable for a scapular displacement procedure [9, 10, 20]. The level of evidence and a summary of recommendations are provided in Tables 30.2 and 30.3, respectively.

References

1. Grogan DP, Stanley EA, Bobechko WP. The congenital undescended scapula. Surgical correction by the Woodward procedure. J Bone Joint Surg Br. 1983;65:598–605.
2. Pinsky HA, Pizzutillo PD, MacEwen GD. Congenital elevation of the scapula. Orthop Trans. 1980;4:288–9.
3. Eulenberg M. Beitrag zur dislocation der scapula. Amtliche Berichte uber die Versammlungen deutscher Naturforscher und Aerzte fur die Jahre. 1863;37:291–4.
4. Sprengel RD. Die angeborene Verschiebung der Schulterblattes nach oben. Archiv fur Klinische Chirurgie. 1891;42:545–9.
5. Horwitz AE. Congenital elevation of the scapula-Sprengel's deformity. Am J Orthop Surg. 1908;6:260–311.

6. Carson WG, Lovell WW. Whitesides TE Jr. Congenital elevation of the scapula. Surgical correction by the Woodward procedure. J Bone Joint Surg Am 1981; 63: 1199–1207.
7. Greitemann B, Rondhuis JJ, Karbowski A. Treatment of congenital elevation of the scapula. 10 (2–18) year follow-up of 37 cases of Sprengel's deformity. Acta Orthop Scand. 1993;64:365–8.
8. Khairouni A, Bensahel H, Csukonyi Z, Desgrippes Y, Pennecot GF. Congenital high scapula. J Pediatr Orthop B. 2002;11: 85–8.
9. Jeannopoulos CL. Congenital elevation of the scapula. J Bone Joint Surg Am. 1952;34:883–92.
10. Cavendish ME. Congenital elevation of the scapula. J Bone Joint Surg Br. 1972;54-B:395–408.
11. Ross DM, Cruess RL. The surgical correction of congenital elevation of the scapula. A review of seventy-seven cases. Clin Orthop. 1977;125:17–23.
12. Farsetti P, Weinstein SL, Caterini R, De Maio F, Ippolito E. Sprengel's deformity: long-term follow-up study of 22 cases. J Pediatr Orthop B. 2003;12:202–10.
13. Borges JL, Shah A, Torres BC, Bowen JR. Modified Woodward procedure for Sprengel deformity of the shoulder: Lon-term results. J Pediatr Orthop. 1996;16:508–13.
14. Woodward JW. Congenital elevation of the scapula: correction by release and transplantation of muscle origins. A preliminary report. J Bone Joint Surg Am. 1961;43-A:219–28.
15. Walstra FE, Alta TD, van der Eijken JW, Willems WJ, Ham SJ. Long-term follow-up of Sprengel's deformity treated with the Woodward procedure. J Shoulder Elbow Surg. 2013;22: 752–9.
16. Siu KK, Ko JY, Huang CC, Wang FS, Chen JM, Wong T. Woodward procedure improves shoulder function in Sprengel deformity. Chang Gung Med J. 2011;34:403–9.
17. Ahmad AA. Surgical correction of severe Sprengel deformity to allow greater postoperative range of shoulder abduction. J Pediatr Orthop. 2010;30:575–81.
18. Green WT. The surgical correction of congenital elevation of the scapula (Sprengel's deformity). J Bone Joint Surg Am. 1957;39:149–51.
19. Andrault G, Salmeron F, Laville JM. Green's surgical procedure in Sprengel's deformity: cosmetic and functional results. Orthop Traumatol Surg Res. 2009;95:330–5.
20. Leibovic SJ, Ehrlich MG, Zaleske DJ. Sprengel deformity. J Bone Joint Surg Am. 1990;72:192–7.
21. Gonen E, Simsek U, Solak S, Bektaser B, Ates Y, Aydin E. Long-term results of modified Green method in Sprengel's deformity. J Child Orthop. 2010;4:309–14.
22. Bellemans M, Lamourex J. Results of surgical treatment of Sprengel deformity by a modified Green's procedure. J Pediatr Orthop B. 1999;8:194–6.
23. Konig F. Eine neue operation des angeborenen sculterblattochstandes. Beitr Klin Chir. 1914;94:530–7.
24. McMurtry I, Bennet GC, Bradish C. Osteotomy for congenital elevation of the scapula (Sprengel's deformity). J Bone Joint Surg Br. 2004;87-B:986–9.
25. Wilkinson JA, Campbell D. Scapular osteotomy for Sprengel's shoulder. J Bone Joint Surg Br. 1980;62-B:486–90.
26. Mears DC. Partial resection of the scapula and a release of the long head of triceps for the management of Sprengel's deformity. J Pediatr Orthop. 2001;21:242–5.
27. Masquijo JJ, Bassini O, Paganini F, Goyeneche R, Miscione H. Congenital elevation of the scapula. Surgical treatment with Mears technique. J Pediatr Orthop. 2009;29:269–74.
28. Zhang ZM, Zhang J, Lu ML, Cao GL, Dai LY. Partial scapulectomy for congenital elevation of the scapula. CORR. 2006;457:171–5.

Part VII

The Elbow

Evidence-Based Treatment of Glenohumeral Dysplasia Caused by Obstetric Brachial Plexus Injuries

31

Philip Holland and Matthew F. Nixon

Abstract

Glenohumeral dysplasia is a disorder that occurs in the growing skeleton following a partial denervation of the muscles around the shoulder due to an obstetric brachial plexus palsy. Typically, there is an unopposed internal rotation force resulting in soft tissue and skeletal abnormalities. Management depends on a number of factors, but in the neonate consists of microsurgical reconstruction of the brachial plexus, and in the older child rebalancing of the muscles around the shoulder in the form of tendon transfers. Once skeletal deformities develop these need addressing in the form of reconstruction of the glenoid and humerus.

Keywords

Obstetric brachial plexus injury • Erb's palsy • Glenohumeral dysplasia • Glenoid retroversion • Humeral retroversion • Neurotisation • Tendon transfer • Osteotomy

Introduction

Obstetric brachial plexus injuries (OBPI) occur in approximately 0.1–0.4 % of births [1–3]. The risk factors for an OBPI include babies that are large for gestational age, multiparous pregnancies, prolonged labour, difficult deliveries and foetal distress [1, 4]). The incidence of OBPIs is falling which in part is thought to be due to advances in identifying and managing these risk factors [1, 4]).

An isolated upper trunk injury is the most common OBPI [4]. This causes denervation of some of the shoulder muscles leading to glenohumeral internal rotation contractures and dysplasia. Hand function is often well preserved. The aim of shoulder surgery is to prevent and reverse the development of glenohumeral contractures and dysplasia.

This chapter discusses the surgical management of the shoulder following isolated upper trunk OBPIs with or without involvement of the middle trunk.

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Methodology

The Medline, PubMed, and Google Scholar databases were searched to identify articles with the key term "obstetric brachial plexus injury." From the articles identified only articles which included one or more of the terms surgery, pathology, classification or shoulder in the title or abstract were included. Trial registries and the Internet were also searched. The references of articles identified were reviewed. Articles were graded according to the Oxford Evidence Based Grading System.

What Is the Natural History of Obpis?

The levels of evidence for published work included in this chapter ranged from II to V; there was no level I evidence.

Patterns of Nerve Injury

The first published descriptions of OBPIs were by Erb, Duchenne and Klumpke (Duchenne 1872 Erb 1874 Klumpke 1885). The terms Erbs Palsy and Klumpkes Palsy are

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commonly used in clinical practice to describe isolated upper trunk injuries and isolated lower trunk injuries respectively. This does not help differentiate between common clinical scenarios because isolated lower trunk injuries are extremely rare [5, 6].

OBPIs are typically caused by lateral neck flexion to the contralateral side during birth. This causes an upper trunk traction injury which in severe cases can extend to affect the middle and lower trunks. Narakas recognised this and produced a classification system that differentiates between the common clinical presentations. Narakas One injuries affect only the upper trunk (C5 to C6); Narakas Two injuries extend to involve the middle trunk (C5 to C7); Narakas Three injuries involve the upper middle and lower trunks (C5 to T1) and Narakas Four injuries involve the upper, middle, lower trunks and the stellate ganglion causing a Horner's Syndrome. The presence of a Horner's Syndrome signifies a preganglionic injury that is a relative contraindication to direct nerve repair [7–9]. Isolated lower brachial plexus injuries are very rare and are not included in the Narakas classification.

The characteristic abnormality at the shoulder in a Narakas One (upper trunk) OBPI is relative weakness of shoulder external rotation leading to an internal rotation contracture. This occurs because subscapularis function is preserved (upper and lower subscapular nerves) but infraspinatus and supraspinatus function is not preserved (suprascapular nerve). Of the posterior cuff muscles only teres minor function is preserved (axillary nerve) although this too is partly denervated. Teres minor is only a week shoulder external rotator when the arm is adducted and cannot compensate for the week infraspinatus.

Abduction weakness is also common in Narakas One (upper trunk) OBPIs, however, this is usually less disabling than the external rotation weakness [7, 9]. Abduction weakness occurs because the strong adductors subscapularis (upper and lower subscapular nerve), teres major (lower subscapular nerve), triceps (radial nerve) and latissimus dorsi (thoracodorsal nerve) have preserved innervation but the strong abductors supraspinatus (suprascapular nerve) and deltoid (axillary nerve) are denervated completely and in part respectively.

Most of loss of the abductor function of deltoid is likely to be because of the loss of the force couples from the rotator cuff rather than direct deltoid denervation. This is based on expert opinion reported in case series which have found deltoid to be well preserved and healthy at the time of surgery [10]. Furthermore, this is supported by experience in adult rotator cuff tears where supraspinatus dysfunction can severely reduce deltoid efficiency [11].

Narakas Two (upper and middle trunk) OBPIs affect the shoulder in a similar way to Narakas One OBPIs but the muscle paralysis is denser causing a more profound internal rotation and adduction contracture. This is because the cross over from C7 (middle trunk) to C5 and C6 (upper trunk) is lost.

A notable difference between a Narakas One (upper trunk) and a Narakas Two (upper and middle trunk) OBPI is that patients with a Narakas Two OBPI may develop a wrist drop. This is because ECRB and ECRL (posterior interosseous nerve) are innervated by C7. Despite this, patients with a Narakas Two OBPI often have good hand function.

Narakas Three (upper trunk, middle trunk and lower trunk) OBPIs and Narakas Four (upper trunk, middle trunk, lower trunk and stellate ganglion) OBPIs often result in poor arm and hand function.

The Natural History of Nerve Recovery

Some studies report that approximately two thirds of patients with OBPIs will have a spontaneous complete recovery. Noetzel et al. [12] followed up 80 patients on a monthly basis for 6 months. They found that a complete recovery occurred in 53 patients (66 %). Similar recovery rates were found by Sjoberg et al. [13] who found that 36 (75 %) of 48 patients made a complete recovery. Jackson et al. [14] followed up 19 patients and found that 15 (79 %) made a full recovery by 12 months. These are not population based cohorts and the methods used to detect persisting weakness are often subjective. This will affect the studies findings; however, most authors agree that between 60 % and 90 % of patients will make a complete recovery [15, 16].

Some research has investigated which patients are likely to make a complete recovery. Bisinella et al. [6] retrospectively reviewed 74 patients with OBPIs at a minimum of 2 years to investigate if the Narakas classification system is able to predict recovery. They found that normal function was achieved by 2 years in 38 % of patients with Narakas One OBPIs and 2 % of patients with Narakas Three OBPIs. Other studies have reported complete recovery in up to 95 % of Narakas One OBPIs [17].

Hoeksma performed a cohort study of 56 patients who were followed up for a mean of 3 years and 10 months. They found that biceps function at three months was the most reliable predictor of complete recovery. They also found that shoulder external rotation was the last function to recover and the least likely to recover [18]. Other studies have also found that antigravity elbow flexion is a good prognostic indicator of complete recovery and that shoulder external rotation is the often last movement to recover [7, 12].

Many patients with OBPIs will not make a full recovery and shoulder external rotation is the movement that is least likely to recover. Careful serial examination of patients with

OBPIs is essential to identify patients with weakness of external rotation. These patients should be followed up closely to identify early any internal rotation contractures or glenohumeral dysplasia.

The Natural History of Glenohumeral Dysplasia

Retroversion of the humeral head occurs because in Narakas One and Two OBPIs the muscle imbalance at the shoulder constantly internally rotates the humeral head. The child therefore externally rotates the arm for functional tasks leading to remodelling into external rotation of the humerus distal to the rotator cuff attachment. This manifests itself as retroversion of the humeral head. As retroversion of the humeral head increases it can sublux or dislocate. Changes in the glenoid occur secondary to this which can be a flat glenoid, a retroverted glenoid or a biconcave glenoid. Alongside the glenoid changes other changes also occur. The coracoid becomes elongated and hooked and the coracohumeral and glenohumeral ligaments become elongated and tight.

Pearl and Edgerton [19] found that 18 (72 %) of the 25 patients they operated on for OBPIs had glenohumeral dysplasia. This is a selected group of patients and it would be reasonable to assume that they would have found a lower incidence of glenohumeral dysplasia if they included patients who did not undergo surgery. Moukoko et al. [20] performed serial examinations on 134 consecutive patients with OBPIs. Those that they suspected had a posterior dislocation underwent an ultrasound scan and they found that 8 % of patients had a posterior dislocation.

Most of the pathological increase in humeral head retroversion occurs after 12 months of age. In an MRI study of children with OBPIs infants younger than 12 months had a mean humeral retroversion of 25.9° in the affected shoulder and 24.4° in the normal shoulder. Children older than 12 months had a mean humeral retroversion of 29.9° in the affected shoulder and 19.6° in the normal shoulder [21]. The difference in humeral head retroversion after 12 months of age was found to be statistically significant. These findings are supported by another MRI study that found that out of 25 children only 7 (28 %) had a congruent glenohumeral joint. In this study some children as young as two had severely advanced dysplasia [19]. It is likely that because infants under 12 months old do not use their arms much there is little remodelling and so no glenohumeral dysplasia develops. After 12 months of age children use their arms much more and dysplasia occurs which can be rapid in some patients [19, 22].

Clinical Assessment of OBPI

Internal rotation contractures are cosmetically disfiguring and functionally impairing. Serial examination can identify infants that are developing an internal rotation contracture early. The mallet score is commonly used in these serial assessments (Fig. 31.1). The Mallet score measures five movements, which are abduction, external rotation, hand to head, hand to back and hand to mouth. Each movement is graded out of five based on the range of movement with 5 being normal and 1 being no active motion.

The components of the Mallet score should recorded separately because some of the components can be normal in the presence of severe contractures. Changes in the individual components of the score are therefore considered a more accurate than the total score [7]). Bae et al. [23] investigated the reliability of the Mallet score. Eighty children with OBPIs were examined by two trained examiners on two occasions. They found good inter-observer and intra-observer reliability.

How to Prevent Glenohumeral Dysplasia?

Studies identified and included in this chapter were of levels III to V.

During the first 24 months of life there is a role for nerve surgery with the primary aim of reinnervating muscles. At the time of nerve surgery concomitant shoulder surgery should be performed if the patient has a structural abnormality [24]. After 24 months of life the primary aim of surgery should be to improve any glenohumeral abnormality.

Nonsurgical Treatment

All patients with an OBPI who have a shoulder that is congruently reduced should undergo physiotherapy to prevent contractures and maintain a functional arm. This requires an experienced therapist and parents who are able to engage with therapy at home. The aim of therapy is to encourage the child to use the affected arm without stressing them. Therapy should focus on external rotation with the shoulder in an adducted position. One technique that is used is to lay the child on the normal side and prompt them to play with a toy with the affected arm. Another way to prevent contractures is to encourage parents to position the arm in an abducted and externally rotated position on a pillow after the child has fallen asleep. Surgery should be considered if despite physiotherapy the patient develops a subluxed shoulder, a dislocated shoulder or has a progressive loss of external rotation [25].

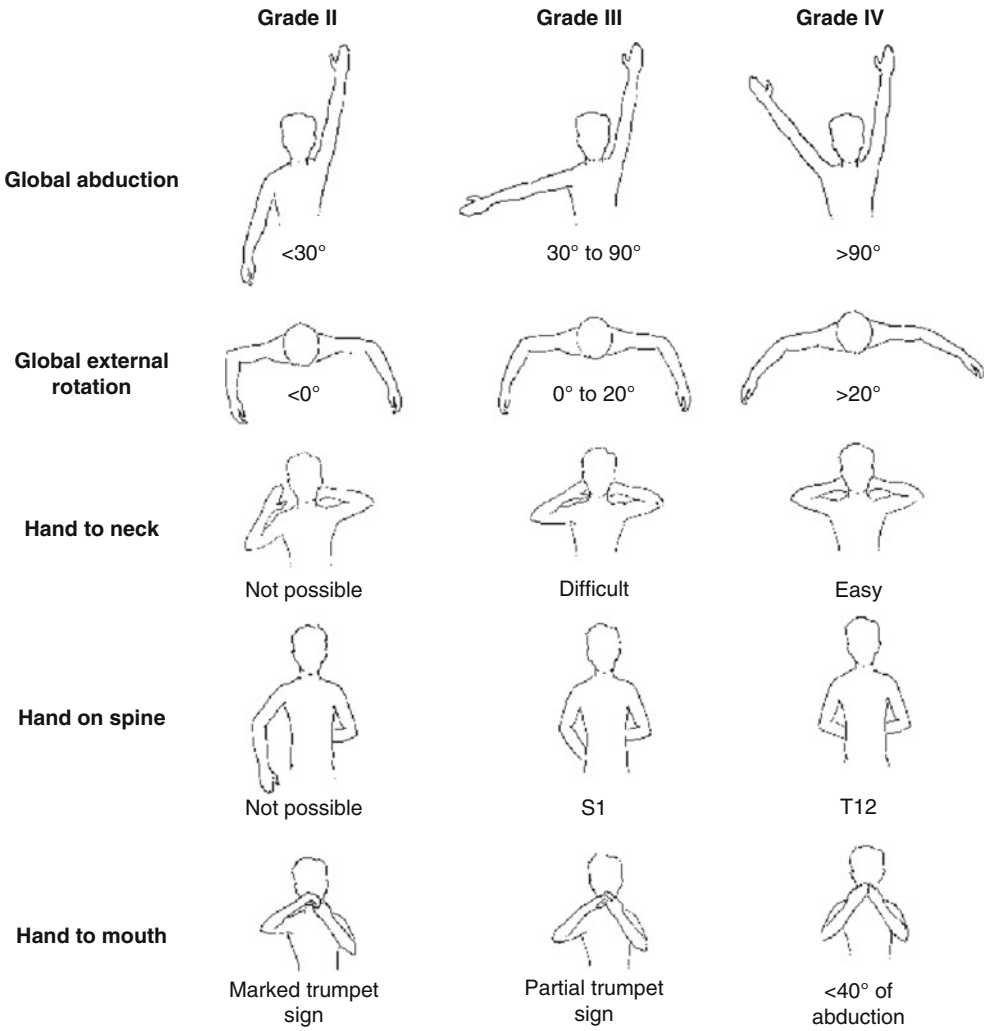

Fig. 31.1 The Mallet score

Subscapularis Release and Tendon Transfers

When nonsurgical treatment has failed and an internal rotation contracture with a congruent joint is present surgical lengthening of subscapularis with or without a tendon transfer to rebalance the external rotators should be considered.

Some surgeons advocate performing a tendon transfer at the time of a subscapularis release. Others advocate a tendon transfer if active external rotation has not been achieved by 2 years of age. A latissimus dorsi transfer to the greater tuberosity is the most commonly used tendon transfer [15, 25, 26]. In 1978 Hoffer et al. [25] wrote one of the earliest descriptions of tendon transfers for OBPIs. They described an open anterior release through a deltopectoral approach combined with a posterior incision to transfer latissimus dorsi to the greater tuberosity. They reported successful surgery on 11 children aged between 2 and 7 years.

Following Hoffer's study others have replicated there results. Thatte et al. [15] reported a series of 150 patients out of 305 patients treated for an OBPI. The surgery differed from that described by Hoffer in that the 150 patients all underwent a transfer of both latissimus dorsi and teres major. Patients were followed up for a mean of 4 years (range 2.5–8 years). They reported that all patients had an improvement in shoulder function and 35 (23 %) made a complete recovery.

Aydin et al. [26] reported a similar series of 46 patients who had a latissimus dorsi transfer and were followed up for a mean of 41 months. They analysed the results of those with severe contractures (<90° abduction) and moderate contractures (>90° abduction) separately. In the group with severe contractures they found a significant increase in abduction from 62.5° to 131.4° and external rotation from 21.4° to 82.6°. In the group with moderate contractures they found a significant increase in abduction from 99.4° to 140° and external rotation from 33.2° to 82.7°. There was no significant difference between the two groups. These study findings have been reproduced by others and demonstrate that even in the presence of severe contractures a latissimus dorsi transfer

can rebalance the shoulder muscles sufficiently to achieve improved external rotation and abduction [27].

Most studies report the short to medium term results of tendon transfers [5]. The longest follow up study we identified was by Pagnotta et al. [28] who followed up 203 patients who had undergone a latissimus dorsi transfer at 1, 3, 6, 10 and 15 years. They found that following a latissimus dorsi transfer abduction began to deteriorate at 6 years, however, external rotation was preserved. More long term studies are required to establish if there is deterioration in function over time and to what extent.

Tendon transfers of the lower trapezius to infraspinatus have been performed. This is usually as a revision procedure when latissimus dorsi and teres major have already been utilised. Using trapezius has the theoretical advantage of utilising a muscle that is not innervated by the brachial plexus. Trapezius also normally acts in phase with the posterior cuff. Bertelli et al. [29] reported the outcome in seven children at 2 and 4 years post surgery. They found the mean increase in external rotation to be 47° at 2 years and 54.3° at 4 years. We could not identify any other papers investigating trapezius transfers however it is a viable option and is worthy of further study.

Several studies have shown that in the short to medium term a latissimus dorsi transfer and anterior release can improve shoulder function.

How to Treat Established Glenohumeral Dysplasia?

When the glenohumeral joint is subluxed or dislocated but the articular surfaces are preserved joint reduction by soft tissue releases and osteotomies can be performed. A step wise approach to achieve this has been described by Di Mascio et al. [30]. They advocate that as little surgery as is necessary to achieve a congruent stable reduction throughout a functional range of motion should be performed. Step one is an open reduction; step two is a lengthening of subscapularis and an anterior capsule release; step three is an internal rotation humeral osteotomy and step four is a glenoplasty. They used this technique to treat 29 patients with a mean age of 5 years and reported the results at a mean follow up of 34 months. They reported a mean increase in abduction of 24° and external rotation of 54°.

Step One – Open Reduction

In Di Mascio's series they used an open reduction through a deltopectoral approach. This enables good access to the anterior structures and also allows access to the humeral neck for an osteotomy if required. None of the 29 patient in Di Mascio's series was treated by open reduction alone. We could not identify any case series reporting the outcome of open reduction without a muscle lengthening procedure in our literature search. An open reduction alone is not therefore considered a standard treatment.

Step Two – Lengthening of Subscapularis and Anterior Capsule Release

The coracoid is often elongated and is a physical barrier to reduction. In this situation a coracoid osteotomy is necessary to reduce the shoulder. This should be done before any other releases because a coracoid osteotomy releases its soft tissue attachments and improves access to other structures. If a coracoid osteotomy is not being performed or if despite a coracoid osteotomy a concentric reduction is not possible a subscapularis and pectoralis major lengthening should be performed. If this does not achieve a concentric reduction throughout the range of motion then the SGHL, MGHL, IGHL, CHL and rotator interval should all be fully released [30]. Brachialis and teres major may also be contracted and require releasing.

Most surgeons advocate that the releases are performed as an open operation, however, an arthroscopic release of the subscapularis and capsule alone may also be considered. This has the advantage of enabling a formal assessment of the glenoid which is difficult to image in infants. Pedowitz et al. [31] reported a series of 22 children undergoing an arthroscopic anterior capsule and subscapularis release to reduce subluxed shoulders. Post operatively they all were placed in spica casts. They found that they were able to reduce the shoulders successfully.

Kozin et al. [32] performed arthroscopic releases in 29 children; 16 of which also underwent tendon transfers. They followed these patients up at one year and found both an improvement in clinical results and a maintained reduction of the humeral head.

Pearl et al. reported similar results in their series of 33 children. The younger cohort of 19 children with a mean age of 1.5 years underwent an anterior release alone. The older cohort of 14 children with a mean age of 6.7 years underwent an anterior release and latissimus dorsi transfer. They found a significant improvement in external rotation in both groups. 18 of these patients' had a preoperative MRI demonstrating a pseudoglenoid and this had improved in 12 of the 15 children who had post operative MRI scans.

Not all series have shown good results using soft tissue surgery alone to treat established glenohumeral dysplasia. Hultgren et al. [33] followed up 118 patients for a minimum of 7 years. All patients had undergone soft tissue procedures alone to reduce subluxed glenohumeral joints. They reported a high failure rate with 66 (56 %) patients developing a subsequent subluxation and 23 (19 %) of these requiring a humeral osteotomy.

Step Three – Internal Rotation Humeral Osteotomy

Di Mascio et al. [30] described performing a derotational humeral osteotomy to restore a more normal humeral neck version when the retroversion is greater than that that which will remodel. Our literature search identified two papers reporting the outcomes of internal rotation humeral osteotomies. Sibinski et al. [34, 35] performed 10 internal rotation osteotomies on 25 patients undergoing a latissimus dorsi transfer. At a mean of 3.8 years there was an improvement in Mallet scores and internal rotation but other movements did not improve significantly.

Kambhampati et al. [10] performed a humeral osteotomy when humeral head retroversion was found to be more than 40° at the time of surgery. This was required in 70 (38 %) of the 183 shoulders that were operated on for glenohumeral dysplasia. They reported the results of all the patients treated and found a mean increase in external rotation of 58° and a mean increase in the Mallet score from 9.4 to 13. They reported 20 failures that required a further surgical reduction and glenoplasty.

Step Four – Glenoplasty

Di Mascio et al. [30] also report good results from a glenoplasty in their series of 29 patients who had a mean increase in external rotation of 54° with a mean loss of internal rotation of 7° at a mean of 34 months. They found no recurrent dislocations.

Kambhampati et al. [10] reported a failure rate of 29 % when a soft tissue procedure and humeral osteotomy was used. These 20 patients went on to have a glenoplasty which was reported to be successful at the time of surgery although the results from this are yet to be published.

There is a paucity of evidence for the outcomes of glenoplasty. There is potential for remodelling without glenoplasty however in some series the results from severe dysplasia are disappointing.

Conclusion

The natural history of OBPIs is well described and most patients with an OBPI will make a full recovery. Where weakness persists it is most likely to be weakness of external rotation. Many, but not all, patients with weakness of external rotation will develop glenohumeral dysplasia. There is no reliable way to predict which patients will develop severe symptomatic glenohumeral dysplasia.

There are numerous case series that demonstrate that patients with progressive glenohumeral dysplasia can maintain good shoulder function with surgery. It is generally accepted that an anterior release with or without a tendon transfer should be performed as early as is practical however the evidence for this is poor.

In the presence of glenohumeral dysplasia a soft tissue procedure alone may be sufficient to reduce the joint. There is then potential for remodelling over time. When severe glenohumeral dysplasia is present there is a high risk of recurrence and a glenoid or humeral osteotomy is likely to be required although the evidence as to which osteotomy should be used is poor.

References

1. Greenwald AG, Schute PC, Shiveley JL. Brachial Plexus birth Palsy: a 10-year report on the incidence and prognosis. J Pediatr Orthop. 1984;4(6):689–92.
2. Foad SL. The epidemiology of Neonatal Brachial Plexus Palsy in the United States. J Bone Joint Surg (Am). 2008;90(6):1258.
3. Pöyhiä TH et al. Brachial plexus birth injury: US screening for Glenohumeral joint instability 1. Radiology. 254(1):253–60.
4. Rayner CR, Hardy AE. Birth injuries of the brachial plexus. Incidence and prognosis. Plast Reconstr Surg. 1981;68(4):653.
5. Abzug JM, Kozin SH. Evaluation and management of brachial plexus birth palsy. Orthop Clin North Am. 2014;45(2):225–32.
6. Bisinella G. Obstetric brachial plexus lesions: a study of 74 children registered with the British paediatric surveillance unit (March 1998–March 1999). J Hand Surg: J Br Soc Surg Hand. 2003;28(1):40–5.
7. Al-Qattan MM et al. Narakas classification of obstetric brachial plexus palsy revisited. J Hand Surg Eur Vol. 2009;34(6):788–91.
8. Birch, R. Birth lesions of the brachial plexus, in surgical disorders of the peripheral nerves. London: Springer Science + Business Media; 1998. pp. 429–81.
9. Narakas A. Brachial plexus surgery. Orthop Clin North Am. 1981;12(2):303–23.
10. Kambhampati SBS. Posterior subluxation and dislocation of the shoulder in obstetric brachial plexus palsy. J Bone Joint Surg. 2006;88-B(2):213–9.
11. Shinozaki N et al. Differences in muscle activities during shoulder elevation in patients with symptomatic and asymptomatic rotator cuff tears: analysis by positron emission tomography. J Shoulder Elbow Surg. 2014;23(3):e61–7.
12. Noetzel MJ et al. Prospective study of recovery following neonatal brachial plexus injury. J Child Neurol. 2001;16(7):488–92.
13. SjÖBerg I, Erichs K, Bjerre I. Cause and effect of obstetric (neonatal) brachial plexus palsy. Acta Paediatr. 1988;77(3):357–64.
14. Jackson ST, Hoffer MM, Parrish N. Brachial-plexus palsy in the newborn. J Bone Joint Surg Am. 1988;70(8):1217–20.
15. Thatte M et al. Clinical outcome of shoulder muscle transfer for shoulder deformities in obstetric brachial plexus palsy: A study of 150 cases. Indian J Plast Surg. 2011;44(1):21.
16. Pondaag W et al. Natural history of obstetric brachial plexus palsy: a systematic review. Dev Med Child Neurol. 2004;46(02):138–44.
17. Foad SL et al. Prognosis following neonatal brachial plexus palsy: an evidence-based review. J Child Orthop. 2009;3(6):459–63.
18. Hoeksma AF et al. Neurological recovery in obstetric brachial plexus injuries: an historical cohort study. Dev Med Child Neurol. 2004;46(02).
19. Pearl ML, Edgerton BW. Glenoid deformity secondary to brachial plexus birth palsy. J Bone Joint Surg Am. 1998;80(5):659–67.

20. Moukoko D et al. Posterior shoulder dislocation in infants with neonatal brachial plexus palsy. J Bone Joint Surg Am. 2004; 86-A(4):787–93.
21. Edelson G. Variations in the retroversion of the humeral head. J Shoulder Elbow Surg. 1999;8(2):142–5.
22. Waters PM. The early effects of tendon transfers and open capsulorrhaphy on glenohumeral deformity in brachial plexus birth palsy. J Bone Joint Surg (Am). 2008;90(10):2171.
23. Bae DS, Waters PM, Zurakowski D. Correlation of pediatric outcomes data collection instrument with measures of active movement in children with brachial plexus birth palsy. J Pediatr Orthop. 2008;28(5):584–92.
24. Grossman JAI et al. Outcome after later combined brachial plexus and shoulder surgery after birth trauma. J Bone Joint Surg (Br). 2003;85(8):1166–8.
25. Hoffer MM, Wickenden R, Roper B. Brachial plexus birth palsies. Results of tendon transfers to the rotator cuff. J Bone Joint Surg Am. 1978;60(5):691–5.
26. Aydin A, Ozkan T, Onel D. Does preoperative abduction value affect functional outcome of combined muscle transfer and release procedures in obstetrical palsy patients with shoulder involvement? BMC Musculoskelet Disord. 2004;5:25.
27. Ozturk K. Reconstruction of shoulder abduction and external rotation with latissimus dorsi and teres major transfer in obstetric brachial plexus palsy. Acta Orthop Traumatol Turc. 2010;44:186–93.
28. Pagnotta A, Haerle M, Gilbert A. Long-term results on abduction and external rotation of the shoulder after Latissimus Dorsi transfer for sequelae of obstetric palsy. Clin Orthop Relat Res. 2004;426: 199–205.
29. Bertelli JA. Lengthening of subscapularis and transfer of the lower trapezius in the correction of recurrent internal rotation contracture following obstetric brachial plexus palsy. J Bone Joint Surg. 2009;91-B(7):943–8.
30. Di Mascio L et al. Glenoplasty for complex shoulder subluxation and dislocation in children with obstetric brachial plexus palsy. J Bone Joint Surg. 2011;93-B(1):102–7.
31. Pedowitz DI et al. Arthroscopic treatment of posterior glenohumeral joint subluxation resulting from brachial plexus birth palsy. J Shoulder Elbow Surg. 2007;16(1):6–13.
32. Kozin SH et al. Arthroscopic treatment of internal rotation contracture and glenohumeral dysplasia in children with brachial plexus birth palsy. J Shoulder Elbow Surg. 2010;19(1):102–10.
33. Hultgren T et al. Surgical correction of shoulder rotation deformity in brachial plexus birth palsy: long-term results in 118 patients. Bone Joint J. 2014;96-B(10):1411–8.
34. Sibiński M, Synder M. Soft tissue rebalancing procedures with and without internal rotation osteotomy for shoulder deformity in children with persistent obstetric brachial plexus palsy. Arch Orthop Trauma Surg. 2010;130(12):1499–504.
35. Sibinski M, Hems TEJ, Sherlock DA. Management strategies for shoulder reconstruction in obstetric brachial plexus injury with special reference to loss of internal rotation after surgery. J Hand Surg Eur Vol. 2012;37(8):772–9.

Evidence-Based Treatments of Paediatric Elbow Fractures

32

Emily J. Mounsey and Andrew Howard

Abstract

Fractures around the elbow are common in children and account for a large proportion of both paediatric fracture clinics and paediatric trauma lists. Many of these injuries can be managed simply with good functional results; however, the potential for disabling consequences of these fractures remains and the broad spectrum of fracture variants and severities can continue to challenge experienced clinicians. In this chapter, we examined the evidence relating to the management of common fractures around the elbow in an attempt to answer some of the important clinical questions, as well as identify those that remain unresolved.

Keywords

Children elbow fracture • Supracondylar fracture • Lateral condyle fracture • Medial condyle fracture • Radial head fracture

Introduction

Injuries around the elbow make up a large proportion of both paediatric fracture clinics and, particularly, paediatric trauma lists. Representing 5–10 % of all fractures in children, many of these injuries can be managed simply with good functional results. The potential for disabling consequences of these fractures remains, and the broad spectrum of fracture variants and severities can continue to challenge experienced clinicians. We will examine the evidence relating to the management of common fractures around the elbow in an attempt to answer some of the important clinical questions, as well as identify those that remain unresolved.

Much of paediatric orthopaedic practice is not evidenced based, and it is likely that some aspects of management will never be vigorously trialled. We include a review of level one and two evidence related to the topic as well as relevant retrospective comparative level three evidence and level four case series. We also comment on two recent guidelines regarding supracondylar fractures. Guidelines are an accessible distillation of best available evidence and ideally clinicians should spend less time with the primary literature in areas where guidelines exist. We will discuss how to evaluate a guideline, and the strengths and limitations of attempts to create guidelines.

Supracondylar Humeral Fractures

Supracondylar humeral fractures represent 50–70 % of all paediatric elbow fractures [1] (Fig. 32.1). An understanding of the unique anatomy of the elbow with its narrow surface area of bone at the medial and lateral columns, as seen on the lateral radiograph, is essential when making management decisions. Any instability of the fracture is likely to result in rotation, with the distal fragment tending to internally rotate then drop into varus misalignment.

Supracondylar fractures are divided into displaced and undisplaced injures. Undisplaced fractures can be treated non-operatively with immobilisation in an above elbow cast or splint [2]. The Cuomo case series includes all consecutively enrolled patients, and documents 100 % treatment success with posterior splinting. Case series are more valid if

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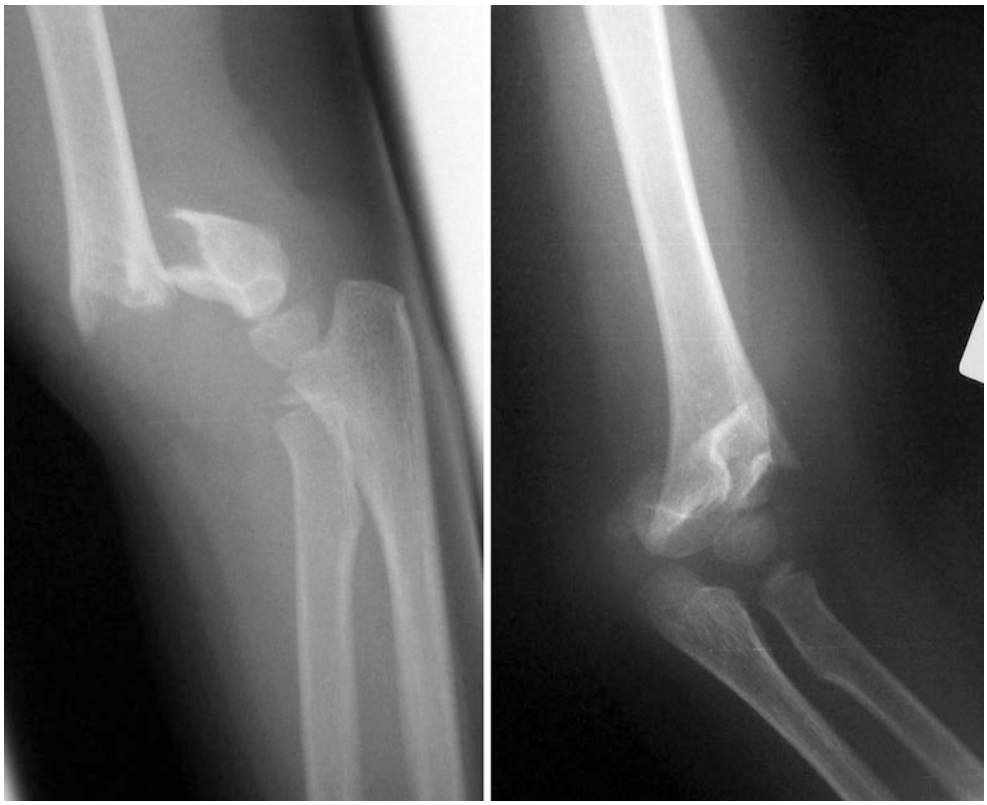

Fig. 32.1 Supracondylar humeral fracture

patients are enrolled consecutively and fully accounted for, and are most useful in assessing 100 % or 0 % outcomes ('penicillin and parachutes' studies).

Closed reduction and percutaneous pinning (CRPP) of displaced fractures yields predictable results with the fewest complications when compared to alternatives such as closed reduction and cast immobilisation or traction. Pirone's classic retrospective comparative study published in 1988 compared 4 methods of management; closed reduction and casting (130 patients), closed reduction and percutaneous K-wire fixation (78 patients), traction (15 patients), and open reduction with internal fixation (7 patients). Of the 130 patients managed in cast, 29 had conversion to K-wire fixation due to circulatory problems experienced due to hyperflexion of the elbow, 14 had varus malunion, 6 had reduced range of movement and 1 patient developed Volkmann's ischaemic contracture. In the group managed with K-wire fixation there were 3 varus malunions and 2 patients who experienced loss of motion. Despite several patients who had initially absent radial pulse with a well-perfused hand, no patient in the K-wire group developed Volkmann's ischaemic contracture. Approximately half had all lateral wires, and half crossed pins, there were no pin-related nerve complications but there were 2 pin infections. At final follow-up 78 % of patients managed with K wires had both carrying angle and range of motion within 5° of the contralateral side, compared to 51 % of the group treated with cast [3]. Although this study has some potential for bias (better, or worse patients may have been selected for the new technique of closed pinning so results may indicate patient selection rather than treatment effectiveness), the differences in important outcomes were compelling and all favoured closed reduction and pinning, which was increasingly adopted following the results of this influential paper. Given the harm (ischemic contracture, malunion) documented in the closed reduction and cast group, it is impractical to propose a randomized trial comparing these treatments at present.

There is a grey zone regarding the management of minimally displaced Gartland type 2 injuries. Care should be taken particularly in those fractures that have medial comminution and/or initial varus displacement. This pattern of injury can be more unstable than the initial radiographs suggest leading to medial collapse and varus malunion if managed non-operatively [4, 5]. A retrospective review of 189 type 2 supracondylar humerus fractures managed operatively concluded that following CRPP there was a high probability of satisfactory outcome with a low complication rate (4 pin tract infections, no loss of reduction) [6]. The American Academy of Orthopaedic Surgeons (AAOS) [7] guidelines of the treatment of paediatric supracondylar humerus fractures advise that all Gartland type 2 and 3 fractures be managed with CRPP. This management is supported by the practice at our institution as well as others [5, 8].

There has been debate regarding the most appropriate placement of percutaneous pins. Initial descriptions of

percutaneous pinning techniques described crossed pins inserted through medial and lateral entries, whereas recent literature emphasizes the importance of all-lateral pinning constructs in avoiding iatrogenic injury to the ulnar nerve. Excellent outcomes have been reported with all lateral pins that are placed in a bi-cortical fashion that avoid crossing, and give adequate spread at the fracture site [9]. Kocher published a small randomized trial showing no difference in outcome between crossed or lateral entry pins either from displacement or ulna nerve injury [10], but the trial was underpowered to detect differences in ulnar nerve injury which is a rare outcome. A systematic review in 2007 reported an iatrogenic nerve injury rate of 1.9 % in all lateral pins and 3.5 % for crossed pins with a loss of reduction of 0.7 % in lateral entry pins and 0 % in crossed pins [11]. A further meta-analysis in 2012 [12] showed a 4.3 times greater risk of ulna nerve injury with crossed pins. Others showed a decrease in ulna nerve injury of 15–2 % by using all lateral pins with selective use of a medial pin if the fracture was deemed unstable [13]. Well placed all lateral insertion of pins yield a construct that is 'strong enough' to prevent displacement and avoids the higher risk of iatrogenic ulna nerve injury. The British Orthopaedic Association Standards for Trauma (BOAST) guidelines advise that 2 mm diameter wires should be used where possible [14]. The AAOS guideline carefully considered all of the published literature reporting on pin placement and nerve outcomes. Both case series and comparative studies were used to provide the most robust estimate of ulnar nerve lesions from medial pin placement. The pooled estimated was a 6 % risk of iatrogenic ulnar nerve injury (49 of 808 patients in published series) and this was sufficient for AAOS to recommend stabilization of displaced supracondylar fractures with two or three laterally inserted wires [7]. Language in the guideline permits the use of medial wires if the specific fracture anatomy cannot be adequately stabilized with lateral pins only – this may occur, for example, if an oblique fracture line runs from proximally medially to distally laterally.

There is a wide variation across the literature regarding the need for open reduction suggesting that we do not yet have a clear statement on what can be called an acceptable reduction. Routine open reduction has been shown to cause increased stiffness at 2-year follow-up and is not advised [15]. There is limited evidence to suggest that using open reduction techniques will improve reduction [7]. Fracture, surgeon, and hospital factors are taken into account and it is possible that a low rate of open reduction, seen in higher volume centres, is not as attainable or desirable in lower volume practices. In evaluating the results of a reduction all would agree that anatomical reduction, if attainable, is optimal. Varus malunion, so called gunstock deformity must be avoided as this is cosmetically unacceptable to many patients and is the most common reason for revision operation. Small amounts of medial and lateral translation, anterior or posterior translation, or angulation in the sagittal plane may be consistent with perfect functional results, but one of the biggest gaps in the literature is clear information about what position short of anatomic reduction is acceptable (Fig. 32.2).

Neither the BOAST nor the AAOS guidelines are able advise on out of hours surgical intervention unless there are indications for urgent surgery. These indications include absent radial pulse, clinical signs of impaired perfusion of the hand and evidence of threatened skin viability [7, 14]. A retrospective study in 2001 showed that supracondylar fractures operated within 8 h had no difference in outcome with regard to conversion to open reduction, pin track injections or iatrogenic nerve injury compared to those operated on after 8 h from injury [16]. Other groups support these findings, with one retrospective study also showing no differences in quality of reduction once united [17, 18]. One study did show an increased need for open reduction when fractures were operated on after 8 h, 33 % open reduction rate in 45 patients. They also had a higher than usual requirement for open reduction for those operated within 8 h, 11.2 % of 126 patients, which brings into question their threshold for open reduction, and whether this is a suitable outcome measure [19]. The BOAST guidelines suggest that supracondylar fractures without indications for urgent surgery require early surgical treatment, ideally on the day of admission but not during the night. This practice is supported in our institution.

Injuries with vascular compromise, specifically those with an absent radial pulse can be divided into two distinct groups. One is the pale, cold pulse-less limb that does not improve after reduction; this type of injury should be explored and the brachial artery identified and repaired if necessary. The second is the perfused pulse-less hand, specifically an absent radial pulse with good capillary refill, good skin colour, and a warm extremity. There is mixed literature on this subject, with two series reaching opposite conclusions from similar clinical data. One study explored all injuries with a pink pulseless hand [20], the other managed all with observation [21]; in both cases patients did well, with no circulatory problems at follow-up. Another study divided cases into those that were explored and had vascular repair, and those that were managed expectantly. At follow-up ten patients were imaged using magnetic resonance angiography, five showed occlusion or re-stenosis of the brachial artery despite repair, all had a radial pulse present [22]. BOAST guidelines suggest that a perfused limb does not require brachial artery exploration whether or not the radial pulse is present. For those injures where the limb remains ischemic after fracture reduction, a surgeon competent to perform small vessel vascular repair should explore the brachial artery [14].

Fig. 32.2 Deformities after supracondylar humeral fractures. *Top* images: extension deformity of the distal humerus. *Bottom left*: valgus deformity of the distal humerus. *Bottom right*: varus deformity of the distal humerus (often called gunstock deformity)

The majority of associated nerve injuries in supracondylar fractures are transient neurapraxias that can be managed expectantly. It is worth considering that if there is neurological compromise pre-operatively and it is not possible to achieve a perfect reduction, then the nerve may be lodged in the fracture site. If there is suspicion of an iatrogenic nerve injury, any medially placed wire should be removed, and consideration should be given to surgical exploration [14].

The duration of immobilisation following percutaneous pinning varies in the literature. The AAOS drew inconclusive recommendations on the optimal time for pin removal and mobilisation. In our institution pins are removed at 3 weeks, and children are allowed to mobilise as tolerated. Physiotherapy is not routinely used in our institution. The AAOS guidelines were unable to recommend either for or against the use of physical therapy as there are no high quality studies documenting any long-term benefit of the intervention. The rapid and near complete return of range of motion experienced by most children suggests that the role of physical therapy might be restricted to the small number of patients who have persistent stiffness a few weeks after pin removal.

The AAOS guideline on paediatric supracondylar fractures was created because this is such a common fracture with the potential for significant associated injuries, and significant complications. The guideline supports closed reduction for displaced fractures, with percutaneous pinning using two or three lateral entry pins. AAOS applies strict criteria to the inclusion and evaluation of evidence with a consistent approach for all the guidelines it creates [23]. An initial meeting of a clinical panel is used to carefully define 15 key questions, which will produce the recommendations. Clinicians from academic and community practices and from all specialties involved in the care are represented.

Expert methodologists guide the process of guideline development. Trained methodologists and medical librarians do literature searches and abstraction. Only the highest level of evidence is considered in formulating a particular recommendation however both quality and quantity criteria apply – so, for example, a small underpowered but randomized trial will not exclude the consideration of larger retrospective studies. The literature review document for this guideline, prepared by the methodologists, ran to some 1200 pages and included 1726 citations, with 1682 articles finally excluded, leaving 44 to be considered for the recommendations [7]. Novel meta-analyses were performed for some questions including the rate of ulnar nerve injury from medial placement of pins. A second meeting of the clinical panel confirmed the application of the literature review to the answers to clinical questions. Level of evidence and quality of evidence are both considered in determining the strength of a particular recommendation. For many sensible clinical questions, there simply is not enough evidence to make evidence based recommendations and an inconclusive statement is required. An exception is in the case of potential life or limb outcomes, in which case a consensus recommendation is permitted – as in the case of a vascular injury. The advantage of adhering strictly to process is that the guideline itself should be without bias. The potential for practice guidelines to be either biased or conflicting has been demonstrated in, for example, discrepancies in recommendations regarding thromboprophylaxis generated by orthopaedic surgeons versus haematologists versus chest physicians working from the same literature [24]. A disadvantage of a strict adherence to methodology is that the resulting guideline may be seen as predictable, unremarkable, indecisive, or difficult to use in the clinical setting. AAOS has addressed this disadvantage by creating appropriate use criteria (AUCs) for supracondylar fractures based on the guideline [7] (Fig. 32.3). An AUC is a web-based tool designed for easy application. The clinical scenario is input into the tool, and the tool presents treatments with a rating of how appropriate each treatment is for that particular situation. The appropriateness is based on a clinical panel voting explicitly on how the guideline applies to that clinical setting. Clinicians should resist the temptation to read and apply individual studies in isolation now that systematic summaries of the literature exist.

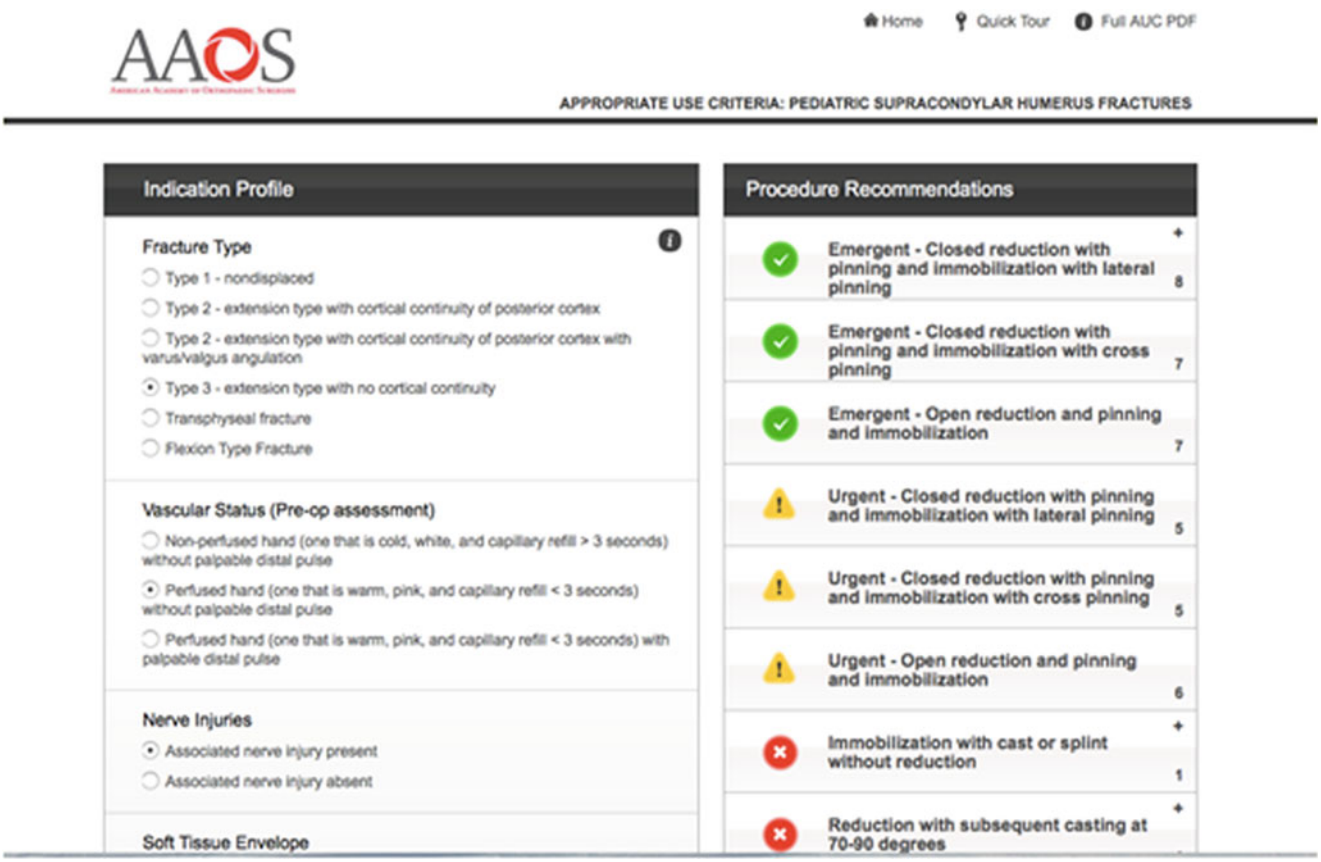

Fig. 32.3 The AAOS clinical practice guideline. The AAOS clinical practice guideline has been turned into an appropriate use criteria or AUC as shown here. The user can select a combination of clinical circumstances, and view treatment recommendations considered most appropriate by an expert panel applying the guideline to each situation

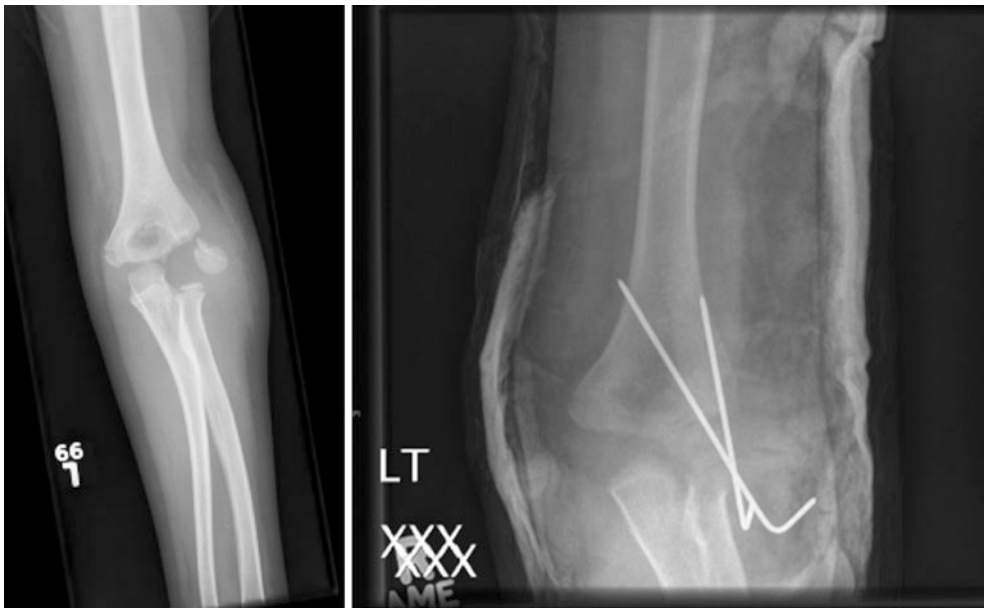

Fig. 32.4 Lateral condyle fracture of the elbow

Lateral Condyle Fractures

Lateral condyle fractures can be difficult to diagnose and have a propensity for late displacement. Compared to other paediatric fractures they have a higher risk of non-union, which, if allowed to occur can result in progressive valgus deformity of the elbow and tardy ulna nerve palsy (Fig. 32.4).

An internal oblique radiograph is the most sensitive view to ascertain the displacement of a lateral condyle fracture of the elbow. This was shown in a prospective cohort of 54 patients where 70 % of patients were noted to have different amounts of displacement of the fracture on an internal oblique view compared to standard antero-posterior and lateral images [25]. For undisplaced or minimally displaced fractures MRI studies can be useful to determine the stability of the fracture. A prospective cohort of 16 patients was imaged with both radiographs and MRI. There were 12 patients who had radiographically unstable (>3 mm displacement) lateral condyle fractures; of these 10 had an intact articular cartilage hinge, none of these displaced, of the 2 who had disrupted articular cartilage, one displaced [26]. Studies regarding the management of 'undisplaced' fractures (less than 2 mm displacement on internal oblique view) have shown mixed reports. One retrospective comparative series showed poor result with cast immobilisation alone; of the 17 patients 5 displaced, 4 required later surgery, 2 malunited and 2 went on to non-union. For the 13 treated with open K-wire stabilisation, 2 lost reduction and malunited [27]. Conversely a large case series reported 95 non-operatively managed lateral condyle fractures that had <2 mm initial displacement; 93 healed and only 2 displaced then went on to unite when treated with open reduction and K wire fixation [28]. Neither of these studies used MRI in their diagnostic algorithm. MRI may have supplied further prognostic information regarding the stability of the fracture and some explanation for the discrepancy in their findings.

It is well reported that displaced lateral condyle fractures require accurate reduction, usually open, then fixation. A biomechanical study showed that greater than 60° divergent pins yielded better stability than parallel pins [29]. Screw fixation is an alternative to pin fixation, which is, advocated in some case series however there is inadequate comparative information to recommend one over the other. Screw fixation has been shown to have increased biomechanical stability in synthetic bone [30]. However a comparative cohort study comparing 44 patients managed with screw osteosynthesis and 33 patients managed with K-wires found that K-wire fixation had comparatively similar outcomes to screw fixation, with no fracture going on to non-union in either group [31]. In our institution patients are usually immobilised post-operatively for 3 weeks then examined clinically and radiographically following pin removal. The rate of non-union after 3 weeks of pin fixation was 1 in a consecutive series of 104 patients [32].

Radial Neck Fractures

There have been a number of retrospective studies that attempt to compare closed versus open reduction of radial neck fractures, however all suffer from selection bias since the majority managed closed are minimally displaced and those that require open reduction tend to be more displaced and fail closed

reduction methods. A retrospective cohort in 1978 [33] reported 43 children with radial neck fractures followed for 8 years. 'Good' outcomes were reported in 19 of the 23 patients managed non-operatively and only 5 of the 14 treated with open reduction. Only one of the children treated non-operatively compared to half of those treated with surgery had displacement of more than 60°. In 1993 another retrospective cohort compared 100 children with radial neck fractures. Excellent and good results were consistently obtained after closed manipulation with disappointing results following open reduction reported. All fractures managed with open reduction had angulation of 30–140° [34]. These and other studies are reassuring that excellent results can be expected when managing minimally displaced radial neck fractures, but do not aid decision-making with regard to fractures that remain displaced despite closed reduction techniques (Fig. 32.5).

In an attempt to improve the understanding of displaced fractures that required open reduction, a retrospective cohort reviewed 24 patients who required open reduction, this represented 12 % of their total radial neck fractures. Fifty-five percent of patients were deemed to have a good outcome at mean follow-up of 7 years, with the remaining having fair or poor results. Their only poor prognostic indicator at the time of the injury was a second associated elbow injury. They concluded, without a comparative group, that they had better outcomes using open reduction than if the fracture had remained non-anatomically reduced, where the radius is completely displaced and without contact with the rim of the metaphysic [35]. A further retrospective comparative study compared 78 children with radial neck fractures. Their treatment methods were dictated by the severity of the case, which they found correlated with outcome. Patients with less complex injuries, managed non-operatively, had higher rates of excellent or good outcomes compared to those with more severe fractures that required operative treatment. Nineteen out of twenty six patients who underwent percutaneous reduction had excellent or good outcomes compared to 6/17 that required open reductions. Interestingly they did not find a correlation between outcome and associated injury [36]. Many of the disappointing outcomes relating to open reductions of displaced fractures are related to stiffness, since capsular dissection is required. There is also a risk of damage to the blood supply of the radial head. For this reason there has been a focus on operative reduction and fixation techniques that avoid open surgery.

In 1992 a case series of 36 consecutive fractures displaced more than 30°, 33 were successfully reduced using percutaneous k-wire techniques with no internal fixation to maintain reduction. Of these, 31 had full range of motion at follow-up [37]. Other case series examining the results of closed or percutaneous reduction techniques, with inter-medullary stabilisation have yielded similarly good results [38–40].

There is general consensus that minimally displaced radial neck fractures can be managed non-operatively with good results. Closed or percutaneous K-wire assisted reduction of displaced fractures, with or without intramedullary stabilisation, is effective where possible. There is little information regarding severely displaced radial neck fractures where percutaneous reduction is not possible. The present literature associates poor results with open reductions. Given the quality of the literature it is not possible to determine whether this association is driven by indications (more severe fractures get open reductions) or by intervention (open reduction itself causes capsular stiffness, slower healing, and potential disruption of blood supply) or whether both indications and intervention contribute to poor results.

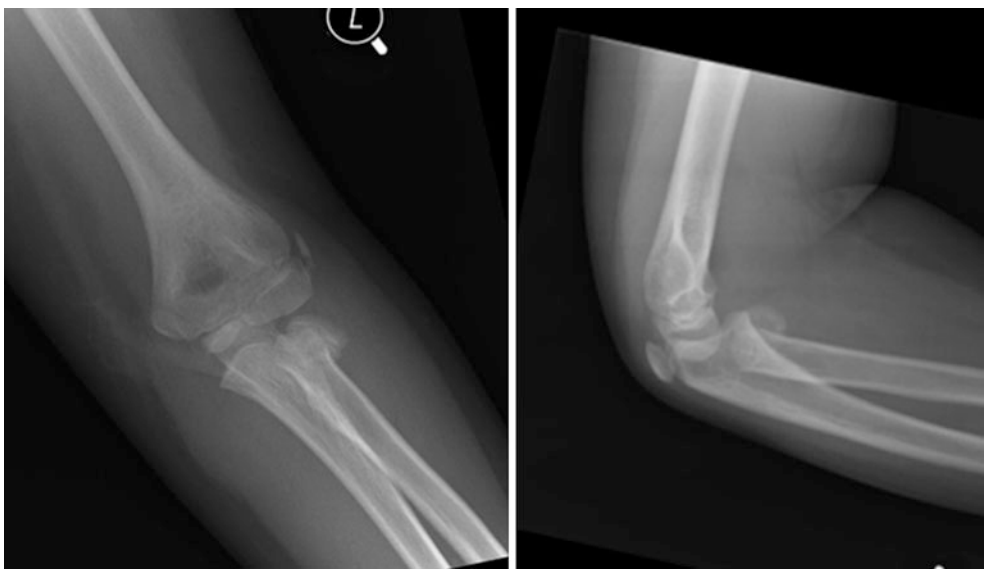

Fig. 32.5 Radial head fracture

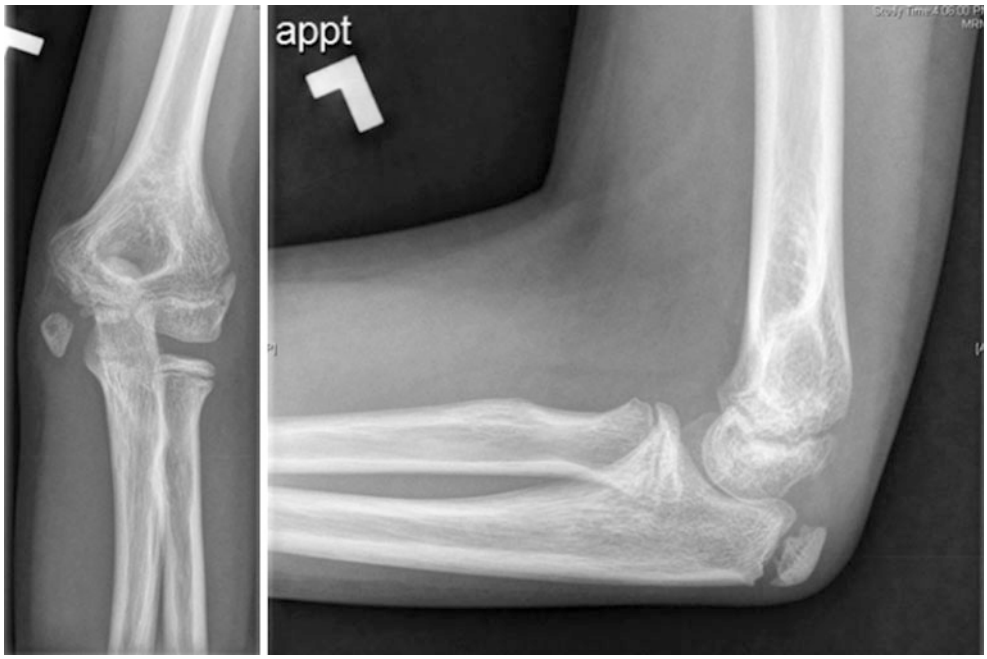

Fig. 32.6 Medial epicondyle fracture

Medial Epicondyle Fractures

Fractures of the medial epicondyle of the distal humerus account for approximately 12 % of all paediatric elbow fractures [41]. They may be associated with elbow dislocation, fracture fragment incarceration, and ulna nerve dysfunction.

Controversy remains regarding the management of medial epicondyle fractures since non-union does not equate to pain, and a united fracture does not mean the patient will be symptom free. A comparative study followed 43 children for 2–5 years. Of those treated closed, 55 % united but only 5 % had symptoms at follow-up. For those that were managed with open reduction, 87 % united but 39 % were symptomatic at follow-up [42]. This was a retrospective study so there may be some selection bias (Fig. 32.6).

Most studies discussing, but not resolving, the question of best treatment for displaced fractures have measured the displacement on an AP radiograph. A CT scan study published by Edmonds calls the traditional literature into question by documenting substantial displacement in the AP direction, even of fractures which appear undisplaced on an AP radiographic projection [43].

There are two scenarios relating to medial epicondyle fractures that do have clear management pathways. First, if the medial epicondyle is trapped within the joint then it should be reduced (Fig. 32.7). Some suggest one attempt at closed reduction techniques [44], with care not to damage the ulna nerve, moving to open methods if unsuccessful. Second, excision of the medial epicondyle yields poor results, which may include instability, pain and paraesthesia in the distribution of the ulna nerve. A comparative series of medial epicondyle fractures displaced 5–15 mm, with average long-term follow-up of 33 years compared closed management, open reduction and internal fixation, and primary excision of the medial epicondyle. For those 19 patients managed with long arm cast, 17 had a radiographic non-union but all had stable elbows to valgus stress, two had minor elbow symptoms. Of the 17 patients treated with open reduction and internal fixation, three had minor symptoms, all had radiographic union and all were stable. For the 6 patients managed with excision of the epicondylar fragment, four had ulnar paraesthesia, elbow instability and constant pain. The authors concluded that despite the high rate of radiographic non-union, the functional results supported non-operative management for medial epicondyle fractures displaced 5–15 mm [45].

Two retrospective reviews compared non-operative and operative management of displaced medial epicondyle fractures in competitive athletes [46, 47]. Patient selection was based on fracture displacement, injury mechanism and elbow instability preoperatively. Both reported return to play following rehabilitation, at the appropriate level, and encouraging patient reported outcomes in both groups. Of some concern, one study did state that 6 of the 14 patients who received operative treatment reported numbness [47]. Both these studies have significant selection bias and as such may not aid with decision-making regarding these injuries.

Overall there is a preference for non-operative management of medial epicondyle fractures, particularly in those

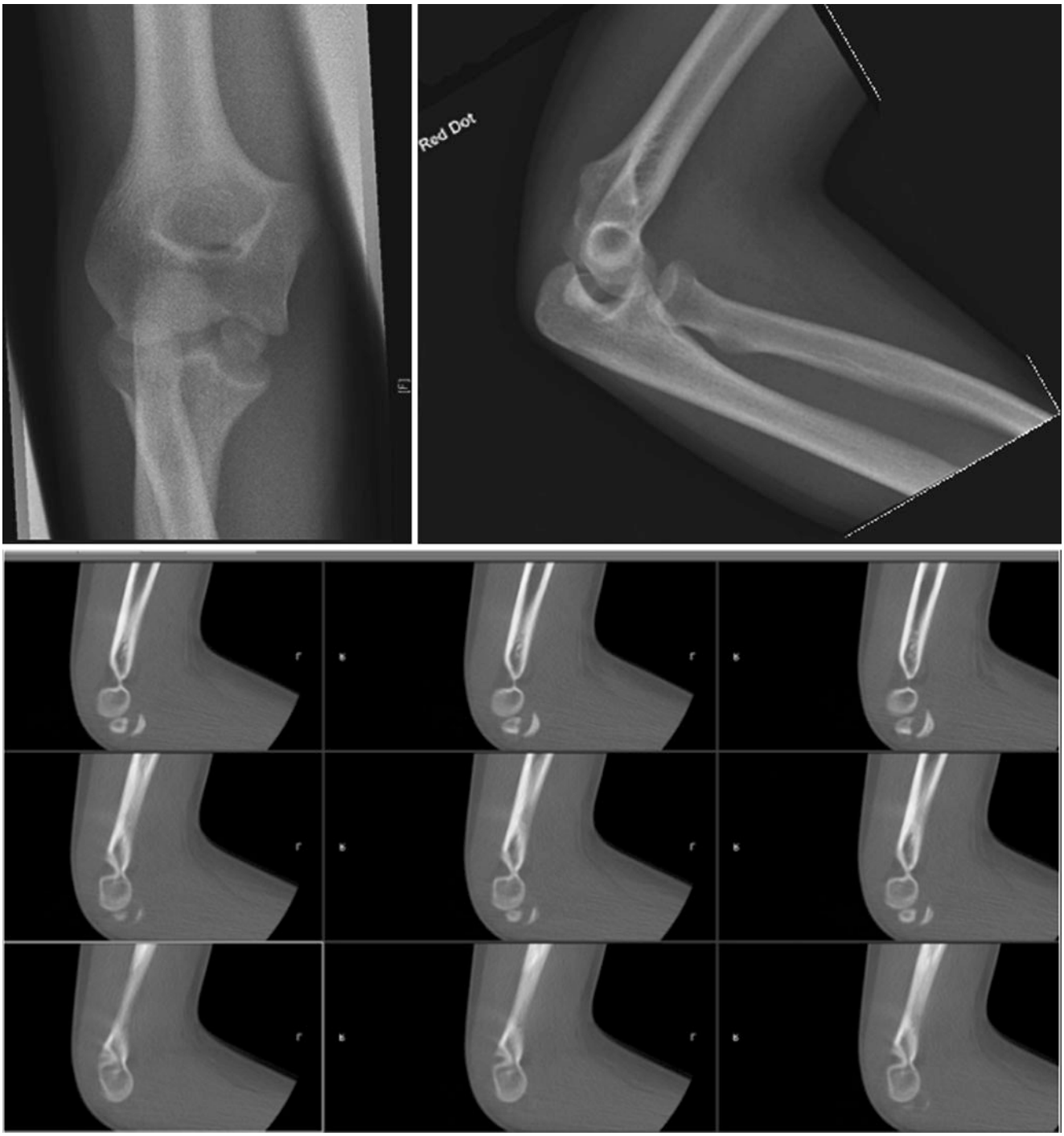

Fig. 32.7 Medial epicondyle fracture entrapped in the elbow joint

which are un-displaced or minimally displaced. There may be a cohort of patients with displaced medial epicondyle fractures who benefit from operative intervention. The present literature cannot define the indications for operative fixation largely because the present literature comparing operative to non-operative management is fraught with selection bias. Severe displaced injuries could be randomised to operative versus non-operative management, but given the small numbers of such injuries a multicentre trial would be required.

Recommendations for treatment of paediatric elbow fractures are listed in Table 32.1.

Table 32.1 Recommendations for treatment of paediatric elbow fractures

Statement	Grade of recommendation
Closed reduction and lateral entry percutaneous pinning is the preferred initial treatment for displaced supracondylar humeral fractures.	B
Delaying treatment of closed supracondylar humeral fractures without neurovascular compromise to more than 8 is not associated with a higher risk.	C
Expectant treatment is accepted option in a viable, pulseless hand after closed reduction and percutaneous pinning of supracondylar humeral fractures.	C
Undisplaced lateral condyle humeral fractures can be treated with an above elbow cast or percutaneous pinning	B
Displaced lateral condyle humeral fractures should be treated with open reduction and stabilisation	B/C
Undisplaced or minimally displaced medial epicondyle fractures can be treated non-operatively	C
Displaced medial epicondyle fractures can be treated operatively	I
If the medial epicondyle is trapped within the joint then it should be reduced	B
Excision of the medial epicondyle yields poor results	B
Undisplaced or minimally displaced radial neck fractures can be treated by no reduction or closed reduction.	B
Displaced radial head fracture needs an open reduction and stabilisation.	I

References

1. Farnsworth CL, Silva PD, Mubarak SJ. Etiology of supracondylar humerus fractures. J Pediatr Orthop. 1998;18(1):38–42.
2. Cuomo AV, Howard A, Hsueh S, Boutis K. Gartland type I supracondylar humerus fractures in children: is splint immobilization enough? Pediatr Emerg Care. 2012;28(11):1150–3.
3. Pirone AM, Graham HK, Krajbich JI. Management of displaced extension-type supracondylar fractures of the humerus in children. J Bone Joint Surg Am. 1988;70(5):641–50.
4. Aitken AL. Supracondylar fractures in children. Am J Surg. 1943;59(2):161–71.
5. Prietto CA. Supracondylar fractures of the humerus. A comparative study of Dunlop's traction versus percutaneous pinning. J Bone Joint Surg Am. 1979;61(3):425–8.
6. Skaggs DL, Sankar WN, Albrektson J, Vaishnav S, Choi PD, Kay RM. How safe is the operative treatment of Gartland type 2 supracondylar humerus fractures in children? J Pediatr Orthop. 2008;28(2):139–41.
7. Howard A, Mulpuri K, Abel MF, Braun S, Bueche M, Epps H, et al. The treatment of pediatric supracondylar humerus fractures. 2011; Available at: http://www.aaos.org/research/guidelines/SupracondylarFracture/SupConFullGuideline.pdf
8. De Boeck H, De Smet P, Penders W, De Rydt D. Supracondylar elbow fractures with impaction of the medial condyle in children. J Pediatr Orthop. 1995;15(4):444–8.
9. Hamdi A, Poitras P, Louati H, Dagenais S, Masquijo JJ, Kontio K. Biomechanical analysis of lateral pin placements for pediatric supracondylar humerus fractures. J Pediatr Orthop. 2010 Mar;30(2):135–9.
10. Kocher MS, Kasser JR, Waters PM, Bae D, Snyder BD, Hresko MT, et al. Lateral entry compared with medial and lateral entry pin fixation for completely displaced supracondylar humeral fractures in children. A randomized clinical trial. J Bone Joint Surg Am. 2007 Apr;89(4):706–12.
11. Brauer CA, Lee BM, Bae DS, Waters PM, Kocher MS. A systematic review of medial and lateral entry pinning versus lateral entry pinning for supracondylar fractures of the humerus. J Pediatr Orthop. 2007;27(2):181–6.
12. Woratanarat P, Angsanuntsukh C, Rattanasiri S, Attia J, Woratanarat T, Thakkinstian A. Meta-analysis of pinning in supracondylar fracture of the humerus in children. J Orthop Trauma. 2012;26(1):48–53.
13. Skaggs DL, Hale JM, Bassett J, Kaminsky C, Kay RM, Tolo VT. Operative treatment of supracondylar fractures of the humerus in children. The consequences of pin placement. J Bone Joint Surg Am. 2001;83-A(5):735–40.
14. Britisth Orthopaedic Association. Supracondylar fractures of the humerus in children. Boast 11 2014.
15. Ozkoc G, Gonc U, Kayaalp A, Teker K, Peker TT. Displaced supracondylar humeral fractures in children: open reduction vs. closed reduction and pinning. Arch Orthop Trauma Surg. 2004;124(8):547–51.
16. Mehlman CT, Strub WM, Roy DR, Wall EJ, Crawford AH. The effect of surgical timing on the perioperative complications of treatment of supracondylar humeral fractures in children. J Bone Joint Surg Am. 2001;83-A(3):323–7.
17. Carmichael KD, Joyner K. Quality of reduction versus timing of surgical intervention for pediatric supracondylar humerus fractures. Orthopedics. 2006;29(7):628–32.
18. Sibinski M, Sharma H, Bennet GC. Early versus delayed treatment of extension type-3 supracondylar fractures of the humerus in children. J Bone Joint Surg Br. 2006;88(3):380–1.
19. Walmsley PJ, Kelly MB, Robb JE, Annan IH, Porter DE. Delay increases the need for open reduction of type-III supracondylar fractures of the humerus. J Bone Joint Surg Br. 2006;88(4):528–30.
20. Schoenecker PL, Delgado E, Rotman M, Sicard GA, Capelli AM. Pulseless arm in association with totally displaced supracondylar fracture. J Orthop Trauma. 1996;10(6):410–5.
21. Garbuz DS, Leitch K, Wright JG. The treatment of supracondylar fractures in children with an absent radial pulse. J Pediatr Orthop. 1996;16(5):594–6.
22. Sabharwal S, Tredwell SJ, Beauchamp RD, Mackenzie WG, Jakubec DM, Cairns R, et al. Management of pulseless pink hand in pediatric supracondylar fractures of humerus. J Pediatr Orthop. 1997;17(3):303–10.
23. Surgeons AA. Introductory packet for Clinical Practice Guidelines (CPG)/Systematic Review (SR Work Group Members. 2015; Available at: http://www.aaos.org/research/guidelines/InteractiveIntroPacket.pdf

24. Warwick D, Dahl OE, Fisher WD. International surgical thrombosis forum. Orthopaedic thromboprophylaxis: limitations of current guidelines. J Bone Joint Surg Br. 2008;90(2):127–32.
25. Song KS, Kang CH, Min BW, Bae KC, Cho CH. Internal oblique radiographs for diagnosis of nondisplaced or minimally displaced lateral condylar fractures of the humerus in children. J Bone Joint Surg Am. 2007;89(1):58–63.
26. Horn BD, Herman MJ, Crisci K, Pizzutillo PD, MacEwen GD. Fractures of the lateral humeral condyle: role of the cartilage hinge in fracture stability. J Pediatr Orthop. 2002;22(1):8–11.
27. Launay F, Leet AI, Jacopin S, Jouve JL, Bollini G, Sponseller PD. Lateral humeral condyle fractures in children: a comparison of two approaches to treatment. J Pediatr Orthop. 2004;24(4):385–91.
28. Bast SC, Hoffer MM, Aval S. Nonoperative treatment for minimally and nondisplaced lateral humeral condyle fractures in children. J Pediatr Orthop. 1998;18(4):448–50.
29. Bloom T, Chen LY, Sabharwal S. Biomechanical analysis of lateral humeral condyle fracture pinning. J Pediatr Orthop. 2011;31(2):130–7.
30. Pondaag W, Malessy MJ, van Dijk JG, Thomeer RT. Natural history of obstetric brachial plexus palsy: a systematic review. Dev Med Child Neurol. 2004;46(2):138–44.
31. Sharma H, Maheshwari R, Wilson N. Lateral humeral condyle fractures in children: a comparative cohort study on screws versus K-wires. J Bone Joint Surg Br. 2006;434.
32. Thomas DP, Howard AW, Cole WG, Hedden DM. Three weeks of Kirschner wire fixation for displaced lateral condylar fractures of the humerus in children. J Pediatr Orthop. 2001;21(5):565–9.
33. Vahvanen V, Gripenberg L. Fracture of the radial neck in children. A long-term follow-up study of 43 cases. Acta Orthop Scand. 1978;49(1):32–8.
34. D'souza S, Vaishya R, Klenerman L. Management of radial neck fractures in children: a retrospective analysis of one hundred patients. J Pediatr Orthop. 1993;13(2):232–8.
35. Falciglia F, Giordano M, Aulisa AG, Di Lazzaro A, Guzzanti V. Radial neck fractures in children: results when open reduction is indicated. J Pediatr Orthop. 2014;34(8):756–62.
36. Basmajian HG, Choi PD, Huh K, Sankar WN, Wells L, Arkader A. Radial neck fractures in children: experience from two level-1 trauma centers. J Pediatr Orthop B. 2014;23(4):369–74.
37. Steele JA, Graham HK. Angulated radial neck fractures in children. A prospective study of percutaneous reduction. J Bone Joint Surg Br. 1992;75(1):169.
38. Stiefel D, Meuli M, Altermatt S. Fractures of the neck of the radius in children. Early experience with intramedullary pinning. J Bone Joint Surg Br. 2001;83(4):536–41.
39. Schmittenbecher PP, Haevernick B, Herold A, Knorr P, Schmid E. Treatment decision, method of osteosynthesis, and outcome in radial neck fractures in children: a multicenter study. J Pediatr Orthop. 2005;25(1):45–50.
40. Wang J, Chen W, Guo M, Su Y, Zhang Y. Percutaneous reduction and intramedullary fixation technique for displaced pediatric radial neck fractures. J Pediatr Orthop B. 2013;22(2):127–32.
41. Beaty JH. Operative treatment of femoral shaft fractures in children and adolescents. Clin Orthop Relat Res. 2005;434:114–22.
42. Wilson NIL. Treatment of fractures of the medial epicondyle of the humerus. Injury. 1988;19(5):342–4.
43. Edmonds EW. How displaced are "nondisplaced" fractures of the medial humeral epicondyle in children? Results of a three-dimensional computed tomography analysis. J Bone Joint Surg Am. 2010;92(17):2785–91.
44. Roberts N. Displacement of the internal epicondyle into the elbow-joint. The Lancet. 1934;224(5788):281.
45. Farsetti P, Potenza V, Caterini R, Ippolito E. Long-term results of treatment of fractures of the medial humeral epicondyle in children. J Bone Joint Surg Am. 2001;83-A(9):1299–305.
46. Osbahr DC, Chalmers PN, Frank JS, Williams RJ3rd, Widmann RF, Green DW. Acute, avulsion fractures of the medial epicondyle while throwing in youth baseball players: a variant of Little League elbow. J Shoulder Elbow Surg 2010;19(7):951-957.
47. Lawrence JT, Patel NM, Macknin J, Flynn JM, Cameron D, Wolfgruber HC, et al. Return to competitive sports after medial epicondyle fractures in adolescent athletes: results of operative and nonoperative treatment. Am J Sports Med. 2013;41(5):1152–7.

Evidence-Based Treatment of Madelung's Deformity

33

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Abstract

Madelung's deformity is a premature growth arrest of the volar ulna aspect of the radial growth plate leading to ulna overgrowth and a progressive wrist deformity. Treatment depends on the skeletal maturity of the patient and can be targeted at either the ulna or the radius. We describe the rationale and evidence behind the treatments available and propose a classification to guide management.

Keywords

Madelung's deformity • Growth arrest • Vicker's ligament • Classification • Corrective osteotomy

Background

Madelung's deformity (MD) is a rare condition caused by abnormal growth arrest at the ulna and volar aspect of the distal radial physis. The characteristic osseous deformity is a volar curvature of the distal radius, a positive ulna variance and proximal subsidence of the lunate. The result is a characteristic aesthetic appearance with associated functional limitation. Two anatomical anomalies appear to have a central role in the development of congenital MD. Firstly, Vickers [27] and then subsequently Cook (Cook et al. [3]) described an abnormal bar that crosses the distal radial physis at its ulnar and volar aspect which is proposed to restrict growth in the ulna and palmer aspect of the physis. This can be either completely or partially ossified or fibrous. Secondly, abnormal volar ligaments are thought to act as tethers during rapid growth exacerbating the problem. Vicker's ligament, a fibrous band running from the ulna border of the radius to the lunate, and a pathological radiotriquetral ligament have both been described. The ligaments cause a compressive force on the distal radial physis leading to premature closure thus contributing to the resultant osseous deformity. The deformity characteristically presents during adolescence following a period of rapid growth [14] (Stehling et al. [24]).

The clinical picture includes pain and a reduction in range of motion, particularly in radial deviation and pronation-supination. A reduction in grip strength has also been reported with a spectrum of severity often seen (McCarroll et al. [17]). No correlation has, however, been established between pain and level of radiographic deformity and function can often be reasonably well maintained (Huguet et al. [12]) Cosmetic concerns, due to dorsal subluxation of the carpus and prominence of the ulna head, can be the trigger for presentation to orthopaedic services and are often cited as an indication for operative intervention (Fig. 33.1)

Multiple operative treatment options have been proposed, principally corrective osteotomies. However, owing to its rarity, the literature is sparse and there is little evidence to support definitive management strategies.

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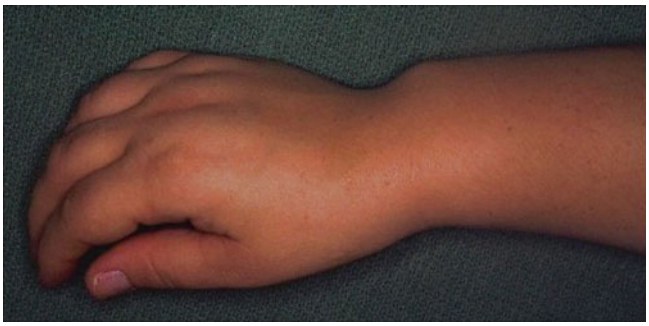

Fig. 33.1 Characteristic aesthetic appearance of Madelung's deformity

What Is the Natural History of Madelung's Deformity?

Several hundred cases of MD have been presented in the literature since its first description. However, no published reports exist on the actual frequency of MD in the population or natural history [8]. This is not a surprise given the wide spectrum of clinical presentation; from completely asymptomatic in some individuals to quite disabling symptoms in others (Zebala et al. [28]). Reports have linked the osseous deformity to progressive ulnocarpal abutment, distal radio-ulna joint instability and progressive radiocarpal arthritis [8]. Three cases of extensor tendon rupture have been reported in severe and chronic cases due to abrasion on the prominent ulna head (Rondier et al. [21]; Ducloyer et al. [6]).

Is There a Role for Nonoperative Treatment?

Optimal treatment of MD is controversial particularly if patients were asymptomatic or not sufficiently symptomatic to warrant surgical treatment. The non-surgical management of patients with MD is analgesia and activity modification [26].

What Are the Operative Options?

Multiple surgical approaches have been described in the literature. Broadly, these can be considered in three categories: those involving the radius or the ulna in isolation and those involving both bones in combination. Surgery to the radius aims to address the primary pathology and/or correct the resultant deformity. Isolated procedures to the ulna treat ulna sided wrist pain that occurs secondary to ulnocarpal impingement. However, there is little clarity and no consensus within the literature to enable the development of a definitive management strategy. Evidence is limited to small case series and frequently the surgical techniques employed are not uniform and therefore drawing definitive conclusions from heterogeneous groups can be difficult. Additionally, the optimal timing of surgical intervention is not known. Surgery before physeal closure may lessen the development of secondary degenerative changes in the carpus and allow some remodelling, whereas intervention after skeletal maturity decreases the likelihood of recurrence.

Surgery to the Radius

Physiolysis

Vicker's and Nielsen describe a physiolysis procedure with release of Vicker's ligament, to address both proposed pathoanatomical causes of the deformity. The procedure described (also referred to as Langenskiöld technique) excises the abnormal physis in the ulna and volar portion of the distal radius with the interposition of fat. Improvements in range of motion and pain were seen in a cohort of 24 wrists in 17 patients who underwent the procedure at a mean age of 12 years. The authors advocate the need for early surgery as remodelling potential is necessary [27]. Subsequently, Ogino (Ogino et al. [19]) reported outcomes of 3 wrists in 2 patients with a mean age of 12 years. However, in 1 patient palmer shift of the carpus and radial inclination worsened. Most recently Paes described the long term (12 year) outcomes of 5 wrists in 3 patients who underwent the procedure at a mean age of 12 years. Results were poor with pain and a restriction in range of motion seen at long term follow up in all patients. Consequently, the author no longer advocates physiolysis questioning the applicability of the Langenskiöld technique, which was initially described for physeal arrest post trauma, to Madelung's where the physis is pathological. Concerns have also been raised regarding distal radio-ulna joint degeneration that results from operating on the ulna side of the distal radius (Paes et al. [7]).

Isolated Radial Osteotomy

Isolated radial osteotomies have been described with the rationale to correct the volar and ulna angulation and lengthen the distal radius. Murphy (Murphy et al. [18]) described an opening wedge osteotomy performed through a volar approach which enabled a biplane deformity correction. Corticocancellous graft from iliac crest was used and the abnormal volar ligaments were excised. The outcomes of 12

wrists in 11 patients who underwent the procedure at a mean age of 16 years were reported. Patients were subjectively pleased with the function and cosmetic appearance of the wrist and ulna variance and radioulnar inclination also improved although no statistical analysis was provided. A subset of the cohort however also underwent an ulna osteotomy or more proximal radial osteotomy. Mallard (Mallard et al. [15]) advocates the use of a reverse wedge osteotomy in skeletally mature patients. A bone wedge is cut from the excess radial and dorsal cortical bone, reversed and placed into an osteotomy on the ulna side of the distal radius. In a case series of 11 wrists, range of motion improved and patients were satisfied with the outcome. However, 30% required an additional ulna osteotomy for ulna-carpal impingement. An llizarov technique has also been described. This facilitates lengthening of the radius in addition to correction of the angular deformity. Houshain reported favourable outcomes with regard to pain, grip strength and range of motion in two patients (Houshian et al. [11]). Similar findings are also reported by de Billy in a case series of 5 wrists (de Billy et al. [4]).

Dome osteotomies have the theoretical advantage over the above procedures of being able to achieve deformity correction in all three planes. Harley performed a radial dome osteotomy with release of Vicker's ligament in 26 wrists from 18 patients at a mean age of 13 years. At a mean follow up of 2 years improvements in supination and extension range of motion were seen in addition to ulna tilt and lunate subsidence (Harley et al. [10]). Steinman reported the long term (11 year) follow up of the above series and concluded that radiographic deformity correction was maintained in addition to a good to excellent functional outcomes (Steinman et al. [25]).

Imai recently reported a case study of a 12 year old female with a bilateral deformity corrected by a novel customised cylindrical distal radius osteotomy. The axis of the deformity and the degree of rotation required to correct it were determined from pre-operative CT scans which allowed customised osteotomy templates to be developed. At 28 month follow up the patient was pain free and both grip strength and ROM had improved (Imai et al. [13]).

Surgery to the Ulna

Epiphysiodesis

Epiphysiodesis of the ulna can be performed in conjunction with a radial osteotomy to prevent ulna positive variance. Bak described a case of a 14 year old female who developed an early acquired Madelung like deformity from repetitive injury during training who was subsequently able to return to high level competition [1].

Isolated Ulna Osteotomy

Previously Darrach advocated resection of the ulna head either alone (or in combination with a radial osteotomy) to manage ulna sided wrist pain. However, subsequent lateral subluxation of the caprus caused concern (Ranawat et al. [20]). Schroven undertook a segmental resection of a portion of ulna shaft, just distal to the distal radio-ulna joint, to create a pseudoarthrosis with the rationale of restoring stability while retaining the ulna head (Schroven et al. [23]).

Isolated osteotomies of the ulna have been presented in the literature. These are indicated to treat ulna sided wrist pain in skeletally mature patients with a mild MD. Ulna head prominence is addressed preventing ulnocarpal abutment. Bruno performed an isolated ulna shortening osteotomy in 9 wrists in adults with symptomatic ulnocarpal impingement. At a mean follow up of 42 months ulna variance was significantly reduced (from 3.3mm to -1.1mm) and all patients were asymptomatic (Bruno et al. [2]). Subsequently, on a similar subset of patients with a mild MD, Glard performed a shortening osteotomy of the ulna with anterior angulation, the aim being to reduce the distal radioulnar joint. Postoperatively, pain and range of motion in pronation and supination improved and all patients were satisfied with the cosmetic effect. The mean volar angulation at ulna osteotomy was 17° (Glard et al. [9]).

Surgery Combined to the Radius and Ulna

Combined corrective osteotomies of the radius and ulna aim to correct the radial deformity while shortening the ulna to achieve reduction of the distal radio-ulna joint. Dos Reis described a wedge subtraction osteotomy of the radius and shortening osteotomy of the ulna. Grip strength and range of motion significantly improved at 53 month follow up in 18 patients who underwent the above procedure at a mean age of 22 years (dos Reis et al. [5]). Similarly, a closing wedge osteotomy of the radial metaphysis and shortening ulna osteotomy with slight volar angulation to reduce distal radio-ulna joint was described by Salon. Improvements in pain and range of motion were seen at 9 year follow up in 11 wrists who underwent the procedure at a mean age of 14 years. Remodelling of the distal radio-ulna joint was seen in all patients (Salon et al. [22]). Recently McCarrol presented encouraging early results from a very distal radial osteotomy with ulna shortening. The distal nature of the radial osteotomy enabled flexibility to independently correct the three plane deformity [16].

A summary of recommendations for MD is listed in Table 33.1.

Table 33.1 Summary of recommendations for Madelung's deformity

Clinical situations	Grade of recommendation
1. What is the natural history of untreated MD?	D
2. Is there a need for treating asymptomatic MD?	C
3. What is the best surgical treatment for MD?	D

Table 33.2 Manchester Madelung's deformity classification

Grade	Features	Management
1: Skeletally immature patients		
1a	Pre growth arrest. Those patients without radiographic deformity, but for whom genetic testing and or family history predict the development of the disease. The "at risk" group	Release of abnormal radio-carpal ligament
1b	Following growth arrest with *adequate* growth potential for remodelling. These patients do not have a well-defined ulna styloid	Release of abnormal radio-carpal ligament and physeal bar plus ulna epiphysiodesis
1c	Following growth arrest with *insufficient* growth potential for remodelling. These patients have a well-defined ulna styloid	Radial dome osteotomy and ulna epiphysiodesis
2: Skeletally mature patients		
2a	Mild coronal plane deformity	Ulna shortening only
2b	Severe coronal and sagittal plane deformity	Radial dome osteotomy (± extension) with ulna shortening
3: Secondary arthritis		
3	Longstanding deformity and sequelae, including distal radio-ulnar joint instability and osteoarthritis	Darrach's procedure or joint fusion

Authors' Preferences

The authors propose the following classification to aid in description of the deformity and plan appropriate treatment (Table 33.2**).**

References

1. Bak K, Boeckstyns M. Epiphysiodesis for bilateral irregular closure of the distal radial physis in a gymnast. Scand J Med Sci Sports. 1997;**7**(6):363–6.
2. Bruno RJ, Blank JE, et al. Treatment of Madelung's deformity in adults by ulna reduction osteotomy. J Hand Surg Am. 2003;**28**(3):421–6.
3. Cook PA, Yu JS, et al. Madelung deformity in skeletally immature patients: morphologic assessment using radiography, CT, and MRI. J Comput Assist Tomogr. 1996;**20**(4):505–11.
4. de Billy B, Gastaud F, et al. Treatment of Madelung's deformity by lengthening and reaxation of the distal extremity of the radius by Ilizarov's technique. Eur J Pediatr Surg. 1997;**7**(5):296–8.
5. dos Reis FB, Katchburian MV, et al. Osteotomy of the radius and ulna for the Madelung deformity. J Bone Joint Surg Br. 1998;**80**(5):817–24.
6. Ducloyer P, Leclercq C, et al. Spontaneous ruptures of the extensor tendons of the fingers in Madelung's deformity. J Hand Surg Br. 1991;**16**(3):329–33.
7. Paes EC, Theunissen CMJ, Sakkers RJB, Schuurman AH. Langenskiöld procedure for Madelung's deformity: case series of late Sequelae. Open J Orthop. 2014;**4**(11):313–20
8. Ghatan AC, Hanel DP. Madelung deformity. J Am Acad Orthop Surg. 2013;**21**(6):372–82.
9. Glard Y, Gay A, et al. Isolated wedge osteotomy of the ulna for mild Madelung's deformity. J Hand Surg Am. 2007;**32**(7):1037–42.
10. Harley BJ, Brown C, et al. Volar ligament release and distal radius dome osteotomy for correction of Madelung's deformity. J Hand Surg Am. 2006;**31**(9):1499–506.
11. Houshian S, Jorgsholm PB, et al. Madelung deformity treated with Ilizarov technique: a report of two cases. J Hand Surg Br. 2000;**25**(4):396–9.
12. Huguet S, Leheup B, et al. Radiological and clinical analysis of Madelung's deformity in children. Orthop Traumatol Surg Res. 2020;**100**(6 Suppl):S349–52.
13. Imai Y, Miyake J, et al. Cylindrical corrective osteotomy for Madelung deformity using a computer simulation: case report. J Hand Surg Am. 2013;**38**(10):1925–32.
14. Kim HK. Madelung deformity with Vickers ligament. Pediatr Radiol. 2009;**39**(11):1251.
15. Mallard F, Jeudy J, et al. Reverse wedge osteotomy of the distal radius in Madelung's deformity. Orthop Traumatol Surg Res. 2013;**99**(4 Suppl):S279–83.
16. McCarroll Jr HR, James MA. Very distal radial osteotomy for Madelung's deformity. Tech Hand Up Extrem Surg. 2010;**14**(2):85–93.
17. McCarroll Jr HR, James MA, et al. Madelung's deformity: quantitative assessment of x-ray deformity. J Hand Surg Am. 2005;**30**(6):1211–20.
18. Murphy MS, Linscheid RL, et al. Radial opening wedge osteotomy in Madelung's deformity. J Hand Surg Am. 1996;**21**(6):1035–44.
19. Ogino T, Ishigaki D, et al. Free fat graft for congenital hand differences. Clin Orthop Surg. 2012;**4**(1):45–57.
20. Ranawat CS, DeFiore J, et al. Madelung's deformity. An end-result study of surgical treatment. J Bone Joint Surg Am. 1975;**57**(6):772–5.
21. Rondier J, Cirotteau Y, et al. Madelung's disease and lesions of the extensor tendons. A case report. Rev Chir Orthop Reparatrice Appar Mot. 1990;**76**(3):204–8.

22. Salon A, Serra M, et al. Long-term follow-up of surgical correction of Madelung's deformity with conservation of the distal radioulnar joint in teenagers. J Hand Surg Br. 2000;**25**(1):22–5.
23. Schroven I, De Smet L, et al. Radial osteotomy and Sauve-Kapandji procedure for deformities of the distal radius. Acta Orthop Belg. 1995;**61**(1):1–5.
24. Stehling C, Langer M, et al. High resolution 3.0 Tesla MR imaging findings in patients with bilateral Madelung's deformity. Surg Radiol Anat. 2009;**31**(7):551–7.
25. Steinman S, Oishi S, et al. Volar ligament release and distal radial dome osteotomy for the correction of Madelung deformity: long-term follow-up. J Bone Joint Surg Am. 2013;**95**(13): 1198–204.
26. Tachdjians. Pediatric Orthopaedics. Philadelphia: Saunders Elsevier; 2008.
27. Vickers D, Nielsen G. Madelung deformity: surgical prophylaxis (physiolysis) during the late growth period by resection of the dyschondrosteosis lesion. J Hand Surg Br. 1992;**17**(4):401–7.
28. Zebala LP, Manske PR, et al. Madelung's deformity: a spectrum of presentation. J Hand Surg Am. 2007;**32**(9):1393–401.

Evidence-Based Treatments of Congenital Radio-Ulnar Synostosis

34

Wei Y. Leong and Matthew F. Nixon

Abstract

Congenital radio-ulnar synostosis (CRUS) is a rare disease characterized by abnormal fusion of radius and ulna resulting in limitations of supination and pronation. There are limited evidence in literature and can be found in the form of case series and case reports consisting of small number of patients. Majority of patients affected has an idiopathic cause, however, some patients do have a familial predisposition or underlying genetic condition. Treatment options are largely based on age, severity of synostosis and severity of functional restriction at presentation. Most cases are treated non-operatively as patients do compensate remarkably well by modifying day-to-day activities. The two main surgical treatments consist of either surgical resection with mobilization of synostosis or surgical derotation of forearm into a more functional position. We recommend that all patients who present with CRUJ be investigated for underlying genetic disorders. Patients should be assessed on a case-to-case basis in terms of suitability for surgical intervention based on their functional requirement and current limitations or symptoms.

Keywords

Congenital radio-ulnar synostosis • CRUS • Synostosis

Background

Congenital radio-ulnar synostosis (CRUS) is a congenital abnormality caused by a failure of segmentation in the embryonic stage causing an abnormal fusion of radius and ulna. The first case was described by Sandifort in his Museum Anatomicus in 1793 [1]. Most patients show proximal radio-ulna fusion with rare occurrence of distal joint involvement. Forearm position can be pathologically fixed in a position ranging from neutral to severe pronation. The etiology is likely secondary to an embryonic insult resulting in failure of normal longitudinal segmentation between the 2 bones during seventh week of development [2]. Radial head dislocation is commonly found as an associated deformity and is thought to be a result of complete proximal coalition causing interference to joint formation. Limited fusion distal to epiphysis of radius could also be the cause of unequal radial head growth [3]. There has been report suggesting that the condition tends to be bilateral in about 50–80 % of cases [4–5].

Searching Methods

Search was performed on Medline with Keywords with Mesh Heading 'Synostosis', 'Congenital', 'Radius', and 'Ulna'. The search method is outlined in Table 34.1 and flow chart of search methodology is presented in Fig. 34.1. Abstract of 283 articles were then examined to exclude unrelated subjects as well as excluding animal subjects. Sixty-five articles were obtained for further review. Due to the rarity of the disease, all of the articles were of level 4 evidence with mainly case reports and small number of case series.

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Table 34.1 Search methodology

Searches	Methods	Results
1	Exp Synostosis/or radio-ulnar synostosis.mp.	7976
2	Congenital.mp. or exp Congenital Abnormalities/	583,195
3	Exp Radius/or radius.mp.	34,880
4	Exp Ulna/or ulna.mp.	9703
5	3 or 4	38,991
6	1 and 2 and 5	403
7	Limit 6 to English language	283

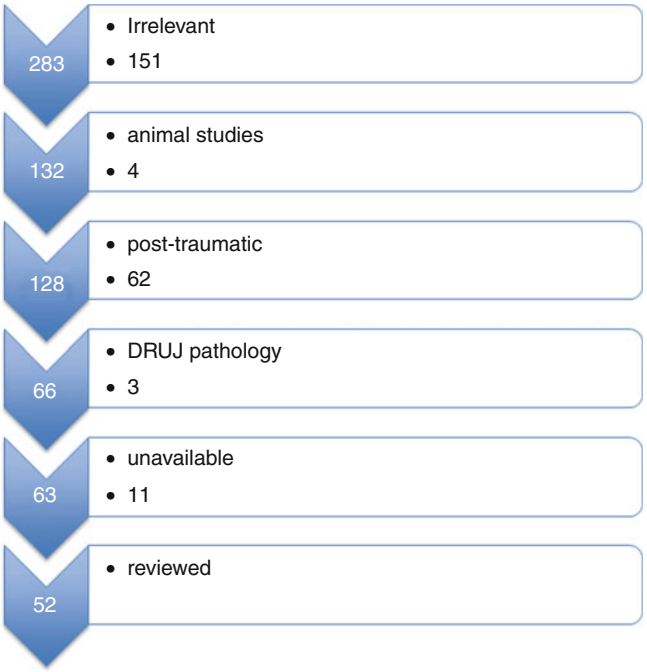

Fig. 34.1 Flow chart of search methodology

What Are the Causes and Natural Progression of Disease?

Embroyology

At 26 days after conception, the upper limb bud arises from the body wall. Elbow is first identifiable at 35 days. Three connected cartilaginous anlagen that will eventually progress to become humerus, radius and ulna are still present at this stage [6]. By 42 days the three bones are separated by condensation of tissue but joints are still yet to form. At this stage, the forearm remains to be in neutral position and pronation occurs by week 8. Longitudinal segmentation with separation starts distally and progress proximally. Various factors either as a result of abnormal genetics or environmental factors can interrupt subsequent morphogenesis of the elbow joint at this point leading to failure of segmentation. The final specific defect is influenced by differences in development timing and can result in complete bony synostosis

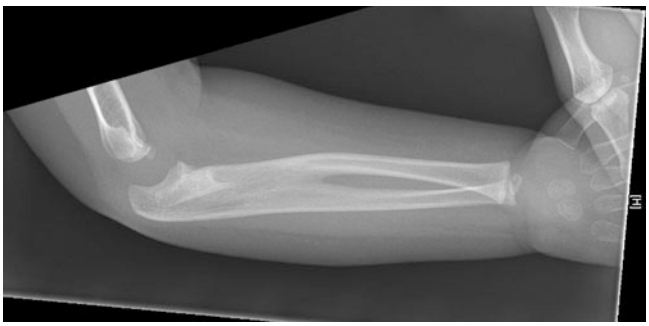

Fig. 34.2 Plain x-ray showing complete bony radio-ulnar synostosis

(Fig. 34.2) or smaller area of synostosis if joint development continues before development arrest [3].

Genetic Associations

The actual etiology of the condition remains unknown. However, a lot of authors had suggested a familial presentation and genetic background. Disorders such as Apert and Klinefelter syndromes have been known to be associated with CRUS.

Catena et al has suggested in their proposed classification system that CRUS to be related to Type IV Poland Anomaly. The classic Poland Anomaly consists of unilateral agenesis/hypoplasia of pectoralis major muscle with associated ipsilateral hand or upper limb anomalies [7].

Gaspar et al also identified an association of CRUS with Giuffre-Tsukahara syndrome (GTS) which appeared to be a X-linked dominant inheritance. The other features of the syndrome included, short stature, microencephaly, scoliosis and mental retardation [8]. This report was also supported by Zhu et al in their paper with a three generation Chinese family with similar associated features suggested to be a variant of GTS due to a lack of microencephaly in the family [9].

There seems to be also multiple reports of CRUS being linked with bone marrow failure. Sola et al and Thompson et al have also identified association with amegakaryocytic thrombocytonpenia as a result of HOXA11 gene mutation. Thompson et al also reported in his case series of two families

that there is an autosomal dominant disorder presenting with CRUS, congenital amegakaryotic thrombocyptopenia, aplastic anemia, clinodactaly, syndactaly, hip dysplasia and sensorineural hearing loss [10–11]. Castillo-caro et al. also reported on a case in infant associated with amegakaryotic thrombocytopenia without HOXA11 mutation [12].

Natural Progression of Disease

There seems to be a male preponderance in the incidence of congenital radio-ulnar synostosis. As previously mentioned, some patients might have a familial predisposition reported to be 13 % or other associated phenotypes suggesting congenital syndromes. Patients commonly presents at an average age of 2 and a half years old with majority noted by parents due to their functional abnormalities [5]. Children tend to compensate well using ipsilateral shoulder and wrist when deformity is mild resulting in minimal noticeable disability. Most patients present with hand fixed in various degrees of pronation (Fig. 34.3). With a bony synostosis (the most common type) there is no true pronation or supination, although adaptive ligamentous laxity at either the wrist or elbow can allow some apparent motion. The position of the wrist is variable ranging from being fixed in full supination to full pronation, with restriction in function dependent on the position. Quite frequently patients modify their activities and adapt to compensate for restricted rotation of forearm and rarely complained about restriction of function. In general an unilateral mid-pronated position is least debilitating, but with functional deficiencies were more commonly mentioned in patients with bilateral involvement and in more severe degrees of hyperpronation, in which some patients try the 'backhanded' technique in getting objects to mouth or receiving change [1–3].

Patients with CRUS can experience sudden onset of acute flexion contracture secondary to hyperflexion injury. Wang et al explained that this could be due to overgrown dislocated radial head becoming trapped under hypertrophied annular ligament-type tissue.

Classification

Wilkie and Davenport et al developed a classification for the condition in 1914. They suggested two different types of synostosis based on radiological appearance with either complete synostosis or partial union just distal to proximal radial epiphysis associated with radial head dislocation [13].

In 1985, Cleary and Omer et al noticed in their case series 4 different types of radiological appearance which conforms with the theory that timing of embryological arrest maybe reflected by both fixed position of forearm and degree of bony development at elbow. However, there were no functional

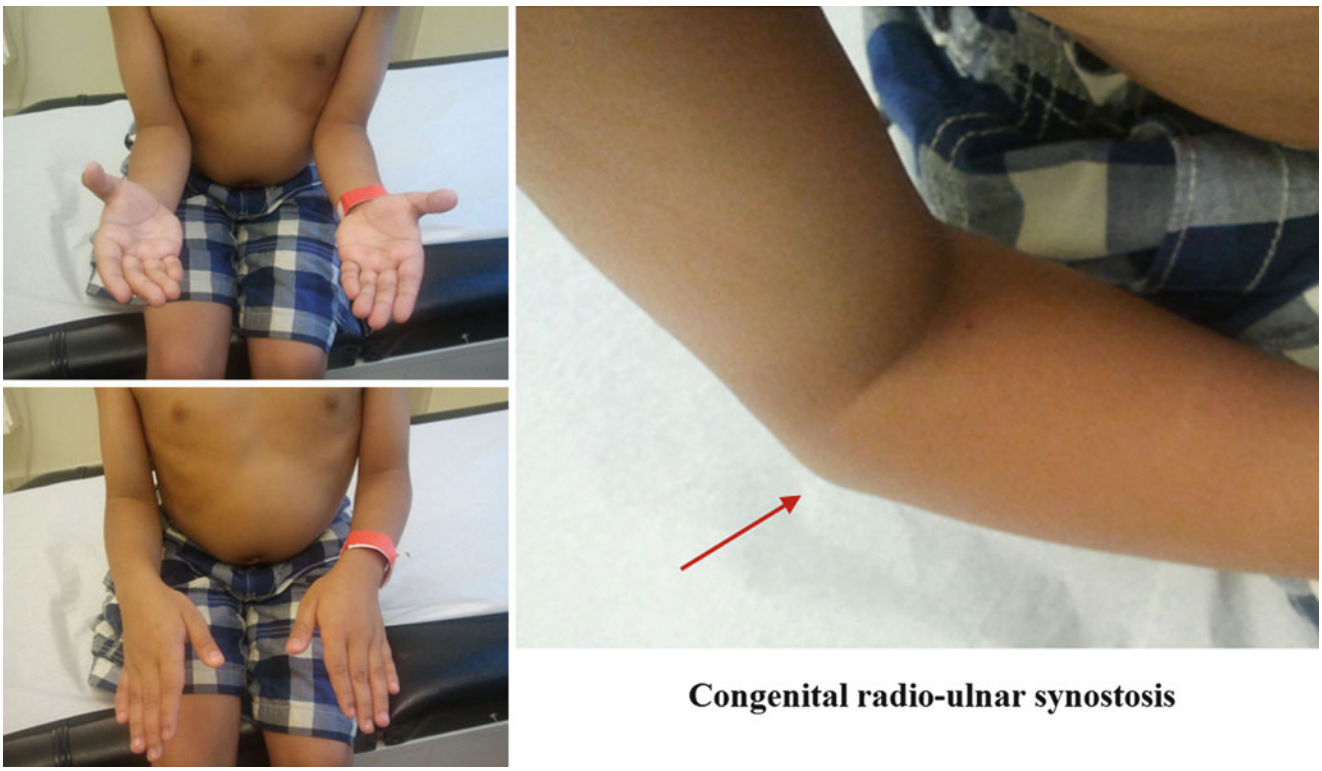

Fig. 34.3 Radio-ulnar synostosis clinical photograph, arrow showing radial head prominence

Table 34.2 Cleary and omer classification

1. Fibrous synostosis
2. Bony synostosis
3. Osseous synostosis associated with posterior dislocation of a hypoplastic radial head
4. Osseous synostosis with associated anterior dislocation of mushroom-shaped radial head

differences noted amongst different radiological appearance hence it was not particularly helpful in determining management of patients (Table 34.2) [14].

What Is the Evidence Behind the Current Treatment Options?

As mentioned before, patients can compensate remarkably well despite deformity and reduction in forearm rotation which explained why the condition remain under diagnosed. Hence, in majority of patients CRUS can be treated non surgically especially if the arm is in a neutral position and there is a reasonable degree of compensatory wrist rotation.

Relative indications for surgery include

- 90° of fixed pronation,
- Bilateral CRUJ to position one hand in mid-pronation and the other in a neutral or slightly supination position.
- Poor function due to other underlying associated symptoms secondary to associated syndrome either causing spasticity or other neurological conditions.
- Painful snapping elbow with or without elbow flexion contracture as a result of recurrent anterior dislocation of radial head due to malformation of radial head.

Surgical options can be split into attempts to release the synostosis and restore forearm rotation and de-rotation osteotomies to reposition a synostosed arm into a more function position.

The Role of Surgical Release of CRUS

Patient Selection

There is little point releasing a synostosis unless the joint remains pain free and there is active pronation and supination. This requires a congruent radiocapitellar joint, the presence of muscles to drive movement and ensuring that reformation of the synostosis does not occur.

Timing of this surgery remains difficult. It was thought that it should be performed on patients between 4 and 12 years old if it is indicated as osteotomy and rigid fixation is tricky for children who are under 3 years old and there is less remodeling potential for radial head dysplasia in older children. Furthermore, microvascular anastomosis, if required, can be quite challenging for younger patient. In older children (>12 years old) supinator and pronator seemed to be more atrophic and additional tendon transfer might be required to restore forearm rotation.

Surgical Technique

Mobilisation of CRUJ with osteotomy and synostosis has been trialed since the 1990s. Concerns about recurrent ankylosis resulted in multiple different surgical techniques to counter this. The early results of mobilization with anconeous muscle and nonvascularised spacers has not been particularly successful due to inadequate soft tissue graft. Interposition with radio-ulnar prosthesis has shown to be a success in post traumatic radio-ulnar synostosis. However, CRUJ is associated with underlying deformity of bone with associated tight and constricted soft tissue, hence functional outcome from these prosthesis tends to be poor.

In 1997, Fuminori Kanaya reported a new method of mobilization of CRUJ and interposition with free vascularized fascio-fat graft. He described technique of separating the synostosis through both anterior and posterior approaches. He separates both synchondrosis between radial head and ulna and synostosis using a steel burr until 1 cm distal to the synostosis. Dislocated radial head which was conical in shape will be trimmed flat. Through anterior approach the release the remaining of attachment of biceps tendon from ulna to improve forearm rotation. Shortening wedge osteotomy of the radius, flexion osteotomy for posterior dislocation and extension for anterior dislocation, due to persistent radial head dislocation during rotation of forearm followed this. A vascular graft was taken from the ipsilateral arm with a skin flap included to allowed monitoring of viability of flap with the profunda humeri vessel used as a donor vessel anastomosed with recurrent radial vessel. The average arc of motion patients achieved with this treatment was 71°. He described no re-ankylosis and maintenance of patients' range of movement on follow-up of all his seven patients in his case series [15–16].

Oka et al confirmed findings that function and range of movement does increase for patients treated by Kanaya procedure with greatest improvement in patients without radial head dislocation in their 3D in vivo analysis [17]. Other techniques of interposition such as using de-epithelialized groin fasciocutaneous graft and de-epitheliazed

anterolateral thigh fasciocutaneous flap was also described by Chen et al. [18].

The Role of Derotation Osteotomy of Radius and Ulna in CRUS

Patient Selection

There are large amount of case series in the literature to support this technique and the results are generally good in restoring and maintaining function. The popularity of this technique could also be due to the fact that osteotomy is a far less challenging operation compared to microvascular surgery requirement of Kanaya technique. There was again a preference for this technique of treatment to be performed in early childhood. It was thought that delay can result in progressive deformity of elbow, forearm and wrist resulting in persistent pain and soft tissue tightness. Horii et al recommended that this technique can be safely used between ages of 4 and 9 years old as robust periosteum at this age can hold the cut radius and facilitate callus formation [19].

Site of Osteotomy

Different authors advocate different site of osteotomy with the most common site being in midshaft radius and ulna [20]. Some surgeons advocate ulna midshaft and distal radius osteotomy as distal radius osteotomy does not require any fixation and any deformity will be corrected by patient's ability to remodel with time [21].

Methods of Fixation

There are various method suggested for fixation of bone following derotation. This ranges from plaster of paris to maintain position, Intramedullary K wire, crossed K wires on proximal ulna and plate fixation. Nonunion is generally very uncommon in children, however Lin et al did noticed in their series of 26 forearms in 23 patients to have 2 non-union which required subsequent bone grafting and plate stabilization in their technique of radius and ulna midshaft derotational osteotomy with plaster immobilization post op. [22]

Position of Forearm

Indication for surgery tends to be extreme pronation and supination. Position of forearm following correction need to be considered very carefully particularly in patients with bilateral involvement. In dominant hand, forearm should be left in approximately 20–30° of pronation to facilitate day to day activities such as writing. In patients with bilateral involvement, this became prudent to ensure that non- dominant hand to 20° supination position to facilitate toiletry hygiene [3].

Result: Function and Range of Movement Achieved

Derotational osteotomy unfortunately does not improve arc of movement. Flexion and extension of elbow joint does not seemed to be affected and patients tend to maintain their preop ability to compensate to certain extend in terms of rotation at the distal radio-ulnar joint. Nonetheless, patients tend to be quite pleased with better functional position of forearm following surgery. Majority of patients do report better function and improvement in sporting activities or work productivity.

Complications

Neurovascular compromise has been reported with surgical management but more common in surgery involving proximal ulna osteotomy or resection of synostosis. Green et al and Simmons et al had both reported cases of Volkman Contracture following resection of synostosis [3, 23]. There were also multiple report of ulnar, radial and median nerve palsy. Malunion due to loss of derotation position and nonunion of osteotomy site has also been reported.

Conclusion

CRUS remained to be a rare condition and under diagnosed likely secondary to excellent ability of patient to adapt their daily function to reduction of forearm rotation. All cases of CRUS should be investigated for an associated genetic cause and other associated malformations. Patients should be assessed on a case-to-case basis in terms of suitability for surgical intervention based on their functional requirement and their current limitations or symptoms.

References

1. Mital MA. Congenital radioulnar synostosis and congenital dislocation of the radial head. Orthop Clin North Am. 1976;7:375–83.
2. Sachar K, Akelman E, Ehrlich MG. Radioulnar synostosis. Hand Clin. 1994;10:399–404.
3. Simmons BP, Southmayd WW, Riseborough EJ. Congenital radioulnar synostosis. J Hand Surg Am. 1983;8:829–38.
4. Elliott A, Kibria L, M. R. The developmental spectrum of proximal radioulnar synostosis. Skeletal Radiol. 2010;39:49–54.
5. Simmons B, Southmayd W, Riseborough E. Congenital radioulnar synostosis. J Hand Surg. 1983;8(6):829–38.
6. Lewis W. The development of the arm in man. Am J Anat. 1901;1:169–83.
7. Catena N, Divizia M, Calevo M. Hand and upper limb anomalies in Poland syndrome: a new proposal of classification. J Pediatr Orthop. 2012;32:722–6.

8. Gaspar H, Albermann K, Baumer A, et al. Clinical delineation of Giuffrè- Tsukahara syndrome: another case with microcephaly and radio-ulnar synostosis with apparent X-linked semi-dominant inheritance. Am J Med Genet A. 2008;146A:1453–7.
9. Zhu Y, Jin K, Mei H, et al. A family with radio-ulnar synostosis, scoliosis and thick Vermilion of lips: a novel syndrome or variant of Giuffrè- Tsukahara syndrome? Am J Med Genet A. 2012;158A:2036–42.
10. Thompson A, Woodruff K, Feig S, et al. Congenital thrombocytopenia and radio-ulnar synostosis: a new familial syndrome. Br J Haematol. 2001;113:866–70.
11. Thompson AA, Nguyen LT. Amegakaryocytic thrombocytopenia and radio-ulnar synostosis are associated with HOXA11 mutation. Nat Genet. 2000;26:397–8.
12. Castillo-Caro P, Dhanrah S, Haut P, et al. Proximal radio-ulnar synostosis with bone marrow failure syndrome in an infant withour a HOXA11 mutation. J Pediatr Hematol Oncol. 2010;32:479–85.
13. Wilkie DPD. Cngenital radio-ulnar synostosis. Br J Surg. 1914;1:366–75.
14. Je C, Omer GE. Congenital proximal radio-ulnar synostosis: natural history and functional assessment. J Bone Joint Surg Am. 1985;67-A:539–45.
15. Fuminori K. Mobilisation of congenital proximal radio-ulnar synostosis: a technical detail. Tech Hand Up Extrem Surg. 1997;1(3):183–8.
16. Kanaya F, Ibaraki K. Molisation of a congenital proximal radioulnar synostosis with use of vascularized Fascio-fat graft. J Bone Joint Surg. 1998;80(8):1186–92.
17. Oka K, Doi K, Suzuki K et al. In vivo three-dimensional motion analysis of the forearm with radioulnar synostosis treated by the Kanaya Procedure. J Orthop Res 2006; 1028-1035
18. Kao HK, Chen HC, Chen HT. Congenital radioulnar synostosis treated using a microvascular free Fascio-fat flap. Chang Gung Med J. 2005;28:117–22.
19. Horii E, Koh S, Hattori T, et al. Single osteotomy at the radial diaphysis for Congenital radioulnar synostosis. J Hand Surg Am. 2014;39(8):1553–7.
20. Murase T, Tada K, Yoshida T, et al. Derotational Osteotomy at the shafts of the radius and ulna for Congenital Radioulnar synostosis. J Hand Surg. 2003;28A:133–7.
21. Ramachandran M, Lau K, Jones DH. Rotational osteotomies for congenital radioulnar synostosis. J Bone Joint Surg. 2005;87-B:1406–10.
22. Lin HH, Strecker W, Manske PR, et al. A surgical technique of radioulnar osteoclasis to correct severe forearm rotation deformities. J Pediatr Orthop. 1995;15:53–8.
23. Green WT, M. M. Congenital radio-ulnar synostosis: surgical treatment. J Bone Joint Surg Am. 1969;51:1042.

Part VIII

The Wrist and Hand

What Is the Best Treatment for Paediatric Trigger Thumb (Acquired Thumb Flexion Contracture)?

35

James S. Huntley

Abstract

Paediatric trigger thumb, better termed *pediatric acquired thumb flexion contracture*, is a common condition, usually presenting at about the age of 2 years with thumb interphalangeal joint fixed flexion (more rarely with triggering), and a palpable nodule in the flexor pollicis longus tendon at the level of the metacarpophalangeal joint. The term 'trigger' thumb is a misnomer in the majority of cases, though it provides a useful term for the retrieval of references, given its propagation through the literature. The general natural history across populations has not been well defined, although two recent studies define it well for Korea. Management options include conservative (observation, exercises, splinting) and surgical (open A1 pulley release or percutaneous A1 pulley release). The evidence is predominantly level III and IV. Many of the study interpretations are vulnerable to bias. Additionally, methodological flaws were common and would result in the downgrading of levels of evidence of some papers. Unsurprisingly therefore, the grade and strength of recommendations are weak.

Keywords

Paediatric trigger thumb • Locked thumb • Notta's nodule • Splinting therapy • Interphalangeal joint • Open release of A1 pulley • Percutaneous release

Introduction

Paediatric "trigger" thumb (Fig. 35.1) is a common (1/2000 [1] – 3.3/1000 [2]; incidence variable by ethnic group [3]) acquired condition [2, 4, 5], usually presenting at about the age of 2 years with a fixed flexion deformity of the thumb interphalangeal joint (IPJ), though sometimes with triggering/snapping [4–8] and even more rarely with the nodule or pain as the primary feature [9]. Slakey & Hennrikus [4] and Moon et al. [10] prospectively examined 4719 and 7700 newborn infants respectively, to determine the congenital incidence of 'trigger thumb' – in these studies no cases were found. Slakey & Hennrikus suggested the condition be termed '*acquired thumb flexion contracture*' rather than '*congenital trigger thumb*'. Clinically, a volar "Notta's nodule" is usually palpable in the flexor pollicis longus tendon, at the level of the metacarpophalangeal joint [11, 12].

The natural history of this condition has recently been characterised well in the Korean population [13–15], but is not well established more generally [16]. Our understanding of the condition across geographic areas and ethnic groups remains incomplete [15].

Management options include observation alone, non-surgical (splintage or extension exercises), and surgical (open release of A1 pulley (± limited release of oblique pulley) or percutaneous release). Although there has been substantial progress over the last decade, the relative indications for conservative or surgical management have not been well-defined [17]; this latter topic was recently the subject of a systematic review (Farr et al. [18]). In this chapter, I have sought to establish the evidence base for the natural history and particular treatments.

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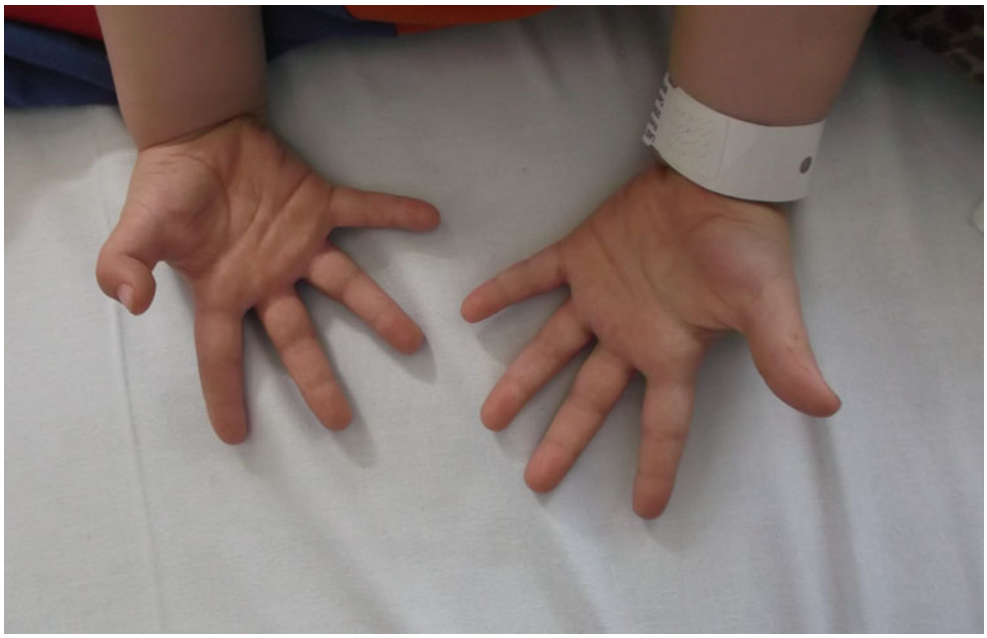

Fig. 35.1 Typical appearance of paediatric acquired thumb flexion contracture (right side)

Questions

The specific questions considered were:

1. What is the natural history of paediatric trigger thumb?
2. What is the best treatment for paediatric trigger thumb?
3. What is the best mode of surgical treatment?
4. What is the best time for surgery?

Search Strategy

A general strategy was developed along **PICO** lines: **P**roblem/**P**opulation: paediatric patients only (>15 years to be excluded) with "trigger" thumb (acquired flexion contracture), **I**ntervention (any – anticipating (i) none (natural history), (ii) exercise, (iii) splinting, (iv) open surgery, (v) percutaneous surgery), **C**omparison (any/none of (i) – (v) above), **O**utcomes (any). Case reports and series of <10 patients were to be excluded.

The Pubmed database was searched using the following strategy (accessed 15/10/2015): (("pediatrics"[MeSH Terms] OR "pediatrics"[All Fields] OR "pediatric"[All Fields]) OR ("pediatrics"[MeSH Terms] OR "pediatrics"[All Fields] OR "paediatric"[All Fields]) OR ("child"[MeSH Terms] OR "child"[All Fields])) AND ("precipitating factors"[MeSH Terms] OR ("precipitating"[All Fields] AND "factors"[All Fields]) OR "precipitating factors"All Fields] OR "trigger"[All Fields]) AND (("thumb"[MeSH Terms] OR "thumb"[All Fields]) OR digit[All Fields]).

This yielded 109 references. Titles and abstracts were reviewed allowing the exclusion of 61; the remaining 48 papers were retrieved in full and analysed, with removal of a further 17. The references from papers in this retrieval allowed access of 4 further relevant papers, not identified in the original search. Ultimately, there were 2 natural history studies, 1 level III systematic review, 1 level II cross-over trial, 6 Level III comparative studies, and 25 Level 4 case series. Searches of the Cochrane controlled trials register and Cochrane database (accessed 11/11/2015) did not provide additional material. The 35 studies are summarized in Table 35.1; country of origin has been included because of the suggestion [35] that there may be geographic/ethnic variation in natural history or response to conservative treatment.

Results

What Is the Natural History of Paediatric Trigger Thumb?

It has been difficult to define the natural history of this condition as the literature is largely dependent on retrospective case series [15], prone to selection and referral biases, with heterogeneous groups followed for variable timescales, and being reported as defining particular spontaneous resolution rates. For instance Skov et al. [27] on the basis of their retrospective series of 37 surgically treated patients, make the statement '*No patient had recovered spontaneously in the interval between the onset of symptoms and operation*'; details of case capture are not given so it is far from clear that the study would have identified such patients; indeed patients might have resolved in other intervals: (i) pre-referral, and (ii) referral to clinic attendance.

Table 35.1 Summary table of included studies

Paper [country]	Design	Number patients	Details/results	Level of evidence
Baek et al. [13] [Korea]	Prospective observational	53 patients; 71 TTs	45/71 (63 %) showed resolution by mean age 5 yrs	Natural history
Baek and Lee [14] [Korea]	Prospective observational – extension of previous study [12]	Extra 25 patients (27 TTs); 1 operated on, 10 lost to FU, leaving 67 children (87 TTs)	FU (median 87.3 mths, range 60–156 mths): 66/87 TTs (75 %) had resolved. All resolutions had occurred by 48 mths	Natural history
Watanabe et al. [19] [Japan]	Case series	48 patients; 60 TTs	Passive exercise programme by mother FU (Mean 44 mths, range 3–90 mths): according to Watanabe stage – 24 had complete recovery (16/20 originally stage 2 (80 %), 8/38 originally stage 3 (21 %); all stage 3 thumbs improved (26/38 progressed to stage 1)	IV
Jung et al. [20] [Korea]	Case series	30 patients; 35 TTs	Extension exercise programme by mother FU (mean 63 mths, range 49–73 mths), 28/35 (80 %) could be fully extended. Five TTs required OR (fully successful)	IV
Forlin et al. [21] [Brazil]	Retrospective case series	11 patients; 13 TTs	Manipulation + home-delivered stretching programme FU (mean 10 yrs, range 5–16 yrs): 7 totally resolved, 3 partly resolved; other 3 required OR (all diagnosed >2 yrs 6 mths)	IV
Nemoto et al. [22] [Japan]	Case series	33 patients; 43 TTs	Custom extension splint night/nap-times 8 patients (10 TTs) dropped out; excluding these, 24/33 TTs recovered completely (73 %) by mean 10 mths. Seven improved; two required OR	IV
Tan et al. [23] [Singapore]	Comparative cohort	115 patients; 138 TTs	Basis of group allocation unclear: (i) '*immediate surgery group*' (59) (ii) '*conservative*' – either splinting or passive stretch (56) FU: minimum 12 mths (a) Splintage success 24/31 (77 %); stretching success 13/25 (52 %). Success of conservative treatment declined with increasing age (b) 13 *ex* 19 failing conservative treatment underwent OR i.e. 72 underwent surgery, only one recurrence (1.4 %)	III
Lee et al. [24] [Taiwan]	Comparative cohort	50 patients *with reducibleTTs*; 62 thumbs – to two groups by parental decision	Group 1 (24 children, 31 TTs – extension splinting) and Group 2 (26 children, 31 TTs) assessed at mean 20 wks FU: cured, improved, non-improved being 12, 10, 9 and 4, 3, 24 in splintage and observation groups respectively	III

(continued)

Table 35.1 (continued)

Paper [country]	Design	Number patients	Details/results	Level of evidence
Koh et al. [25] [Japan]	Comparative cohort	64 patients; 87 TTs (all locked: Watanabe stage 3)	Allocation to splintage group if parents agreed: (i) coil splintage group (32), (ii) observation group (32). Six (19 %) defaulted from splint to observation group Mean FU: (i) 26 mths, (ii) 66 mths Outcomes: resolution rates (i) 24/26, (ii) 23/38	III but *analysis **not** on intention-to-treat basis*
Marriott and Basu [26] [Scotland]	Retrospective case series	80 patients; 103 TTs	94 thumbs operated; FU incomplete – only 57 had post-op visit; alleged 7/94 recurrences (of these, 5 had reoperation – all satisfactory)	IV
Dinham and Meggitt [7] [England]	Retrospective case series	105 patients; 131 TTs	Variety treatments – conservative (observation) and OR: (i) 8/26 of the 'congenital group' resolved by 12 mths (ii) 13/107 resolved during observation (up to 6 mths) (iii) 100/105 operated TTs had 'full movements' at FU (unspecified time-frame); 3 (3 %) had residual fixed flexion deformity; 2 required reoperation	IV
Skov et al. [27] [Denmark]	Retrospective case series	37 patients (from series of 42; 5 lost to FU) with TT	All underwent OR For extended FU (mean 69 mths, range 18–130 mths), 31 recalled to department, 6 contacted by phone. All had full movement and 'normal' thumb strength. Ten (25 %) had bow-stringing of 1–2 mm (previously unnoticed by patients)	IV
Ger et al. [1] [USA]	Retrospective case series	41 patients; 53 TTs	Case capture details not given (i) <6 mths age; 13 patients (19 TTs) observed for mean 44 mths, range 9–139 mths. No SR observed (ii) >6 mths age; only 9 watched for 6 mths before surgery – no SR All 53 TTs came to OR – at one yr FU: no residual contracture	IV
Slakey and Hennrikus [4] [USA]	Case series	15 patients; 17 TTs	All treated by OR FU (mean 12 mths, range 6–35 mths): 16/17 (94 %) had full ROM compared to contralateral	IV
Hierner and Berger [28] [Germany]	Retrospective case series	34 patients with TTs	All treated by OR FU (8 lost to FU; 26 available at 1 yr): 24/26 (92 %) had full ROM (active and passive) compared to contralateral Complications: (i) nerve damage (1; 4 %) (ii) secondary procedure for inadequate release (2; 8 %)	IV
Hudson et al. [29] [South Africa]	Retrospective case series	49 patients; 60 TTs	All treated by OR FU (mean 44 mths): 58/60 (97 %) had full ROM; 2 (3 %) had residual fixed flexion requiring reoperation	IV

(continued)

Table 35.1 (continued)

Paper [country]	Design	Number patients	Details/results	Level of evidence
Mulpruek and Prichasuk [30] [Thailand]	Retrospective case series	42 patients; 54 TTs	Stratified by age onset; SR within 3 mths presentation for 10/42 (predominantly the 8/20 in the age group >6 mths); 32 OR with no residual deformity/recurrence	IV
Dunsmuir and Sherlock [31] [Scotland]	Retrospective case series	192 patients; 227 TTs	After consultation, parents selected (i) observation (53 children; median age 25 mths) or (ii) listed for OR (139 children; median age 30 mths). Outcomes: SR occurred in 26/53 (49 %) from the observation group at a mean of 7 mths (range 1–23 mths). The remaining 27 transferred to surgical group (166 patients), all treated with OR (recurrence rate 3.5 %)	III
Moon et al. [10] [Korea]	Retrospective case series	33 patients; 35 TTs	Protocol: observation if <1 yr, either observation or OR if 1 – 3 yrs, OR if > 3 yrs Outcomes: (i) 12/35 resolved at mean 5 mths (ii) 23 cases treated by OR – no complications (FU unspecified)	IV
McAdams et al. [32] [USA]	Retrospective case series	21 patients; 30 TTs	OR for all (mean operative age 3.3 yrs, range 8 mths–12 yrs) FU long-term: mean 15.1 yrs (range 2–40 yrs) Outcomes: (i) no recurrence/nodules (ii) loss of relative IPJ flexion (23 %) (iii) MCPJ hyperextension (17.6 %) (iv) all seven patients with longitudinal scar had concerns over its appearance	IV
Herdem et al. [33] [Turkey]	Retrospective case series	36 patients	Initial conservative treatment (3 mths stretching exercises) in 26/36 aged <3 yrs uniformly unsuccessful. OR performed for all FU (mean 7 yrs, range 5–15 yrs): all free from contracture with normal ROM	IV
Kuo and Rayan [8] [USA]	Retrospective case series	39 patients; 50 TTs	OR (A1 pulley and 50 % release oblique pulley) of all FU (24 patients, 28 TTs – 15 lost and 6 only by phone) mean 79 mths (range 3–228 mths): (i) no bowstringing (ii) 19/20 resolution Notta's node (iii) 100 % parent/patient satisfaction (iv) 5 thumbs noted to have 15 degrees less IPJ flexion	IV

(continued)

Table 35.1 (continued)

Paper [country]	Design	Number patients	Details/results	Level of evidence
Han et al. [34] [Korea]	Retrospective case series	23 patients, all >5 yrs at operation	Clinical FU (mean 2 yrs 3 mths, range 9 mths–4 years 7 mths): all achieved full extension; no complaint of weakness or functional limitation	IV
Leung et al. [9] [Hong Kong]	Retrospective case series	180 patients; 209 TTs	All treated by OR FU (mean 5 mths, range 3–29 mths): 193/209 (95 %) full ROM, 9/209 (4 %) residual contracture, 5 (2 %) required reoperation	IV
Marek et al. [35] [USA]	Retrospective case series	173 patients; 217 TTs	All treated by OR Clinical FU (mean 27 days, range 2–840 days): all achieved full extension with no major complication; 5 thumbs had minor skin complications that resolved with conservative management	IV
Chalise et al. [36] [Nepal]	Case series	45 patients	Treatment according to Watanabe stage [(0–2): 29 treated by minimum 9 months stretching exercises; (3) treated by OR] FU (minimum 12 mths): (i) conservative treatment successful in 21/29 (72 %), 82 % in <1 yr, 50 % in > 3 yr age (ii) 2/24 recurrence rate for surgically treated group	IV
Farr et al. [18] [Austria]	Systematic review of open surgery *vs* non-operative treatment for paediatric TT	17 retrospective studies, 1 prospective study identified with at least 12 mths mean FU	OR (634 children, 759 thumbs), splinting (115 children, 138 thumbs), passive exercising (89 children, 108 thumbs). Full IPJ motion restored in 95 % children undergoing OR, 67 % children splinted, 55 % after passive exercising	III
Wang and Lin [37] [Taiwan]	Case series (2 comparative groups for type anaesthaesia: GA *vs* LA)	33 patients, 40 TTs	All treated by PR FU was mean 4.7 yrs, range 16 mths–13 yrs 1 mth: Overall success rate 36/40 (96 %); 15/16 and 21/24 in GA and LA groups respectively. No neurological complication	III as regards GA *vs* LA**, IV for procedure per se**
Wang and Lin [38] [Taiwan]	Case series	28 patients, 32 TTs	All treated by OR, forming an historical control group for the PR series in [32] (above) Results: no complications over unspecified FU period	IV**
Ruiz-Iban et al. [39] [Spain]	Case series	23 patients, 27 locked TTs	All treated by PR FU (1TT lost) at mean 3 yrs (range 1–7.2 yrs): 25/26 excellent results. One recurrence at one mth (4 % failure rate) treated by uncomplicated OR	IV

(continued)

Table 35.1 (continued)

Paper [country]	Design	Number patients	Details/results	Level of evidence
Ramirez-Barragan [40] [Spain]	Comparative cohort study	108 patients, 135 TTs	135 TTs: 92 OR *vs* 43 PR (group allocation by surgeon's preference) FU (mean 24 mths, range 4–60 mths): (i) No operative complications (ii) Higher recurrence for PR: 15/43 (35 %) *vs* 6/92 (7 %)	III
Fuentes et al. [41] [Spain]	Comparative cohort study	159 patients, 176 TTs	176 TTs: 124 OR *vs* 52 PR (group allocation by surgeon's preference) FU time unclear. Results: (i) No operative complications (ii) Lower mean operative time for PR: 14.56 *vs* 33.49 mins ($p < 0.01$) (iii) Higher recurrence for PR: 6/52 (11.5 %) *vs* 7/124 (5.4 %)	III
Sevancan et al. [42] [Turkey]	Case series	26 patients, 31 TTs	All treated by PR FU (mean 2.5 yrs, range 1–4.5 yrs): no complications and 1 recurrence at post-op wk 3 (treated successfully by OR)	IV
Amrani et al. [43] [Morocco]	Retrospective case series	52 patients, 63 TTs	All underwent PR FU (mean 28 mths, range 12–30 mths), 50 thumbs available to review: 48/50 good; 2 (4 %) recurrences requiring OR. No neurovascular complications	IV
Masquijo et al. [44] [Argentina]	Prospective cross-over comparative study	15 patients with 20 locked TTs	All underwent PR followed by staged OR Results: 16/20 tendon laceration by PR; distance between PR and digital nerve 2.45 mm (range 1–4 mm). Trial stopped early because of perceived risks	II

Key: ** = largely duplicate of earlier manuscript; *FU* follow-up, *TT* trigger thumb, *PR* percutaneous release, *OR* open release, *ROM* range of movement, *SR* spontaneous resolution, *IPJ* interphalangeal joint, *MCPJ* metacarpophalangeal joint

Baek et al. [13] reported a prospective observational study on 53 children (71 thumbs; mean age at presentation was 2 years, median follow-up was 48 months) with trigger thumb (mean flexion contracture 26°). Forty-five *ex* 71 thumbs (63 %) showed resolution by an average age 5 years, though some had limitations in IPJ hyperextension by contralateral comparison. In 2011, they extended their earlier study by enrolling 25 more children (27 thumbs), producing a group of 98 thumbs in 78 children, and elongating the observation period to a minimum of 5 years [14]. There was no conservative treatment such as passive stretching or splinting. One child was operated on at parental request and ten were lost to follow-up, leaving a group of 87 trigger thumbs in 67 children (43 boys, 24 girls), to be prospectively followed on a six monthly/annual basis, before/after resolution respectively. At final follow-up (median 87.3 months, range 60–156 months), 66 ex 87 (76 %) had resolved. All these resolutions had occurred by 48 months. In the remaining 21 thumbs, though there was no resolution, the flexion deformities continued to improve beyond 48 months.

Marek et al. [35] highlighted the large range in trigger thumb spontaneous recovery rates in the literature and sought to define a difference between the likelihood of resolution without surgery, between Western and Southeast Asian papers. Of note the studies by Baek et al. are confined to the Korean population. Though Marek et al.'s treatment of the subject was superficial (for instance, no consideration being given to the very large variation in recovery rates within the Western studies selectively identified), it is possible that different geographic or ethnic groups have a different natural history, or indeed responsiveness to splintage, stretching or exercise.

The high resolution rates in the conservative arms of some surgical studies (whether these be based on pathology

(eg Watanabe stage) or parent selection based; [31]; Moon et al. 2001; [36]) support the contention for a period of observation/conservative management. The two studies by Baek et al. [13, 14], supported by the resolution and/or improvement rates in the conservative treatment (exercises or splintage) studies below ([19, 22–25]; Jung et al. [20, 21]) support the contention that a patient maybe safely observed for several years, and further that there is no evidence that such delay compromises subsequent surgical treatment/outcomes. However prolonged observation/exercise/splintage may be more problematic for parents and patients than an operation.

What Is the Best Treatment for Paediatric Trigger Thumb?

Exercise Programme

Four key studies (all level IV, with no control group) have sought to define the role of an exercise programme in the treatment of this condition:

(1) Watanabe et al. [19] reported a case series of conservative management (passive exercise by the mother – flexion and extension of the affected thumb IPJ, with the metacarpophalangeal joint held in extension) of 48 children (19 boys, 29 girls; mean age at diagnosis 26 months, range 0–48 months; mean follow-up 44 months, range 3–90 months) with paediatric trigger thumb (60 thumbs; 12 bilateral). Presentations were divided into 4 stages (see Table 35.2 for description of the Watanabe classification), and in this series, 38 and 22 thumbs (63 % and 37 %) were initially assigned to stages 3 and 2, respectively. Two patients (2 thumbs, both stage 2) excluded themselves during the course of treatment. Twenty-four thumbs (16/20 originally stage 2 (80 %), and 8/38 originally stage 3 (20 %) had complete recovery (mean follow-up 62 months, range 12–90 months). All stage 3 thumbs improved (8/38 became normal; 26/38 progressed to stage 1; 4/38 became stage 2) (Table 35.2).

(2) Herdem et al. ([33]; see below – reported under surgical series, as all patients came to surgery) reported a retrospective case series of 36 children (18 boys, 18 girls; mean age 34 months, range 9 months–13 years) with trigger thumb (47 thumbs) treated surgically by open release, in whom 3 months passive stretching exercises in all patients diagnosed under 3 years age (26 patients) was uniformly unsuccessful.

(3) Jung et al. [20] performed a prospective analysis on 30 patients (35 thumbs: 10 right, 15 left, 5 bilateral) with paediatric locked/trigger thumb presenting at a mean age of 28 months (range 11–50 months). Treatment was by an extension exercise programme (10–20 x per day) delivered by the mother (compliance unknown). The Watanabe classification (Table 35.2) for severity was used at all assessment points, with the modification that stage 0 was split into A/B, being 'Extension beyond 0 degrees without inducing triggering' and 'Extension to 0 degrees without inducing triggering', respectively. At follow-up (mean 63 months; range 49–73 months), 80 % of all thumbs could be fully extended, and none deteriorated (all except one improved). Five thumbs (in four patients) required surgical treatment for residual triggering at follow-up; all had "recovered fully" by 6 months minimum post-operative follow-up. In light of a spontaneous resolution rate of 80 % by a minimum of 4 years after presentation, the authors suggested that '*if there are no risk factors that suggest a poor prognosis, a feasible approach is to wait until school age (7 years of age) because the chance of spontaneous resolution is so high and delayed surgery does not adversely affect the outcome*.'

(4) Forlin et al. [19] reported a single surgeon retrospective case series on children (11 patients; 7 boys, 4 girls; mean age at first consultation 26.3 months, range 11–36 months) with trigger thumb (13 thumbs; 2 bilateral) treated by manipulation and home-delivered stretching programme. At follow-up (mean 10 years, range 5–16 years), 10/13 were "satisfactory", 7 being totally resolved and 3 partly resolved. All three thumbs requiring surgery were initially diagnosed after the age 2 years 6 months.

Three of four studies (all without a control arm) suggested high response rate to an exercise programme over protracted time-periods; conversely, the one remaining study [33], with

Table 35.2 Watanabe classification of paediatric trigger thumb [19]

Stage	Condition
0	Normal
1	Locking in flexion or extension
	Active movement with a triggering
2	Locking in flexion or extension
	Passive movement with a triggering
3	Locked in flexion or extension

26 patients <3 years, suggested no resolution in response to a stretching programme over a short time period (3 months).

Splintage

Four key studies (3 level III with comparative groups; 1 level IV, with no control group) have sought to define the role of splintage in the treatment of this condition:

(1) Nemoto et al. [22] reported a case series of 33 patients with 43 trigger digits (40 thumbs and 3 fingers) treated with a custom extension splint at night and nap-times, the IPJ being stabilised in maximum extension by volar polyethylene with a dorsal strap. Eight patients (10 digits) dropped out of treatment; 24 digits '*recovered completely*' (73 % cure rate) by an average of 10 months, seven improved and two digits required surgery. The authors concluded that '*splint therapy is effective in treating trigger thumbs*'. However, given that there was no control group, this study did not establish whether splintage has a better resolution rate than would have occurred with observation alone.

(2) Tan et al. [23] reported results of 138 trigger thumbs (23 bilateral) from 115 children (57 boys, 58 girls). Fifty-nine children were treated surgically (the '*immediate surgery group*'; mean age presentation 26.5 months); 56 patients were treated '*conservatively*' (mean age presentation 19 months), either by splinting or being taught a passive stretching routine. It is not clear as to how the allocation between groups was made. There was a success rate of 66 % after conservative treatment. The mean period of successful conservative treatment was 6 months (range 3–9 months), and minimum follow-up was 12 months. Splintage success was 24/31 (77 %) and parental stretching success was 13/25 (52 %). Success of conservative treatment decreased with increasing age, being 85 % (17/20) in <1 year, 61 % (11/18) in 1–2 years, and 50 % (9/18) in >2 years. Thirteen (*ex* 19) patients who failed conservative treatment went on to surgery – meaning that 72 patients underwent surgery, with one recurrence (1.4 %).

(3) Lee et al. [24] reported a comparative cohort study on 50 children (mean age 1 year 11 months, range 0–4 years; 20 male, 35 female) with Watanabe stage 1 or 2 thumbs (i.e. reducible at presentation; see Table 35.2; over the same time period 16 children with 16 irreducible thumbs were treated surgically and excluded from the study), allocated to one of two groups: (i) extension splinting (24 children, 31 thumbs, splinted for mean 11.7 weeks, range 4–24 weeks), or (ii) observation alone (26 children, 31 thumbs), on the basis of parental selection after detailed information. Baseline data were presented and were broadly similar between the two groups. At variable follow-up (mean of 18.1 and 22.1 weeks for the splinted and observation groups, respectively; last follow-up was by telephone interview), thumbs were classed as 'cured' (full range of movement with no snapping), 'improved' (full range of movement with minimal snapping, <one episode/week) or 'nonimproved' (persistent or recurrent flexion contracture), these results being 12, 10, 9 in the splintage group and 4, 3, 24 in the observation group. Subsequent A1 pulley release in non-improved thumbs still had excellent results. This level III study applies only to those thumbs that were initially reducible (a relatively small subset of pediatric trigger thumbs).

(4) Koh et al. [25] reported a retrospective comparative study of 64 children (28 boys, 36 girls; at first visit, mean age 2 years 6 months, range 3 months to 7 years 10 months) with trigger thumb locked at the IPJ (Watanabe stage 3). Puzzlingly, they alluded to 16 further patients who were excluded because the follow-up period was less than 2 years without complete resolution, though the splintage group had follow-up of mean 26 months (and standard deviation 25 months). Patients were offered a custom-made coil night-splint, extending the interphalangeal joint, but preventing metacarpophalangeal hyperextension, and allocated to the splintage group if patients and parents agreed. Originally 32 were allocated to the splintage group but 6 (19 %) dropped out to the observation group, leaving 26 in the splintage group, and 38 in the observation group. The follow-up periods were markedly different, being (months; mean ± SD) 26 ± 25 and 66 ± 43, for the splint and observation groups respectively. The complete resolution rates were documented as 24/26 and 23/38. Importantly, this analysis was not made on an *intention-to-treat* basis.

Two of the studies were from Japan [22, 25], one from Singapore [23] and one from Taiwan [24]. It may be that results from these regions are not generalizable to other geographic or ethnic areas/groups. Although the response rates to treatment seem encouraging, there are major reservations in their interpretation: [22] was level IV, with no control group; [24] applies only to a small subset of thumbs; the analysis of [25] was not performed on an intention-to-treat basis. There is, therefore, minimal evidence that splintage is better than observation alone. In addition, conservative treatment may be harder to endure and convince for patients and parents, than an operation.

Surgery

Eighteen key studies (1 level III with comparative groups [31]; 1 level III systematic review (Farr et al. [18]); 16 level IV; listed and considered chronologically) have sought to define the role of open surgery (studies ostensibly on percutaneous ± open surgery are considered in a later section) in the treatment of this condition:

(1) Marriott and Basu [26] reported their observations on a retrospective case series of 80 patients (34 male, 46 female) with trigger thumb (23 bilateral, 103 thumbs), presenting over an 18-year period, 1946–1963. Only 6/80 presented at younger than 3 months; 52 *ex* 80 presented at 1–3 years age. Of 80, 57 were followed up with a post-op visit, and of these, 45 were followed 1 month later. From 94 operations, there were 7 recurrences (7.4 %); 5 of these underwent reoperation, all with satisfactory results.

(2) Dinham and Meggitt [7] reported a retrospective case series of 105 children (54 boys, 51 girls) with paediatric trigger thumb (26 bilateral; 131 thumbs; diagnosed at mean age of 2 years, range birth – 11 years; 19 patients were deemed 'congenital' and 7 of these were bilateral i.e. 26 thumbs in this group) treated by observation or surgical release. Eight (ex 26; 31 %) of the 'congenital' group resolved spontaneously by 12 months. In the whole group of 131 thumbs, 107 were '*watched for up to 6 months*', 13 recovered spontaneously. It is difficult to define the 'non-congenital group' in this manuscript, but the authors assert that for '*trigger thumbs in children first noticed between the age of six to thirty months*', it is safe to watch 'for six months because there is an expected spontaneous recovery rate of about 12 %'. Given that 81 *ex* 107 thumbs underwent operation after 6 months, the cadre left for analysis was prohibitively small as to whether further resolution might be possible. For 105 operated thumbs at follow-up (period unspecified), 100 (95 %) had 'full movements', 3 (3 %) had a residual 15 degree fixed flexion deformity, and 2 required reoperation.

(3) Skov et al. [27] reported a retrospective surgical case series of 37 patients (17 boys, 20 girls) from an identified case load of 42 (i.e. loss to follow-up of 5 patients), operated on (mean operative age 38 months, range 10–144 months) for trigger thumb at <15 years. For extended follow-up (mean 69 months, range 18–130 months), 31 patients were examined in the department and 6 were contacted by telephone. All had full movement, no nerve damage, and 'normal' thumb strength. Ten thumbs (25 %) had bow-stringing of 1–2 mm, all previously unnoticed by patients.

(4) Ger et al. [1] reported a retrospective case series of 41 surgically treated patients with pediatric trigger thumb (53 thumbs), collated over a 30-year period (1959 to 1989) at the AI du Pont Institute, '*a major regional referral center for pediatric orthopaedics*.' The case capture details were not given. Group 1 (<6 months age; 13 patients; 19 thumbs) had a mean observation period of 44 months (range, 9–139 months). No thumbs resolved without surgery. In group 2 (>6 months age; 28 patients; 34 thumbs; mean age at diagnosis 42 months, range 10–148 months), only 9 patients were watched for 6 months before surgery, but again there were no cases of resolution without surgery. No thumb had any residual contracture when examined at one year post-operatively.

(5) Slakey and Hennrikus [4] reported a case series of 15 children (8 boys, 7 girls; mean age at diagnosis 24 months, range 3–49 months) with paediatric trigger thumb (2 bilateral; 17 thumbs) treated by surgical release (mean operative age 30 months, range 15–51 months). At follow-up (mean 12 months, range 6–35 months), 16 (94 %) had a full range of movement, equivalent to the contralateral side; 1 thumb had a ten degree hyperextension deficit compared with the other side.

(6) Hierner and Berger [28] reported a retrospective case series of 34 patients with trigger thumb, treated by open release of A1 pulley; 26 patients were available for follow-up (at 1 year) free active and passive movement comparable to the contralateral side was achieved in 24 (92 %). One case (4 %) had nerve damage, and two (8 %) cases required a secondary procedure due to inadequate release.

(7) Hudson et al. [29] reported a retrospective case series of 49 children (mean age 1.9 years) with paediatric trigger thumb (11 bilateral; 60 thumbs) treated by surgical release. At follow-up (mean 44 months), 58 (97 %) had a full range of movement, 2 (3 %) had a residual fixed flexion deformity requiring reoperation.

(8) Mulpruek and Prichasuk [30] reported a retrospective case series of 42 children (mean age at initial visit 2.6 years, range 1–6 years) classified into 3 groups according to noted age onset: group 1 (<6 months; 14 cases), group 2 (>6 months; 20 cases), group 3 (uncertain; 8 cases) with paediatric trigger thumb (54 thumbs; 12 bilateral) in whom surgical release was offered for all. Spontaneous recovery occurred in 10 out of 42 (1 *ex* 14 in group 1, 8 ex 20 in group 2, 1 *ex* 8 in group 3; 24 % overall) within 3 months of the initial visit. It was noted that no bilateral case had spontaneous resolution. The 32 operative cases had a mean operative age of 35 months (range 18–72 months); there were no complications. At follow-up (mean 40 months, range 8–65 months), there were no residual/recurrent fixed flexion deformities.

(9) Dunsmuir and Sherlock [31] reported a retrospective case series (with two comparative groups) of 192 children with trigger thumb (227 thumbs; 93 boys, 99 girls). At the presentation consultation, information was given to the parents who then selected either observation or surgery: 53 children (57 thumbs; 28 %; median age 25 months, range 1–156 months) were observed, and 139 children (170 thumbs; 72 %; median

age 30 months, range 7–108 months) were listed for surgery (open release A1 pulley). In the observation group, spontaneous recovery occurred in 26 patients (49 %) with no subsequent recurrence and no residual flexion deformity; the mean period of observation was 7 months (range 1–23 months). Twenty-seven patients from the observation "deferred" group came to surgery i.e. 166 patients had surgery at a median of 46 days after listing (recurrence rate post surgery was 3.5 %).

(10) Moon et al. [10] reported a retrospective case series of 33 children with trigger thumb (35 thumbs; 2 bilateral; 33 locked and only 2 triggering). At presentation, 4 (thumbs) cases were <1 year, 26 were aged 1–3 years, and 5 were >3 years. The protocol was: observation of those presenting in first year of life until their first birthday, either observation (until 3 years) or surgical release for children aged 1–3 years at presentation, and surgical release if > 3 years age. Twelve out of 35 cases resolved (at a mean of 5 months observation, range 1–24 months). Twenty-three cases were operated on (mean operative age 29 months, range 13–57 months) without complication (follow-up period and mode of assessment unspecified).

(11) McAdams et al. [32] reported a retrospective case series of 21 children (9 boys, 12 girls) with trigger thumb (30 thumbs, 9 bilateral) with long-term follow-up (mean 15.1 years, range 2–40 years), having previously undergone surgical release of the A1 pulley (mean operative age 3.3 years, range 8 months–12 years). No recurrences and no triggering/nodules were found – nor was there any functional deficit. However 23 % had a loss of relative IPJ motion, and 17.6 % had metacarpophalangeal joint hyperextension, this being unrelated to the age at which surgery had been performed. Seven *ex* 21 had a longitudinal scar, and all these had concerns over the scar appearance. No information was given as to the adequacy of case capture/retrieval, leaving a concern as to the representativeness of this series.

(12) Herdem et al. [33] reported a retrospective case series of 36 children (18 boys, 18 girls; mean age 34 months, range 9 months–13 years) with trigger thumb (47 thumbs; 11 bilateral) treated surgically by open release. Conservative treatment (passive stretching exercises) was instituted in all patients under 3 years age (26 ex 36 patients; 3 months programme). This was unsuccessful in all patients, so open release was performed for all with mean follow-up 7 years (range, 5–15 years); all patients were free from contracture and nodule and had a normal range of movement.

(13) Kuo et al. (2010) reported a retrospective case series of 39 consecutive patients (25 boys, 14 girls) with paediatric trigger thumb (50 thumbs; 11 bilateral) presenting at a mean age of 24 months [range: birth – 54 months], with (26 *ex* 28) mean IPJ flexion contracture of 49 degrees (range 10–90 degrees) and treated surgically by release of the annular *pulley and partial (50 %) release of the oblique pulley*. Fifteen patients were lost to follow-up, and a further six were assessable only by telephone interview. Of 24 patients (28 thumbs), at follow-up (mean 79 months; range 3–228 months), there was: (i) no bowstringing, (ii) Notta's node resolution (19/20), and (iii) 100 % parent/patient satisfaction. Five thumbs were noted to have a mean of 12 degrees less IPJ flexion.

(14) Han et al. [34] reported a retrospective case series of 23 consecutive children (9 boys, 14 girls) with paediatric trigger thumb (8 bilateral; 31 thumbs) treated by surgical release *at age >5 years* (mean operative age 7.5 years). There were no post-operative complications. At clinical follow-up (mean 2 years 3 months, range 9 months–4 years 7 months) all had regained full extension, and there was no complaint of grip weakness or functional limitation.

(15) Leung et al. [9] reported a retrospective case series of 180 children (88 boys, 92 girls; mean age at onset 19 months, range 1 month–6 years, and 20 % diagnosed at less than 1 year i.e. in infantile group) with paediatric trigger thumb (29 bilateral; 209 thumbs; 48 infantile, 161 childhood) treated by surgical release. At follow-up (mean 5 months, range 3–29 months), 193 (95 %) had a full range of movement with '*scarcely apparent scar*', 9 (4 %) had a residual fixed flexion deformity, 3 (1 %) had a hypertrophic scar, and 5 (2 %) required reoperation. Of the 9 thumbs with a residual flexion deformity, 4 (*ex* 48; 8 %) and 5 (*ex* 161; 3 %) were from the infantile and childhood groups, respectively; these 9 were subjected to a physiotherapy regime, but 3 from each group were adjudged failures (6 % and 2 %, respectively).

(16) Marek et al. [35] reported a retrospective case series of 173 patients (101 boys, 72 girls) with trigger thumbs (217 thumbs; 66 unilateral left, 65 unilateral right and 43 bilateral – *NB this would equate to 174 rather than the reported 173 patients*) treated by surgical release, the indication being a locked trigger thumb for at least 6 months. Allegedly the commonest history at the time of presentation was intermittent triggering with the thumb eventually becoming locked in flexion. Age of onset of symptoms was 25 months (range 0–78 months), and time to surgery was mean 11 months (range 2–90 months) – this means that there were patients in whom the 6 month stipulation for chronicity was not observed. At clinic follow-up (mean 27 days, range 2–840 days), all 217 thumbs achieved full extension; there were no digital nerve injuries or other major complications. The authors supplemented the clinical

study with a practice pattern survey of pediatric hand surgeons. In their abstract, they concluded that surgical release '… *is the treatment of choice for a locked pediatric trigger thumb.*' This far-reaching conclusion is justifiable on the data presented by the authors only in so far as it reflects the practice/opinion of the hand surgeons surveyed.

(17) Chalise et al. [36] documented a case series of 45 patients (24 boys, 21 girls; mean age at presentation 28.5 months, range 3 months–7 years) in which management was stratified according to Watanabe stage: (i) stages 0, 1, 2 (i.e IPJ flexion contracture reducible by gentle manipulation): 29 patients initially treated by stretching exercises, and (ii) stage 3 (i.e irreducible locked IPJ): 16 patients treated by surgical release of the A1 pulley. If 9 months of conservative treatment was ineffective, patients were offered surgery. Minimum follow-up was 12 months. Conservative treatment was successful in 21/29 (72 %), but decreased from 82 % (9 cases) in the <1 year age, to 50 % (3 cases) in >3 year age. Surgically treated cases were successful in 22/24, with recurrence of flexion contracture in 2/24 (8.3 %).

(18) Farr et al. [18] performed a systematic review comparing open surgery and non-operative treatments, limited to follow-up data of mean at least 12 months. They identified seventeen retrospective studies and one prospective study, reporting on open surgery (634 children, 759 thumbs), splinting (115 children, 138 thumbs) and passive exercising (89 children, 108 thumbs). Whilst acknowledging the low levels of evidence available their guarded conclusion was that '*open surgery resulted in more reliable and rapid outcomes compared with non-operative treatment.*'

As explicitly noted by Leung et al. [9], hospital-based case series data are substantially vulnerable to referral bias, which itself are likely to vary substantially between health systems and across cultures. Nevertheless, the series presented support the conclusions of the systematic review outlined above (Farr et al. [18]): (i) '*Full interphalangeal joint motion without residual triggering was achieved in 95 % of all children undergoing surgery*' (though it is worth emphasising that only open release was considered as a form of surgery), and (ii) '*open surgery results in more reliable and rapid outcomes compared with nonoperative treatment.*' Conversely, given the improvement of thumbs with conservative management, and no evidence of a decline in the excellent results with time to surgery (especially Han et al. 2002), a period of observation may be prudent and acceptable to parents/child. Additionally, limited data [32] suggest that a transverse incision would be preferred to a longitudinal one for open release.

What Is the Best Mode of Surgical Treatment?

Percutaneous releases have been considered as an alternative to open surgery, largely as an extension from this approach for trigger digits from the adult literature [45–47]. Eight studies were identified ([37–40]; Fuentes et al. [41–44]), 5 level IV, 2 level III and 1 level II. These studies are outlined below:

(1) Wang and Lin [37] reported a case series of 33 children (19 boys, 14 girls) with trigger thumb (40 thumbs; 7 bilateral; mean age onset 2.4 years) treated by percutaneous release (mean operative age 2.5 years, range 10 months–8 years 9 months) with a 19 guage hypodermic needle. Patients were allocated to two groups for anaesthaesia, according to "*tolerance of the child and parents*": (i) general anaesthaesia (13 patients), local anaesthaesia (20 patients). At follow-up (mean 4.7 years, range 16 months–13 years 1 month), successful releases were obtained in 15 *ex* 16 and 21 *ex* 24 thumbs, for the general and local anaesthetic groups, respectively i.e overall success rate of 36/40 (90 %). Examination for nerve injury did not indicate any adverse sequelae. A satisfactory release was obtained in 30 patients (91 %) with one recurrence at postoperative week 3 (subsequently treated by open release), 1 having a hypertrophied pulley too thick for percutaneous release, and 2 patients becoming uncooperative during the procedure under local anaesthaesia to a sufficient degree for parents to refuse further management.

(2) Wang and Lin [38] produced a largely duplicate publication (without citation of their 2004 paper) with substantial results and textual homologies to the earlier paper concerning the series with percutaneous release; there is a puzzling alteration in the average length of follow-up of this cohort – this being 4.7 years in the first paper and 5.7 years in the second, although the range remains the same. The additional data in this paper were from a historical 'control' group of 28 patients (16 boys, 12 girls; mean age at presentation 11 months, range 0–50 months) who had open releases for trigger thumbs (32 thumbs; 4 bilateral) at a mean operative age of 28 months (range 8–60 months), and had uniformly (100 %) satisfactory outcomes over an unspecified time period.

(3) Ruiz-Iban et al. [39] reported a prospectively collated case series of 23 children (stated as 5 boys and 17 girls (albeit this indicates 22 children!)) under 6 years of age

(mean age 2.7 years, range 14 months–5.6 years) with trigger thumb (27 thumbs; 4 bilateral; all locked in flexion i.e. Watanabe stage 3) treated by percutaneous release (PR) of the A1 pulley. One child was lost to follow-up, leaving 22 children (26 thumbs) at follow-up (mean 3.0 years, range 1–7.2 years), which involved assessment of IPJ and MCPJ range, pinch strength, sensibility (two-point-discriminator) and triggering (with comparison of contralateral side): 25 *ex* 26 (96 %) had excellent results with no residual triggering (two of these had a mild MCPJ extension deficit at one month post-operatively, which resolved with exercises). One child had a recurrent locked thumb at the one month visit (representing a 4 % failure rate) and underwent uncomplicated open release one month further on. In no patient was there a sensibility defect, loss of strength, IPF motion loss or MCPJ hyperextension deformity. On the basis of these data, the conclusion drawn by the authors was that the technique is "safe", a conclusion from this small sample size, that this reviewer believes is too far reaching – as often exemplified by the problem with small (or zero) numerators [48].

(4) Ramirez-Barragan et al. [40] reported a retrospective study on 108 children (51 boys, 57 girls) with trigger thumb (135 thumbs; 27 bilateral) comparing open (92 cases) and percutaneous (43 cases) release surgeries with both groups being performed under general anaesthaesia, and case allocation being by surgeon's preference. At follow-up (mean 24 months, range 4 months–60 months), there were no complications of lack of sensitivity, residual pain or alteration of thumb mobility in either group. However, there were recurrence rates of 6 *ex* 92 (7 %) and 15 *ex* 43 (35 %) in the open and percutaneous groups respectively. In light of the high recurrence rate following percutaneous surgery (a six times higher recurrence risk), the authors made a preferential recommendation for the open technique.

(5) Fuentes et al. [41] reported a retrospective study on 159 children with trigger thumb (176 thumbs; 17 bilateral; 86 boys, 90 girls) comparing open (124 cases) and percutaneous (52 cases) release surgeries, with the open group performed under general anaesthaesia, the percutaneous group under general sedation, and case allocation and subsequent check-ups were by treating surgeon's preference (follow-up was *'during the post-operative period'* for the open release, and *'3 times every week'* for the percutaneous group; the lengths and assessors of follow-up are not given). Allegedly, there were no *'complications any type: infection, vascular injury, or neurological*'. For the open surgery group, the mean operative age was 2.65 years, the mean hospital stay 0.9 days, and the mean operative time 33.49 mins (range 10–65 mins). Conversely, for the percutaneous surgery group, the mean operative age was 2.45 years, the mean hospital stay 0.21 days, and the mean operative time 14.56 mins (range 5–25 mins), this latter parameter being significantly reduced ($p < 0.01$). However, there were recurrence rates of 7 *ex* 124 (5.6 %) and 6 *ex* 52 (11.5 %) in the open and percutaneous groups respectively; all recurrences were treated by open release *'with 100 % success'*. The authors asserted (without supportive evidence being presented) that *'most of the relapses following percutaneous polectomy occurred in patients who had not done the home flexion-extension exercises or who had done so improperly'*.

(6) Sevancan et al. [42] reported a case series of 26 children (12 boys, 14 girls) with trigger thumb (31 thumbs; 5 bilateral; 28 rigid type, 1 passive trigger, 2 active trigger) treated by percutaneous release (mean operative age 2.6 years, range 14 months–5 years) with a well described technique involving an 18 guage needle attached to 10 cc saline-filled syringe. At follow-up (mean 2.5 years, range 1–4.5 years), examination for nerve injury did not indicate any sequelae. A satisfactory release was obtained in 30 (97 %) with one recurrence at postoperative week 3 (subsequently treated by open release).

(7) Amrani et al. [43] reported a case series of 52 children (29 boys, 23 girls) with trigger thumbs (63 thumbs, 11 bilateral) treated by percutaneous release of the A1 pulley (at mean operative age 32 months, range 12 months–5 years). At follow-up (mean 28 months, range 12–30 months), 50 thumbs were available for review: 48 (96 %) had a good result, but 2 (4 %) had recurrent triggering, requiring subsequent open release. No thumb demonstrated any clinical neurovascular deficit.

(8) Masquijo et al. [44] reported a level II prospective, cross-over comparative (internally controlled) study, with 15 patients (8 boys, 7 girls; mean operative age 3.2 years, range 2–7 years) with locked trigger thumbs (number = 20) underwent percutaneous release (PR) followed by a staged open release (OR). Outcomes were (i) thumb extension preoperative (-45.2 ± 21.7°) / after PR (-4 ± 8°) / after OR (0°), (ii) extent of A1 release at PR (100 % in 4/20, >75 % in 2/20, 50–75 % in 14/20), (iii) iatrogenic nerve (0/20) / vessel (0/20) / tendon (16/20 laceration) injury, and (iv) distance between PR and digital nerve (mean 2.45 ± 0.9 mm; range 1–4 mm). Though no iatrogenic nerve injuries were encountered, the trial was stopped early because of the perceived risks. In conclusion, the authors did *'not recommend percutaneous release in the pediatric thumb, given the risk of neurovascular iatrogenic injury or incomplete A1 pulley release'*.

Table 35.3 Summary of grades of recommendations

Recommendation	Level of evidence	Grade
In certain populations, where there is a high spontaneous resolution rate, a conservative approach may be advocated	III, IV	B
Exercise/splintage may be better than observation alone	II, IV	C
Surgery can be safely deferred until at least age 3 years	III, IV; consistent	B
Open surgery (release A1 pulley) reliably and reproducibly (>95 % effective) solves the flexion contracture associated with pediatric trigger thumb	III; consistent	B
For open surgery a transverse skin incision is preferred to longitudinal	IV	C
Open release is the preferred surgical modality (as opposed to percutaneous release)	II, III	B

Excluding the largely duplicate study [38], the four level IV case series [37, 39, 42, 43] report on percutaneous release in 33, 23, 26, and 52 children respectively (134 pooled), with 91 %, 96 %, 97 %, and 96 % success rates. However, the two level III studies ([40]: 92 cases open *vs* 43 cases percutaneous; [41]: 124 cases open *vs* 52 cases percutaneous), incorporating a comparative cohort of patients undergoing open release, have much higher recurrence rates in the percutaneous release groups: recurrence rates (open release *vs* percutaneous release) were 7 % *vs* 35 %, and 5.6 % *vs* 11.5 %, respectively.

The well-designed cross-over study of Masquijo et al. ([44]; 15 patients, 20 thumbs) documented the incompleteness of release performed percutaneously, as well as the associated flexor tendon injury (80 %) and proximity between percutaneous release and the digital nerve; the study was halted early because of the perceived risk

At the current time, the higher level (II – III) evidence concerning the percutaneous technique is averse to the apparently encouraging results reported in the four case series. Despite possible benefits in terms of operative time, the safety and efficacy of this approach are far from proven. Open release remains the standard of surgical care.

What Is the Best Time for Surgery?

The difficulties in defining the natural history of this condition, particularly in the *inter*-study heterogeneity in spontaneous resolution rates [35], have been alluded to earlier. The series of Baek et al. [13, 14] in the Korean population suggested a 76 % resolution rate with observation alone, albeit over an observation period of 4 years. The case series and comparative cohorts on conservative management support the contention that surgery may be deferred with reasonable expectation of resolution.

Dinham and Meggitt [7] documented a 31 % resolution rate (8/26) in their 'congenital group' by 12 months – but markedly less in the older age-group. Chalise et al. [36] found a 82 % resolution rate with conservative management in the <1 year group, dropping to 50 % effectiveness in the >3 year age group. Conversely: (i) Ger et al. [1] documented that all 13 patients in their <6 months group (followed for mean 44 months) failed to resolve spontaneously, and (ii) Mulpruek and Prichasuk [30] found a greater resolution rate in the >6 month age-group (40 %) than the <6 months age-group (1/14; 7 %).

The case series of open surgery do not indicate worse results of surgery with older operative age, in particular: (i) [27]: mean operative age 3 years 2 months (range 10–144 months), (ii) [32]: mean operative age 3.3 years (range 8 months–12 years), (iii) [34]: all operative ages >5 years, with mean operative age 7.5 years.

The best timing of surgery remains dependent on patient and parent specific factors, including: (i) age/size (particularly operative field/anaesthetic risk), (ii) the responsiveness to observation/conservative treatment, and (iii) the 'reasonableness' in *the particular patient population* of observation / conservative treatment.

Conclusions

The discussion and conclusions to this review have been discussed under the appropriate research questions. Grades of recommendations are given in Table 35.3.

References

1. Ger E, Kupcha P, Ger D. The management of trigger thumb in children. J Hand Surg Am. 1991;16:944–7.
2. Kikuchi N, Ogino T. Incidence and development of trigger thumb in children. J Hand Surg Am. 2006;31:541–3.
3. Ashford JS, Bidic SM. Evaluation of paediatric trigger thumb in the Hispanic population at a southwest urban medical center. Plast Reconstr Surg. 2009;124:1221–4.
4. Slakey JB, Hennrikus WL. Acquired thumb flexion contracture in children: congenital trigger thumb. J Bone Joint Surg Br. 1996;78:481–3.

5. Rodgers WB, Waters PM. Incidence of trigger digits in newborns. J Hand Surg Am. 1994;19:364–8.
6. Bauer AS, Bae DS. Pediatric trigger digits. J Hand Surg Am. 2015;40:2304–9.
7. Dinham JM, Meggitt BF. Trigger thumbs in children: a review of the natural history and indications for treatment in 105 patients. J Bone Joint Surg Br. 1974;56:153–5.
8. Kuo M, Rayan GM. Complete annular and partial oblique pulley release for pediatric locked trigger thumb. Hand (NY). 2010;5:408–14.
9. Leung OY, Ip FK, Wong TC, Wan SH. Trigger thumbs in children: results of surgical release. Hong Kong Med J. 2011;17:372–5.
10. Moon WN, Suh SW, Kim IC. Trigger digits in children. J Hand Surg Br. 2001;26B:11–2.
11. Clapham PJ, Chung KC. A historical perspective of the Notta's node in trigger fingers. J Hand Surg Am. 2009;34:1518–22.
12. Notta A. Recherches sur une affection particuliere des gaines tendineuses de la main, caracterisee par le developement de une nodosite sur la trajet des tendons flechisseurs des doigts et par l'empechment de leur mouvements. Archives Generales de Medicine. 1850;4(24):142–62.
13. Baek GH, Kim JH, Chung MS, Kang SB, Lee YH, Gong HS. The natural history of pediatric trigger thumb. J Bone Joint Surg Am. 2008;90:980–5.
14. Baek GH, Lee HJ. The natural history of pediatric trigger thumb: a study with a minimum of five years follow-up. Clin Orth Surg. 2011;3:157–60.
15. Shah AS, Bae DS. Management of pediatric trigger thumb and trigger finger. J Am Acad Ortho Surg. 2012;20:206–13.
16. Bae DS. Pediatric trigger thumb. J Hand Surg Am. 2008;33A:1190–1.
17. Ogino T. Trigger thumb in children: current recommendations for treatment. J Hand Surg Am. 2008;33:982–4.
18. Farr S, Grill F, Ganger R, Girsch W. Open surgery versus nonoperative treatments for paediatric trigger thumb: a systematic review. J Hand Surg Eur. 2014;39:719–26.
19. Watanabe H, Hamada Y, Toshima T, Nagasawa K. Conservative treatment for trigger thumb in children. Arch Orthop Trauma Surg. 2001;121:388–90.
20. Jung HJ, Lee JS, Song KS, Yang JJ. Conservative treatment of pediatric trigger thumb: follow-up for over 4 years. J Hand Surg Eur. 2012;37:220–4.
21. Forlin E, Kaetsu EY, Vasconcelos JEE. Success of conservative treatment of trigger thumb in children after minimum follow-up of five years. Rev Bras Ortop. 2012;47:483–7.
22. Nemoto K, Nemoto T, Terada N, Amako M, Kawaguchi M. Splint therapy for trigger thumb and finger in children. J Hand Surg Br. 1996;21:416–8.
23. Tan AH, Lam KS, Lee EH. The treatment outcome of trigger thumb in children. J Pediatr Orthop B. 2002;11:256–9.
24. Lee ZL, Chang CH, Yang WY, Hung SS, Shih CH. Extension splint for trigger thumb in children. J Pediatr Orthop. 2006;26:785–7.
25. Koh S, Horii E, Hattori T, Hiroishi M, Otsuka J. Pediatric trigger thumb with locked interphalangeal joint: can observation or splinting be a treatment option? J Pediatr Orthop. 2012;32:724–6.
26. Marriott FPS, Basu S. Trigger thumb in infancy and childhood. A survey of 80 patients. Ulster Med J. 1967;36:53–61.
27. Skov O, Bach A, Hammer A. Trigger thumbs in children: a follow-up study of 37 children below 15 years of age. J Hand Surg Br. 1990;15:466–7.
28. Hierner R, Berger A. Tendovaginitus stenosans of the thumb in small children (pollex flexus congenitus). The authors' results and review of the literature. Chirurg. 1997;68:1190–3.
29. Hudson DA, Grobbelaar AO, Bloch CE. Trigger thumb in children – results of simple surgical treatment. S Afr J Surg. 1998;36:91–2.
30. Mulpruek P, Prichasuk S. Spontaneous recovery of trigger thumbs in children. J Hand Surg Br. 1998;23B:255–7.
31. Dunsmuir RA, Sherlock DA. The outcome of treatment of trigger thumb in children. J Bone Joint Surg Br. 2000;82:736–8.
32. McAdams TR, Monheim MS, Omer GEJ. Long-term follow-up of surgical release of the A(1) pulley in childhood trigger thumb. J Pediatr Orthop. 2002;22:41–3.
33. Herdem M, Bayram H, Togrul E, Sarpel Y. Clinical analysis of the trigger thumb of childhood. Turk J Pediatr. 2003;45:237–9.
34. Han SH, Yoon HK, Shin DE, Song DG. Trigger thumb in children: results of surgical treatment in children above 5 years of age. J Pediatr Orthop. 2010;30:710–4.
35. Marek DJ, Fitoussi F, Bohn DC, Van Heest AE. Surgical release of the pediatric trigger thumb. J Hand Surg Am. 2011;36:647–52.
36. Chalise PK, Mishra AK, Shah SB, Adhikari V, Singh RP. The treatment of trigger thumb in children: conservative or surgical? Nepal Med Coll J. 2012;15:122–4.
37. Wang HC, Lin GT. Percutaneous release for trigger thumb in children undergeneral and local anaesthaesia. Kaohsiung J Med Sci. 2004;20:546–51.
38. Wang HC, Lin GT. Retrospective study of open versus percutaneous surgery for trigger thumb in children. Plast Reconstr Surg. 2005;115:1963–70.
39. Ruiz-Iban MA, Gonzalez-Herranz P, Mondejar JA. Percutaneous trigger thumb release in children. J Pediatr Orthop. 2006;26:67–70.
40. Ramirez-Barragan A, Martinez-Caballero I, Epeldegui-Torre T. Open versus percutaneous surgery for trigger thumb in children. Rev Ortop Traumatol (Madr). 2007;51:25–9.
41. Fuentes LM, Carmona FJD, Diaz AT, Moreno EM, Rueda DF. Trigger thumb in children: comparison of results between percutaneous release and open surgery. Rev Esp Cir Ortop Traumatol. 2010;54:351–6.
42. Sevancan A, Innan U, Kose N, Omeroglu H, Seber S. Percutaneous release for trigger thumbs in children: improvements of the technique and results of 31 thumbs. J Pediatr Orthop. 2010;30:705–9.
43. Amrani A, Dandane MA, Alami ZF. Percutaneous release of trigger thumb in children: 63 cases. Chir Main. 2011;30:102–4.
44. Masquijo JJ, Ferreyra A, Lanfranchi L, Torres-Gomez A, Allende V. Percutaneous trigger thumb release in children: neither effective nor safe. J Paed Orthop. 2014;34:534–6.
45. Eastwood DM, Gupta KJ, Johnson DP. Percutaneous release of the trigger finger: an office procedure. J Hand Surg Am. 1992;17:114–7.
46. Gilberts EC, Wereldsma JC. Long-term results of percutaneous and open surgery for trigger fingers and thumbs. Int Surg. 2002;87:48–52.
47. Ha KI, Park MJ, Ha CW. Percutaneous release of trigger digits. J Bone Joint Surg Br. 2001;83:75–7.
48. Hanley JA, Lippman-Hand A. If nothing goes wrong, is everything all right? Interpreting zero numerators. JAMA. 1983;249:1743–5.

Evidence-Based Treatment of Forearm Fractures in Children

36

Oluwarantimi O Ayodele and Alwyn Abraham

Abstract

Paediatric forearm fractures constitute 5.4 % of fractures in children under the age of 16. They are more common in boys and seem to be increasing in frequency. Although the majority of these fractures can be treated non-operatively there has been an increasing trend towards operative intervention. There is no consensus on which fractures should be managed operatively and what the best modality of operative management is. There is no Level I or II studies looking at these two modalities and therefore clinical decisions are guided by published observational studies. These studies do little to suggest a conclusive answer with regards to best practice for this fracture. The orthopaedic community is therefore in a genuine state of equipoise and there is a need for clinically relevant RCTs to help clearly differentiate between these available modalities.

Keywords

Forearm • Radius • Ulna • Fracture • Elastic nails • TENS • Flexible nails • Refracture • Monteggia • Galeazzi • Close reduction • Open reduction

Introduction

"Forearm fractures" refer to fractures of one or both of the radius and ulna. In children, the mechanism of injury is usually a fall onto an outstretched hand. Whilst fractures of the forearm as a whole constitute 4.5 % of paediatric fractures, the majority of these are distal radius and distal ulna [1]. Both bone diaphyseal fractures constituted 6.5 % of fractures in children aged 12 and under in a large epidemiological carried out in Nottingham in 1981 [2]. Boys were twice as likely to sustain these fractures and the average age was 7.8 years. A number of studies from Europe, United States and Asia suggest an upwards trend in the incidence of forearm fractures [3–6]. The reason for this remains unclear although in Scandinavia the accelerating increase in forearm fractures was found to be associated with trampolines [7]. Diaphyseal forearm fractures are the most common site for refracture in the paediatric population. They are also the most common open fracture of the upper limb.

Fracture of the ulna shaft can occur with dislocation of the radial head proximally at the elbow (Monteggia fracture). Reduction of the ulna deformity allows relocation of this dislocation. A similar dislocation of the distal radio-ulnar joint can occur with fractures of the distal radius (Galeazzi fracture) [8].

Fractures of the forearm are acutely painful and at initial presentation require prompt analgesia and plaster cast splintage. Plaster cast management not only provides analgesic after injury but helps to prevent increasing deformity whilst the fracture unites. Pain reduces gradually over the course of two weeks. Once fracture union occurs and motion at the fracture site ceases, splintage can be discontinued and function of the forearm is gradually restored.

The forearm functions as a joint allowing for internal rotation (pronation: palm faces posterior) or external rotation (supination: palm faces anterior) [9]. This ability to pronate

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and supinate depends, amongst other things, on the shape of the radial diaphysis i.e. the radial bow. Interventions for radius and ulna diaphyseal fractures which restore the pre-fracture shape of these bones should therefore result in better forearm pronation-supination function.

Children have significant potential for bone remodelling [10–12]. There are a number of factors that come into play with regards to the potential of a particular fracture to remodel [11]. These include the age of the child, the distance of the fracture from the physis, whether there is any rotational deformity, and the direction of the plane of the fracture relative to the adjacent joints. Rotational deformities in general cannot remodel [13]. Nonetheless the potential to remodel means the majority of fractures; forearm or elsewhere, can be successfully managed non-operatively. Observational studies have suggested that over 90 % of both bones forearm fractures can be managed non-operatively [12, 14]. A recent randomised controlled trial demonstrated good results with casts that can be removed at home without further fracture clinic appointments [15].

The remodelling potential in the forearm is less in the diaphyseal region as compared to the distal one third of the radius and ulna due to increasing distance from the physis. The majority of the remodelling potential in a paediatric long bone originates from the physis. In fact distal radial fractures have been shown to be able to remodel at a rate of 0.9° per month because of the proximity to the physis [13]. Conversely diaphyseal fractures remodel less well and are associated with higher rates of malunion. This is increasingly so with increasing age of the child. They are also associated with significant rates of redisplacement [16].

Operative intervention is therefore required for some fractures either to facilitate indirect reduction (Elastic stable Intramedullary nailing – ESIN) or to stabilise direct reduction (Open Reduction Internal fixation – ORIF). There is no clear consensus on the amount of angulation or malrotation that is acceptable. It is generally accepted that below the age of 10 children have the greatest bone modelling potential. Therefore, less deformity could be accepted in children above the age of 10. Where a child of less than 10 years old could reasonably be expected to remodel up to 20° of diaphyseal angulation this is not as certain in the older child. After the age of 10 years accepting more than 10° of diaphyseal angulation could result in unacceptable results. The decision to operate must therefore be multifactorial taking into account, the fracture morphology and location as well as the age of the child. In addition the ability or not to reduce the fracture and maintain it in a stable fashion will further determine the need for and method of surgery. The risk of redisplacement and therefore the stability of a fracture is a factor of the location of the fracture along the length of the radius or ulna, the quality of the reduction and the quality of the applied plaster cast [16–19] (Figs. 36.1 and 36.2).

Surgical intervention is aimed at reduction followed by stabilisation of the fracture. Fracture reduction reduces the deformity following a fracture by normalising the alignment of the radius and ulna. Reduction can either be closed (externally by manipulation under X-ray control) or open (by surgical exposure, visualisation and instrumentation). Surgical stabilisation of the fracture involves holding the reduction such that the restored alignment following reduction is maintained. Devices for surgical stabilisation can be either internal to the forearm or external such as an external fixator. Internal devices can either be elastic stable intramedullary nails (ESIN) or a combination of plates and screws. Whilst in adults ORIF has been established as the optimal surgical intervention for diaphyseal forearm fractures, in children the options of ORIF and ESIN are both widely used and equally popular.

Not only is there an increase in the incidence of forearm fractures but there is a trend towards surgical stabilisation [3, 7, 14]. An 11 year period from 1998 saw the proportion of fractures being managed with ESIN triple from 10 % to 30 % [7]. The increasing popularity of ESIN can be attributed to the less invasive surgical technique of ESIN with the possibility (perceived) of less complications, the resulting reduced risk of redisplacement postoperatively [20, 21] and the attraction of a prosthesis that is relatively easier to remove than the traditional plates and screws.

The trend has generally been to remove orthopaedic implants once the operated bone has fully healed. This is usually undertaken a year or so after operation before new bone formation buries the implant completely. Removal of ESINs is much easier than plates or other implants that are fixed to the bone. Access to the ends of the ESIN is usually in safe zones, where neurovascular structures (nerves and blood vessels) are not at risk. Conversely plate removal requires a surgical dissection though inter-nervous or inter-muscular planes. These planes are often scarred and less discrete following previous open reduction and internal fixation. Thus repeat surgery, for the removal of plates and screws, requiring dissection may endanger neurovascular structures.

Surgical stabilisation prevents motion at the fracture site, thereby preventing displacement and recurrence of deformity at the fracture site. Anatomical restoration should help to maximise function of the forearm once rehabilitation from the injury is complete.

Generally intramedullary nailing consists of a manipulation (closed reduction) with percutaneous insertion (through a small incision in the skin) of the Elastic Stable Intramedullary Nail (ESIN). This technique allows for the formation of callus from the fracture haematoma which subsequently ossifies. The fracture ends are manipulated to approximate the shape of the pre-fracture radial shaft before being secured using an ESIN.

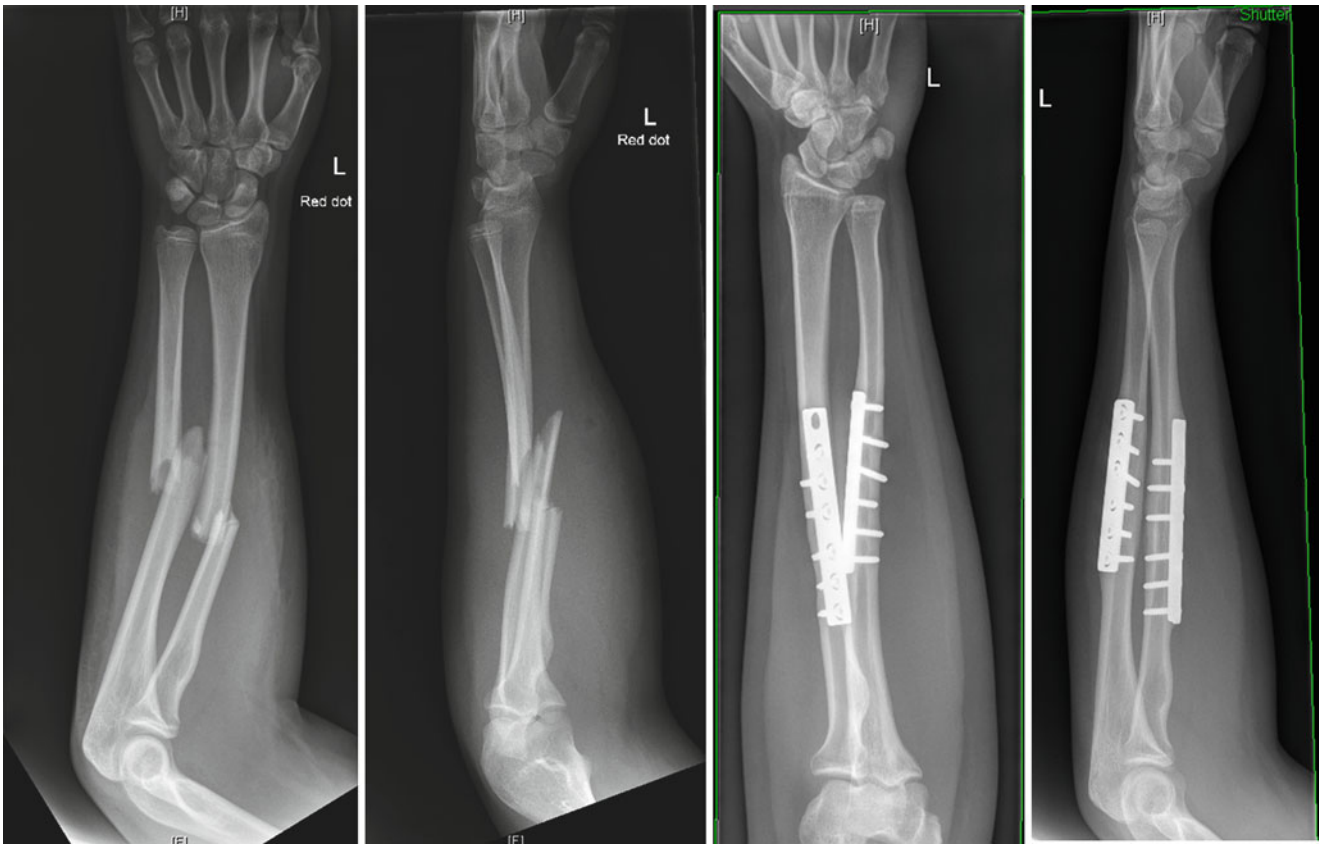

Fig. 36.1 Forearm bones fractures treated with open reduction and internal fixation

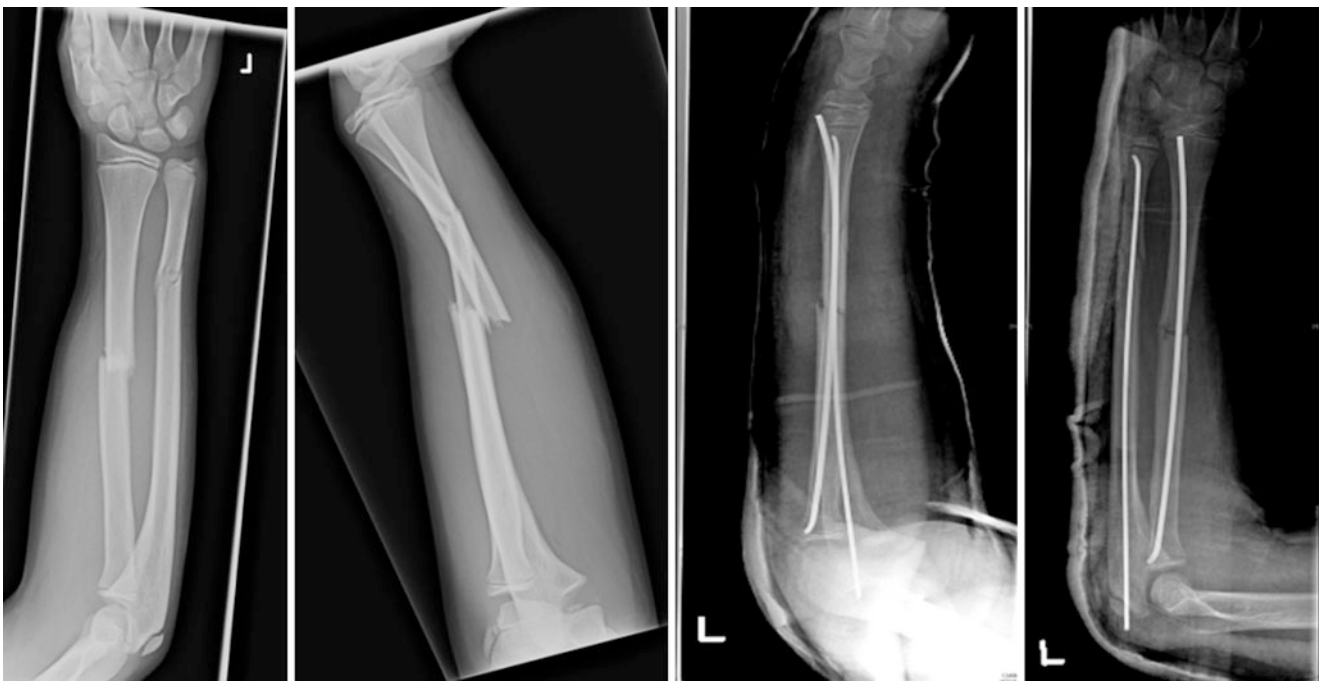

Fig. 36.2 Forearm bones fractures treated with elastic nailing

Open reduction and internal fixation (ORIF) involves surgical dissection and exposure of the fracture. The two ends of the fractured bone are perfectly matched like a jigsaw. This type of reduction is both macroscopic and microscopic allowing for the restoration of the microscopic, cellular and matrix bone anatomy, without the need for bridging callus. The reduced bone is then stabilised using an implant, typically a plate with screws.

The indications for surgical interventions, for diaphyseal fractures of the radius and ulna in children, are not clear. Similarly, the relative effects of various methods of surgery, particularly intramedullary nailing and plate fixation are not established.

There is paucity of Level I and II data that compares either surgical versus non-surgical interventions, or different surgical interventions for the fixation of fractures of the forearm in children. A review of the literature reveals a handful of Level III or IV retrospective studies looking at a comparison of IM nailing compared to ORIF. Some of these studies are of historic value, using implants (e.g. Rush Pin) which are no longer in vogue. There are two recent systematic reviews assessing the literature on operative intervention for forearm fractures [22, 23]. Baldwin et al. [22] conducted a systematic review and meta-analysis of 12 (525 patients); all were level III studies. Primary outcome was union rate. Patel et al. [23] conducted a systematic review of 8 studies; 446 participants (264 IMN and 182 plating). Primary outcome was the functional range of motion. The main summary of both demonstrates little difference in outcomes between ESIN and ORIF in paediatric patients. They also demonstrate that this genuine point of clinical equipoise now warrants randomised control trials to determine the better of the two methods.

Whilst there is no Level I and II evidence, there are a number of observational studies that must be noted [12, 13, 14, 24–28]. The studies most relevant to the management of forearm fractures vary in size from 31 to147 participants. The average age in each study also differed, as well as the average age between groups in each study. The average age of the ESIN groups (9.3–13.3) trended to be lower than the ORIF group (9.5–14.5) in all the studies (though not statistically significantly). None of these papers demonstrated any statistically significant differences in functional outcome between ORIF and ESIN. In addition differences with regards to complications between the two groups were not consistent between the studies. The systematic review with the largest number of studies found that non-union and delayed union was relatively uncommon in all series [22]. The incidence of non-unions requiring revision surgery in the meta-analysis was 1.7 % for ORIF vs. 2.4 % for ESIN. The incidence of delayed union in the IMN group was 5.5 % compared to 0.8 % in the ORIF group. However, this difference was not found to be statistically significant. This same review split complications into major and minor and found major complications to be 7.2 % in the ORIF group and 9 % in the IMN group with the complications being non-union, hardware failure, deep infection, compartment syndrome, unintended return to theatre in the immediate postoperative period, refracture, nerve injury, neuropathy, CRPS and arterial injury. All other complications (e.g. superficial infection, hypertrophic scar) were considered minor. As may be expected duration of surgery was significantly shorter in ESIN (or its equivalent) in the studies in which operating time was measured [27]. Cosmesis was significantly better in the ESIN group, though only measured in two studies [25, 27]. Compartment syndromes were reported in more than one study [14, 26] and in both groups. However, none of the papers indicated if the detected cases were of pre or postoperative compartment syndromes. Of note one paper that found compartment syndrome in their ESIN group did not have any cases of compartment syndrome in the ESIN group treated more than 24 h after the fracture was sustained [14].

A few authors published a small case series of using a single bone fixation [28–30]; however, their series probably included a recognised pattern of fractures where one of the bones is minimally displaced and relatively stable fracture (Fig. 36.3).

An additional relevant factor is removal of metal work. In the author's opinion, variation in the elective removal of implants is likely to reflect differences in the local healthcare economy. In the UK there is little appetite for the removal of all metal work routinely as this presents another burden of operative need on a publicly funded service. This may not be the case in nations where medical intervention has a financial incentive. Removal of metal work raises the concerns surrounding refracture. In the studies examined by the authors, practitioners tended to advocate routine removal of all nails in most studies. Three studies reported the effects of removal of metalwork [14, 26, 31]. Refractures were reported in both ESIN and ORIF groups. Operative decision making around removal of metal work needs to be better understood and studied.

A thorough search of the medical literature has failed to reveal any prospective trials comparing ORIF and ESIN for the treatment of radius and ulna diaphyseal fractures in children. As such we are unable to further advise orthopaedic surgeons on the relative merits of ORIF and ESIN. It is unlikely that alternative prospective, comparative studies on this topic have gone unnoticed in our search.

We would like to emphasise the lack of evidence available, to inform the available choices of surgical stabilisation for diaphyseal fractures of the radius and ulna in children. The two choices of ORIF and ESIN are both acceptable with neither having proven advantages of lower complication rates or better radiological and functional outcomes. It is likely that orthopaedic surgeons will make choices based on their training and expertise and given the current level of

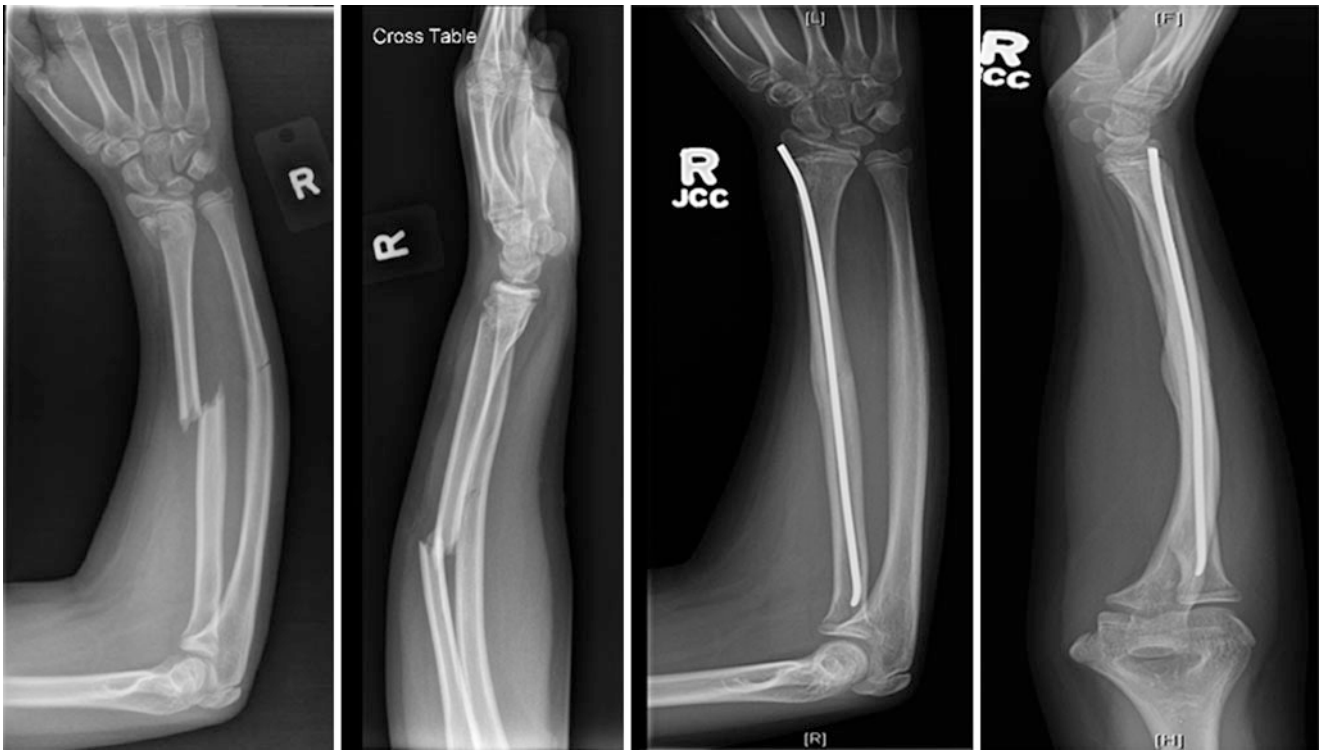

Fig. 36.3 Forearm bones fractures treated with a single elastic nail

available evidence we feel this is an acceptable surgical algorithm.

A lower refracture rate and easy removal of metal implants is a desirable outcome for many orthopaedic surgeons treating forearm fractures in children. These outcomes are difficult to comment on given the shortage of evidence, but in this regard, the attractiveness of ESIN over ORIF is shared by the authors. That ESIN splints the whole length of the long bone, without mechanical foci of stress concentrations and is thought to reduce the risk of refracture, however the literature does not indicate that this is necessarily the case. In addition, percutaneous insertion of ESIN in safe zones, distant to nerves and vessels, facilitates easier removal compared to ORIF implants.

Diaphyseal fractures in children are common and we accept the need for studies that help surgeons make the best choices for patients suffering these injuries. Ideally studies would be randomised, prospective trials with 3 year follow up to assess for healing, functional recovery, refracture rates and complications associated with removal.

References

1. Rennie L et al. The epidemiology of fractures in children. Injury. 2007;38(8):913–22.
2. Worlock P, Stower M. Fracture patterns in Nottingham children. J Pediatr Orthop. 1986;6(6):656–60.
3. Helenius I. Operative treatment of fractures in children is increasing <sbt aid = "1338974"> a population-based study from Finland</sbt>. J Bone Joint Surg (Am). 2009;91(11):2612.
4. Donaldson LJ et al. The epidemiology of fractures in England. J Epidemiol Community Health. 2008;62(2):174–80.
5. Hagino H et al. Increasing incidence of distal radius fractures in Japanese children and adolescents. J Orthop Sci. 2000;5(4):356–60.
6. Khosla S et al. Incidence of childhood distal forearm fractures over 30 years. JAMA. 2003;290(11):1479.
7. Pokka T, Serlo W, Sinikumpu J-J. The changing pattern of pediatric both-bone forearm shaft fractures among 86,000 children from 1997 to 2009. Eur J Pediatr Surg. 2009;23(04):289–96.
8. Alshryda S, Jones S, Banaszkiewicz P. Postgraduate paediatric orthopaedics: the candidate's guide to the FRCS (Tr and Orth) examination. Cambridge: Cambridge University Press; 2014.
9. Gray H. Anatomy of the human body. Philadelphia: Lea & Febiger, 1918. Online edition Bartleby.com. 2000.
10. Friberg KSI. Remodelling after distal forearm fractures in children II: the final orientation of the distal and proximal epiphyseal plates of the radius. Acta Orthop. 1979;50(6):731–9.
11. Blount WP. Forearm fractures in children. Clin Orthop Relat Res. 1967;&NA(51):93–108.
12. Smith VA et al. Treatment of pediatric both-bone forearm fractures. J Pediatr Orthop. 2005;25(3):309–13.
13. Friberg KSI. Remodelling after distal forearm fractures in children III: correction of residual angulation in fractures of the radius. Acta Orthop. 1979;50(6):741–9.
14. Flynn JM et al. Eleven years experience in the operative management of pediatric forearm fractures. J Pediatr Orthop. 2010;30(4):313–9.
15. Hamilton TW et al. The treatment of stable paediatric forearm fractures using a cast that may be removed at home: comparison with traditional management in a randomised controlled trial. Bone Joint J. 2013;95-B(12):1714–20.

16. Bhatia M, Housden PH. Redisplacement of paediatric forearm fractures: role of plaster moulding and padding. Injury. 2006;37(3):259–68.
17. Yang J-J et al. Redisplacement of diaphyseal fractures of the forearm after closed reduction in children: a retrospective analysis of risk factors. J Orthop Trauma. 2012;26(2):110–6.
18. Iltar S et al. The value of the three-point index in predicting redisplacement of diaphyseal fractures of the forearm in children. The Bone & Joint Journal. 2013;95-B(4):563–7.
19. Colaris JW et al. Risk factors for the displacement of fractures of both bones of the forearm in children. Bone Joint J. 2013;95-B(5):689–93.
20. Van der Reis WL et al. Intramedullary nailing versus plate fixation for unstable forearm fractures in children. J Pediatr Orthop. 1998;18(1):9–13.
21. Shoemaker SD et al. Intramedullary Kirschner wire fixation of open or unstable forearm fractures in children. J Pediatr Orthop. 1999;19(3):329–37.
22. Baldwin K et al. Both bone forearm fractures in children and adolescents, which fixation strategy is superior – plates or nails? A systematic review and meta-analysis of observational studies. J Orthop Trauma. 2014;28(1):e8–e14.
23. Patel A, Li L, Anand A. Systematic review: functional outcomes and complications of intramedullary nailing versus plate fixation for both-bone diaphyseal forearm fractures in children. Injury. 2014;45(8):1135–43.
24. Shah AS et al. Stabilization of adolescent both-bone forearm fractures: a comparison of intramedullary nailing versus open reduction and internal fixation. J Orthop Trauma. 2010;24(7):440–7.
25. Teoh KH et al. An age – and sex-matched comparative study on both-bone diaphyseal paediatric forearm fracture. J Child Orthop. 2009;3(5):367–73.
26. Reinhardt KR et al. Comparison of intramedullary nailing to plating for both-bone forearm fractures in older children. J Pediatr Orthop. 2008;28(4):403–9.
27. Fernandez FF et al. Unstable diaphyseal fractures of both bones of the forearm in children: plate fixation versus intramedullary nailing. Injury. 2005;36(10):1210–6.
28. Houshian S, Bajaj SK. Forearm fractures in children. Single bone fixation with elastic stable intramedullary nailing in 20 cases. Injury. 2005;36(12):1421–6.
29. Alnaib M et al. Radius-only intramedullary nailing for both-bones diaphyseal forearm fractures in children. Acta Orthop Belg. 2011;77(4):458–63.
30. Bader M, Sanz L, Waseem M. Forearm fractures in children: single bone fixation with elastic stable intramedullary nailing in 20 cases. Injury. 2006;37(9):923–4 .author reply 924–5
31. Kose O et al. Open intramedullary Kirschner wire versus screw and plate fixation for unstable forearm fractures in children. J Orthop Surg (Hong Kong). 2008;16(2):165–9.

Evidence-Based Treatment of Wrist Fractures in Children

37

Assad A. Qureshi and Alwyn Abraham

Abstract

Distal radial fractures are a common injury pattern in the paediatric population. The management of displaced metaphyseal fractures introduces several controversies in management. Questions arise regarding the optimal method of stabilising these injuries following successful reduction under anaesthesia. There is a paucity of clinical evidence investigating comparative treatment effects of different interventions with only six trials being included in a Cochrane review on this topic. Trials investigating above elbow versus below elbow casts have found no difference in the re-displacement rate but demonstrated less functional restriction, earlier return to activities and reduced elbow stiffness in below elbow casts. Percutaneous wiring is associated with a lower re-displacement rate compared with cast alone treatment but does not yield a difference in functional outcome at 3 months when comparing the two treatments. Further work is required to elucidate the influence of factors such as specific fracture pattern, ulna involvement and age of the patient on long term clinical and functional outcomes following treatment to help identify which patients might benefit from surgical intervention.

Keywords

Paediatric • Distal radius fracture • Above elbow cast • Below elbow cast • Percutaneous wiring

Background

Distal radius and ulna fractures account for a large proportion of paediatric injuries with a fall on the outstretched hand being the most common mechanism [1]. The annual incidence of these injuries in the UK is estimated to be 16 per 1000 children. Six morphologic subtypes have been described: (1) buckle, (2) green stick, (3) metaphyseal, (4) distal radial and (5) ulna physeal injuries in addition to (6) galeazzi fracture patterns [2].

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Buckle fractures of the distal radius and ulna constitute incomplete fractures. Their hallmark characteristic is compression of the outer circumference of the cortex with preservation of length and alignment. Treatment is aimed at splintage for pain relief with discontinuation based on symptomatic improvement. Green stick fractures are similar to buckle injuries with partial cortical disruption, but different in that plastic deformation occurs in the un-fractured cortex at this level. The extent of plastic deformation will determine the magnitude of the resulting angular deformity in the fractured bone and the need for reduction prior to immobilisation.

When the magnitude of the deforming force exceeds the capacity for plastic deformation in the intact cortex, bicortical disruption occurs resulting in a metaphyseal

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fracture. These are usually displaced and require reduction and immobilisation. Fractures occurring through the distal radial physis are more common in older children and if displaced, require reduction and immobilisation. Distal ulna physeal injuries are quite rare as are Galeazzi fractures where there is a fracture of the distal radius with concurrent disruption of the distal radio-ulna joint. The latter fracture type assumes clinical importance as reduction of the distal radial fracture is mandatory to restore the congruity of the distal radio-ulna joint which is crucial to preserve normal forearm rotation.

An important consideration in the management of distal radial injuries is the limits of acceptance with respect to the extent of fracture displacement and the need for reduction. If displaced, one must question whether the resulting deformity would be acceptable to both the patient and parents, whether it would impede function and whether it could be reasonably expected to remodel with growth. Once these limits have been defined, there is a need to distinguish stable injury patterns which might require cast immobilisation alone from relatively unstable injuries requiring surgical fixation. When cast immobilisation is preferred, we need to ask how long to continue immobilisation, the position of the arm and whether the elbow and wrist need to be immobilised or can be left free. This review will focus on these areas of uncertainty and the evidence basis supporting clinical decision making.

A Cochrane review has been published by our group detailing the current evidence base for treating distal radius fractures in the paediatric population [3]. The main aims of the review were to determine the influence of modifiable treatment factors in these injuries on outcome with a specific emphasis on function, re-displacement, residual deformity, other complications and patient/parent satisfaction. The modifiable treatment parameters investigated were the type of cast; with respect to forearm position (pronation/supination), inclusion of the wrist and elbow and the use of percutaneous wires compared with cast immobilisation. The effects of patient variables such as type of fracture and age of the child were also sought. The review principally looked at level 1 randomised or quasi randomised study designs comparing different treatment arms. The methodological quality of the studies was assessed using a modified version of the Cochrane Bone, Joint and Muscle Trauma Group's quality assessment tool [3].

Trial Characteristics

Six trials were included with a total of 478 participants, most of whom were male with age ranging from 8 to 12 years. All six trials specified fractures of the distal radial metaphysis as the event of interest. Three studies [4–6] further defined specific inclusion criteria for their population of displaced fractures while three did not [7–9]. One study excluded patients with concurrent ulna fractures [4]. Three studies included patients with distal ulna fractures [5, 7, 8]. These six studies were grouped according to the comparison between different treatment modalities following reduction of a displaced fracture (see Table 37.1).

Assessment of Methodological Quality of Included Trials

Participant allocation was concealed in one trial [8] but not in the two studies utilising quasi randomised methods [4, 9]. In the Miller study [6], nine additional non-randomised patients were included. Blinding of treatment providers and participants was not feasible in the included trial designs. In all of the included studies, assessment of outcomes was unblinded despite attempts in one study to blind radiographic assessment [8]. The reporting of the baseline characteristics of the patient population was poor with only two studies [4, 5] fully satisfying this criteria. The Boyer trial [7] reported insufficient data to judge study quality in this regard. In two studies [7, 8], provision of comparable care programs was deemed highly likely. Poor reporting of the comparability of care programs in both study arms other than the trial intervention yielded poor quality scores in the remaining studies. All of the included trials provided sufficient information on the inclusion and exclusion criteria defining the study population as well as the interventions being compared. Outcome instruments were adequately described in all included trials. Three trials did not assess function as an outcome measure

Table 37.1 Study treatment comparisons, authors and participant numbers

Comparison	Studies	No of participants
Below elbow cast vs Above elbow cast	Bohm et al. [8]	229
	Webb et al. [9]	
Neutral vs Pronated vs Suppinated forearm position in above elbow cast	Boyer et al. [7]	109
Percutaneous wire fixation + above elbow cast vs above elbow cast alone	Gibbons et al. [4]	125
	McLauchlan et al. [5]	
	Miller et al. [6]	

[4, 7, 8]. Set time points for follow up were defined in all but one study [7] but no study reported outcomes at or beyond 1 year.

Comparison of Treatment Effect in Reported Trials

Below Elbow Versus Above Elbow Cast Immobilisation

Two studies reported outcomes in a total of 229 participants [8, 9]. The differing criteria may have contributed to the moderate statistical heterogeneity (I^2 = 48.1 %). However, both trials found a trend towards a reduced risk of re-displacement during immobilisation in a below-elbow cast. The analysis is distorted by the absolute numbers of re-manipulations required in each study, four in the Bohm trial [8] and nil in the Webb trial [9], suggesting that the higher re-displacement rate in the above elbow group does not impact on the need for re-manipulation. The below elbow cast group demonstrated significantly fewer limitations and reduced need for help in activities of daily living during cast treatment while the above elbow cast group missed, on average, one extra day of school.

Following cast removal, the above elbow cast group in the Webb study [9] demonstrated significantly reduced elbow range of motion compared with the below elbow group (mean difference 28.7°). Children in the below elbow group regained elbow range of motion 10 days earlier compared with the above elbow group. Comparison of the final elbow range of motion between the two groups did reveal a statistically significant difference. However, the clinical relevance of a three to four degree difference between treatment arms in the context of inter- and intra- observer error in measurements, is of questionable significance.

Most complications were cast related with similar numbers in the two groups having their cast changed or reinforced due to cast weakening or loosening [8]. Five participants requested change from an above elbow to a below elbow cast to reduce discomfort.

Although suggestive of differences in outcome between the two treatments, both studies findings are hampered by methodological limitations. The absence of functional outcomes in the Bohm trial [8] does not yield insight into the function of re-displaced fractures which did not require re-manipulation. This study also demonstrated a significant imbalance in fracture types. The Webb study [9] reported insufficient information on the study population's baseline characteristics in addition to employing quasi randomised methods. Both studies reported on cast fit [10] which provided some assurance of correct application although failed to describe the position of the forearm in the above elbow cast.

Above Elbow Cast: Forearm Pronated Versus Neutral Versus Supinated

Comparisons in forearm position in above elbow cast were investigated in one study [7] with 109 participants treated for displaced or angulated fractures following reduction under general anaesthesia. Ten children were excluded due to insufficient radiographic assessment. One child in each of the supination and pronation groups required repeat manipulation due to an unacceptable loss of alignment but demonstrated satisfactory outcomes at final follow up. At final follow up, at or beyond 6 weeks, there was no significant difference when comparing the different groups with respect to residual angular deformity (mean 7°). This study demonstrated significant methodological flaws with a quasi-randomised design, varied follow up and an absence of reported population baseline characteristics. These limitations permit very few conclusions regarding which, if any, forearm position is preferable.

Percutaneous Wire Fixation and Above Elbow Cast Versus Above Elbow Cast Alone

Three trials compared these treatments following reduction of a displaced fracture in a total of 125 children [4–6]. Cumulative analysis revealed all re-displacements, with the exception of one case, occurred in the cast alone group. The single instance case of re-displacement in the wires group [5] occurred secondary to wire migration. In the Gibbons study [4] all of the re-displacements were re-manipulated. Of the seven re-displacements in the McLauchlan study [5], four underwent surgical re-manipulation. Only one of the six children with a re-displacement in the Miller study [6] underwent a secondary procedure to improve the position. There was no significant difference in the rate of other complications between the wire fixation and cast only group. There were more planned secondary procedures in the wire fixation group reflecting the routine removal of wires following fracture healing. There were a significantly higher number of unscheduled secondary procedures in the cast only group. However, these results were heterogenous (I^2 = 49.5 %) and reported significance was only marginal when using a random effects model [3]. The Miller study supported a reduction in the cost of wire fixation compared with cast alone (US $3347 versus $3831) when treating these fractures due to the costs of unplanned secondary procedures.

Clinical outcome in the wire fixation and cast alone groups was comparable in all three studies. McLauchlan [5] reported no significant difference in grip strength or range of motion following treatment when comparing the two groups. Five children out of 56 reviewed at 3 months complained of minor pain after strenuous activity although none had any functional deficit. Radiographic assessment at 3 months demonstrated greater residual coronal (mean difference – 3°) and sagittal deformity (mean difference – 5.1°) in the cast alone group compared with the wires group. Miller reported no limitations in function, strength or range of motion at a mean follow up of 2.8 years [6]. There was no residual cosmetic deformity in the study group with all 25 fractures remodelling to anatomical alignment.

All three trials consistently reported a lower re-displacement rate with wire fixation and unplanned secondary procedures compared with cast immobilisation alone. This effect was most marked in the Gibbons study [4], where the distal ulna was intact in all cases. The reporting of routine wire removal being a scheduled secondary procedure distinct from unscheduled secondary procedures in the cast alone group lends itself to bias. Amongst other biases, major selection bias could not be ruled out for two of the three included studies [4, 6]. The lack of long term outcomes limits the findings of the McLauchlan study [5] although no child demonstrated a functional deficit at 3 months in this study.

Discussion

Despite the large number of children sustaining such injuries, there is a paucity of level 1 evidence comparing outcomes from different treatment modalities. The most recent Cochrane review [3] included 6 trials from either North America or the UK involving 478 children. Cumulative review of these 6 studies does not appear to favour any particular treatment over another. Although the risk of re-displacement may differ between treatments, there does not appear to be any significant difference in other complications or functional outcome at final follow up. The suggested relationship between re-displacement and the need for re-manipulation is imprecise and requires further evaluation. The available evidence suggests a low incidence of complications or poor outcomes following treatment stressing the importance of appropriately powered studies designed to detect differences between treatment arms. Treatment effect may also be masked in trials where different types of displaced radial fracture have been included. The propensity for certain fracture types to re-displace has not been assessed by any of the current randomised trials. Although one study investigating the influence of forearm position in above elbow cast demonstrated no difference, any potential treatment effect may exist as a function of optimal forearm position being dictated by the injury characteristics. The influence of an intact ulna on the outcomes of specific treatments remains unanswered. Future level 1 studies must be directed at identifying the important injury morphological characteristics conferring improved outcomes of one particular treatment over another.

The inherent remodelling potential of these injuries does question whether short term outcomes before remodelling is complete assume equal importance to outcomes assessed at final follow up when any residual deformity will have remodelled to anatomical alignment. The age of the child is an important determinant of remodelling potential and any prospective study design should consider appropriate inclusion criteria to objectify the influence of re-displacement on the need for re-manipulation. The precise relationship between radiographic deformity and clinical deformity is uncertain and highlights the need for patient centred outcome measures to assess the impact of perceived limb alignment following treatment on quality of life. The included studies demonstrate the historic bias towards surgeon measured outcomes where assessment is focussed on range of motion, grip strength and radiographic alignment. Notably of the six included studies, three studies [4, 7, 8] did not assess functional outcomes. Appropriate patient centred outcomes are an integral recommendation for any future trials investigating outcomes from these injuries.

Current Conclusions

The current evidence suggests that for displaced distal radius fractures which have been reduced, below elbow casts are not associated with a higher re-displacement rate compared with above elbow casts. Improved functional outcomes have been reported with below elbow casts. These arguments render any question of the optimal forearm position in an above elbow cast superfluous. The limited evidence investigating forearm position as a variable dictating outcome does not rebut this opinion. Displaced fractures stabilised with percutaneous wires are more likely to maintain their position compared with cast treatment alone despite no difference in reported functional outcomes at 3 months.

References

1. Worlock P, Stower M. Fracture patterns in Nottingham children. J Pediatr Orthop. 1986;6(6):656–60.
2. Herring JA. Tachdjian's pediatric orthopaedics. Vol 3, 3rd ed. London: Saunders; 2002.
3. Abraham A, Handoll HH, Khan T. Interventions for treating wrist fractures in children. Cochrane Database Syst Rev. 2008;16(2): CD004576. doi: 10.1002/14651858.CD004576.pub2.

4. Gibbons CL, Woods DA, Pailthorpe C, Carr AJ, Worlock P. The management of isolated distal radius fractures in children. J Pediatr Orthop. 1994;14(2):207–10.
5. McLauchlan GJ, Cowan B, Annan IH, Robb JE. Management of completely displaced metaphyseal fractures of the distal radius in children. A prospective, randomised controlled trial. J Bone Joint Surg Br. 2002;84B(3):413–7.
6. Miller BS, Taylor B, Widmann RF, Bae DS, Snyder BD, Waters PM. Cast immobilization versus percutaneous pin fixation of displaced distal radius fractures in children. A prospective, randomized study. J Pediatr Orthop. 2005;25(4):490–4.
7. Boyer BA, Overton B, Schrader W, Riley P, Fleissner P. Position of immobilization for pediatric forearm fractures. J Pediatr Orthop. 2002;22(2):185–7.
8. Bohm ER, Bubbar V, Yong Hing K, Dzus A. Above and below-the-elbow plaster casts for distal forearm fractures in children – A randomized controlled trial. J Bone Joint Surg Am. 2006;88A(1):1–8.
9. Webb GR, Galpin RD, Armstrong DG. Comparison of short and long arm plaster casts for displaced fractures in the distal third of the forearm in children. J Bone Joint Surg Am. 2006;88A(1):9–17.
10. Chess DG, Hyndman JC, Leahey JL, Brown DC, Sinclair AM. Short arm plaster cast for distal pediatric forearm fractures. J Pediatr Orthop. 1994;14(2):211–3.

Part IX

Neuromuscular Diseases

The Value of Gait Analysis in Decision Making About Surgical Treatment of Cerebral Palsy

38

Clare Carpenter and Alfie Bass

Abstract

Gait analysis provides detailed objective quantitative measurements with regards to locomotion. The process of gait analysis involves the evaluation of a combination of data. This includes the visual observation of gait, a standardized clinical examination and instrumented analysis. Biomechanical data including joint movements and forces can be quantified and define the segmental movements of the limbs. This allows an accurate quantification of the deviations away from normal gait parameters and using that data to understand deformity and pathology, plan surgical strategy and objectively measure outcomes subsequent to intervention. Whether gait analysis leads to significant improvement in structural abnormalities, gait, function, quality of life, and cost-effectiveness is still yet to be determined.

Keywords

Cerebral Palsy • Gait • Mobility • Ambulation • Instrumented • Video • Surgery • Outcome • Walking • Analysis • Measures

Introduction

Gait analysis (GA) has been central to the understanding of disordered gait in children with neuromuscular disease. It has enhanced our understanding of the natural history of these gait patterns and provided objective analysis of both non-surgical and operative intervention. There has been the development of a language to allow better communication amongst health care professionals involved with these children. Criticisms of the technique are based on technical aspects of the procedure wherein biological systems are modeled on certain mathematical assumptions, which may introduce inaccuracies. Human bias in the interpretation and translation of the data throws into question the reproducibility of the data between different laboratories and within the same laboratory on different occasions.

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Historically, the mainstay of management of children with cerebral palsy (CP) was to maintain and improve ambulation. More recently the focus of intervention has been around functionality of the child whether the child may be ambulant or not. This has been driven by the International Classification of Functioning, Disability and Health (ICF) framework to include structure, function and also activity and participation. This has shifted our focus somewhat and widened the spectrum of musculoskeletal intervention for these children. More recently GA has expanded towards evaluation of the upper limb. It must be remembered that GA provides only information with regards the structure and function domain of the ICF framework for disability.

Defining Gait

Gait involves repetitious and cyclical movements of the foot from one position of support to the next, allowing the body to travel. There is a sequence of muscle actions that relates to three major tasks being accomplished during each gait cycle: weight acceptance, single-limb support, and limb

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advancement [1]. Because the moment of floor contact is the most readily defined event in the sequence of movements, this action is conventionally chosen to mark the beginning of the gait cycle. A complete gait cycle, or stride, begins when one foot strikes the ground and ends when the same foot strikes the ground again.

Understanding normal walking patterns underpins the appreciation of pathological gait. Jacquelin Perry was a pioneer in this area and defined eight functional phases of the normal gait cycle, divided into two major phases: stance (60 %) and swing (40 %). Stance is the period of time when the foot is in contact with the ground and supports the weight of the body. It begins with *initial contact*, which in normal gait is with the heel, and ends at *toe-off*, when swing phase begins. Swing is defined as the period of time when the foot is off the ground and moves forward [1]. This sequence of events is reliant on the integrity of the musculoskeletal system and neurological control; the bones act as rigid levers arms converting forces into movement.

Prerequisites to Gait in Cerebral Palsy

The primary neurological impairment caused by the brain injury in CP affects muscle tone, selective motor control, strength, co-ordination and balance. There is a gradual development of muscle contracture and bony deformity with growth of the child, which results in 'lever-arm dysfunction' and abnormal gait. Gage noted that the five main prerequisites to normal gait [1] were abnormal in children with CP, contributing to their functional disabilities [2]:

1. Stability in stance
2. Foot prepositioning
3. Adequate stride length
4. Clearance in swing
5. Energy consumption

Stability in the stance phase is necessary to support the body and allow advancement of the limb in swing, maintain balance, aid propulsion and allow limb prepositioning. Deformities, such as a planovalgus foot, with an externally rotated tibia can lead to instability in stance. Foot preposition allows the hindfoot to be prepared for initial contact in order to achieve a heel strike; clearance in swing is required to avoid foot drag. An adequate step length is required for efficient movement, limiting energy expenditure. Depending on severity, the energy demands of a child with CP can be 1.5 to 3 times higher due to an increased cadence and spasticity-related muscular co-contraction of (both agonist and antagonistic) muscles around the knee and ankle joints. The energy produced by co-contracting muscle forces are wasted and lead to subsequent abnormal gait deviations.

Gait Patterns in Cerebral Palsy

There are characteristic abnormal gait patterns observed in ambulatory children with spastic hemiplegia and diplegia [3, 4]. Winters et al. described four basic patterns of the hemiplegic gait [3]. There is a predominance of equinus during the stance phase due to the gastrocnemius spasticity, as a powerful plantar-flexor very early on in the young child's development, with relative weakness of tibialis anterior. The classification is a continuum and there may be an overlap between types. As the child grows, knee and hip flexion become more prominent. (Table 38.1)**.**

Historically, many authors have classified sagittal gait patterns in spastic diplegia [4–6]. Using gait analysis, Rodda et al. further qualified the transverse and coronal plane abnormalities [5], which subsequently demonstrated excellent intra-rater reliability [6]. As with hemiplegic patterns, these are not static and may evolve over time with growth of the child and changing spasticity within the dominant muscle groups. The first two patterns are seen in younger children. As the natural history proceeds there is decreasing equinus with increasing proximal contracture (Table 38.2).

Coronal plane abnormalities are spasticity or contracture of the hip adductors, limb length discrepancy and hip subluxation. Transverse plane abnormalities include pelvic rotation, femoral anteversion, external tibial torsion and foot deformities.

These classifications may be an over-simplification of the many gait deviations that occur in the ambulatory child and a recent international consensus has used IGA data from all three planes to expand existing classifications into twelve recognized patterns [7]. What is unknown currently is whether all these patterns are pathological or indeed amenable to surgical intervention.

Why Is Gait Analysis Required in the Surgical Management of Cerebral Palsy?

It is appreciated that the human eye cannot capture all the details of a real-time gait assessment. Over the years many modalities have been used to try and capture gait and accurately assess some of the finer details that may contribute to pathological gait patterns.

Observational gait analysis utilizes a low cost system of video gait analysis, which can then be viewed and usually scored against validated outcome tools, including: the Observational Gait scale, Salford Gait tool, Observational gait analysis, Edinburgh visual gait score and Physician rating scale. These scales do not contain objective kinematic and kinetic data and have limited objectivity when applied to surgical decision-making and outcomes post intervention [8].

Table 38.1 Types of hemiplegic cerebral palsy

Type	Deformity	Management considerations	Orthosis
I	Functional equinus in swing, normal dorsiflexion in stance		Hinged AFO
II	Ankle equinus in swing and stance	Gastrosoleus release	Hinged AFO
III	Type II including decreased knee movement in swing and stance	Gastrosoleus release +/– Medial hamstring release and rectus femoris transfer	Hinged AFO
IV	Type III and limited hip movement in swing and stance In association with an anteverted femur and an increased adduction moment, these children may also exhibit late onset hip subluxation	As for Type III +/– Soft tissue hip releases and a VDRO	Solid AFO

Data from Roddy and Graham [5]

Table 38.2 Types of spastic diplegia

Type	Deformity	Management considerations	Orthosis
True equinus	This is due to calf spasticity in the young child. May be associated with knee recurvatum	BtxA – Gastrosoleus Surgery – gastrosoleus lengthening	Hinged AFO
Jump gait (+/– Stiff knee)	Fixed equinus in addition to a flexed knee and hip	BtxA – Gastrosoleus and hamstrings Surgery – Gastrosoleus and hamstring release +/– RF transfer	Hinged AFO
Apparent equinus	This is frequently mistaken as true equinus; the increased hip and knee flexion may give the impression that the equinus is real	BtxA – Hamstring and Psoas Surgery – Multilevel contracture release	Solid or Ground Reaction AFO
Crouch	Excessive dorsiflexion in stance with increased knee and hip flexion	Surgery – Multilevel contracture release Foot & Bony torsional abnormalities	Solid or Ground Reaction AFO

Data from Roddy and Graham [5]
A fifth group was defined as asymmetrical where each lower limb may demonstrate one of the basic patterns above

Instrumented gait analysis (IGA) is the gold standard assessment and involves the placement of markers, or arrays, on predefined anatomical areas of the child. The multidirectional video cameras detect the position of the arrays and the data are input to a computer-based mathematical model of limb movement. The simplest view of a model of the human body assumes that the lower limbs move as seven rigid segments: the pelvis, two thighs, two shanks (or legs) and two feet. The output data are presented as a series of three-dimensional kinematic (joint movements) and kinetic (forces) graphs in the coronal, sagittal and transverse planes. Additional measurements may include electromyography (muscle activation), oxygen consumption and pedobarography (plantar foot pressures).

Gait Scoring Systems

The gait deviation index (GDI) and the gait profile score (GPS) are indices generated by combining all the raw kinematic and kinetic data obtained from IGA. The advantage of this summary statistic is to provide a rapid simple value that can be used in clinical practice to indicate the normality of gait. Both are used within the literature, with good intra-rater reliability [9]. The GPS is stated to be the user-friendliest score and uses a mathematical equation to summarise all the raw data to a single number. The larger the number the greater the gait deviates from a normal control population [10, 11]; the minimal clinically important difference for the GPS is 1.6° [12]. These scoring systems allow complex data to be simplified and for groups of patients to be compared before and after complex interventions.

Multilevel Surgery

It is appreciated that IGA has contributed to our understanding of pathological gait yet many institutions continue to operate on children with CP without access to this facility. Studies have tried to quantify the usefulness of IGA and when available it is adhered to 75 % of the time [13, 14] and altered management in 40–90 % of cases [15]. It was the

fine-tuning of soft tissue releases that were mainly altered, with the large bony corrections having good initial agreement [16, 17]. A randomized control trial in 2011 confirmed the efficacy of gait analysis on altering surgical decision-making but did not report in whether this translated into more beneficial patient outcomes [18]. The literature fails to determine whether the use of IGA is cost-effective, although Wren et al. have reported that pre-operative gait analysis decreased the reoperation rate in ambulatory children with cerebral palsy from 40 % to 20 % by 5 years after the index procedure, without any increase in overall cost [19, 20].

With the use of IGA, surgeons have increased ability to accurately assess their patients' multilevel needs. As a result, single event multilevel surgery (SEMLS) has largely replaced repeated single level surgery in children with CP. This involves bilateral simultaneous correction of all bony and soft tissue deformities that have been identified preoperatively. Numerous studies using pre- and post-operative gait analysis confirm successful outcomes 5 years following SEMLS at 5 years [21]. There are many components to the multilevel deformity correction and we will consider these in turn.

The Internally Rotated Lower Limb

An internally rotated gait pattern with a negative foot progression angle can lead to lever arm dysfunction and problems with knee (foot) clearance. This can be a multilevel problem with an internally rotated femur, internal tibial torsion and pes varus; IGA has identified that multiple causes usually co-exist and need to be addressed [22].

An 'internally rotated femur' may reflect both (i) static excessive femoral anteversion, (which persists as a failure of developmental remodeling mechanisms, due to the lack of a maintained upright posture and hip extension), and (ii) the dynamic contribution from muscular imbalance, crouch gait, and spasticity of the hip internal rotators. Femoral derotation osteotomy alone has shown to improve hip rotation and foot progression long-term [23, 24] and more 'normal' gait parameters can be achieved when a femoral derotation osteotomy is included within the surgical prescription [25]. There is no difference in outcomes, when this is done either proximally or distally [26] – albeit that the site has largely been surgeons' preference. If there is associated hip subluxation, proximal femoral osteotomy is indicated to address the additional deformities.

The accuracy with regards to the amount of derotation required to normalize gait can be addressed objectively with IGA and is associated with superior postoperative outcomes in comparison to clinical examination alone [27]. The difficulty lies with the surgeon intraoperatively, accurately quantifying the correction. Two studies based on pre- and postoperative IGA data demonstrated that only one- half to two-thirds of the amount of correction was obtained intraoperatively, thereby for a derotation of 20 ° of desired correction on post-operative gait analysis, the surgeon should plan to rotate the femur 30–40 ° at surgery [26, 28].

Knee Flexion Deformity

Clinical examination and reliance on the popliteal angle is misleading to quantify functional hamstring length. Musculoskeletal modeling of the hamstrings suggests that the hamstring length may be normal or long [29] – thereby a release or lengthening can potentially be detrimental [30]. Surgical options for the hamstrings are numerous. The concept of 'dose of surgery' aims to titrate the amount of surgery on the severity of the flexion deformity [31]. A medial hamstring lengthening is suggested for children with a fixed flexion deformity (FFD) <15 °. Between 15 ° and 25 ° a combined medial hamstring lengthening and semitendinosus transfer to the adductor tubercle is preferred. Postoperatively this is associated with improved sagittal plane knee kinematics reduced knee flexion at initial contact, knee extension approximated to normal during loading and reduced peak knee flexion in swing, with increased range of knee movement. The semitendinosus transfer avoids over lengthening of the hamstrings with less effect on anterior pelvic tilt [32].

Knee deformities greater than 25 ° require associated bony surgery, which may include either anterior distal femoral guided-growth or distal femoral extension osteotomy. There are few studies relating to outcomes of anterior stapling (or 8-pating) in children with CP, although there are small case series demonstrating their effectiveness for improving knee flexion deformity in growing children [33]. Distal femoral osteotomy can improve knee range of movement and gait indices. However, when performed for crouch gait, postoperative walking ability is improved when combined with a patellar advancement procedure to address the quadriceps insufficiency. A combined procedure is also associated with less recurrence of the knee deformity [34].

Stiff-Knee Gait

A stiff-knee gait can lead to problems of foot clearance, falls and stair climbing for children with CP [35]. Reliance on clinical observation and the Duncan-Ely test for preoperative assessment can be misleading [36]. IGA demonstrates a decreased magnitude and delayed timing of peak knee flexion in conjunction with EMG evidence of over activity of the Rectus Femoris (RF) during swing phase.

Transferring the distal stump of the RF to the medial hamstrings [37] is shown to reduce out of phase extensor RF activity and increase peak knee flexion during the swing phase,

improving foot clearance [38, 39]. Distal RF transfer is beneficial in GMFCS I-III [40] and is frequently included as a component of single event multi-surgery (SEMLS). Consistently, there are demonstrable improvements in knee kinematics, yet translating these findings into positive clinical outcomes is variable [41, 42]. It is debatable whether the transferred RF actually improves knee flexion or rather simply defunctions the dysphasic extensor activity [43]. Comparison of distal RF resection versus transposition, have revealed equivalent results; it is thought that the transposed tendon scars to the fascial bed and fails to act as a knee flexor [43]. A prospective randomized control trial comparing SEMLS with and without distal RF transfer demonstrated that significant preoperative stance phase knee flexion was associated with poorer outcomes [43]. There are a small number of studies demonstrating favourable short-term outcomes with comparable kinematic improvements with distal RF resection [44].

Release of the proximal part of RF was attractive as this also plays a part in hip flexion and anterior pelvic tilt. However, kinematic data have never shown this to be effective alone [45] or in combination with a distal release [46].

Ankle Equinus

Ankle equinus has been reported in 61 % of children with CP who present to a gait lab [47]. Clinical observation alone tends to overestimate the amount of equinus [48]. IGA is more accurate in quantifying the magnitude of the functional equinus and where the deformity is unresponsive to non-surgical management, can aid surgical planning.

Over-lengthening of the gastrosoleus complex can lead to deterioration in gait, resulting in crouch. Borton et al. [49] claimed that the highest rates of calcaneal gait are seen in diplegic children who had undergone percutaneous Achilles lengthening and recommended that this technique should be avoided in children with bilateral CP.

There are a large number of lengthening techniques described within the literature for the surgeon to choose from [50]. Understanding the functional anatomy can aide surgical decision-making and cadaveric studies have defined three main zones; Zone 1 from the origin of the gastrocnemius muscle to the most distal fibres of the medial belly, Zone 2 from the distal extent of the medial gastrocnemius muscle belly to the most distal fibres of the soleus. Zone 3 is the Achilles tendon. Increased gain in length is achieved, the more distal the lengthening procedure [51].

Zone 1 and 2 gastrosoleus procedures have been popularised for equinus in diplegic gait since the advent of IGA [52]. Firth et al. [53] reported no cases of crouch gait, with only a 2.5 % rate of calcaneus at more than 7 years post-operatively in children with gastrocnemius recession as part of SEMLS. There was a recurrent equinus in 35 %, though mild recurrent equinus was well tolerated, and relatively easily treated by additional procedures (conversely, calcaneus may lead to crouch gait and can be very difficult to salvage).

Zone 3 lengthening should be reserved for severe equinus deformities in hemiplegics as they produce unacceptable rates of crouch in bilateral CP [49, 50]. A tight gastrosoleus complex results in the calcaneum driven into either varus or valgus. Any of the associated combinations exist although equinovarus is more commonly associated with unilateral CP and equinovalgus with bilateral involvement.

Equinovarus results in an internal foot progression angle and classically was thought to be associated with tibialis posterior spasticity. In fact, IGA and EMG studies have shown abnormal activity in tibialis anterior in one-third of patients, the tibialis posterior in one-third, and both muscles in the remaining third [54].

Botulinum Toxin (BtxA)

BtxA has moderately good reported outcomes in the management of lower limb spasticity and a positive effect on gait pattern [55]. When assessing spasticity and outcomes form BtxA there is a discrepancy between static clinical evaluation and IGA. As spasticity is velocity-dependent, the effects of spasticity and its inhibition are more pronounced in the biarticular muscles at higher walking speeds [56]. This may account for some of the ambiguity in the early literature where IGA was not routinely used to report outcomes, and the effects seen were marginal.

The effect of BtxA is greatest when used at a younger age, 5–10 years where gait may be more flexible [57]. It has been suggested that BtxA may delay the need for and reduce the frequency of surgical intervention [55, 58].

Selective Dorsal Rhizotomy (SDR)

This technique was first described by Peacock in 1960. More recently, its popularity has increased for permanent tone reduction in some children with CP. The surgical technique involves direct sequential stimulation of the nerves and aims to section 50–75 % of dorsal nerve rootlets. Reported benefits include: improvement in mobility, hygiene and self care issues, pain and lower limb spasms, improved bladder control and reduced upper limb tone. Many of these positive effects have not been objectively quantified within the literature.

IGA can aid preoperative selection for the procedure, fine-tuning of orthotics and direct postoperative physiotherapy regimens [59]. Post SDR analysis has demonstrated significant improvements in sagittal hip, knee and ankle kinematics with a predominantly sacrum up posture [60].

Table 38.3 Table of recommendations

Statement	Level of evidence	Grade of evidence
Instrumented gait analysis provides more objective data than observational analysis when assessing gait in children with Cerebral Palsy	I	A
Instrumented gait analysis alters surgical decision making when planning multilevel surgery	I	B
Improved postoperative outcomes are associated with children who underwent preoperative evaluation with instrumented gait analysis	III/IV	B
Instrumented gait analysis can decrease the reoperation rate in ambulatory children with cerebral palsy	III	B
Perioperative gait analysis is cost-effective	III	I

The Oswestry experience has demonstrated improvements in preswing knee flexion, walking speeds, step-length and normalised step length [59, 61].

Conclusion

IGA has been central to our understanding of the lower-limb during gait and the impacts of surgery. The literature supports gait analysis as requirement for the accurate identification of biomechanical abnormalities, and it appears that this is clinically beneficial to fine-tune surgical strategy. Whether there are demonstrable cost benefits over observational analysis alone is yet to be determined. However, it does provide information, which informs us, with, regards changes to the structural aspect of the structure/function domain of the ICF framework and must be combined with functional measures to accurately determine the outcome of our interventions. Table 38.3 lists the current recommendations regarding gait analysis.

References

1. Perry J, Burnfield J. Gait analysis: normal and pathological function. 2nd revised edition. SLACK: Thorofare; 2010.
2. Gage JR. Gait analysis: an essential tool in the treatment of cerebral palsy. Clin Orthop Relat Res. 1993;288:126–34.
3. Winters TF, Gage JR, Hicks R. Gait patterns in spastic hemiplegia in children and young adults. J Bone Joint Surg (Am). 1987;69:437–41.
4. Sutherland DH, Davids JR. Common gait abnormalities of the knee in cerebral palsy. Clin Orthop. 1993;288:139–47.
5. Rodda J, Graham HK. Classification of gait patterns in spastic hemiplegia and spastic diplegia: a basis for a management algorithm. Eur J Neurol. 2001;8:S98–108.
6. Rodda JM, Graham HK, Carson L, Galea MP, Wolfe R. Sagittal gait patterns in spastic diplegia. J Bone Joint Surg Br. 2004;86(2):251–8.
7. Nieuwenhuys A, Õunpuu S, Van Campenhout A, Theologis T, De Cat J, Stout J, Molenaers G, De Laet T, Desloovere K. Identification of joint patterns during gait in children with cerebral palsy: a Delphi consensus study. Dev Med Child Neurol. 2015. doi:10.1111/dmcn.12892.
8. Rathinam C, Bateman A, Peirson J, Skinner J. Observational gait assessment tools in paediatrics – a systematic review. Gait Posture. 2014;40(2):279–85.
9. Rasmussen HM, Nielsen DB, Pedersen NW, Overgaard S, Holsgaard-Larsen A. Gait deviation index, gait profile score and gait variable score in children with spastic cerebral palsy: intra-rater reliability and agreement across two repeated sessions. Gait Posture. 2015;42(2):133–7.
10. Baker R, McGinley JL, Schwartz MH, Beynon S, Rozumalski A, Graham HK, Tirosh O. The gait profile score and movement analysis profile. Gait Posture. 2009;30:265–9.
11. Schwartz MH, Rozumalski A. The gait deviation index: a new comprehensive index of gait pathology. Gait Posture. 2008;28(3):351–7.
12. Baker R, McGinley JL, Schwartz M, Thomason P, Rodda J, Graham HK. The minimal clinically important difference for the gait profile score. Gait Posture. 2012;35:612–5.
13. Wren TA, Woolf K, Kay RM. How closely do surgeons follow gait analysis recommendations and why? J Pediatr Orthop B. 2005;14(3):202–5.
14. Wren TA, Elihu KJ, Mansour S, Rethlefsen SA, Ryan DD, Smith ML, et al. Differences in implementation of gait analysis recommendations based on affiliation with a gait laboratory. Gait Posture. 2013;37(2):206–9.
15. Cook RE, Schneider I, Hazlewood ME, Hillman SJ, Robb JE. Gait analysis alters decision-making in cerebral palsy. J Pediatr Orthop. 2003;23(3):292–5.
16. Kay RM, Dennis S, Rethlefsen S, Reynolds RA, Skaggs DL, Tolo VT. The effect of preoperative gait analysis on orthopaedic decision making. Clin Orthop Relat Res. 2000;372:217–22.
17. DeLuca PA, Davis RB, Ounpuu S, Rose S, Sirkin R. Alterations in surgical decision making in patients with cerebral palsy based on three-dimensional gait analysis. J Pediatr Orthop. 1997;17(5):608–14.
18. Wren TA, Otsuka NY, Bowen RE, Scaduto AA, Chan LS, Sheng M, Hara R, RM K. Influence of gait analysis on decision-making for lower extremity orthopaedic surgery: baseline data from a randomized controlled trial. Gait Posture. 2011;34(3):364–9.
19. Wren TA, Kalisvaart MM, Ghatan CE, Rethlefsen SA, Hara R, Sheng M, et al. Effects of preoperative gait analysis on costs and amount of surgery. J Pediatr Orthop. 2009;29(6):558–63.
20. Kay RM, Dennis S, Rethlefsen S, Skaggs DL, Tolo VT. Impact of postoperative gait analysis on orthopaedic care. Clin Orthop Relat Res. 2000;374:259–64.
21. Thomason P, Baker R, Dodd K, Taylor N, Selber P, Wolfe R, Graham HK. Single-event multilevel surgery in children with spastic diplegia: a pilot randomized controlled trial. J Bone Joint Surg Am. 2011;93(5):451–60.
22. Rethlefsen SA, Healy BS, Wren TA, Skaggs DL, Kay RM. Causes of intoeing gait in children with cerebral palsy. J Bone Joint Surg Am. 2006;88(10):2175–80.

23. Dreher T, Wolf SI, Heitzmann D, Swartman B, Schuster W, Gantz S, Hagmann S, Döderlein L, Braatz F. Long-term outcome of femoral derotation osteotomy in children with spastic diplegia. Gait Posture. 2012;36(3):467–70.
24. Ounpuu S, DeLuca P, Davis R, MJ R. Long-term effects of femoral derotation osteotomies: an evaluation using three-dimensional gait analysis. J Pediatr Orthop. 2002;22(2):139–45.
25. Wren TA, Lening C, Rethlefsen SA, Kay RM. Impact of gait analysis on correction of excessive hip internal rotation in ambulatory children with cerebral palsy: a randomized controlled trial. Dev Med Child Neurol. 2013;55(10):919–25.
26. Pirpiris M, Trivett A, Baker R, Rodda J, Nattrass GR, Graham HK. Femoral derotation osteotomy in spastic diplegia. Proximal or distal? J Bone Joint Surg Br. 2003;85(2):265–72.
27. Niklasch M, Dreher T, Döderlein L, Wolf SI, Ziegler K, Brunner R, Rutz E. Superior functional outcome after femoral derotation osteotomy according to gait analysis in cerebral palsy. Gait Posture. 2015;41(1):52–6.
28. Kay RM, Rethlefsen SA, Hale JM, Skaggs DL, Tolo VT. Comparison of proximal and distal rotational femoral osteotomy in children with cerebral palsy. J Pediatr Orthop. 2003;23(2):150–4.
29. Laracca E, Stewart C, Postans N, Roberts A. The effects of surgical lengthening of hamstring muscles in children with cerebral palsy – the consequences of pre-operative muscle length measurement. Gait Posture. 2014;39(3):847–51.
30. Delp SL, Arnold AS, Speers RA, Moore CA. Hamstrings and psoas lengths during normal and crouch gait: implications for muscle-tendon surgery. J Orthop Res. 1996;14:144–51.
31. Young JL, Rodda J, Selber P, Rutz E, Graham HK. Management of the knee in spastic diplegia: what is the dose? Orthop Clin North Am. 2010;41(4):561–77.
32. Ma FY, Selber P, Nattrass GR, Harvey AR, Wolfe R, Graham HK. Lengthening and transfer of hamstrings for a flexion deformity of the knee in children with bilateral cerebral palsy: technique and preliminary results. J Bone Joint Surg Br. 2006;88(2):248–54.
33. Klatt J, Stevens PM. Guided growth for fixed knee flexion deformity. J Pediatr Orthop. 2008;28(6):626–31.
34. Stout JL, Gage JR, Schwartz MH, Novacheck TF. Distal femoral extension osteotomy and patellar tendon advancement to treat persistent crouch gait in cerebral palsy. J Bone Joint Surg Am. 2008;90(11):2470–84.
35. Gage JR, Perry J, Hicks RR, Koop S, Werntz JR. Rectus femoris transfer to improve knee function of children with cerebral palsy. Dev Med Child Neurol. 1987;29(2):159–66.
36. Kay RM, Rethlefsen SA, Kelly JP, Wren TA. Predictive value of the Duncan-Ely test in distal rectus femoris transfer. J Pediatr Orthop. 2004;24(1):59–62.
37. Perry J. Distal rectus femoris transfer. Dev Med Child Neurol. 1987;29(2):153–8.
38. Saw A, Smith PA, Sirirungruangsarn Y, Chen S, Hassani S, Harris G, Kuo KN. Rectus femoris transfer for children with cerebral palsy: long-term outcome. J Pediatr Orthop. 2003;23(5):672–8.
39. Fox MD, Reinbolt JA, Ounpuu S, Delp SL. Mechanisms of improved knee flexion after rectus femoris transfer surgery. J Biomech. 2009;42(5):614–9.
40. Rethlefsen SA, Kam G, Wren TA, Kay RM. Predictors of outcome of distal rectus femoris transfer surgery in ambulatory children with cerebral palsy. J Pediatr Orthop B. 2009;18(2):58–62.
41. Dreher T, Wolf S, Maier M, Hagmann S, Vegvari D, Gantz S, Heitzmann D, Wenz W, Braatz F. Long-term results after distal rectus femoris transfer as a part of multilevel surgery for the correction of stiff-knee gait in spastic diplegic cerebral palsy. J Bone Joint Surg Am. 2012;94(19):e142.
42. Dreher T, Gotze M, Wolf SI, Hagmann S, Heitzmann D, Gantz S, Braatz F. Distal rectus femoris transfer as part of multilevel surgery in children with spastic diplegia – a randomized clinical trial. Gait Posture. 2012;36(2):212–8.
43. Riewald SA, Delp SL. The action of the rectus femoris muscle following distal tendon transfer: does it generate knee flexion moment? Dev Med Child Neurol. 1997;39(2):99–105.
44. Presedo A, Megrot F, Ilharreborde B, Mazda K, Penneçot GF. Rectus femoris distal tendon resection improves knee motion in patients with spastic diplegia. Clin Orthop Relat Res. 2012; 470(5): 1312-1319.
45. Sutherland DH, Santi M, Abel MF. Treatment of stiff-knee gait in cerebral palsy: a comparison by gait analysis of distal rectus femoris transfer versus proximal rectus release. J Pediatr Orthop. 1990;10(4):433–41.
46. Végvári D, Wolf S, Heitzmann D, Klotz MC, Dreher T. Does proximal rectus femoris release influence kinematics in patients with cerebral palsy and stiff knee gait? Clin Orthop Relat Res. 2013;471(10):3293–300.
47. Wren TA, Rethlefsen S, Kay RM. Prevalence of specific gait abnormalities in children with cerebral palsy: influence of cerebral palsy subtype, age, and previous surgery. J Pediatr Orthop. 2005;25(1):79–83.
48. Wren TA, Rethlefsen SA, Healy BS, Do KP, Dennis SW, Kay RM. Reliability and validity of visual assessments of gait using a modified physician rating scale for crouch and foot contact. J Pediatr Orthop. 2005;25(5):646–50.
49. Borton DC, Walker K, Pirpiris M, Nattrass GR, Graham HK. Isolated calf lengthening in cerebral palsy. Outcome analysis of risk factors. J Bone Joint Surg Br. 2001;83(3):364–70.
50. Shore BJ, White N, Kerr Graham H. Surgical correction of equinus deformity in children with cerebral palsy: a systematic review. J Child Orthop. 2010;4(4):277–90.
51. Firth GB, McMullan M, Chin T, Ma F, Selber P, Eizenberg N, Wolfe R, Graham HK. Lengthening of the gastrocnemius-soleus complex: an anatomical and biomechanical study in human cadavers. J Bone Joint Surg Am. 2013;95(16):1489–96.
52. Tinney A, Thomason P, Sangeux M, Khot A, Graham HK. The transverse Vulpius gastrocsoleus recession for equinus gait in children with cerebral palsy. Bone Joint J. 2015;97-B(4):564–71.
53. Firth GB, Passmore E, Sangeux M, Thomason P, Rodda J, Donath S, Selber P, Graham HK. Multilevel surgery for equinus gait in children with spastic diplegic cerebral palsy: medium-term follow-up with gait analysis. J Bone Joint Surg Am. 2013;95(10):931–8.
54. Michlitsch MG, Rethlefsen SA, Kay RM. The contributions of anterior and posterior tibialis dysfunction to varus foot deformity in patients with cerebral palsy. J Bone Joint Surg Am. 2006;88(8):1764–8.
55. Desloovere K, Molenaers G, De Cat J, Pauwels P, Van Campenhout A, Ortibus E, Fabry G, De Cock P. Motor function following multilevel botulinum toxin type A treatment in children with cerebral palsy. Dev Med Child Neurol. 2007;49(1):56–61.
56. Van Campenhout A, Bar-On L, Aertbeliën E, Huenaerts C, Molenaers G, Desloovere K. Can we unmask features of spasticity during gait in children with cerebral palsy by increasing their walking velocity? Gait Posture. 2014;39(3):953–7.
57. Molenaers G, Desloovere K, Fabry G, De Cock P. The effects of quantitative gait assessment and botulinum toxin a on musculoskeletal surgery in children with cerebral palsy. J Bone Joint Surg Am. 2006;88(1):161–70.
58. Molenaers G, Fagard K, Van Campenhout A, Desloovere K. Botulinum toxin A treatment of the lower extremities in children with cerebral palsy. J Child Orthop. 2013;7(5):383–7.
59. Roberts A, Stewart C, Freeman R. Gait analysis to guide a selective dorsal rhizotomy program. Gait Posture. 2015;42(1):16–22.
60. Abel MF, Damiano DL, Gilgannon M, Carmines D, Kang HG, Bennett BC, Laws Jr ER. Biomechanical changes in gait following selective dorsal rhizotomy. J Neurosurg. 2005;102(2 Suppl):157–62.
61. Cole GF, Farmer SE, Roberts A, Stewart C, Patrick JH. Selective dorsal rhizotomy for children with cerebral palsy: the Oswestry experience. Arch Dis Child. 2007;92(9):781–5.

The Evidence Base for Botulinum Toxin Injection for the Treatment of Cerebral Palsy–Related Spasticity in the Lower Limb: The Long-Term Effects

39

James S. Huntley and Lyndon J. Bradley

Abstract

Botulinum toxin is a potent neurotoxin that is widely used (and has been for many years) for the treatment of focal spasticity. Its short-term effects have been well documented. There remains considerable ambivalence over its long-term effects, and some concern over side-effects. A scoping assessment of the literature defines the paucity of the evidence base concerning long-term effectiveness. Only three level 1 and two level 2 studies could be identified, on lower limb effects of botulinum toxin in children with cerebral palsy *at a time interval greater than 1 year*. Furthermore, our interpretation of the results *of these studies* is that there is no evidence of clinical benefit. We hope this analysis and summary can guide clinicians in counseling parents and patients as regards long-term expectations, and reservations/implications of management.

Keywords

Botulinum toxin • Botulinum • BoNT-A • BtA • BT • Botox • Dysport • Cerebral palsy • Spasticity • Lower limb • Contracture • Hip subluxation • Hip dislocation

Introduction

Botulinum toxin is a potent neurotoxin produced by *Clostridium botulinum* which acts presynaptically, blocking the release of acetylcholine at the neuromuscular junction [1] inducing a flaccid weakness [2]. It is widely used for the treatment of spasticity and has been recommended for such use in multiple guidelines and systematic reviews [3–11]. However, there are concerns over its use, including toxicity [1, 7], response to treatment [12], decreasing effects with repeated administration [9], irreversible damage to motor endplates [13], continuing muscle stiffness [14], irreversibility of certain effects on muscle architecture [15, 16], development of antibodies after repeated treatments [17], actions other than at the neuromuscular junction - in particular secondary central changes [18], negative effects at non-target muscles [19] and retrograde axonal transport [20, 21].

Criticism has also been levied at the tendency for studies to concentrate on short-term outcome measures such as range of motion and spasticity, rather than activity and participation [22]. Whilst accepting the short-term beneficial effects on spasticity and range of movement, other commentators *more than a decade ago* highlighted the lack of evidence of long-term benefits, for instance on function, mobility or muscle growth [23]. More recently, a systematic review on the efficacy of botulinum toxin A in children with cerebral palsy (GMFCS IV and V) concluded that the poor levels of evidence of included studies precluded the feasibility of conclusions [24].

Our aim was to assess the evidence of longer term effects (>1 year) of botulinum toxin in the treatment of lower limb spasticity in children with cerebral palsy. We addressed this question in a brief review in 2014 [25], and have reused that search strategy in an attempt to update the material available

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for review. In the event, no further references (complying with all inclusion/exclusion criteria) were found.

Question

They key question is: *For paediatric patients with spastic cerebral palsy affecting the lower limbs, are there long-term beneficial effects (outcomes) from botulinum toxin injection?*

Search Strategy

The previously utilized search strategy [25] was applied for MEDLINE, PubMed and Cochrane databases (date accessed: 20/09/2015), using a PICO format: {'cerebral palsy'} (**P**), {'botulinum toxin', 'Botulinum', 'BoNT-A', 'BoNT-B', 'BtA', 'BtB', 'BT', 'Botox' and 'Dysport'} (**I**). Age was limited to 0–18 years, and randomized controlled trials (**C, O**). The titles and abstracts (and if needed, full article) were again reviewed, so that only studies pertaining to the lower limbs (i.e. excluding those considering dystonia, sialorrhea, autonomic dysfunction and upper limb pathology) were included. Studies with follow-up of 1 year or less were excluded, leaving only five studies for detailed consideration (Table 39.1).

Hawamdeh et al. [26] reported a study of the effects of three successive injections (intervals of 3–4 months) of botulinum toxin A (either Botox or Dysport) on calf muscle tone (modified Ashworth score (MAS)), passive ankle dorsiflexion range and gross motor function (GMF), at 3 months and 18 months (after last injection) in children with spastic diplegia, aged 3–15 years. Patients were excluded from the trial if they had fixed contractures or deformities, '*previous surgery to the lower limb at least 1 year before the start of the study*', previous botulinum toxin treatment, treatment with phenol,

Table 39.1 Summary table of studies included and characteristics in this evidence-based review

Results				
Paper	Design	Number of patients and treatment	Follow-up, details, results	Level of evidence
Hawamdeh et al.[a] [26]	RCT	60; 20 control, 40 treatment (botox injection to calves; 3 × 6 monthly)	Follow-up periods 1 and 2 years: (i) allegedly improved MAS, (ii) statistically significant (but possibly not clinically relevant) increase in mean passive ankle 7dorsiflexion (12 vs 15 degrees), (iii) increase in median GMFM	2[a]
Graham et al. [27]	Multicentre RCT	91; 44 control, 47 treatment (botox injections to hamstrings/adductors + hip abduction brace)	Follow-up: 3 years. On the basis of change in RMP, there was no evidence of a clinically important effect on rate or amount of hip subluxation	1
Moore et al. [28]	Placebo controlled RCT	64; 32 control, 32 treatment (6 monthly botox injections lower limb muscles for 2 years)	Follow-up periods 1 and 2 years: no evidence of cumulative or persistent change in GMFM or PEDI scales or subscales	1
Tedroff et al.[a] [29]	RCT	15; 9 control, 6 treatment (botox injections to gastrocnemius 6 months apart)	Follow-up periods 1 and 3.5 years: (i) plantarflexor tone, hamstring MAS '*significant change from baseline*', (ii) NSD: (a) ROM, (b) gait analysis, (c) GMFM, (d) PEDI	2[a]
Willoughby et al. [30]	RCT	46 (single centre subset of Graham et al. [27]); 23 control, 23 treatment (botox injections to hamstrings/ adductors + hip abduction brace)	Follow-up mean 10yrs 10mths: NSD in ultimate RMP, hip morphology, *delay to* or *need for* preventative or reconstructive hip surgery	1

NSD no significant difference, *RCT* randomised controlled trial, *MAS* modified ashworth score, *GMFM* gross motor function measure, *RMP* reimers' migration percentage, *ROM* range of movement, *PEDI* paediatric evaluation disability index

[a]Subject to bias/methodological concerns sufficient to downgrade level of evidence further

intrathecal baclofen or other antispasticity agent. There were 60 patients: 40 in the treatment group, and 20 in the control group. At 18 months, the study reported the treatment group to have (i) better muscle tone (modified Ashworth score), involving a manoeuvre of questionable validity to convert categorical to numerical data, (ii) a statistically significant (but clinically insubstantial) improvement in passive ankle dorsiflexion range (12 vs 15 degrees), and (iii) an increase in median gross motor function from grade 4 "*reasonably useful non-ambulant locomotion and/or walking when assisted*" to 5 "*walking with a walking aid*" (both control and treatment groups improved from a baseline of grade 3 – it is not clear how many more patients improved by how much more to result in the observed difference in median GMF score).

The authors alleged the study to be randomized and single-blind. This study was not placebo-controlled, so patients and parents were aware of the treatment received; the two groups had their ancillary physical treatments under different physiotherapists on different days. It is questionable therefore whether physiotherapists would be 'blind' to the nature of treatment of their group (as alleged). Furthermore "*All outcome measurements were performed by an experienced senior paediatric physiotherapist who knew whether a child belonged to the treatment or the control group*". It would appear therefore that the study was not blinded as to either participants or assessor, and should not be classed as 'single blind' by the conventional meaning for this term. Additionally, there is concern about the mode of local randomization '*by flipping a coin*', apparently dividing subjects into two homogeneous groups: treatment group (40) and control group (20). This study is subject to major bias, sufficient to downgrade the level of evidence accorded, and the results should be treated/interpreted with major reservation.

Moore et al. [28] reported a single-center randomized, placebo-controlled, parallel group, double-blind trial of the effect of botulinum toxin A (Dysport) injection (repeated 3 monthly, if clinically indicated; i.e. 8 injection cycles) on motor function (Gross Motor Function Measure (GMFM) and Paediatric Evaluation of Disability Index (PEDI)) at 2 years (a 'trough' rather than 'peak' effect), on children with cerebral palsy aged 2–6 years (or 2–8 years, if targeting the hamstring muscles), with clinically significant spasticity of one or both lower limbs, and excessive involuntary muscle activity. Patients were excluded from the trial if they had *'a disease, handicap or situation likely to make treatment too difficult or dangerous or to invalidate or prevent assessments*', or had been treated with botulinum toxin previously. There were 64 patients, with 32 in each group (placebo and botulinum toxin). There was no difference in adverse events between the two groups. The study found no evidence of cumulative or persisting benefit (mean change scores for both GMFM and PEDI, for either total or subscale values) from treatment at either 1 or 2 years. They commented that (i) the ceiling effects of the scales used may have limited responsiveness, and (ii) that '*the small numbers may have caused a type 2 error although, if anything, the trends suggest that placebo is better'*.

Tedroff et al. [31] reported a single center study on the effect of botulinum toxin A (Botox – two injections, 6 months apart, in gastrocnemius) on muscle tone (modified Ashworth score), development of contractures and gait (gait analysis, GMFM-66 and PEDI) in 15 young children (six in the botox treatment group, and nine in the control group; mean age 16 months) with spastic cerebral palsy (either unilateral or bilateral), after 1 and 3.5 years. The treatment phase was initiated when children were '*pulling to stand'*. Both groups received a daily stretching programme. Unfortunately, the groups allegedly contained heterogeneous GMFCS proportionality at the start of the study (the botox group: GMFCS I (4), GMFCS II (2); the control group: GMFCS I (4), GMFCS II (4), GMFCS III (1)), though GMFCS is hard to apportion at <2 years age. Furthermore, although the baseline characteristics of the two groups were held as showing no statistical significant difference except for plantarflexor tone, there were differences in mean values (eg ankle dorsal extension 10 degrees in the botulinum group and 17.8 degrees in the control group) with large standard deviations (unsurprisingly given the small numbers in each group). The authors hold that plantarflexor tone was significantly reduced after 3.5 years in the treatment group. Furthermore, they found the 'change-score in knee flexion muscle tone' to be significantly different between the two groups after 3.5 years. There was no significant difference between the groups in (i) changes in ankle dorsiflexion range at any time, (ii) gait analysis, (iii) GMFM-66, or (iv) PEDI. The limitations of this study are profound, including small sample size, initial assessment of subjects, heterogeneity of patients within groups, differences in baseline characteristics between groups, lack of blinding of either treatment or assessment (except 3D-gait analysis). These concerns, together with the difficulty in deriving meaningful values from the presented data (much is presented as 'change from baseline' or 'effect sizes', themselves "*calculated by dividing the absolute mean difference between the groups by the SD for the control group at the baseline*"), largely negate any value of the study.

Graham et al. [27] reported a multicenter (5 centers) randomized controlled trial of the effect of 6 monthly intramuscular botulinum toxin A injection (adductors and hamstrings) and SWASH hip abduction orthosis (6 h per day) on hip displacement (Reimers' migration percentage (RMP) on radiography, 6 monthly) over 3 years, on children with bilateral spastic cerebral palsy (including hip adductor reactivity), aged 1–5 years, and with hips at risk (RMP 10–40 %). Patients were excluded from the trial if they had pseudobulbar palsy, previous hip surgery, hip

Table 39.2 Summary of grades of recommendation

Recommendation	Level of evidence	Grade
The use of botulinum toxin does not prevent the long term development of contractures	1	B
The use of botulinum toxin does not prevent the long term development of hip subluxation or requirement for hip surgery	1	B
The use of botulinum toxin does not result in long term improvement of joint range of motion	2	C
The use of botulinum toxin improves the long term function and participation of the child	–	I
The use of botulinum toxin prevents the long term development of muscle stiffness	–	I

flexion contracture > 30 degrees, or scoliosis with Cobb angle > 20 degrees. There were 91 patients, at a 98 % recruitment rate; 44 and 47 patients were allocated to control and treatment groups, respectively. Of note, there were 12 and 33 adverse events (major and minor, respectively) after 204 injection episodes (incidences of 6 and 16 %). Two modes of analysis were used to compare the treatment and control groups – one showing no statistically significant effect, the other suggesting a small decrease in the rate of displacement for the treatment group (3.1 % unweighted; 1.4 % weighted), indicative of a possible delay in the time to surgery, but not a difference on the number requiring surgery ultimately. The authors summarized: *"There was no evidence of a beneficial effect of treatment that was clinically important."*

Willoughby et al. [30] reported the long-term outcome data (hip morphology and surgery requirements) for 46 patients (23 in each of the two groups – control and treatment (3 year programme botulinum toxin and hip abduction orthosis); 31 male, 15 female; 36 quadriplegic, 10 diplegic; mean follow-up after recruitment was 10 years and 10 months) from the major single centre in the trial outlined above [27]. There was no significant difference between the treatment and control groups in ultimate mean migration percentage, hip morphology (Melbourne Cerebral Palsy Hip Classification System (MCPHCS)), need for preventive surgery (21 ex 23, and 19 ex 23 in treatment and control groups respectively) and reconstructive surgery (10 ex 23, 8 ex 23). Furthermore, for children requiring surgery, '*there was no statistically significant difference between the groups in mean age at either preventive or reconstructive surgery.*' i.e. there was no long-term benefit from the treatment, either in terms of reducing/delaying the need for surgery or improving later hip morphology.

Conclusions

The controlled evidence for using botox treatment for spasticity is based on short-term studies [5, 28, 32]. In a systematic review [5]) on the effectiveness of botulinum toxin for treatment of lower limb spasticity in children with cerebral palsy, 15 studies (6 level I and 9 level II) were analysed, culminating in the suggestion that botulinum toxin is effective in reducing spasticity and providing a '*time limited improvement in function in the upper and lower limbs for children with CP*'. They also asserted that '*Most of the previous trials cover short observation periods, ranging from 6–24 weeks only. They are, therefore, insufficient to capture long-term effects of repeated botulinum toxin injection such as the prevention of contractures and secondary pain.*'

In a more recent systematic review [33], concerning treatment effects on walking of children with leg spasticity due to cerebral palsy, 8 trials were included, with follow-up time for assessment ranging from 1 week to a maximum of only 12 months. An abundance of lower level evidence (III/IV) has been held to suggest long-term effectiveness of botulinum toxin injection [31, 34–39].

It is disappointing that despite over 20 years of clinical use, we were able to find only five level I and II studies on the long-term (>1 year) effects of botulinum toxin on the lower limbs of children with cerebral palsy. As previously [25], we would affirm that '*the use of intramuscular injection of botulinum toxin A for spasticity management in the lower limbs of children with cerebral palsy has not been shown to have clinically relevant effect beyond one year*'. Additionally, it is of concern that about half of studies on the effect of botulinum toxin A in cerebral palsy are performed with industry sponsorship, and further that the quailitative conclusions of industry-associated studies are more favourable to the use of this modality than those without funding links [40].

A summary for current grades of recommendation for the use of botulinum toxin is provided in Table 39.2.

References

1. Ramachandran M, Eastwood DM. Botulinum toxin and its orthopaedic applications. J Bone Joint Surg Br. 2006;88:981–7.
2. Sesardic D, Jones RGA, Leung T, Alsop T, Tierney R. Detection of antibodies against botulinum toxins. Mov Disord. 2004;18(S8):S85–91.
3. Eames NW, Baker R, Hill N, Graham K, Taylor T, Cosgrove A. The effect of botulinum toxin A on gastrocnemius length: magnitude and duration of response. Dev Med Child Neurol. 1999;41:226–32.
4. Graham HK, Aoki KR, Autti-Ramo I, Boyd RN, Delgado MR, Gormley ME, Guyer BM, Heinen F, Holton AF, Matthews D, Molenaers G, Motta F, Garcia Ruiz PJ, Wissel J. Recommendations for the use of botulinum toxin type A in the management of cerebral palsy. Gait Posture. 2000;11:67–79.

5. Lukban MB, Rosales RL, Dressler D. Effectiveness of botulinum toxin A for upper and lower limb spasticity in children with cerebral palsy: a summary of evidence. J Neural Transm. 2009;116: 319–31.
6. Esquenazi A, Novak I, Sheean G, Singer BJ, Ward AB. International consensus statement for the use of botulinum toxin treatment in adults and children with neurological impairments – introduction. Eur J Neurol. 2010;17(S2):1–8.
7. Love SC, Novak I, Kentish M, Desloovere K, Heinen F, Molenaers G, O'Flaherty SO, Graham HK. Botulinum toxin assessment, intervention and after-care for lower limb spasticity in children with cerebral palsy: international consensus statement. Eur J Neurol. 2010;17(S2):9–37.
8. NICE. 2012. NICE clinical guidelines. Report no.: CG145.
9. Molenaers G, Fagard K, van Camoenhout A, Desloovere K. Botulinum toxin A treatment of the lower extremeties in children with cerebral palsy. J Child Orthop. 2013;7:383–7.
10. Novak I, Mcintyre S, Morgan C, Campbell L, Dark L, Morton N, Stumbles E, Wilson SA, Goldsmith S. A systematic review of interventions for children with cerebral palsy: state of the evidence. Dev Med Child Neurol. 2013;55:885–910.
11. Strobl W, Theologis T, Brunner R, Kocer S, Viehweger E, Pascual-Pascual I, Placzek R. Best clinical practice in botulinum toxin treatment for children with cerebral palsy. Toxins. 2015;7:1629–48.
12. Koog YH, Min BI. Effects of botulinum toxin on calf muscles in children with cerebral palsy: a systematic review. Clin Rehabil. 2010;24:685–700.
13. Druschel C, Althuizes HC, Funk JF, Placzek R. Off label use of botulinum toxin in children under two years of age: a systematic review. Toxins (Basel). 2013;5:60–72.
14. Alhusaini A, Crosbie J. No change in calf muscle passive stiffness after botulinum toxin injection in children with cerebral palsy. Dev Med Child Neurol. 2011;53:553–8.
15. Fortuna R, Horisberger M, Vaz MA, Herzog W. Do skeletal muscle properties recover following repeat onabotulinum toxin A injections? J Biomech. 2013;46:2426–33.
16. Fortuna R, Vaz MA, Sawatsky A, Hart DA, Herzog W. A clinically relevant BTX-A injection protocol leads to persistent weakness, contractile material loss, and an altered mRNA expression phenotype in rabbit quadriceps muscles. J Biomech. 2015;48:1700–6.
17. Eleopra R, Tugnoli V, Quatrale R, Rossetto O, Montecucco C. Different types of botulinum toxin in humans. Mov Disord. 2004;19(S8):S53–9.
18. Curra A, Trompetto C, Abbruzzese G, Berardelli A. Central effects of botulinum toxin type A: evidence and supposition. Mov Disord. 2004;19(S8):S60–4.
19. Fortuna R, Vaz MA, Youssef AR, Longino D, Herzog W. Changes in contractile properties of muscles receiving repeat injections of botulinum toxin (Botox). J Biomech. 2011;44:39–44.
20. Boroff DA, Chen GS. On the question of permeability of the blood-brain barrier to botulinum toxin. Int Arch Allergy Appl Immunol. 1975;48:495–504.
21. Gracies JM. Physiological effects of botulinum toxin in spasticity. Mov Disord. 2004;19:S120–8.
22. Baird MW, Vargus-Adams J. Outcome measures used in studies of botulinum toxin in childhood cerebral palsy: a systematic review. J Child Neurol. 2010;25:721–7.
23. Gough M, Fairhurst C, Shortland AP. Botulinum toxin and cerebral palsy: time for reflection? Dev Med Child Neurol. 2005;47: 709–12.
24. Pin TW, Elasry J, Lewis J. Efficacy of botulinum toxin A in children with cerebral palsy in gross motor function classification system levels IV and V: a systematic review. Dev Med Child Neurol. 2013;55:304–13.
25. Bradley LJ, Huntley JS. Is there any long-term benefit from injecting botulinum toxin-A into children with cerebral palsy? Arch Dis Child. 2014;99:392–4.
26. Hawamdeh ZM, Ibrahim AI, Al-Qudah AA. Long-term effect of botulinum toxin (A) in the management of calf spasticity in children with diplegic cerebral palsy. Eura Medicophys. 2007;43: 311–8.
27. Graham HK, Boyd R, Carlin JB, Dobson F, Lowe K, Nattrass G, Thomason P, Wolfe R, Reddihough D. Does botulinum toxin A combined with bracing prevent hip displacement in children with cerebral palsy and "hips at risk"? A randomized, controlled trial. J Bone Joint Surg. 2008;90:23–33.
28. Moore AP, Ade-Hall RA, Tudur Smith C, Rosenbloom L, Walsh HPJ, Mohamed K, Williamson PR. Two-year placebo-controlled trial of botulinum toxin A for leg spasticity in cerebral palsy. Neurology. 2008;71:122–8.
29. Tedroff K, Granath F, Forssberg H, Haglund-Akerlind Y. Long-term effects of botulinum toxin A in children with cerebral palsy. Dev Med Child Neurol. 2009;51:120–7.
30. Willoughby K, Ang SG, Thomason P, Graham HK. The impact of botulinum toxin A and abduction bracing on long-term hip development in children with cerebral palsy. Dev Med Child Neurol. 2012;54:743–7.
31. Tedroff K, Lowing K, Haglund-Akerlind Y, Gutierrez-Farewik E, Forssberg H. Botulinum toxin A treatment in toddlers with cerebral palsy. Acta Paediatr. 2010;99:1156–62.
32. Ade-Hall RA, Moore AP. Botulinum toxin type A in the treatment of lower limb spasticity in cerebral palsy. Cochrane Database Syst Rev. 2000;2:CD001408.
33. Ryll U, Bastiaenen C, de Bie R, Staal B. Effects of leg muscle botulinum toxin A injections on walking in children with spasticity-related cerebral palsy: a systemiatic review. Dev Med Clin Neurol. 2011;53:210–6.
34. Reddihough DS, King JA, Coleman GJ, Fosang A, McCoy AT, Thomason P, Graham HK. Functional outcome of botulinum toxin A injections to the lower limbs in cerebral palsy. Dev Med Child Neurol. 44:820–7.
35. Hagglund G, Andersson S, Duppe H, Lauge-Pedersen H, Nordmark E, Westborn L. Prevention of severe contractures might replace multilevel surgery in cerebral palsy: results of a population-based health care programme and new techniques to reduce spasticity. J Pediatr Orthop B. 2005;14:269–73.
36. Molenaers G, Desloovere K, Fabry G, de Cock P. The effects of quantitative gait assessment and botulinum toxin A on musculoskeletal surgery in children with cerebral palsy. J Bone Joint Surg Am. 2006;88:161–70.
37. Scholtes VA, Dallmeijer AJ, Knol DL, Speth LA, Maathuis CG, Jongerius PH, Becher JG. The combined effect of lower-limb multilevel botulinum toxin type a and comprehensive rehabilitation on mobility in children with cerebral palsy: a randomized clinical trial. Arch Phys Med Rehabil. 2006;87:1551–8.
38. Fattal-Valevski A, Domenievitz D, Giladi N, Wientroub S, Hayek S. Long-term effect of repeated injections of botulinum toxin in children with cerebral palsy: a prospective study. J Child Orthop. 2008;2:29–35.
39. Colovic H, Dimitrijevich L, Stankovic I, Nikolic D, Radovic-Janosevic D. Estimation of botulinum toxin type A efficacy on spasticity and functional outcome in children with spastic cerebral palsy. Biomed Pap Med Fac Univ Palacky Olomouc Czech Repub. 2012;156:41–7.
40. Sung KH, Chung CY, Lee KM, Lee YK, Lee J, Choi IH, Cho TJ, Yoo WJ, Park MS. Conflict of interest in the assessment of botulinum toxin A injections in patients with cerebral palsy: a systematic review. J Pediatr Orthop. 2013;33:494–500.

Evidence-Based Treatment for Feet Deformities in Children with Neuromuscular Conditions

40

Emmanouil Morakis and Anne Foster

Abstract

Foot deformities are very common in neuromuscular diseases. In these conditions, the muscles themselves or their control by the nervous system are affected with subsequent spasticity, weakness or both. In general, spasticity is a result of an upper motor neurons injury (e.g. cerebral palsy). Weakness commonly develops in injuries of the lower motor neurons (e.g. Charcot-Marie-Tooth disease).

Muscles imbalance because of spasticity, weakness, or both can lead to the development of foot deformities. These can take the form of equinus, varus, valgus, cavus, planus or combinations of these. In this chapter we summarised the common foot deformities encountered in clinical practice with particular reference to cerebral palsy (CP) and spina bifida. Charcot-Marie-Tooth disease (CMT) is covered in chapter.

Keywords

Foot deformities • Neuromuscular diseases • Cerebral palsy • Equinus • Cavo-varus • Plano-valgus • Equino-valgus • Cavus • Planus • Spina bifida

Introduction

Cerebral palsy has been defined as "group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior; by epilepsy, and by secondary musculoskeletal problems" [1].

Cerebral palsy can be classified according to the main movement disorder, the topographical distribution and the gross motor function. The most common movement disorder is the spastic type followed by the dyskinetic and mixed types. Less common forms include the ataxic and hypotonic. Topographical involvement in a patient differentiates into monoplegia, hemiplegia, diplegia, triplegia and quadriplegia. Often differentiation between diplegia and quadriplegia can be difficult. The Gross Motor Function Classification System (GMFCS) distinguishes five levels of motor function with decreasing level of function and independent walking and increasing use of assistive devices [2]. It has been found to be valid, reliable, useful and stable over time [3].

Abnormal tone, spasticity, muscle imbalance and impaired motor control create a dynamic deformity of the foot. Over time, the development of soft tissue contractures, bone deformities and joint instability transforms the flexible deformity into a rigid one [4]. Most patients with cerebral palsy will develop a flexible or rigid deformity of their feet [5]. The most common foot deformities in patients with cerebral palsy include equinus, equino-plano-valgus and equino-cavo-varus [5].

Foot deformity in children with cerebral palsy can cause pain with ambulation, orthosis intolerance and frequent tripping. It also affects their standing and walking ability. It causes gait dysfunction, as the foot deformity compromises the stability in stance, foot clearance in swing and prepositioning in terminal swing.

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Foot deformities in children with cerebral palsy can be managed with non-operative and surgical methods. Non-operative methods include orthotics, tone management medication (focal or global) and physiotherapy. The goals of treatment depend on the ambulatory capacity of the children. In non-ambulatory patients treatment goals include comfortable footwear and orthotic use, avoiding skin irritation and allowing therapeutic standing. In patients that walk, the main goal is a painless and efficient gait.

The basic principles of surgical treatment in children with cerebral palsy and foot deformity include the correction of the bone deformities, the stabilization of unstable joints and rebalancing of the muscles. In ambulatory patients, foot deformity correction is usually part of a single event multi level surgical procedure.

Equinus Deformity

Equinus deformity is the most common foot deformity in children with cerebral palsy (Fig. 40.1). It is the result of the overactive gastrosoleus complex against the ground reaction force. It impairs gait function; causes difficulty with footwear and can cause painful feet. Non-operative treatment strategies are being used in younger children, when the equinus deformity is usually flexible. These include the use of orthotics, tone management medication (botulinum toxin) and physiotherapy. Surgical treatment is indicated when non-operative measures fail and the deformity becomes rigid. This should be performed with caution, especially in children with diplegia. Over lengthening of the gastrosoleus complex can lead to weakness and crouch gait.

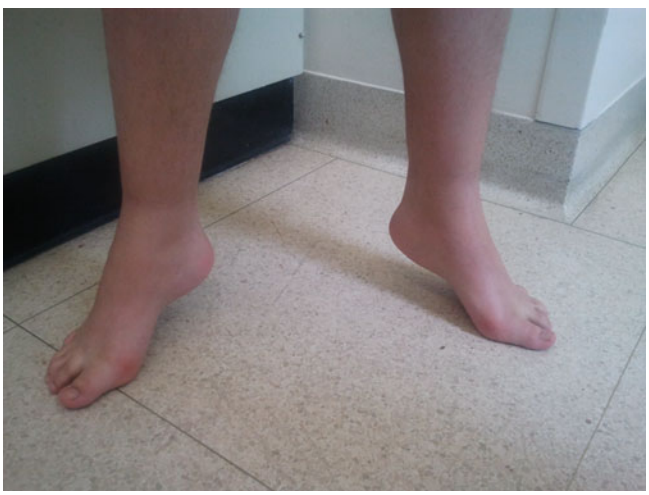

Fig. 40.1 Equinus feet in a child with cerebral palsy

Does Botulinum Toxin Injection Correct Spastic Equinus Deformity and Improve Gait or Function?

Botulinum toxin is produced by the anaerobic bacterium Clostridium botulinum and selectively blocks the release of acetylcholine at the cholinergic nerve terminal of a neuromuscular junction. This causes a temporary reduction in muscular activity in the specific muscles [6]. Botulinum toxin intramuscular injection has been used to reduce the spasticity of specific muscle groups in children with cerebral palsy. The reduction of spasticity would augment the effect of interventions (physiotherapy, serial casting, orthotics) that prevent the formation of contractures, enhance motor ability and improve function [7].

Botulinum toxin has been used extensively to address the spastic equinus foot deformity. The types of botulinum toxin, dosage, number of sites of injection and use of adjunct interventions (casting, physiotherapy) vary significantly in the literature. Current evidence supports the use of botulinum toxin to correct the spastic equinus deformity in ambulatory children with cerebral palsy. It has been shown that it improves gait and may improve function.

In a randomized double-blind control study, Koman et al. evaluated the effect of botulinum toxin in 12 ambulatory children with cerebral palsy and dynamic equinus contracture [8]. They assessed gait and function using observational gait analysis, a physician rating scale and a parent/guardian questionnaire. The children that were treated with botulinum toxin had significant improvement in their gait pattern compared to the placebo group.

In a multi-center randomized double-blind control study, Koman et al. evaluated the effect of botulinum toxin in 114 ambulatory children with cerebral palsy and dynamic equinus using an observational gait analysis scale (Physician Rating Scale) [9]. The patients were evaluated in five occasions up to 12 weeks post treatment. Again, the group of patients treated with botulinum toxin demonstrated improved gait pattern comparing to the placebo group. The positive effect on gait pattern lasted up to 12 weeks post treatment.

Sutherland et al. used instrumented gait analysis to objectively evaluate the gait of children with dynamic equinus [10]. In this prospective, randomized, double blind clinical trial, 20 children were evaluated before and 8 weeks following treatment. The children that were treated with botulinum toxin showed improved gait kinematics (ankle dorsiflexion in stance and swing) in the short term.

Twelve matched pairs of children with spastic hemiplegia and dynamic equinus contracture were used in a pair randomized controlled clinical trial to measure the effect of botulinum toxin in functional ability [11]. Love et al. measured motor function using the Gross Motor Function Measure (GMFM) on enrolment and at 1, 3 and 6 months post injection. The children that had treatment with botulinum

toxin demonstrated reduced spasticity and improved motor function that lasted up to 6 months.

Bjornson et al. reported similar findings [12]. In a randomized double masked placebo controlled study, they measured the gross motor skills (GMFM) and energy expenditure in 33 ambulatory children with spastic diplegia without fixed equinus contractures. Seventeen participants were randomized to receive bilateral gastrocnemius botulinum toxin injections and 16 received saline injections. The group of children that received botulinum toxin demonstrated improved gross motor skills 6 months following treatment.

The effect of botulinum toxin on walking was assessed with a video gait analysis scale by Ubhi et al. [13]. In a randomised double blind placebo controlled trial, the gait and function of forty ambulatory children with cerebral palsy and spastic equinus deformity was assessed in four occasions (baseline, 2,4 and 12 weeks post treatment). The children that were treated with botulinum toxin demonstrated improved gait pattern at 6 and 12 weeks; improved walking function (GMFM) at 12 weeks post treatment.

In another randomized control study, El-Etribi et al. compared 20 children with spastic diplegia and dynamic equinus that were treated with botulinum toxin injections and physiotherapy with a similar group of children that were treated with physiotherapy alone [14]. The authors assessed spasticity, ankle range of motion and gait pattern using an observational gait analysis scale (Physician Rating Scale) pre and post treatment. The follow-up of these patients was 12 weeks after treatment. The children that were treated with botulinum toxin injections had reduced spasticity and improved gait pattern that lasted at least for 12 weeks.

Different modalities have been used as an adjunct to botulinum toxin injections, in order to enhance its beneficial effect. The use of serial casting, orthotics and physiotherapy has been suggested to augment the effects of botulinum toxin injections.

Two randomized control studies evaluated the adjunctive effect of casting following botulinum toxin injection in children with dynamic equinus. Bottos et al. compared five children with spastic diplegia that were treated for dynamic equinus with botulinum toxin injections alone with five that had casting for 3 weeks post injection [15]. The addition of casting provided longer lasting effect of reduced spasticity and improved function (GMFM) at 4 months post treatment and increased stride length.

In a multicenter randomized study, 39 ambulatory children with cerebral palsy and dynamic equinus were divided into three treatment groups [16]. One group was treated with botulinum toxin injections alone, the second with botulinum toxin injections and casting and the third with placebo injections and casting. The two groups of patient that were treated with casting showed improvements in ankle kinematics, spasticity, passive ankle range of motion and dorsiflexion strength. No significant differences were identified on the patients that were treated with botulinum toxin injections alone.

Physiotherapy programs are commonly used in children with cerebral palsy. These though differ a lot in terms of the modalities used and their intensity. There is good evidence that physiotherapy programs such as strengthening programs are effective in children with cerebral palsy [17]. Physiotherapy has been part of all studies that evaluated the effectiveness of botulinum toxin injections. There is insufficient evidence that can support or refute the use of physiotherapy programs as an adjunct to botulinum toxin injections to improve dynamic equinus in children with cerebral palsy. Most experts would suggest that physiotherapy should be always be used when treating dynamic equinus deformities in children with cerebral palsy using botulinum toxin injections. There is consensus among experts that physiotherapy modalities, such as stretching, strengthening, targeted motor training should be used routinely as adjuncts to treat dynamic equinus with botulinum toxin injections [18, 19].

Three systematic reviews assessed the efficacy of botulinum toxin for the management of dynamic equinus in patients with cerebral palsy. All three found evidence supporting its efficacy in improving gait over placebo in the short term [20–22].

Does Surgical Treatment of Equinus Deformity Improve Gait and Function in Ambulatory Children with Cerebral Palsy?

Dynamic equinus deformity in children with cerebral palsy progresses with time into a static contracture of the gastrosoleus complex. Equinus contracture is usually part of a constellation of lower limb deformities that affects the gait of children with cerebral palsy. Equinus contracture can impair gait efficiency, lead to foot pain and difficulties with orthotic use and footwear. When non-operative measures fail to control and correct an equinus contracture, surgical treatment is indicated.

Various surgical techniques to lengthen the gastrosoleus complex have been described. Their main difference is the level of lengthening of the gastrosoleus complex: it can be at the muscle belly of gastrocnemius, at the gastrocnemius aponeurosis and soleus fascia or at the Achilles tendon. Lengthening at the level of gastrocnemius muscle belly is indicated when only this muscle is involved. When both the gastrocnemius and soleus are affected, more distal lengthening is performed.

With procedures that are more distal, more lengthening can be achieved, but there is higher risk of over-lengthening and muscle weakness. The outcome of surgical correction of an equinus contracture is more predictable as a child gets older [23]. Children with hemiplegia are more prone to recurrence of the equinus contracture, while children with

diplegia are more prone to over-lengthening and developing a calcaneus deformity [24, 25].

Surgical correction of equinus contracture in children with cerebral palsy improves gait and function in the short and long term. It is usually part of a multi level surgical intervention (single event multilevel surgery) to address all possible dysfunctions that affect gait and function. Davids et al. evaluated the results of surgical lengthening of the gastrocnemius-soleus complex of 53 children with cerebral palsy in a retrospective, cohort study [26]. The mean time between the initial and postoperative follow-up study was 2 years and 3 months, while the follow-up assessment was performed between 1 and 3 years following surgery for most of the children. Significant improvements in swing phase kinematics (mean and peak ankle dorsiflexion) were recorded with three-dimensional gait analysis following surgery.

Saraph et al. evaluated the outcome of 22 children with spastic diplegia that had Baumann gastrosoleus recession, as part of a single event multilevel surgery [27]. The function of the ankle showed significant improvement, as this was assessed by clinical examination and gait analysis before and at 2 years after the operation.

Forty children with spastic diplegia were followed-up for a mean of 7.5 years after gastrocnemius recession or differential gastrocnemius-soleus complex lengthening, as part of single-event multilevel surgery [28]. There was statistically significant and clinically relevant improvement in ankle kinematics and kinetics following surgery. There was a significant improvement in gait function, as this is measured with the Gait Profile Score. The authors reported a rate of recurrence of 35 % and overcorrection of 2.5 %.

In a similar study, Dreher et al. reported on the outcomes of 44 children that had gastrocnemius-soleus recession surgery as part of a single event multi-level surgery [29]. The mean follow up assessment of these children was 8.6 years following the index procedure. The authors reported 24 % recurrence rate and 11 % overcorrection. Kinematic and kinetic data showed significant improvements that were maintained long-term. The maximum ankle power in stance phase was reduced 1 year after surgery, but returned at 3 years follow-up and remained at the 9 years follow-up. Most of the gait parameters showed a tendency for deterioration between the 1-year and 9-year assessments, although these differences did not reach statistical significance.

Svehlik et al. also reported this tendency for deterioration with long-term follow-up [30]. Eighteen children with spastic diplegia that underwent Baumann procedure to correct equinus gait were evaluated clinically and with instrumented gait analysis 10 years following their surgery. There was a significant improvement of ankle kinematics and kinetics following surgery. Despite the tendency for deterioration with time, these remained significantly improved.

Shore et al. performed a systematic review of the literature [31]. They found very poor levels of evidence. Most studies were level 4 quality of evidence, leading to only grade C recommendation. The true of recurrence and overcorrection rate is difficult to detect, since literature includes heterogeneous groups of patients with short-term follow-up. The literature indicates greater incidence of recurrent equinus in children with hemiplegia regardless of procedure, and greater incidence of overcorrection in children with diplegia, particularly following procedures on the Achilles tendon.

There are few studies comparing the different surgical techniques used to correct an equinus contracture in children with cerebral palsy. Most of them are retrospective studies, not randomized, with short follow-up and non-standardized selection criteria. The choice of surgical technique depends on the involvement of the gastrocnemius only or the whole gastrosoleus complex in the equinus deformity. Decision can be made using the Silfverskiöld's test and the instrumented gait analysis data. In children with hemiplegia, there is usually contracture of both the gastrocnemius and soleus. Percutaneous and open Achilles tendon lengthening has been shown to have equally satisfactory outcomes [25]. More distal procedures should be used in children with spastic diplegia, where only the gastrocnemius is usually contracted and the natural history is a progression to a calcaneus deformity [32]. They have a higher risk of recurrence but a low risk of overcorrection [29]. It is always easier to correct a recurrent equinus contracture than to manage a calcaneus deformity.

Equino-Cavo-Varus Deformity

Equinovarus and equinocavovarus deformities are common foot deformities in children with cerebral palsy (Fig. 40.2). The deformity consists of heel varus and equinus with midfoot adduction and supination. Cavus may be also part of the deformity, as the forefoot is plantar-flexed in relation to the hindfoot varus. These deformities are believed to be the result of the muscle overactivity of the tibialis posterior, tibialis anterior or both. This deformity is common in younger children with cerebral palsy. It usually progresses to a fixed deformity with time in children with spastic hemiplegia, while in diplegic or quadriplegic children it usually overcorrects into valgus [33].

This deformity can cause difficulties with shoe wear, brace intolerance and affect walking. As the deformity progresses and become more rigid, children stand on the lateral border of the foot. This results into painful callus formation or even fifth metatarsal stress fractures.

Orthotics can be used in flexible deformities in young children, since these are usually stable in hemiplegic children and may overcorrect into valgus in diplegic or quadriplegic patients [33]. In older children, with flexible or rigid

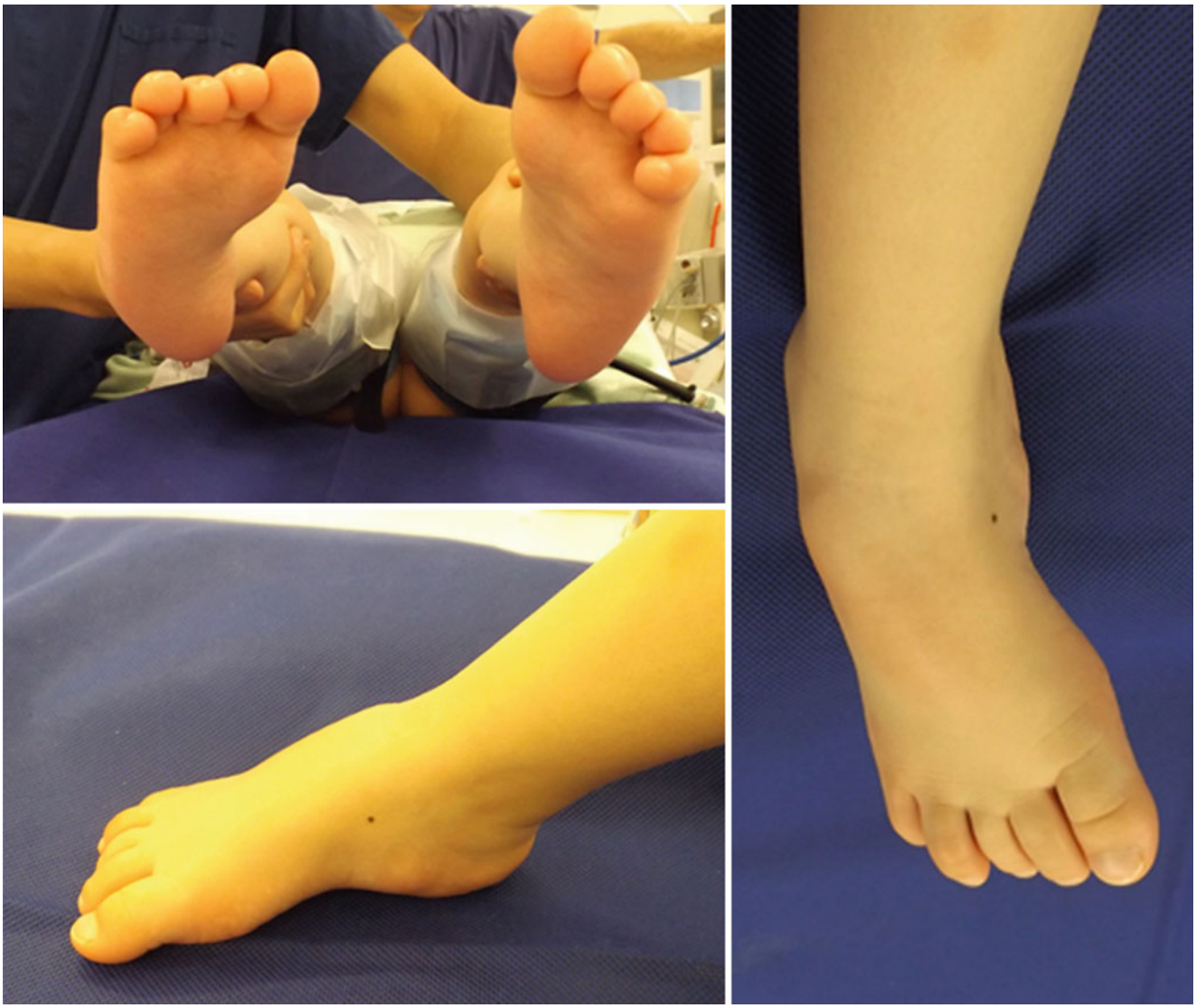

Fig. 40.2 Equino-cavo-varus deformity

deformities that affect gait, cause pain or interfere with bracing, operative treatment is usually indicated.

Does Surgical Treatment of Equinovarus Deformity Improve Gait in Ambulatory Children with Cerebral Palsy? Which Is the Best Surgical Technique?

The evidence for operative treatment of equinovarus foot deformities in children with cerebral palsy in unfortunately poor. It consists mainly of case series with short follow-up (Level 4) and review articles from experts (Level 5). There are no randomized or cohort comparative studies.

Flexible deformities are usually addressed with soft tissue surgery (soft tissue release, tendon lengthening or tendon transfer), while rigid deformities require bony surgery (osteotomies or joint fusion).

Flexible deformities can be corrected with tibialis posterior lengthening, split tibialis posterior transfer, split tibialis anterior transfer or combination of these. Tendon transfer of the whole tendon should be avoided, as there is a high risk of overcorrection, especially in diplegic and quadriplegic patients [34–36]. Lengthening of the tibialis posterior can be performed either proximally as an intramuscular lengthening or distally as Z-lengthening of the tendon [37, 38].

Split transfer of the lateral part of the tibialis anterior or posterior tendon into the lateral part of the foot is preferred [39, 40]. There is lower risk of overcorrection and the muscle does not lose its power. The choice depends on which muscle is the main contributor of the deformity. Michlitsch et al. reviewed the gait analysis data of 78 patients with cerebral palsy and varus foot deformity [41]. The varus foot deformity was a result of the dysfunction of the tibialis anterior in

34 %, tibialis posterior in 33 % and both in 31 %. In the majority of the cases varus occurred during both stance and swing phase of the gait. The timing of varus during the gait is a poor predictor of the associated muscle dysfunction. The use of dynamic electromyography and kinematic data can assist to determine the main contributing muscle to the foot deformity [41, 42].

Most of the studies demonstrate satisfactory outcomes in the short and medium term [40, 43–46]. Studies with longer follow-up show that the outcomes seem to deteriorate with time (recurrence or overcorrection). Kling et al. reported the outcomes of 37 split tibialis posterior transfer in 31 patients with spastic cerebral palsy [40]. The mean follow up of these patients was 8 years. They reported three poor results.

Eighteen children that had split tibialis posterior transfer through the interosseous membrane [47]. The mean follow up was 8.4 years. The authors reported just one poor result.

Chang et al. have reported the largest series of children with spastic equinovarus foot deformities [38]. One hundred and eight children (140 feet) with a mean follow up of 7.3 years were assessed. The surgical techniques used were split tibialis posterior tendon transfer or Z-plasty of the tendon at the level of the medial malleolus or intramuscular lengthening. Sixty-five patients were considered as failures of treatment (recurrence or overcorrection). The factors that had an effect on surgical outcome were involvement of cerebral palsy, age at operation and preoperative ambulation status. Hemiplegic patients, children older than 8 years at the time of operation and ambulatory status have more favorable outcome comparing to diplegic/quadriplegic involvement, younger children and non-ambulators. None of the surgical techniques used was found to give better outcomes.

Flexible equinovarus deformities can progress into rigid deformities with time. Fixed deformities require bony surgery; this can be in the form of osteotomies for mild deformities or arthrodesis for severe ones. Fixed hindfoot deformities can be addressed with calcaneal sliding or closing wedge osteotomy. Cavus and rigid supination deformities can be corrected with midfoot (medial cuneiform, cuboid) osteotomies or joint fusions.

For severe rigid deformities, a triple fusion provides the most reliable long-term outcomes. Tenuta et al. reported on ten patients with cerebral palsy that had triple fusion for a rigid equinovarus foot deformity [48]. The mean follow-up was 16.1 years. Nine patients had no residual deformity; three patients were dissatisfied with the outcome, six reported foot pain and five had ankle degenerative arthritis findings on x-ray. In three patients, their ambulatory capacity improved, while the others remained stable.

Satisfactory outcomes following triple fusion in adult patients with cerebral palsy were also reported by Trehan et al. [49]. Seven of the 21 patients had a foot varus deformity. The mean follow-up was 22 years. Twenty patients were satisfied with the outcome, eight patients reported pain with ambulation, 10/26 feet had residual deformity and three patients had x-ray findings of ankle arthritis (only one symptomatic).

Current evidence shows satisfactory outcomes with surgical correction of equinovarus foot deformities in children with cerebral palsy in the short and mid-term. Satisfactory outcomes should be expected in older children with hemiplegia. Gait analysis studies with the use of dynamic electromyography can help to distinguish if the tibialis posterior, anterior or both are dysfunctional. The appropriate surgical technique can be used to rebalance the foot. Fixed deformities should be corrected first with appropriate osteotomies or joint fusions. In severe rigid deformities, triple arthrodesis can provide the most reliable long-term outcomes.

Equino-Valgus and Plano-Valgus Deformity

This is probably the commonest foot deformity in children with cerebral palsy and the severity can range from a very subtle collapse of medical arch to a very severe deformity where the forefoot pointing outward (Fig. 40.3). The natural history of this deformity overlaps with the natural history of flat feet in early childhood and the progress and future developments are extremely unpredictable before 8 years old [33, 50].

Do Orthoses Improve the Outcomes of Equinovalgus Deformity in Children with Cerebral Palsy?

Tight gastrosoleus complex play an important aetiological role in the development of equinovalgus foot; hence treating this tightness may reduce the incidence and or the severity of the equinovalgus deformity. This has been covered in earlier sections and the values of orthoses have been highlighted.

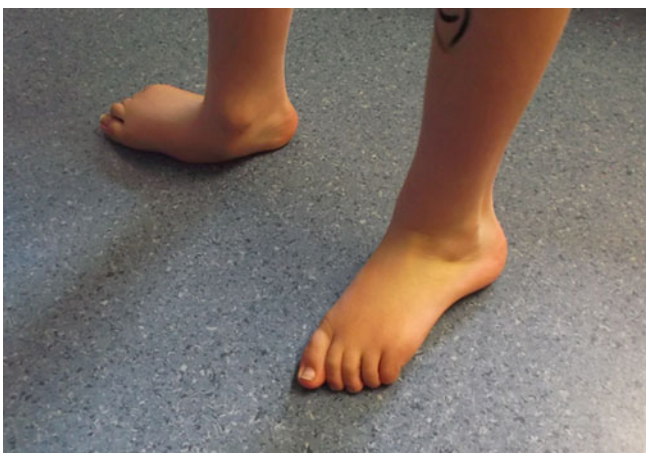

Fig. 40.3 Severe equino-plano-valgus feet deformity

Moreover, the natural history of equinovalgus foot is unpredictable in early childhood (under the age of 8), therefore temporizing interventions such as orthoses are valuable while the clinical pictures are getting clearer [33, 50].

The evidence behind orthotic treatments in plano-valgus feet in normal children was discussed in Chap. 19 and concluded that they may have detrimental effects. However, in children with cerebral palsy the contrary is true. Several studies showed that ankle and foot orthoses (AFO) improve symptoms, slow deterioration and increase gait efficiency [51–54]. However, the best type of AFO has been contested. Kadhim and Miller [55] recommended a pragmatic approach to the AFO choices in children with equinovalgus deformity admitting the lack of the scientific documentation behind such treatment. They recommend a solid AFO until the child begins to walk. Afterward, a hinged (articulated) AFO is used to allow for ankle dorsiflexion and to prevent ankle plantarflexion. Two exceptions are when the child has a severe crouch posture or severe equinovalgus deformity. In the former a solid AFO (or even GRAFO in heavier children >25 kg) is more useful to improve crouch posture. In the latter, hinged AFO is less likely to provide enough support and often causes rubbing and skin irritation. They also recommended supramalleolar AFO for children who can control ankle plantarflexion and dorsiflexion.

Does Surgical Treatment of Equinovalgus Deformity Improve Gait in Ambulatory Children with Cerebral Palsy? Which Is the Best Surgical Technique?

Surgery is indicated when AFOs fail to achieve the desired purposes of keeping the feet symptoms free, providing stability for transfer and mobility. There is a natural selection in treating this deformity when ambulatory patients (GMFCS levels I, II, III) usually present early with a less severe deformity in comparison to non ambulatory patients (GMFCS level IV and V). The aim of treatment in ambulatory patients is to restore normal alignment while preserving joints mobility. Several operations and techniques have been described using a combined soft tissue and bony surgery to restore anatomy to normal or near normal. These include calcaneal lengthening (often called lateral column lengthening) (Fig 40.4), triple C osteotomy (includes medial calcaneal slide, cuboid opening wedge, cuneiform planar closing wedge), talonavicular fusion, subtalar joint fusion and triple arthrodesis. These are often combined with soft tissue procedures such as gastrocnemius muscle lengthening, peroneus brevis tendon lengthening, and tibialis posterior advancement [50, 55–61]. There is no convincing evidence that any is superior to the others. This is not unexpected, given the spectrum of the deformity and the functional demand of patients. Although some studies tried to compare different combination of surgical interventions in treating the calcaneo-valgus feet, their comparison was undermined by the fact the two comparators were not identical.

Mosca [62] reported on 31 patients (26 were neuromuscular) with severe, symptomatic valgus deformities of the hindfoot who were corrected with a modification of the calcaneal lengthening osteotomy. Additionally, an opening-wedge osteotomy of the medial cuneiform was used to correct the deformities of both the hindfoot and the forefoot in the patients who had a skew foot. Satisfactory clinical and radiographic correction of all components of the deformity of the hindfoot was achieved in all but the two most severely deformed feet although these two feet had sufficient correction to eliminate the symptoms.

Ettl et al. [58] reviewed the outcome of calcaneal lengthening for the treatment of planovalgus foot deformity in 19 children (28 feet) with cerebral palsy. There were 14 ambulating (19 feet) and 5 non ambulating children (9 feet). They found satisfactory results in 75 % of the feet. They found no overcorrection but a relapse of the deformity in seven cases.

Sung et al. [57] showed in a study of 71 patients with cerebral palsy that for patients with greater than 23° AP talus-first metatarsal angle, 36° lateral talus-first metatarsal angle, and 72 % naviculocuboid overlap (Fig. 40.5), additional procedures for medial stabilization, such as tibialis posterior tendon reefing and talonavicular arthrodesis, should be considered as a result of the possibility of under correction with calcaneal lengthening alone.

Kim et al. [59] compared the clinical and radiographic results between calcaneal lengthening (18 patients, 28 feet) and triple C osteotomy (20 patients, 32 feet). The aetiology of the foot deformity was idiopathic in 16 feet and cerebral palsy in 44 feet. Non ambulatory patients were excluded. All operations were performed by a single surgeon. It was stated that the choice of surgery was random. In the calcaneal lengthening group, 19 of the 28 feet (68 %) showed a satisfactory outcome and 9 (32 %) an unsatisfactory outcome. In the triple C osteotomy group, 28 of the 32 feet (88 %) showed a satisfactory outcome and 4 (12 %) an unsatisfactory outcome. They concluded that triple C osteotomy is a more effective procedure than calcaneal lengthening for the correction of planovalgus deformity, especially severe pes planovalgus deformities.

Kadhim et al. [50] reported a long term follow up on 24 patients (43 feet); 15 were treated with calcaneal lengthening (mostly GMFCS levels I and II) and 28 with subtalar fusion (mostly GMFCS levels III and IV). Improvement was observed in both groups but additional surgery was required more among patients who were treated with subtalar fusion. Interestingly, residual foot pain was less common among children with poor functional abilities and patients who underwent subtalar fusion.

De Coulon et al. [60] questioned the reliability of other bony surgery in correcting severe planovalgus feet and they

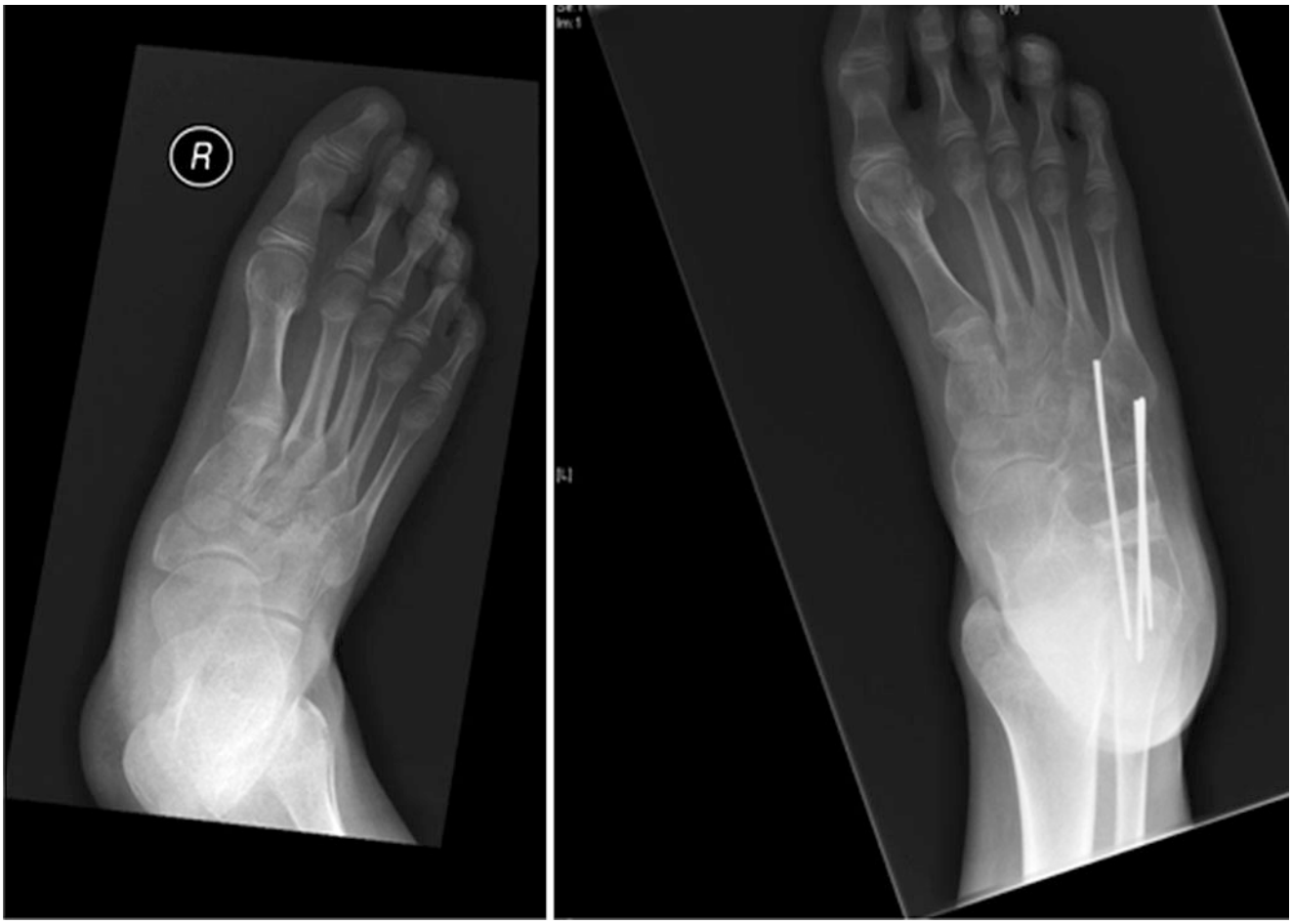

Fig. 40.4 Lateral column lengthening in a child with cerebral palsy

investigated the talo-navicular fusion which is used in adult practice but not in children practice. He reviewed 29 feet who were treated with talonavicular arthrodesis and reported satisfactory outcome in 28 feet, whereas 1 had unsatisfactory results according to the Yoo clinical outcome scoring scale.

Guven et al. [61] in a small series of 9 patient reported a 78 % satisfaction rates after Grice extra articular subtalar joint fusion in spite of the mean AOFAS hindfoot score increased by 15 points only from 53 (range 41–81) to 68.4 (range 51–96).

The above various case series indicate that restoring the anatomy and maintaining the correct anatomy is a key for a successful outcome. Having critically analyzed the above evidence and other excluded studies, it is impossible to favor one operation over other. Even in our hands, we do not use a single approach for all but rather tailor the treatment to individual patient and foot criteria. The deformity is multidimensional involving the hindfoot, midfoot and forefoot. For optimal result, a holistic approach is essential. We correct the hind foot first, then reassess the foot. There is often an associated supination deformity which requires correction. Muscles balance should then be restored to prevent recurrence. We prefer fusion surgery in severe deformity in non ambulatory children.

Foot Deformity in Children with Spina Bifida

The incidence of Spina bifida has significantly decreased in recent years (1.9/10,000 births), nonetheless it is still the cause for chronic disability in 70,000–100,000 persons in the United States with approximately 1500 new pregnancies affected per year [63, 64].

Foot deformities are extremely common in patients with Spina Bifida. Most of the children have or will develop a foot deformity whose severity is mainly related to the level of the lesion [65–67]. These deformities are particularly difficult to manage, because of the combination of motor paralysis/spasticity and sensory loss. Deformities can be present at birth in the form of Equino-Cavo-Varus (CTEV) or vertical talus, or can develop during childhood due to muscle imbalance, forces applied to the foot by weight bearing or gravity, or

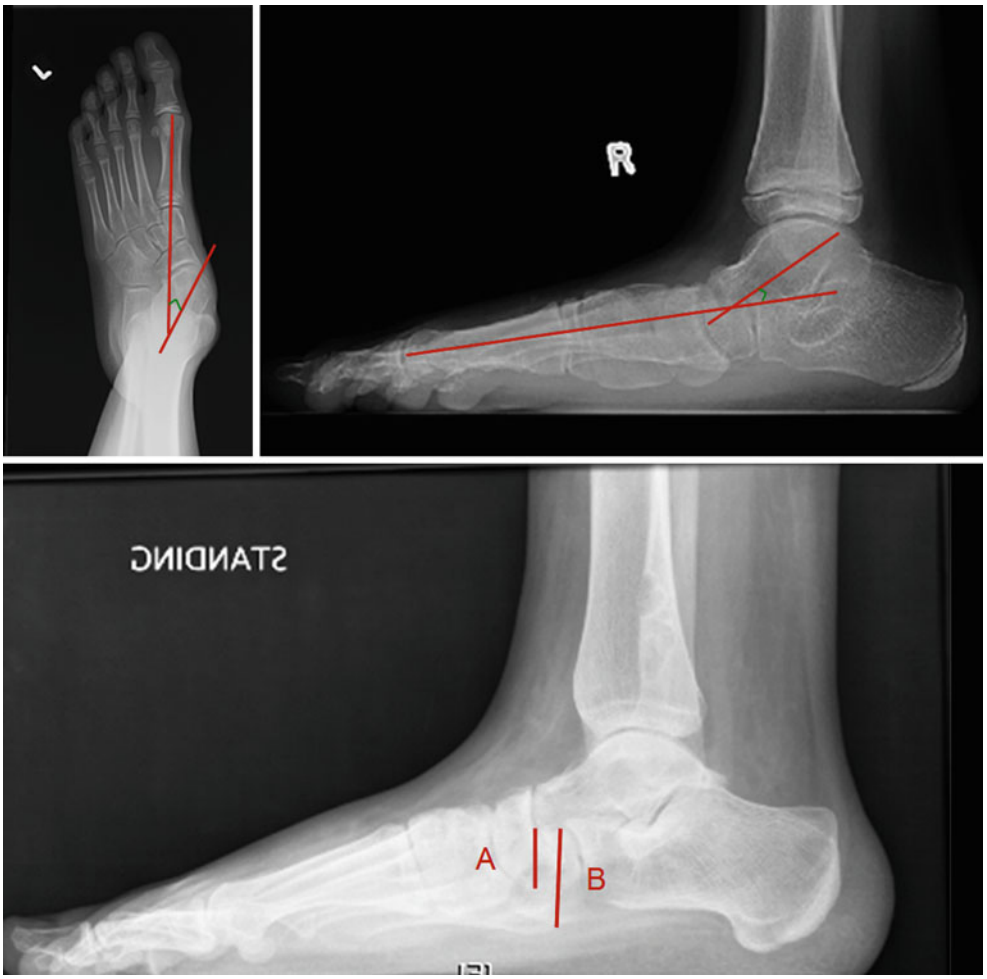

Fig. 40.5 Predictors for under correction using calcaneal lengthening alone. *Top left* image: AP talus-first metatarsal angle, *top right* image: 36° lateral talus-first metatarsal angle, and *bottom* image: naviculocuboid (A/B) which is the overlapped portion of the navicular and cuboid divided by the vertical height of the cuboid

changes in the neurological function. Nearly all these children require some sort of treatment for their foot deformity during their lives. The goal of treatment in ambulatory patients is to provide the child with a splintable, plantigrade, supple foot able to facilitate ambulation. In non-ambulatory patient, the aim of treatment is to allow satisfactory shoe wear and positioning of the foot on the wheelchair [64, 67].

What Is the Best Treatment of Foot Deformities in Spina Bifida?

Foot deformities in Spina Bifida may present as calcaneus, equinus, varus, valgus, or a combination of deformities (Fig. 40.6). Clubfoot and vertical talus are also quite common [64–67]. Early intervention with casting, bracing, or surgical treatment may prevent fixed bony deformities. Patients with spina bifida may require several corrective procedures in order to achieve a plantigrade more functional and braceable foot [64, 68]. Roach et al. have recently reviewed 84 adults with myelomeningocele and found surgeries to maintain a plantigrade foot were helpful, because even those with low-level spinal lesions and good strength had trouble ambulating on deformed feet. However, a plantigrade foot still had a substantial risk of developing pressure sores, many progressing to deep infections and occasional amputations [69].

Equinus Deformity

It has been described in all levels of involvement but it is mostly common in patients with no activity below the knee, i.e. thoracic and high lumbar level of involvement [65–67, 70, 71]. Unopposed gravity or the spastic activity of the triceps surae seems to be the leading causes for this type of deformity [65, 70].

Sharrard et al. suggested regular passive stretching combined with night-time splints starting at birth in an attempt to prevent the deformity in a flail foot. Authors noticed that an

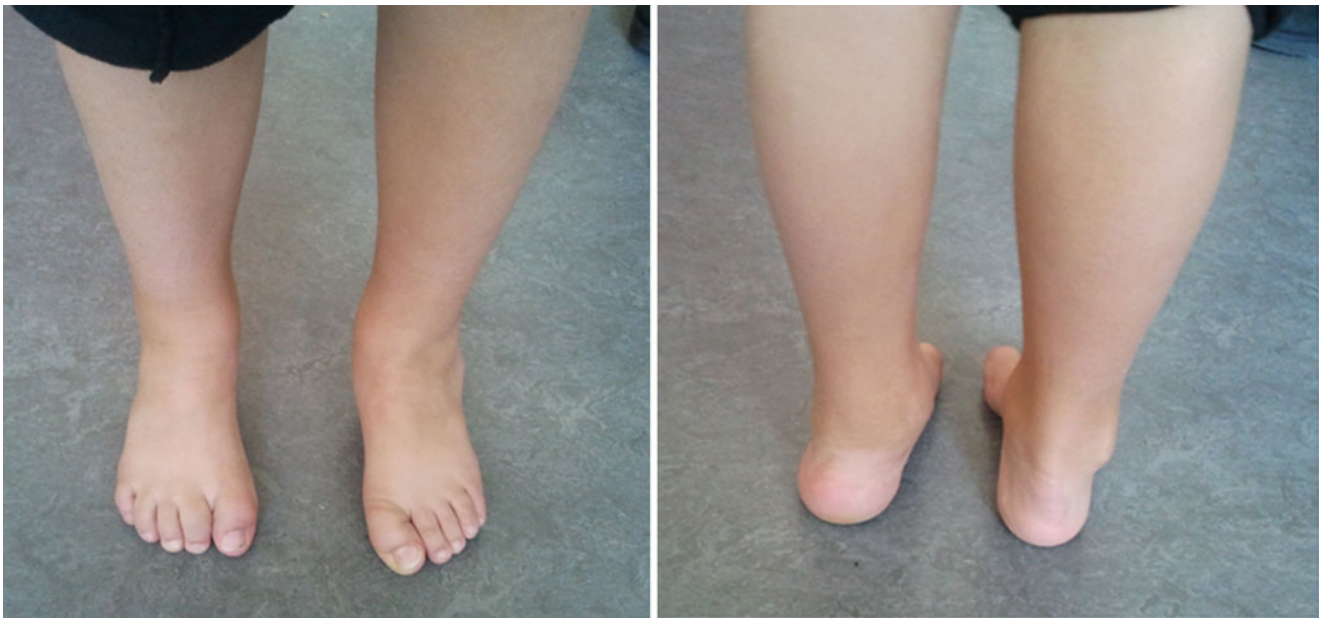

Fig. 40.6 Feet deformity in a child with spina bifida. This young boy with low level spina bifida developed right cavo-varus foot and left plano-valgus foot

equinus deformity with a short tight Achilles tendon developed if stretching was abandoned. In these cases they recommended a percutaneous Achilles tenotomy to correct the deformity and the position was kept with crepe bandage rather than cast or splints. They described rapid recurrence of the deformity in some cases, often associated with a flexion deformity of the toes due to associated shortness of the flexor hallucis longus and flexor digitorum tendons. In resistant cases, they described the split-transfer of the Achilles tendon to the dorsum of the foot, the excision of the inferior tibio-fibular ligament and the excision of the talus. In their series, Authors included 64 feet with equinus deformity, which required a total of 80 procedures (77 soft-tissue operations, 1 tendon transfer and 2 bony procedures) with an overall recurrence rate of 14 % [70].

More recently, other authors reported good results after Achilles tendon lengthening, passive stretching exercises and AFOs. Achilles tendon resection and posterior release has been described for refractory cases to achieve full correction maintained by AFOs during the day and night [64, 68, 72].

The consensus is that a regime of passive stretching combined with splints should be the first line treatment for equinus deformity in spina bifida. Surgery should be considered for an unbraceable foot if skin breakdown or positioning is a problem, or to achieve a plantigrade, braceable foot in a patient with the potential for ambulation. The type of surgical procedure depends on the severity of the deformity and should be tailored on the each patient [64, 68].

Equino-Cavo-Varus Deformity

It is the most common foot deformity in spina bifida. The incidence varies with the neurologic level of involvement; in fact, it has been reported to occur in 30–50 % of patients with sacral level involvement and up to 90 % of patients with thoracic or lumbar levels of involvement [66, 67, 70, 73, 74]. Many factors may contribute to the development of clubfoot in patients with spina bifida. Sharrard noted that the most severe presentations were seen in patients with level L4 of involvement with accompanied spasticity of the triceps surae and both tibialis muscles in combination with the functional absence of the peroneal muscles [70, 71].

Relapse rates of equino-cavo-varus deformity in spina bifida are high, ranging from 22 % to 68 %, regardless of the treatment method [70, 71, 73, 75–77].

Ponseti Method Traditionally the treatment of such a rigid and severe deformity has been extensive soft-tissue release surgery. It was common opinion that non-surgical treatment using splinting, serial casting, and stretching was unsuccessful and at considerable risk of complications including skin breakdown complicated by infection and early recurrence [70, 71]. In more recent years, the Ponseti Method consisting of manipulation and serial casting initially described for the treatment of idiopathic clubfeet [78], has been used to treat neuromuscular and syndromic clubfeet including spina bifida [75, 76, 79, 80].

Janicki et al. reported their results on the treatment of syndromic and idiopathic equino-cavo-varus feet using the

Ponseti Method with a minimum follow-up of 1 year. Five patients for a total of nine feet in their cohort, had spina bifida. All nine feet were corrected using the Ponseti Method. Mean number of casts for correction was 4.2 per foot; only two of them did not require a percutaneous Achilles tenotomy. In these nine feet, there were five recurrences two of which were treated with serial casting only and three of which needed a postero-medial release. The recurrence rate in patients with myelomeningocele was 55 % compared to 13 % in idiopathic clubfeet. Major surgical release was required in 33 % of the spina bifida feet and only in 6.4 % of the non-syndromic feet (significant difference) [76].

Gerlach et al. followed prospectively 16 consecutive patients with myelomeningocele (28 clubfeet) and twenty consecutive patients with idiopathic clubfeet (35 clubfeet) managed with the Ponseti Method. The average duration of follow-up was 34 months for the myelomeningocele group and 37 months for the idiopathic group, respectively. The deformity at presentation was significantly more severe in patients with spina bifida according to the Dimeglio system [81]. The Ponseti Method was successful in all the idiopathic feet and in 27 clubfeet (96.4 %) in the myelomeningocele group. A significant difference was observed in the relapse rate of the two groups, which was 68 % in the spina bifida group and 26 % in the idiopathic group, respectively. All the recurrences were initially treated with the Ponseti Method and only four of the clubfeet in the myelomeningocele group (14 %) and one of the clubfeet in the idiopathic group (3 %) required extensive soft tissue release (no statistical difference). Complications, including blister and iatrogenic distal tibia fractures, were significantly higher in the spina bifida group [75].

Gurnett et al. and Moroney et al. reported their results on the treatment of non-idiopathic clubfeet including spina bifida with the Ponseti method. Authors did not parse their results by specific diagnosis, but the investigators noted that the non-idiopathic clubfeet had a significantly higher relapse rate [79, 80]. Over the 5-year follow-up, Moroney et al. observed that 37 % of non-idiopathic feet required extensive surgical release compared with 2 % in the idiopathic group. Despite this significant difference, authors found the Ponseti method beneficial because it certainly determined an improvement of the deformity with a huge advantage intraoperatively [79].

The evidence available for the use of Ponseti method in the treatment of equino-cavo-varus deformity in spina bifida is fair and limited to a short follow-up.

Soft-Tissue Release

Traditionally an extensive postero-medial release, including complete tenotomies and/or excisions of tendons rather than lengthening, was the preferred treatment for equino-cavo-varus deformity in myelomeningocele. Although the outcome seems to vary with the motor level of involvement, overall, good results have been reported in 61–83 % of patients after surgical release [70, 73, 74, 82].

Sharrard and Grosfield treated 78 clubfeet with spina bifida with an "extensive medial release". In more than half the patients medial release of soft tissues alone with tendon transfer was sufficient. The overall revision rate was 22 % and some feet required up to four soft-tissue procedure (122 soft-tissue releases in total). Seven of the 52 patients developed skin breakdown, and another five progressed to superficial infection, despite the use of a V-Y incision. Authors recommended completing the correction by a calcaneal osteotomy if adequate correction of the deformity by soft-tissue release without excessive skin tension could not be obtained at the first operation, in order to avoid further soft-tissue release through scarred, contracted and adherent tissues [70].

De Carvalho Neto et al. performed a "radical postero-medial release" (PMLR) with excision of tendons, through a Cincinnati incision [83] in 63 equino-cavo-varus feet. In 21 feet, the talus was derotated in the ankle mortise with a temporary K-wire while 57 feet required a plantar fascia release through a separate plantar incision. Outcomes were good 63 %, fair in 14 %, and poor in 23 % of the feet. In 21 feet in which the talar derotation with a K-wire was performed, 76 % of the feet had a good result, 14 % fair and 10 % poor result. Fifty percent of the feet with thoracic/high lumbar level of involvement had poor results. Combining the low-lumbar and sacral-level groups, only 10 % of the feet had poor results [82].

Flynn et al. reported their outcome on 72 feet with 8 years of follow-up that had undergone a radical circumferential release. They observed 61 % good results, 26 % fair results and 13 % poor results. Authors did not observe any correlation between age at surgery, neurosegmental level and the final outcome. Twenty-six feet (36 %) required further surgery, including repeat releases, talectomies, osteotomies, and arthrodesis, with an overall success rate of 67 % [73].

Akbar et al. reviewed 167 equinovarus deformities in 123 patients with myelomeningocele who had a circumferential release through a Cincinnati incision [83]. A plantigrade foot was achieved in 83 % of the patients, with 17 % requiring a second surgical procedure. Muscular imbalance greatly affected success rates, with the highest success rates in the high- or low-level lesions (95 % success for thoracic to L2 level, 83 % for L5 to sacral level), and the least success for the mid-lumbar level lesions (64 %) [74].

Talectomy

It is considered a salvage procedure for rigid equino-cavo-varus deformity aiming to correct both equinus and heel varus deformities without tension. A successful outcome varies between 47 % and 75 % [84–86].

Sherk and Ames published their results on 11 children with spina bifida who underwent 20 talectomies for clubfoot deformities, at an average of 5.3 years of age. Their longest follow-up was only 5 years, and they noted a 25 % recurrence rate complicating brace and shoe wear due to a reduced foot height. Failure was mostly related to incomplete removal of the talus because of extensive scarring and fibrosis due to prior infection or surgery and/or severe concomitant external rotational contracture of the hips in patients with high spinal lesions [84].

In a subsequent follow-up study, Sherk et al. evaluated 19 patients with spina bifida who underwent a total of 31 talectomies between the ages of 4 and 39 years, with an average follow-up of 12 years. Authors observed that all feet were corrected enough for brace/shoe wear. Overall, 16 feet (52 %) appeared plantigrade, 14 feet (45 %) had a relapse of the deformity with different degrees of varus and equinus, and in 3 feet (10 %) the talectomy converted an equino-varus into a cavo-varus deformity. Finally, force plates analysis revealed that only 6 plantigrade feet (38 %) were actually biomechanically plantigrade with uniformly distributed ground reaction forces [85].

Trumble et al. reported their results for talectomies performed to correct resistant equino-cavo-varus deformity in 17 myelomeningocele feet at an average age of 3.5 years and an average 7-year follow-up. Correction of the hindfoot was good in 15 feet and poor in 2 feet. Severe residual forefoot deformity in 9 feet compromised remarkably the functional result of the operation and the overall success rate was only 47 % [86].

Studies on talectomies that grouped multiple diagnoses together but did not sparse the results according to the diagnosis are difficult to interpret and a definite conclusion on the outcome of the procedure on patients with spina bifida cannot be extrapolated [87–90].

Triple Fusion

The role of surgical arthrodesis in Spina Bifida is controversial. The stiffness resulting from the fusion in combination with an insensate foot can result in the development of neuropathic skin changes [91].

Olney and Menelaus reported their results of a triple arthrodesis in 18 feet with spina bifida at an average follow-up of 10 years. Each foot had multiple operations before the fusion. Fifteen of 18 feet (83 %) had a satisfactory outcome, two had recurrent planovalgus deformities and one a painful pseudarthrosis. Three feet had required revision of the triple arthrodesis, and there was one postoperative infection. No patient had lost ambulatory status because of foot problems and eight of the 10 patients who previously needed callipers were able to discard them or to use lighter ones [92].

Other Procedures: Osteotomies

Relapses after extensive soft-tissue release, very often present in the form of an adduction deformity with a relative shortened medial column and a relative elongated lateral column. For such cases, a "double osteotomy" consisting of a closing wedge osteotomy of the cuboid and an opening wedge osteotomy of the medial cuneiform similarly to those performed in patients with idiopathic clubfoot [93], has been suggested [64].

To date no publication is available on the correction of residual forefoot deformity after equino-cavo-varus deformity in patients with myelomeningocele.

Based on the current literature on equino-cavo-varus foot deformity in spina bifida, the evidence is that:

- The Ponseti method is effective at a short follow-up. The recurrence rate after Ponseti method is significantly higher when compared to idiopathic clubfeet [75, 76, 79] (Level of Evidence II/III).
- Although the outcome seems to vary with the motor level of involvement, overall, good results have been reported in 61–83 % of patients after extensive soft-tissue release [7, 10, 11, 19] (Level of Evidence IV).
- Talectomy is a salvage procedure. Successful outcomes vary between 47 % and 75 %. Relapse is not infrequent. Ground reaction force is poorly distributed leading to recurrent pressure sore [84–86] (Level of Evidence IV).
- Triple arthrodesis in a spina bifida patient is a demanding operation and may require revision, but once the deformity is corrected and a solid fusion obtained, the results do not deteriorate with time. However, the stiffness resulting from the fusion in combination with an insensate foot can result in the development of neuropathic skin changes [91, 92] (Level of Evidence IV).

Calcaneus Deformity

Calcaneus deformity occurs in approximately 17–35 % of patients with myelomeningocele and it is most common in L4 or L5 level of involvement. It is due to the strength or spasticity of the ankle dorsiflexors combined with weak or absent plantar flexors. The heel can be either in varus or in valgus. It is a progressive deformity and it leads to difficult brace and shoe wear, pressure sores and heel ulceration, and impaired gait [71, 74, 94, 95]. The gait peculiarity is the loss of toe-off, in fact, during the stance phase the heel has contact with the floor but the forefoot remains elevated with poor balance. In order to get the forefoot to contact the floor, the knees have to flex resulting in crouch gait [96].

In the early phases, the foot is not usually rigid and there is no fixed bony deformity. Early treatment consists of passive stretching, serial casting and bracing aiming to prevent a

severe rigid calcaneus foot. Once the child becomes old enough, early surgical intervention has been recommended because prolonged weight bearing may lead to calcaneovalgus deformities and external tibial torsion. The goal of surgery is to prevent a rigid deformity by eliminating the deforming forces, possibly augmenting the ankle plantarflexors, and balancing the force acting on the foot [94, 95, 97, 98].

Tendon Transfers

Transfer of tendons to restore, or at least improve, the poor ankle plantarflexion are the most commonly described procedures, usually a transfer of the anterior tibialis posteriorly through the interosseous membrane to the calcaneal tuberosity.

Bliss and Menelaus reviewed 25 patients with calcaneus deformity who had had 46 anterior tibialis transfers at an average follow-up age of 22 years. Only 10 patients had a plantigrade foot with no residual deformity and did not require any further surgery. The remaining 36 patients required a total of 66 surgeries (40 bony procedures). At the last follow-up, five of them had a plantigrade foot, 25 patients had a recurrence of the deformity with associated valgus, while in six patients the transfer never proved to work. The overall success rate in this series was 22 % [99].

Janda et al. reviewed 12 feet of six patients who had undergone transfers and found that 50 % of the feet had non-functioning transfers but they were plantigrade.

Paradoxically, of the six functioning transfers, five developed secondary deformities (planovalgus, equinovalgus, equinovarus), and three of them required surgical correction. The overall success rate was 50 % and Authors did not find any correlation between age at time of surgery and outcome [100].

Fraser et al. reported their experience of tibialis anterior transfer for calcaneus deformity in 46 feet of 26 ambulant patients with myelomeningocele. At an average follow-up of 8.4 years, 89 % of the patients had satisfactory results and 64 % of them were able to stand on their toes. Pre-operative trophic ulceration of the heel increased from 3.2 % to 33 % if surgery was delayed. Secondary deformities developed in 76 % of feet. In total 33 of the 46 feet required subsequent operations with an actual success rate of 28 %. Authors could not find any correlation between age at the time of surgery and outcome. The neurological level seemed to affect the outcome; in fact 80 % of patient with neurological level L5 or S1 results were good, while 90 % of patient with L5 level of involvement had poor results [101].

The above studies show a poor correlation between the correction of the calcaneus deformity and a successful transfer. Subsequent dynamic electromyographic studies found a continuous anterior tibialis activity throughout the gait cycle. This electromyographic pattern likely represents a spastic muscle, which acts primarily as a tenodesis and cannot prevent excessive ankle dorsiflexion [100, 102, 103]. Authors have therefore postulated that it would be better to transect the tendon rather than transfer it [40, 41], and they all recommended continued use of postoperative orthotic support [100, 102, 103].

Bradley et al. suggested that the anterior tibialis tendon is simply not strong enough to replace the triceps surae [104]. Other authors augmented the transfer of the tibialis anterior with other procedures reporting better outcomes.

Wenz et al. transferred tibialis anterior and posterior, peroneus brevis and longus, extensor digitorum and hallucis longus to the Achilles but also performed the equivalent of a triple arthrodesis (Inverse Lambrinudi) which elevated the talus to function as a bone block against the anterior distal tibia, limiting dorsiflexion. They reported the outcome of nine patients, average age 17.5 years, at a follow-up of 32 months. Eight out if the nine patients could walk without splints. Only in 2 patients they observed an overcorrection of the deformity that required further treatment [105].

Georgiadis et al. [106] and Park et al. [95] showed that adjunctive procedure like further tendon transfers/releases, osteotomies and or arthrodesis aiming to correct all the deforming forces and residual deformities improved the outcome remarkably. In fact, both authors observed no deformity recurrence or progression, or development of secondary deformities and an overall success rate of 95 % and 100 % respectively at an average follow-up of 6 and 4 years, respectively [95, 106].

Soft-Tissue Release

Rodrigues and Dias have described a complete anterolateral release of the ankle including sectioning the ankle dorsiflexors, toe extensors, the peroneus brevis and longus. They reported their result on 76 patients. Sixty-two had a plantigrade and braceable foot with a success rate of 82 %. The remaining 14 that failed required further treatment, i.e. six required a repeat procedure, and eight underwent Achilles release for equinus deformity [94].

Swaroop and Dias recently recommended to address long-standing rigid deformities with an anterolateral release, combined with a posteriorly based closing wedge osteotomy of the calcaneal tuberosity to shift the tuberosity more posterior and proximal [64]. These procedures have been described for patients with poliomyelitis but no data is yet available on patients with myelomeningocele [107, 108].

Based on the current literature on calcaneus foot deformity in spina bifida, the evidence is that

- Anterior tibialis tendon transfer is successful in 22–64 % of cases but there is a poor correlation between the correction of the calcaneus deformity and a successful transfer.

Orthotic support after surgery is always recommended [99–103] (Level of Evidence IV).

- Adjunctive procedure (tendon transfers/releases, osteotomies and or arthrodesis) aiming to correct the residual deformities can increase the overall success rate up to 95–100 % [95, 106] (Level of Evidence IV).
- A complete anterolateral release of the ankle including sectioning the ankle dorsiflexors, toe extensors, the peroneus brevis and longus is successful in 82 % of the patients [94] (Level of Evidence IV).

Vertical Talus

Congenital vertical talus is a rare but complex foot deformity in children with spina bifida. It has been reported in 4–5 % of children with spina bifida and represents 10 % of foot deformities in these patients [65, 66, 70].

It can present as a congenital deformity, more commonly, or a developmental deformity. Drennan and Sharrard in pathological anatomy study concluded that it is the result of the imbalance between a weak tibialis posterior and strong dorsiflexors and evertors [109].

The mainstay of treatment is operative. As with other foot deformities in spina bifida patients, the goal of treatment is a plantigrade and splintable foot. The best evidence available includes small case series of patients with idiopathic vertical talus and other syndromic conditions, including spina bifida, with short-term follow-up.

Sharrard and Grosfield used different soft tissue and bony procedures (soft tissue release on the outer side of the foot, reduction of the talonavicular dislocation and fixation with a Kirschner wire, transplantation of dorsiflexors and peroneal tendons to restore muscle balance, calcaneal osteotomy, extra-articular subtalar fusion) to correct this deformity with a high failure rate (37 %) [70].

Single or two-stage soft tissue release approaches have been described. In the first part of the two-stage approach the contracted dorsolateral tendons and joint capsules are released and talonavicular and subtalar are reduced. The second stage consists of lengthening the Achilles and peroneal tendons and a release of the posterolateral joint capsule release [110]. The authors of this technique reported satisfactory outcomes in 12/15 feet treated. Among these patient there was a child with spina bifida.

Kodros and Dias reported on the result of a single stage approach using the Cincinnati incision [111]. The cohort of patients included 32 patients (18 patients with neural tube defects – 21 feet) with a follow-up of 7 years. The results in the patients with neural tube defects were good in 18 feet and fair in 3 feet. There were no excellent or poor results. On patient developed equinovarus deformity and two a cavus deformity.

Mazzocca et al. compared two approaches, one dorsal and one posteromedial more extensive [112]. They concluded that the dorsal approach provided better correction with fewer complications.

A minimally invasive approach has been described by Dobbs et al. [113, 114]. This consists of serial manipulation and casting followed by temporary stabilization of the talonavicular joint by K-wire and an Achilles tenotomy. The results of this approach were compared with the traditional more extensive soft tissue release [115]. All patients had minimum follow-up of 5 years (mean 7 years). The minimally invasive treatment method resulted in better ankle range of motion and pain scores. The good outcomes using this approach have been reported by other centers [116, 117].

The cohort of the previous studies included children with both idiopathic congenital vertical talus and syndromic (including spina bifida). Chalayon et al. reported on the outcomes of 15 patients (25 feet) with non-idiopathic deformities (4 with spina bifida) [118]. All of the myelomeningocele patients were successfully corrected without recurrences. In the short-term follow-up, the results were excellent in four patients, good in ten and fair in one. In this cohort of patients, the percentage of feet that achieved full reduction of the talonavicular joint with casting alone was smaller than in the groups of patients with idiopathic congenital vertical talus. Many patients required release of the talonavicular joint and the anterior aspect of the subtalar joint.

In a prospective cohort study, the outcomes of the minimally invasive technique were compared between children with idiopathic and syndromic congenital vertical talus [119]. The authors reported higher relapse rates (10/21 feet) at a mean follow up of 36 months. No statistically significant differences between the two groups were found (recurrence rate, pain/comfort score, correction score), despite the recurrence rate of the syndromic group of patients was higher (67 % vs 33 %).

In older children, the deformity becomes rigid. Surgery that is more extensive is required to correct these deformities. Combination of soft tissue release with bony surgery is required. This can be medial column shortening with lateral column lengthening, extra-articular subtalar arthrodesis and triple arthrodesis [120, 121, 92].

- A minimally invasive technique, as described by Dobbs, gives promising and replicable good results in the short term. The risk of recurrence seems higher comparing to idiopathic vertical talus. (Level of Evidence IV)
- In older children, a single stage dorsal approach of soft tissue releases can give good outcomes in the mid-term. (Level of Evidence IV)
- A triple arthrodesis can be used as a salvage procedure. (Level of Evidence IV)

Hindfoot Valgus

Hindfoot valgus deformity in children with spina bifida develops as the child matures and starts to ambulate [122]. Mild and supple deformities can be controlled with rigid ankle-foot orthosis. As the deformity progresses it can cause skin problems, difficulties with bracing or pain. Appropriate clinical and radiologic assessment to distinguish hindfoot valgus from ankle valgus is important.

Hindfoot valgus can be corrected with a calcaneal sliding osteotomy. Parsons et al. reported unsatisfactory outcomes with sliding or medial closing wedge osteotomy [123]. Eight out of ten feet of seven patients had recurrence of the deformity at 6.5 years of follow-up. Good outcomes were reported only in patients that had previous extra-articular subtalar arthrodesis.

Torosian et al. reported more favorable outcomes using the calcaneal sliding osteotomy in 38 feet of 27 patients with spina bifida [122]. Good results were reported in 31 feet (82 %) with more than 5 years of follow-up.

Extra-articular subtalar or triple fusion has been used to correct this deformity in children with spina bifida [92, 124]. These should be used as salvage procedures only. A rigid and insensate foot is at risk of developing skin ulceration and breakdown [64, 91].

References

1. Rosenbaum P, Paneth N, Leviton A, Goldstein M, Bax M, Damiano D, et al. A report: the definition and classification of cerebral palsy April 2006. Dev Med Child Neurol Suppl. 2007;109:8–14.
2. Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Dev Med Child Neurol. 1997;39(4):214–23.
3. Morris C, Bartlett D. Gross motor function classification system: impact and utility. Dev Med Child Neurol. 2004;46(1):60–5.
4. Mosca VS. The child's foot: principles of management. J Pediatr Orthop. 1998;18(3):281–2.
5. O'Connell PA, D'Souza L, Dudeney S, Stephens M. Foot deformities in children with cerebral palsy. J Pediatr Orthop. 1998; 18(6):743–7.
6. Aoki KR, Guyer B. Botulinum toxin type A and other botulinum toxin serotypes: a comparative review of biochemical and pharmacological actions. Eur J Neurol. 2001;8(Suppl 5):21–9.
7. Placzek R, Siebold D, Funk JF. Development of treatment concepts for the use of botulinum toxin a in children with cerebral palsy. Toxins (Basel). 2010 Sep;2(9):2258–71.
8. Koman LA, Mooney JF, Smith 3rd BP, Goodman A, Mulvaney T. Management of spasticity in cerebral palsy with botulinum-A toxin: report of a preliminary, randomized, double-blind trial. J Pediatr Orthop. 1994;14(3):299–303.
9. Koman LA, Brashear A, Rosenfeld S, Chambers H, Russman B, Rang M, et al. Botulinum toxin type a neuromuscular blockade in the treatment of equinus foot deformity in cerebral palsy: a multicenter, open-label clinical trial. Pediatrics. 2001;108(5):1062–71.
10. Sutherland DH, Kaufman KR, Wyatt MP, Chambers HG, Mubarak SJ. Double-blind study of botulinum A toxin injections into the gastrocnemius muscle in patients with cerebral palsy. Gait Posture. 1999;10(1):1–9.
11. Love SC, Valentine JP, Blair EM, Price CJ, Cole JH, Chauvel PJ. The effect of botulinum toxin type A on the functional ability of the child with spastic hemiplegia a randomized controlled trial. Eur J Neurol. 2001;8(Suppl 5):50–8.
12. Bjornson K, Hays R, Graubert C, Price R, Won F, McLaughlin JF, et al. Botulinum toxin for spasticity in children with cerebral palsy: a comprehensive evaluation. Pediatrics. 2007;120(1): 49–58.
13. Ubhi T, Bhakta BB, Ives HL, Allgar V, Roussounis SH. Randomised double blind placebo controlled trial of the effect of botulinum toxin on walking in cerebral palsy. Arch Dis Child. 2000; 83(6):481–7.
14. El-Etribi MA, Salem ME, El-Shakankiry HM, El-Kahky AM, El-Mahboub SM. The effect of botulinum toxin type-A injection on spasticity, range of motion and gait patterns in children with spastic diplegic cerebral palsy: an Egyptian study. Int J Rehabil Res. 2004;27(4):275–81.
15. Bottos M, Benedetti MG, Salucci P, Gasparroni V, Giannini S. Botulinum toxin with and without casting in ambulant children with spastic diplegia: a clinical and functional assessment. Dev Med Child Neurol. 2003;45(11):758–62.
16. Ackman JD, Russman BS, Thomas SS, Buckon CE, Sussman MD, Masso P, et al. Comparing botulinum toxin A with casting for treatment of dynamic equinus in children with cerebral palsy. Dev Med Child Neurol. 2005;47(9):620–7.
17. Park EY, Kim WH. Meta-analysis of the effect of strengthening interventions in individuals with cerebral palsy. Res Dev Disabil. 2014;35(2):239–49.
18. Love SC, Novak I, Kentish M, Desloovere K, Heinen F, Molenaers G, et al. Botulinum toxin assessment, intervention and after-care for lower limb spasticity in children with cerebral palsy: international consensus statement. Eur J Neurol. 2010;17(Suppl 2): 9–37.
19. Dumas HM, O'Neil ME, Fragala MA. Expert consensus on physical therapist intervention after botulinum toxin a injection for children with cerebral palsy. Pediatr Phys Ther. 2001;13(3):122–32.
20. Boyd RNHR, Bach T, Morris M. Botulinum toxin A (BTX-A) for the management of equinus gait–a meta-analysis of randomized clinical trials. Gait Posture. 2002;16(suppl 1):123.
21. Cardoso ES, Rodrigues BM, Barroso M, Menezes CJ, Lucena RS, Nora DB, et al. Botulinum toxin type A for the treatment of the spastic equinus foot in cerebral palsy. Pediatr Neurol. 2006; 34(2):106–9.
22. Koog YH, Min BI. Effects of botulinum toxin A on calf muscles in children with cerebral palsy: a systematic review. Clin Rehabil. 2010;24(8):685–700.
23. Joo SY, Knowtharapu DN, Rogers KJ, Holmes Jr L, Miller F. Recurrence after surgery for equinus foot deformity in children with cerebral palsy: assessment of predisposing factors for recurrence in a long-term follow-up study. J Child Orthop. 2011;5(4):289–96.
24. Sharrard WJ, Bernstein S. Equinus deformity in cerebral palsy. A comparison between elongation of the tendo calcaneus and gastrocnemius recession. J Bone Joint Surg Br. 1972;54(2):272–6.
25. Borton DC, Walker K, Pirpiris M, Nattrass GR, Graham HK. Isolated calf lengthening in cerebral palsy. Outcome analysis of risk factors. J Bone Joint Surg Br. 2001;83(3):364–70.
26. Davids JR, Rogozinski BM, Hardin JW, Davis RB. Ankle dorsiflexor function after plantar flexor surgery in children with cerebral palsy. J Bone Joint Surg Am. 2011;93(23):e1381–7.
27. Saraph V, Zwick EB, Uitz C, Linhart W, Steinwender G. The Baumann procedure for fixed contracture of the gastrosoleus in

cerebral palsy. Evaluation of function of the ankle after multilevel surgery. J Bone Joint Surg Br. 2000;82(4):535–40.
28. Firth GB, Passmore E, Sangeux M, Thomason P, Rodda J, Donath S, et al. Multilevel surgery for equinus gait in children with spastic diplegic cerebral palsy: medium-term follow-up with gait analysis. J Bone Joint Surg Am. 2013;95(10):931–8.
29. Dreher T, Buccoliero T, Wolf SI, Heitzmann D, Gantz S, Braatz F, et al. Long-term results after gastrocnemius-soleus intramuscular aponeurotic recession as a part of multilevel surgery in spastic diplegic cerebral palsy. J Bone Joint Surg Am. 2012;94(7): 627–37.
30. Svehlik M, Kraus T, Steinwender G, Zwick EB, Saraph V, Linhart WE. The Baumann procedure to correct equinus gait in children with diplegic cerebral palsy: long-term results. J Bone Joint Surg Br. 2012;94(8):1143–7.
31. Shore BJ, White N, Kerr GH. Surgical correction of equinus deformity in children with cerebral palsy: a systematic review. J Child Orthop. 2010;4(4):277–90.
32. Segal LS, Thomas SE, Mazur JM, Mauterer M. Calcaneal gait in spastic diplegia after heel cord lengthening: a study with gait analysis. J Pediatr Orthop. 1989;9(6):697–701.
33. Sees JP, Miller F. Overview of foot deformity management in children with cerebral palsy. J Child Orthop. 2013;7(5):373–7.
34. Root L. Tendon surgery on the feet of children with cerebral palsy. Dev Med Child Neurol. 1976;18(5):671–4.
35. Bisla RS, Louis HJ, Albano P. Transfer of tibialis posterior tendon in cerebral palsy. J Bone Joint Surg Am. 1976;58(4):497–500.
36. Schneider M, Balon K. Deformity of the foot following anterior transfer of the posterior tibial tendon and lengthening of the Achilles tendon for spastic equinovarus. Clin Orthop Relat Res. 1977;125:113–8.
37. Ruda R, Frost HM. Cerebral palsy. Spastic varus and forefoot adductus, treated by intramuscular posterior tibial tendon lengthening. Clin Orthop Relat Res. 1971;79:61–70.
38. Chang CH, Albarracin JP, Lipton GE, Miller F. Long-term follow-up of surgery for equinovarus foot deformity in children with cerebral palsy. J Pediatr Orthop. 2002;22(6):792–9.
39. Hoffer MM, Reiswig JA, Garrett AM, Perry J. The split anterior tibial tendon transfer in the treatment of spastic varus hindfoot of childhood. Orthop Clin North Am. 1974;5(1):31–8.
40. Kling Jr TF, Kaufer H, Hensinger RN. Split posterior tibial-tendon transfers in children with cerebral spastic paralysis and equinovarus deformity. J Bone Joint Surg Am. 1985;67(2):186–94.
41. Michlitsch MG, Rethlefsen SA, Kay RM. The contributions of anterior and posterior tibialis dysfunction to varus foot deformity in patients with cerebral palsy. J Bone Joint Surg Am. 2006; 88(8):1764–8.
42. Scott AC, Scarborough N. The use of dynamic EMG in predicting the outcome of split posterior tibial tendon transfers in spastic hemiplegia. J Pediatr Orthop. 2006;26(6):777–80.
43. Green NE, Griffin PP, Shiavi R. Split posterior tibial-tendon transfer in spastic cerebral palsy. J Bone Joint Surg Am. 1983;65(6): 748–54.
44. O'Byrne JM, Kennedy A, Jenkinson A, O'Brien TM. Split tibialis posterior tendon transfer in the treatment of spastic equinovarus foot. J Pediatr Orthop. 1997;17(4):481–5.
45. Barnes MJ, Herring JA. Combined split anterior tibial-tendon transfer and intramuscular lengthening of the posterior tibial tendon. Results in patients who have a varus deformity of the foot due to spastic cerebral palsy. J Bone Joint Surg Am. 1991;73(5): 734–8.
46. Synder M, Kumar SJ, Stecyk MD. Split tibialis posterior tendon transfer and tendo-Achillis lengthening for spastic equinovarus feet. J Pediatr Orthop. 1993;13(1):20–3.
47. Saji MJ, Upadhyay SS, Hsu LC, Leong JC. Split tibialis posterior transfer for equinovarus deformity in cerebral palsy. Long-term results of a new surgical procedure. J Bone Joint Surg Br. 1993;75(3):498–501.
48. Tenuta J, Shelton YA, Miller F. Long-term follow-up of triple arthrodesis in patients with cerebral palsy. J Pediatr Orthop. 1993;13(6):713–6.
49. Trehan SK, Ihekweazu UN, Root L. Long-term outcomes of triple arthrodesis in cerebral palsy patients. J Pediatr Orthop. 2015; 35(7):751–5.
50. Kadhim M, Holmes Jr L, Miller F. Long-term outcome of planovalgus foot surgical correction in children with cerebral palsy. J Foot Ankle Surg. 2013;52(6):697–703.
51. Danino B, Erel S, Kfir M, Khamis S, Batt R, Hemo Y, et al. Influence of orthosis on the foot progression angle in children with spastic cerebral palsy. Gait Posture. 2015;42(4):518–22.
52. Danino B, Erel S, Kfir M, Khamis S, Batt R, Hemo Y, et al. Are gait indices sensitive enough to reflect the effect of ankle foot orthosis on gait impairment in cerebral palsy diplegic patients? J Pediatr Orthop. 2016;36(3):294–8.
53. Ries AJ, Novacheck TF, Schwartz MH. The efficacy of ankle-foot orthoses on improving the gait of children with diplegic cerebral palsy: a multiple outcome analysis. PM R. 2015;7(9):922–9.
54. Figueiredo EM, Ferreira GB, Maia Moreira RC, Kirkwood RN, Fetters L. Efficacy of ankle-foot orthoses on gait of children with cerebral palsy: systematic review of literature. Pediatr Phys Ther. 2008;20(3):207–23.
55. Kadhim M, Miller F. Pes planovalgus deformity in children with cerebral palsy: review article. J Pediatr Orthop B. 2014;23(5): 400–5.
56. Horton GA, Olney BW. Triple arthrodesis with lateral column lengthening for treatment of severe planovalgus deformity. Foot Ankle Int. 1995;16(7):395–400.
57. Sung KH, Chung CY, Lee KM, Lee SY, Park MS. Calcaneal lengthening for planovalgus foot deformity in patients with cerebral palsy. Clin Orthop Relat Res. 2013;471(5):1682–90.
58. Ettl V, Wollmerstedt N, Kirschner S, Morrison R, Pasold E, Raab P. Calcaneal lengthening for planovalgus deformity in children with cerebral palsy. Foot Ankle Int. 2009;30(5): 398–404.
59. Kim JR, Shin SJ, Wang SI, Kang SM. Comparison of lateral opening wedge calcaneal osteotomy and medial calcaneal sliding-opening wedge cuboid-closing wedge cuneiform osteotomy for correction of planovalgus foot deformity in children. J Foot Ankle Surg. 2013;52(2):162–6.
60. de Coulon G, Turcot K, Canavese F, Dayer R, Kaelin A, Ceroni D. Talonavicular arthrodesis for the treatment of neurological flat foot deformity in pediatric patients: clinical and radiographic evaluation of 29 feet. J Pediatr Orthop. 2011;31(5):557–63.
61. Guven M, Eren A, Akman B, Unay K, Ozkan NK. The results of the Grice subtalar extra-articular arthrodesis for pes planovalgus deformity in patients with cerebral palsy. Acta Orthop Traumatol Turc. 2008;42(1):31–7.
62. Mosca VS. Calcaneal lengthening for valgus deformity of the hindfoot. Results in children who had severe, symptomatic flatfoot and skewfoot. J Bone Joint Surg Am. 1995;77(4):500–12.
63. Swaroop VT, Dias L. Orthopedic management of spina bifida. Part I: hip, knee, and rotational deformities. J Child Orthop. 2009;3(6):441–9.
64. Swaroop VT, Dias L. Orthopaedic management of spina bifida-part II: foot and ankle deformities. J Child Orthop. 2011;5(6): 403–14.
65. Broughton NS, Graham G, Menelaus MB. The high incidence of foot deformity in patients with high-level spina bifida. J Bone Joint Surg Br. 1994;76(4):548–50.
66. Frawley PA, Broughton NS, Menelaus MB. Incidence and type of hindfoot deformities in patients with low-level spina bifida. J Pediatr Orthop. 1998;18(3):312–3.

67. Frischhut B, Stockl B, Landauer F, Krismer M, Menardi G. Foot deformities in adolescents and young adults with spina bifida. J Pediatr Orthop B. 2000;9(3):161–9.
68. van Bosse HJ. Syndromic feet: arthrogryposis and myelomeningocele. Foot Ankle Clin. 2015;20(4):619–44.
69. Roach JW, Short BF, Saltzman HM. Adult consequences of spina bifida: a cohort study. Clin Orthop Relat Res. 2011;469(5): 1246–52.
70. Sharrard WJ, Grosfield I. The management of deformity and paralysis of the foot in myelomeningocele. J Bone Joint Surg Br. 1968;50(3):456–65.
71. Sharrard WJ. The orthopaedic surgery of spina bifida. Clin Orthop Relat Res. 1973;92:195–213.
72. Hayes JT, Gross HP, Dow S. Surgery for paralytic defects secondary to myelomeningocele and myelodysplasia. J Bone Joint Surg Am. 1964;46:1577–97.
73. Flynn JM, Herrera-Soto JA, Ramirez NF, Fernandez-Feliberti R, Vilella F, Guzman J. Clubfoot release in myelodysplasia. J Pediatr Orthop B. 2004;13(4):259–62.
74. Akbar M, Bresch B, Seyler TM, Wenz W, Bruckner T, Abel R, et al. Management of orthopaedic sequelae of congenital spinal disorders. J Bone Joint Surg Am. 2009;91(Suppl 6):87–100.
75. Gerlach DJ, Gurnett CA, Limpaphayom N, Alaee F, Zhang Z, Porter K, et al. Early results of the Ponseti method for the treatment of clubfoot associated with myelomeningocele. J Bone Joint Surg Am. 2009;91(6):1350–9.
76. Janicki JA, Narayanan UG, Harvey B, Roy A, Ramseier LE, Wright JG. Treatment of neuromuscular and syndrome-associated (nonidiopathic) clubfeet using the Ponseti method. J Pediatr Orthop. 2009;29(4):393–7.
77. Segev E, Ezra E, Yaniv M, Wientroub S, Hemo Y. V osteotomy and Ilizarov technique for residual idiopathic or neurogenic clubfeet. J Orthop Surg (Hong Kong). 2008;16(2):215–9.
78. Ponseti IV, Smoley EN. Congenital club foot: the results of treatment. J Bone Joint Surg Am. 1963;45(2):261–344.
79. Moroney PJ, Noel J, Fogarty EE, Kelly PM. A single-center prospective evaluation of the Ponseti method in nonidiopathic congenital talipes equinovarus. J Pediatr Orthop. 2012;32(6):636–40.
80. Gurnett CA, Boehm S, Connolly A, Reimschisel T, Dobbs MB. Impact of congenital talipes equinovarus etiology on treatment outcomes. Dev Med Child Neurol. 2008;50(7):498–502.
81. Dimeglio A, Bonnet F, Mazeau P, De Rosa V. Orthopaedic treatment and passive motion machine: consequences for the surgical treatment of clubfoot. J Pediatr Orthop B. 1996;5(3): 173–80.
82. de Carvalho NJ, Dias LS, Gabrieli AP. Congenital talipes equinovarus in spina bifida: treatment and results. J Pediatr Orthop. 1996;16(6):782–5.
83. Crawford AH, Marxen JL, Osterfeld DL. The Cincinnati incision: a comprehensive approach for surgical procedures of the foot and ankle in childhood. J Bone Joint Surg Am. 1982;64(9):1355–8.
84. Sherk HH, Ames MD. Talectomy in the treatment of the myelomeningocele patient. Clin Orthop Relat Res. 1975;110:218–22.
85. Sherk HH, Marchinski LJ, Clancy M, Melchonni J. Ground reaction forces on the plantar surface of the foot after talectomy in the myelomeningocele. J Pediatr Orthop. 1989;9(3):269–75.
86. Trumble T, Banta JV, Raycroft JF, Curtis BH. Talectomy for equinovarus deformity in myelodysplasia. J Bone Joint Surg Am. 1985;67(1):21–9.
87. Menelaus MB. Talectomy for equinovarus deformity in arthrogryposis and spina bifida. J Bone Joint Surg Br. 1971;53(3): 468–73.
88. Legaspi J, Li YH, Chow W, Leong JC. Talectomy in patients with recurrent deformity in club foot. A long-term follow-up study. J Bone Joint Surg Br. 2001;83(3):384–7.
89. Dias LS, Stern LS. Talectomy in the treatment of resistant talipes equinovarus deformity in myelomeningocele and arthrogryposis. J Pediatr Orthop. 1987;7(1):39–41.
90. Cooper RR, Capello W. Talectomy. A long-term follow-up evaluation. Clin Orthop Relat Res. 1985;201:32–5.
91. Maynard MJ, Weiner LS, Burke SW. Neuropathic foot ulceration in patients with myelodysplasia. J Pediatr Orthop. 1992;12(6): 786–8.
92. Olney BW, Menelaus MB. Triple arthrodesis of the foot in spina bifida patients. J Bone Joint Surg Br. 1988;70(2):234–5.
93. Lourenco AF, Dias LS, Zoellick DM, Sodre H. Treatment of residual adduction deformity in clubfoot: the double osteotomy. J Pediatr Orthop. 2001;21(6):713–8.
94. Rodrigues RC, Dias LS. Calcaneus deformity in spina bifida: results of anterolateral release. J Pediatr Orthop. 1992;12(4): 461–4.
95. Park KB, Park HW, Joo SY, Kim HW. Surgical treatment of calcaneal deformity in a select group of patients with myelomeningocele. J Bone Joint Surg Am. 2008;90(10):2149–59.
96. Galli M, Albertini G, Romei M, Santambrogio GC, Tenore N, Crivellini M. Gait analysis in children affected by myelomeningocele: comparison of the various levels of lesion. Funct Neurol. 2002;17(4):203–10.
97. Dias LS. Ankle valgus in children with myelomeningocele. Dev Med Child Neurol. 1978;20(5):627–33.
98. Dias LS. Valgus deformity of the ankle joint: pathogenesis of fibular shortening. J Pediatr Orthop. 1985;5(2):176–80.
99. Bliss DG, Menelaus MB. The results of transfer of the tibialis anterior to the heel in patients who have a myelomeningocele. J Bone Joint Surg Am. 1986;68(8):1258–64.
100. Janda JP, Skinner SR, Barto PS. Posterior transfer of tibialis anterior in low-level myelodysplasia. Dev Med Child Neurol. 1984;26(1):100–3.
101. Fraser RK, Hoffman EB. Calcaneus deformity in the ambulant patient with myelomeningocele. J Bone Joint Surg Br. 1991; 73(6):994–7.
102. Banta JV, Sutherland DH, Wyatt M. Anterior tibial transfer to the os calcis with Achilles tenodesis for calcaneal deformity in myelomeningocele. J Pediatr Orthop. 1981;1(2): 125–30.
103. Stott NS, Zionts LE, Gronley JK, Perry J. Tibialis anterior transfer for calcaneal deformity: a postoperative gait analysis. J Pediatr Orthop. 1996;16(6):792–8.
104. Bradley GW, Coleman SS. Treatment of the calcaneocavus foot deformity. J Bone Joint Surg Am. 1981;63(7):1159–66.
105. Wenz W, Bruckner T, Akbar M. Complete tendon transfer and inverse Lambrinudi arthrodesis: preliminary results of a new technique for the treatment of paralytic pes calcaneus. Foot Ankle Int. 2008;29(7):683–9.
106. Georgiadis GM, Aronson DD. Posterior transfer of the anterior tibial tendon in children who have a myelomeningocele. J Bone Joint Surg Am. 1990;72(3):392–8.
107. Mitchell GP. Posterior displacement osteotomy of the calcaneus. J Bone Joint Surg Br. 1977;59(2):233–5.
108. Badelon O, Bensahel H. Subtalar posterior displacement osteotomy of the calcaneus: a preliminary report of seven cases. J Pediatr Orthop. 1990;10(3):401–4.
109. Drennan JC, Sharrard WJ. The pathological anatomy of convex pes valgus. J Bone Joint Surg Br. 1971;53(3):455–61.
110. Walker AP, Ghali NN, Silk FF. Congenital vertical talus. The results of staged operative reduction. J Bone Joint Surg Br. 1985;67(1):117–21.
111. Kodros SA, Dias LS. Single-stage surgical correction of congenital vertical talus. J Pediatr Orthop. 1999;19(1):42–8.
112. Mazzocca AD, Thomson JD, Deluca PA, Romness MJ. Comparison of the posterior approach versus the dorsal approach in the

treatment of congenital vertical talus. J Pediatr Orthop. 2001; 21(2):212–7.
113. Alaee F, Boehm S, Dobbs MB. A new approach to the treatment of congenital vertical talus. J Child Orthop. 2007;1(3):165–74.
114. Dobbs MB, Purcell DB, Nunley R, Morcuende JA. Early results of a new method of treatment for idiopathic congenital vertical talus. J Bone Joint Surg Am. 2006;88(6):1192–200.
115. Yang JS, Dobbs MB. Treatment of congenital vertical talus: comparison of minimally invasive and extensive soft-tissue release procedures at minimum five-year follow-up. J Bone Joint Surg Am. 2015;97(16):1354–65.
116. Eberhardt O, Fernandez FF, Wirth T. Treatment of vertical talus with the Dobbs method. Z Orthop Unfall. 2011;149(2):219–24.
117. Aslani H, Sadigi A, Tabrizi A, Bazavar M, Mousavi M. Primary outcomes of the congenital vertical talus correction using the Dobbs method of serial casting and limited surgery. J Child Orthop. 2012;6(4):307–11.
118. Chalayon O, Adams A, Dobbs MB. Minimally invasive approach for the treatment of non-isolated congenital vertical talus. J Bone Joint Surg Am. 2012;94(11):e73.
119. Wright J, Coggings D, Maizen C, Ramachandran M. Reverse Ponseti-type treatment for children with congenital vertical talus: comparison between idiopathic and teratological patients. Bone Joint J. 2014;96-B(2):274–8.
120. Marciniak W. Results of surgical peritalar release with elongation of the lateral column of the foot in treatment of congenital veritcal talus. Chir Narzadow Ruchu Ortop Pol. 1993;58(3): 189–93.
121. Napiontek M. Congenital vertical talus: a retrospective and critical review of 32 feet operated on by peritalar reduction. J Pediatr Orthop B. 1995;4(2):179–87.
122. Torosian CM, Dias LS. Surgical treatment of severe hindfoot valgus by medial displacement osteotomy of the os calcis in children with myelomeningocele. J Pediatr Orthop. 2000;20(2): 226–9.
123. Parsons SW, Duckworth T, Betts RP, Rowley DI. Os calcis osteotomy in the management of deformities of the hindfoot in spinal dysraphism. Z Kinderchir. 1986;41(Suppl 1):30–2.
124. Gallien R, Morin F, Marquis F. Subtalar arthrodesis in children. J Pediatr Orthop. 1989;9(1):59–63.

Upper Limb Problems in Children with Cerebral Palsy

41

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Abstract

The decision regarding the management of upper limb spasticity in cerebral palsy is multifactorial. Intervention ranges from therapy and splinting through to multilevel surgery utilising tendon transfers and arthrodesis procedures. This article presents the current evidence behind treatment choice. A literature search has been conducted on each topic with a subsequent summary of the evidence. The use of Botulinum toxin type A in cerebral palsy has been the focus of several randomised trials. There exists good evidence of efficacy when used in conjunction with therapy however currently there is no clear consensus on optimal dose, frequency and post injection therapy regime. There exists good evidence that the surgical treatment of thumb in palm deformity has beneficial outcomes. The efficacy of surgical intervention for other upper limb deformities is less well understood. Each patient should be thoroughly assessed and surgical plans tailored to their specific needs. Surgical intervention should be delayed until the appropriate age to avoid recurrence with growth and the child should have voluntary motor control of the limb and reasonable cognition in order to comply with post-operative therapy.

Keywords

Upper limb • cerebral palsy • Botox • BoNT-A • Botulinum • Tendon transfer

Introduction

Cerebral palsy (CP) is a non-progressive upper motor neuron disease due to injury to the immature brain. It is the commonest cause of neurological disability in children and a common condition seen by the paediatric orthopaedic surgeon. Upper limb involvement, particularly in those patients with spastic hemiplegic CP, can have a significant impact on function. Generally, flexors exhibit increased tone over extensors. Pectoralis major, biceps brachii, forearm flexors and pronator teres are often hypertonic leading to shoulder flexion, adduction and internal rotation with elbow flexion, forearm pronation, wrist flexion with ulnar deviation. In the hand, finger and thumb flexion is seen along with thumb adduction giving the classic thumb-in-palm deformity. MCPJ dislocations and swan neck deformities can significantly affect opposition and therefore grasp. Patient factors such as cognitive capacity and limb sensation will significantly affect the management and therefore intervention must be tailored to each individual.

Non-invasive management strategies include physiotherapy and occupational therapy aimed at maximising function. Newer modalities are subject to extensive investigation and include bimanual therapy, constraint-induced movement therapy, action observation therapy, virtual reality and robot-assisted therapy. Functional splinting aims to improve motor function by supporting joints in the optimal position during use. There is however a paucity of evidence to support their efficacy. Non-functional splinting provides prolonged stretch with the aim of preventing contraction. These devices provide a small improvement in hand function that is not maintained Jackman [1]. Splinting in the younger age group is not well tolerated.

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Common discussions encountered in the clinic focus on the use of Botulinum toxin type A (BoNT-A) and surgical intervention. We therefore set out to answer key questions on each subject.

Botulinum Toxin A

A literature review was undertaken using the PubMed database. All randomised controlled trials (RCTs) comparing BoNT-A injection or BoNT-A injection and occupational therapy in the upper limb with other types of treatment (including no treatment or placebo) in children with CP were included. The bibliographies of all retrieved trials were reviewed to identify further studies. Fifteen studies meeting the inclusion criteria were identified, including two meta-analyses [2, 3].

Key Questions

What are the indications and aims of BoNT-A use?
What evidence is there to support its use?
What is the optimal dose, frequency and post injection therapy?
What are the complications and side effects?

What Are the Indications and Aims of BoNT-A Use?

Botulinum toxin type A acts by presynaptic blockade of cholinergic receptors at the neuromuscular junction. It is used as an adjunct to standard therapies aimed at improving range of movement and function through the reduction of muscle tone and spasticity. There has been much in the way of investigation into the different facets of BoNT-A administration chiefly dose, duration of effect, repetition and timing of standard therapeutic intervention after injection. The long-term outcome is also under investigation and is understandably of some interest, as the period of clinically useful relaxation appears to be between two and six months following administration [4–6]. The overall aim of BoNT-A injection is to produce a reduction in tone hence allow splinting, casting or therapy for increased muscle length with the hope of improving motor function and therefore reducing, or at least delaying, the need for surgery. NICE guidelines updated in September 2014 list the indications for the use of BoNT-A in children and young people with focal spasticity of the upper limb. These include spasticity impeding fine motor function, compromising care and hygiene, causing pain, impeding tolerance of other treatments such as orthoses or causing cosmetic concerns to the patient [7]. Contraindications include severe muscle weakness, previous adverse reaction or if the patient is receiving aminoglycoside treatment.

What Evidence Is There to Support Its Use?

The use of BoNT-A in the upper limb in children with cerebral palsy was the subject of a Cochrane review in 2010 [8]. They report the pooled data of seven randomized controlled trials and three unpublished trials aimed at understanding the effectiveness of intramuscular BoNT-A versus BoNT-A and occupational therapy [5, 6, 9–14]. They report a significant decrease in spasticity following BoNT-A administration, however a resultant functional gain that is sustained, was not proven. Moderate evidence exists that BoNT-A alone is not effective compared to placebo or indeed no treatment at all. High-level evidence was presented supporting the use of BoNT-A as an adjunct to concomitant occupational therapy. BoNT-A and occupational therapy was shown to be more effective than occupational therapy alone in reducing impairment, improving activity level and goal achievement based on the chosen functional outcome scoring methods. Interestingly however, no improvement in quality of life or perceived self-competence was found.

Supporting the findings of this review is a further RCT by Ferrari et al. [15] comparing BoNT-A and therapy with placebo and therapy. They found that both groups demonstrated an improvement in function with a significantly better improvement in assisted hand assessment (AHA) in the BoNT-A group. However Rameckers et al. [16] report an RCT looking at the benefit of BoNT-A and intensive therapy over intensive therapy alone. Over six months no significant differences were reported between groups with regard to functional measures. Muscle force decreased following BoNT-A injection but returned during the therapy period. The therapy group displayed significantly higher increase in force and accuracy relative to the group receiving BoNT-A [16]. Van Heest et al [17] also report a lack of functional improvement after BoNT-A injection relative to therapy alone. They again observed a significant reduction in grip strength following BoNT-A injection. In this RCT however the reduction in strength continued for six months after the final injection was administered [28, #11568].

What Is the Optimal Dose, Frequency and Post Injection Therapy?

There is no recognized, superior dose regime recommended in the current literature. Doses used in controlled trials to date range from 2 to 12 U/kg body weight per session with a maximum does of 400 units. Forearm muscles have received 0.3–2 U/kg increasing in the upper arm to 4 U/kg body weight per muscle [5]. The commonly employed concentration is between 50 and 100 U/ml [3].

Lowe in 2006 demonstrated better longer-term outcomes with a low volume, higher concentration BoNT-A injection (200 U/ml saline) [10]. However, similar results were not recreated by Kawamura et al. who specifically compared the effects of low dose BoNT-A with a high dose (double

strength) BoNT-A on outcome [11]. In this study however, muscle localization was performed by palpation without stimulation or imaging. Further studies using adults also suggests the high-volume, low concentration injections achieve greater neuromuscular blockade, spasticity reduction and greater range of elbow extension than low-volume, high concentration injections [17]. With a paucity of evidence to support the use of higher doses of BoNT-A in the upper limb of patients with CP, currently there is no reason to deviate from those recommendations contained in the paediatric upper limb hypertonicity BoNT-A evidence based guidelines for intervention and aftercare published in 2010 [5].

This consensus report outlines a pragmatic and evidence based approach to calculating the dose of BoNT-A dependent on muscle groups injected and aims of treatment. Due to the small size of forearm muscles that are in much closer proximity than other areas, lower doses with less dilution are recommended if functional gains are the aim [10, 13, 18].

In terms of concentration the smaller the volume injected, the lower the diffusion within and beyond the muscle [19, 20]. If the treatment is to improve function, it is recommended that the calculated dosage be distributed in a small amount of normal saline in the muscle at one to two injection sites, depending on the age and size of the child. At the forearm, a volume exceeding 0.5 ml is likely to diffuse into neighbouring muscles. A preparation of 100 U/ml saline dilution is therefore recommended dependent on the product used. In children with larger muscles or if the aim of intervention is to improve appearance, tolerance of orthoses, or facilitate care, then a higher volume would be appropriate such as 100 units in 2 mls.

The therapeutic effect of BoNT-A lasts approximately 2–6 months. There is a risk of antibody formation with BoNT-A use and in order to minimise this, intervals between injections should exceed 3 months [21, 22]. In practice it seems sensible to prolong intervals between injections as long as clinically justifiable without jeopardising treatment. This approach will help to allow adaptation to reduce tone and avoid excessive weakness through repeated injections.

No study was found investigating the effect of repeat BoNT-A injections against single injections. Olesch et al. performed an RCT into repeat BoNT-A injections and therapy vs therapy alone. They report progressively reduced spasticity and improved parental perception of performance in the BoNT-A group [23]. They included 22 children without fixed contractures. Three BoNT-A injections were administered no more frequent than every 16 weeks. They prescribed 6 weeks of intensive therapy (twice weekly sessions) followed by routine therapy until the next injection. This study was underpowered however and the only significant difference between the groups was seen in areas where the assessors were not blinded.

In a recent RCT, Lidman et al. compared repeat BoNT-A injections and therapy with therapy alone on hand function. Their injection protocol included BoNT-A administered 2 weeks prior to 8 weeks of therapy. Thereafter parents went back to normal therapy with a second injection at 6 months. They report improvements in ROM and goal performance in both groups. The BoNT-A group demonstrated a significantly better AHA. They included all functional levels; however there were clinical differences at baseline that may have affected functional outcomes. This was a small study (20 participants) with no power calculation and only partial blinding [24].

An investigation into multi-sessional intramuscular injections of BoNT-A performed at weeks eight and 20 following initial injection to muscles still exhibiting marked spasticity was performed by Koman et al. [25]. This prospective, double-blind RCT concluded that after these three injections, children receiving BoNT-A displayed a clinically meaningful improvement in function at 26 weeks compared to those who received placebo. Their cohort included children with varying levels of functional impairment some of whom were not surgical candidates. Follow-up was to 26 weeks but grip strength was not assessed.

Using a similar protocol with injections at initiation, 12 and 24 weeks followed by a supervised therapy regime and continued home therapy; Van Heest et al. did not reproduce these findings. They performed a final assessment 6 months after last injection and report no significant improvements in measurements of bodily impairment, activity limitations or participation restrictions in the BoNT-A group or those treated with ongoing therapy alone at 12 months. Furthermore, they report a significant reduction in grip strength in the BoNT-A group up to 6 months following last injection.

There is great variation in therapy regimes following BoNT-A injection in the literature. To date there is little high quality evidence that any is superior. Hoare et al. report an RCT into whether modified constraint-induced movement therapy (mCIMT) leads to superior gains compared with bimanual occupational therapy (BOT) in young children with unilateral CP following BoNT-A injections. They conclude that there is no benefit to mCIMT despite increased intensity of the home programme over standard BOT [2]. Most regimes utilise static splinting between therapy sessions to maintain any improvement in spasticity achieved.

What Are the Complications and Side Effects?

The 2010 Cochrane review reported excessive grip weakness as the most common adverse event. Several studies report the difference in strength reduction in the smaller muscles of the upper limb compared to the larger muscles of the lower limb. BoNT-A use in the forearm can result in a 90 % reduction in the amplitude of the EMG after electrical stimulation of the corresponding nerve compared to 20 % in

the gastrocnemius muscle [26, 27]. The impact of BoNT-A on grip strength has been investigated by Van Heest et al. They report that the reduction in grip and pinch is not only present immediately after administration but persists for at least 6 months after last injection [28]. Other less commonly reported complaints include nausea, vomiting, flu like symptoms, coughing, soreness at injection site, respiratory infections and headache.

Future Research

There are still many unanswered questions when it comes to the use of BoNT-A in children with CP, not least of all the fundamental question as to whether it is beneficial to functional outcome at all. Key areas for future research include:

1. The impact of BoNT-A dose, concentration, volume and delivery methods to the muscles
2. The impact of repeat injections with detail of the optimal timing and cumulative effects
3. The long-term effect on function investigated using robust outcome measures
4. The optimal frequency, timing and intensity of therapy following BoNT-A injection

Surgical Intervention

The majority of the literature on this topic comprises of retrospective case series. There were no level I studies identified. A meta-analysis of case series was performed in 2005 on the management of thumb-in-palm deformities [29].

One prospective RCT of level II quality was subsequently published comparing surgical intervention, BoNT-A injection and therapy alone [28]. Both the tendon transfer and BoNT-A group received the same standardised therapy protocol as the therapy group. They report that children undergoing surgical treatment demonstrated greater improvement, of modest magnitude, than BoNT-A injections or regular, ongoing therapy at twelve months of follow-up. The surgical protocol that they followed addressed the classical deforming factors in the upper limb in children with CP. This included a transfer of the flexor carpi ulnaris to the extensor carpi radialis brevis, pronator teres release, and extensor pollicis longus rerouting with adductor pollicis release. They report a significant increase in pinch strength and the movement domain of the Pediatric Quality of Life Inventory relative to BoNT-A injection or therapy alone. However there was no significant difference in the other four domains or indeed in activity limitations. Patient reported satisfaction was greater following tendon transfer in the absence of a significant improvement in performance.

Key Questions

1. What are the indications for surgery?
2. When should you operate?
3. What are the common deformities and how are they addressed?

What Are the Indications for Surgery?

The goal of surgery needs to be clearly defined. Aims include improved function, to facilitate hygiene or improve appearance. NICE guidelines on consideration for operative intervention in spasticity and co-existing motor disorders include limited limb function (for example getting dressed) by unfavourable posture or pain, as a result of muscle shortening, contractures or bony deformities; contractures of the shoulder, elbow, wrist or hand cause difficulty with skin hygiene; the cosmetic appearance of the upper limb causes significant concern for the child [30]. The goals of surgical intervention in upper limb spasticity are to reduce the contractile effect of the spastic muscles by release or fractional lengthening, increase the strength of weak antagonists through tendon transfer or improve joint stability through arthrodesis or tenodesis.

The overall condition of the patient affects the decision making process. The ideal candidate to achieve functional improvements should demonstrate adequate cognition, sensibility and motor control of the limb. If these prerequisites are not met, surgical intervention will likely result in improved appearance and hygiene alone [31]. The aim of management prior to reconstruction should be to maintain joint ROM and reduce contractures. It is unwise to attempt tendon transfers in a joint lacking passive movement and post-operative active ROM will not be greater than pre-operative passive ROM. If intensive therapy cannot overcome joint and soft tissue contractures, release and lengthening procedures need to be incorporated into the surgical plan prior to tendon transfer.

When Should You Operate?

The age of surgical intervention remains controversial. Ideally reconstruction should be delayed until at least the age of four to six as at this age the cerebral defect is mature therefore the deformities are easily assessed. The child also stands a better chance of cooperating with rehabilitation regimes [32, 33]. Older age is not a contraindication as long as the patient has a degree of voluntary hand control. A Cochrane review of surgical treatment for thumb-in-palm deformity included nine studies with a mean age ranging from 7.5 to 15 years [29].

Care should be taken however in performing tendon transfers in younger patients as late deformities may occur related to growth spurts. Patterson et al. report that of 24 patients receiving an FCU to ECRB transfer, 12 developed

late deformity between 10 and 105 months postoperatively [34]. Nine required revision surgery commonly for extension deformity. Of the 12 patients, nine were younger than 13 years old at time of index procedure.

What Are the Common Deformities and How Are they Addressed?

Forearm Pronation, Wrist Flexion with Ulnar Deviation

Forearm pronation is a deformity caused by a combined spasticity of both the pronator teres and pronator quadratus muscle with wrist flexion a result of spasticity of the FCU tendon with or without involvement of the long finger flexors. This deformity can seriously limit hand function. The aim of reconstruction is to restore active supination, without compromising the existing pronation movement, and achieve a neutral wrist position to improve grip. This can be achieved using a combination of muscle releases and tendon transfers.

FCU to ECRB (Green's) transfer is the classical procedure used to increase wrist extension with the benefit of gaining active supination [35, 36]. The Green transfer removes FCU as a spastic wrist flexor and augments wrist extension power, thus improving position [37]. The range of flexion extension arc may not improve significantly however centralising this arc at neutral results in improved function and cosmetic appearance [38]. Concomitant procedures including pronator quadratus myotomy, pronator teres myotomy, lengthening or rerouting act to reduce pronation deformity. An average gain in supination of 54° with pronator teres tenotomy and 78° with rerouting has been reported [39].

However it should be borne in mind that operating results in improved extension at the price of wrist flexion [40, 41] and that the Green transfer tensioned past neutral can result in excessive loss of wrist flexion creating extensor habitus [42, 43]. This may be particularly pertinent if the patient has marked spasticity in the long finger flexors as part of the deformity that may lead to difficulty when attempting to open the hand.

Thumb-in-Palm Deformity

A Cochrane review was conducted in 2005 and although studies into this area are of relatively poor design, modest but significant improvements in ROM, pinch grip and hand function were found following operative intervention for thumb-in-palm deformity [44]. There is great difficulty in drawing firm conclusion from published studies due to the heterogeneity of the patients, differing eligibility criteria for surgical intervention and lack of standardised functional assessment tools.

In this review no consensus on the selection criteria for eligibility to undergo surgical intervention was reached. No study provided an evidence base for inclusion however some voluntary control appeared to be necessary to achieve functional improvement. Furthermore there was no standardised method for evaluating operative data either prior to or following intervention. With few exceptions, most assessment tools were not validated. It is not possible on the evidence presented to make judgement about the efficacy of one particular intervention as different procedures were used within most studies along with a variety of outcome measures.

The Cochrane review tells us that surgical intervention can result in correction of thumb-in-palm deformity leaving the thumb out of the palm [31, 41, 45, 46], improved grip and pinch [41, 45–49], function [41, 46, 50, 51], appearance [45, 47–49], stereognosis [41] and quality of life [31, 45].

Although intervention varied widely, common combinations include release of the spastic adductor pollicis from its origin with EPL rerouting through the first dorsal retinacular compartment. This technique reduces thumb adduction and augments the weak extension-abduction force [50, 52, 53].

The thumb-in-palm posturing is a complex problem with multiple muscle involvement combining to produce the resultant deformity. There are several surgical procedures that can be employed when dealing with thumb-in-palm deformity and a combination of procedures is often required. Each patient must therefore be thoroughly assessed and surgical plans tailored to their specific needs.

Future Research

Due to small numbers of procedures being undertaken in such a heterogenous group, robust studies into the efficacy of specific surgical procedures for CP affecting upper limb is fraught with difficulty. Certainly progress has been made with more recent studies comparing surgical intervention to conventional use of BoNT-A and therapy with some benefit being reported in the former group. Key areas for future research include:

1. Further study of the efficacy of tendon transfers relative to conventional intervention
2. The long-term outcome following tendon transfers to quantify the risk of recurrent deformity
3. The optimal timing for surgical intervention to improve function specifically in younger patients

Authors' Recommendations

These patients are very heterogeneous and the management of their spasticity varies according to a number of factors. In general we would recommend spasticity management using botulinum and hand therapy in the younger patients followed by single event multi-level surgery when they are at the appropriate age.

Age

The very young patients (under 2 years) need proactive hand therapy (including techniques such as constraint induced therapy) and carer education to maintain passive range of motion.

Patients up to the age of 10 years are at risk of deformity recurrence or over correction with tendon transfer surgery are better treated with botulinum toxin. These patients may also be suitable for selective peripheral neurotomy which reduces spasticity to a similar degree but on a permanent basis

Older children (over 10 years) generally have lower levels of spasticity are suited to definitive surgery such as tendon transfers or joint arthrodesis. Higher functioning adolescents often have concerns regarding their body-image and this needs to be taken into consideration when deciding whether to offer surgery

Differentiation Between Fixed Flexion and Spasticity

Spasticity is dynamic and the deformity can be corrected with sustained pressure – these patients are suitable for botulinum toxin and joint rebalancing transfers.

Fixed flexion is static and due to muscle and joint fibrosis with secondary osseous changes. These deformities will not fully correct without appropriate soft tissue releases/joint stabilisations.

Degree of Active Control and Level of Cognition

High functioning patient with good active control but poor dynamic posturing would function well with tendon rebalancing whereas lower functioning patients with fixed contractures may benefit from a stabilisation such as wrist arthrodesis) if they are having issues with hand hygiene/dressing etc.

Type of Cerebral Palsy

High tone spasticity is most common and usually presents and behaves in a fairly typical fashion.

Dystonic cerebral palsy has a much higher risk of overcorrection deformity and are better treated with judicious use of botulinum toxin and joint stabilisation procedures rather than tendon re-balancing

Non-cerebral palsy types of neuromuscular pathology also need treating with caution as their natural history may be quite different, particularly if it is a progressive condition.

References

1. Jackman M, Novak I, Lannin N. Effectiveness of hand splints in children with cerebral palsy: a systematic review with meta-analysis. Dev Med Child Neurol. 2014;56(2):138–47.
2. Hoare B et al. Intensive therapy following upper limb botulinum toxin A injection in young children with unilateral cerebral palsy: a randomized trial. Dev Med Child Neurol. 2010;55(3):238–47.
3. Sakzewski L, Ziviani J, Boyd RN. Efficacy of upper limb therapies for unilateral cerebral palsy: a meta-analysis. Pediatrics. 2014;133(1):e175–204.
4. Graham HK. Botulinum toxin A in cerebral palsy: functional outcomes. J Pediatr. 2000;137(3):300–3.
5. Fehlings D et al. Botulinum toxin assessment, intervention and follow-up for paediatric upper limb hypertonicity: international consensus statement. Eur J Neurol. 2010;17:38–56.
6. Fehlings D et al. An evaluation of botulinum-A toxin injections to improve upper extremity function in children with hemiplegic cerebral palsy. J Pediatr. 2000;137(3):331–7.
7. NICE. Botulinum toxin type A for treating children and young people with spasticity. 2014. http://pathways.nice.org.uk/pathways/spasticity-in-children-and-young-people/botulinum-toxin-type-a-for-treating-children-and-young-people-with-spasticity.
8. Hoare BJ et al. Botulinum toxin A as an adjunct to treatment in the management of the upper limb in children with spastic cerebral palsy (UPDATE). Cochrane Database of Syst Rev. 1996, Wiley-Blackwell.
9. Speth L et al. Botulinum toxin A and upper limb functional skills in hemiparetic cerebral palsy: a randomized trial in children receiving intensive therapy. Dev Med Child Neurol. 2005;47(7):468–73.
10. Lowe K, Novak I, Cusick A. Low-dose/high-concentration localized botulinum toxin A improves upper limb movement and function in children with hemiplegic cerebral palsy. Dev Med Child Neurol. 2006;48(03):170.
11. Kawamura A et al. A randomized controlled trial comparing botulinum toxin A dosage in the upper extremity of children with spasticity. Dev Med Child Neurol. 2007;49(5):331–7.
12. Wallen M, O'Flaherty SJ, Waugh M-CA. Functional outcomes of intramuscular botulinum toxin type A and occupational therapy in the upper limbs of children with cerebral palsy: A randomized controlled trial. Arch Phys Med Rehabil. 2007;88(1):1–10.
13. Russo RN et al. Upper-limb botulinum toxin A injection and occupational therapy in children with hemiplegic cerebral palsy identified from a population register: a single-blind, randomized, controlled trial. Pediatrics. 2007;119(5):e1149–58.
14. Corry IS et al. Botulinum toxin A in the hemiplegia upper limb: a double-blind trial. Dev Med Child Neurol. 1997;39(3):185–93.
15. Ferrari A et al. A randomized trial of upper limb botulimun toxin versus placebo injection, combined with physiotherapy, in children with hemiplegia. Res Dev Disabil. 2014;35(10):2505–13.
16. Rameckers EAA et al. Botulinum toxin-A in children with congenital spastic hemiplegia does not improve upper extremity motor-related function over rehabilitation alone: a randomized controlled trial. Neurorehabil Neural Repair. 2008;23(3):218–25.
17. Goldfarb CA et al. Epidemiology of congenital upper limb anomalies in a midwest United States population: an assessment using the Oberg, Manske, and Tonkin classification. J Hand Surg Am. 40(1):127–32, e1–2.
18. Sätilä H et al. Low- and high-dose botulinum toxin A treatment: a retrospective analysis. Pediatr Neurol. 2006;34(4):285–90.
19. Borodic GE et al. Histologic assessment of dose-related diffusion and muscle fiber response after therapeutic botulinum a toxin injections. Mov Disord. 1994;9(1):31–9.

20. Shaari CM, Sanders I. Quantifying how location and dose of botulinum toxin injections affect muscle paralysis. Muscle Nerve. 1993;16(9):964–9.
21. Dressler D. Clinical presentation and management of antibody-induced failure of botulinum toxin therapy. Mov Disord. 2004;19(S8):S92–S100.
22. Herrmann J et al. Clinical impact of antibody formation to botulinum toxin A in children. Ann Neurol. 2004;55(5):732–5.
23. Olesch CA et al. Repeat botulinum toxin-A injections in the upper limb of children with hemiplegia: a randomized controlled trial. Dev Med Child Neurol. 2010;52(1):79–86.
24. Lidman G et al. Botulinum toxin A injections and occupational therapy in children with unilateral spastic cerebral palsy: a randomized controlled trial. Dev Med Child Neurol. 2015;57(8):754–61.
25. Koman LA, et al. Upper extremity spasticity in children with cerebral palsy: A randomized, double-blind, placebo-controlled study of the short-term outcomes of treatment with botulinum A toxin. J Hand Surg. 2013; 38(3):435–46.e1.
26. Koman LA et al. Botulinum toxin type A neuromuscular blockade in the treatment of lower extremity spasticity in cerebral palsy: A randomized, double-blind, placebo-controlled trial. J Pediatr Orthop. 2000;20(1):108.
27. Autti-Ramo I et al. Management of the upper limb with botulinum toxin type A in children with spastic type cerebral palsy and acquired brain injury: clinical implications. Eur J Neurol. 2001;8(s5):136–44.
28. Van Heest AE et al. Tendon transfer surgery in upper-extremity cerebral palsy is more effective than botulinum toxin injections or regular, ongoing therapy. J Bone Joint Surg Am. 2015;97(7):529–36.
29. Smeulders M, Coester A, Kreulen, M. Surgical treatment for the thumb-in-palm deformity in patients with cerebral palsy. In The Cochrane database of systematic reviews (protocol). Wiley-Blackwell; 1996.
30. NICE. NICE. Spasticity in children and young people with non-progressive brain disorders: management of spasticity and co-existing motor disorders and their early musculoskeletal complications. 2012. https://www.nice.org.uk/guidance/cg145?unlid=11007647120151122153056.
31. Rayan GM, Saccone PG. Treatment of spastic thumb-in-palm deformity: A modified extensor pollicis longus tendon rerouting. J Hand Surg. 1996;21(5):834–9.
32. Zancolli EA, Zancolli ER. The infantile spastic hand surgical indications and management. Ann Chir Main. 1984;3(1):66–75.
33. Skoff H, Woodbury DF. Management of the upper extremity in cerebral palsy. J Pediatr Orthop. 1985;5(5):627.
34. Patterson JMM, Wang AA, Hutchinson DT. Late deformities following the transfer of the flexor carpi ulnaris to the extensor carpi radialis brevis in children with cerebral palsy. J Hand Surg. 2010;35(11):1774–8.
35. Green WT, Banks HH. Flexor carpi ulnaris transplant and its use in cerebral palsy. J Bone Joint Surg Am. 1962;44-A:1343–430.
36. Cheema TA et al. Biomechanic comparison of 3 tendon transfers for supination of the forearm. The Journal of Hand Surgery. 2006;31(10):1640–4.
37. de Roode CP, James MA, Van Heest AE. Tendon transfers and releases for the forearm, wrist, and hand in spastic hemiplegic cerebral palsy. Tech Hand Up Extrem Surg. 2010;14(2):129–34.
38. Beach WR et al. Use of the green transfer in treatment of patients with spastic cerebral palsy. J Pediatr Orthop. 1991;11(6):731–6.
39. Strecker WB et al. Comparison of pronator tenotomy and pronator rerouting in children with spastic cerebral palsy. J Hand Surg. 1988;13(4):540–3.
40. Wenner SM, Johnson KA. Transfer of the flexor carpi ulnaris to the radial wrist extensors in cerebral palsy. J Hand Surg. 1988;13(2):231–3.
41. Dahlin L, Komototufvesson Y, Salgeback S. Surgery of the spastic hand in cerebral palsy. Improvement in stereognosis and hand function after surgery. J Hand Surg. 1998;23(3):334–9.
42. Wolf T, Clinkscales C, Hamlin C. Flexor carpi ulnaris tendon transfers in cerebral palsy. J Hand Surg. 1998;23(3):340–3.
43. Thometz JG, Tachdjian M. Long-term follow-up of the flexor carpi ulnaris transfer in spastic hemiplegic children. J Pediatr Orthop. 1988;8(4):407–12.
44. Acosta D et al. Measurement of prompt charm meson production cross sections in pp collisions at square root s = 1.96 TeV. Phys Rev Lett. 2003;91(24):241804.
45. Roth J et al. Functional outcome of upper limp tendon transfers performed in children with spastic hemiplegia. J Hand Surg. 1993;18(3):299–303.
46. Tonkin M. Surgery for cerebral palsy part 3: classification and operative procedures for thumb deformity. J Hand Surg. 2001;26(5):465–70.
47. Inglis AE, Cooper W, Bruton W. Surgical correction of thumb deformities in spastic paralysis. J Bone Joint Surg Am. 1970;52(2):253–68.
48. Lee BS, Horstmann H. The brachioradialis for restoration of abduction and extension of spastic thumb in children. Orthopedics. 1984;7(9):1445–8.
49. Filler BC, Stark HH, Boyes JH. Capsulodesis of the metacarpophalangeal joint of the thumb in children with cerebral palsy. J Bone Joint Surg Am. 1976;58(5):667–70.
50. House JH. A dynamic approach to the thumb-in-palm deformity in cerebral palsy. J Pediatr Orthop. 1981;1(3):357.
51. Nylander G, Carlstrom C, Adolfsson L. 4.5 year follow-up after surgical correction of upper extremity deformities in spastic cerebral palsy. J Hand Surg Br. 1999;24(6):719–23.
52. Manske PR. Redirection of extensor pollicis longus in the treatment of spastic thumb-in-palm deformity. J Hand Surg. 1985;10(4):553–60.
53. Van Heest AE. Surgical technique for thumb-in-palm deformity in cerebral palsy. J Hand Surg. 2011;36(9):1526–31.

Part X

Musculoskeletal Diseases

Evidence-Based Treatment for Musculoskeletal Infection

42

Megan Mignemi, Lawson Copley, and Jonathan Schoenecker

Abstract

Musculoskeletal infection (MSKI) in children is a common cause of hospitalization in the pediatric population worldwide. Severity of disease is dependent upon the amount and type of tissue involved. Though common, pediatric MSKI diagnosis and treatment can be challenging. In this chapter, we have critically reviewed and summarized the evidence that underpin the current practice in the developed world. Evidence-based treatment guidelines applied by a multidisciplinary team resulted in better care of children with MSKI (Grade B). Non-contrasted MRI is the diagnostic modality of choice to distinguish isolated septic arthritis from adjacent infection (Grade B). Septic arthritis is the most common diagnosis with synovial fluid WBC counts between 25,000 and 75,000 cells/mm3 and can be adequately treated by both arthroscopy or traditional arthrotomy (Grade B). Acute haematogenous osteomyelitis (AHO) can be treated with short course of IV antibiotic, followed by oral antibiotic for 3–4 weeks (Grade B).

Keywords

Musculoskeletal infection • Osteomyelitis • Pyomyositis • Septic arthritis

Background

Musculoskeletal infection (MSKI) in children is a common cause of hospitalization in the pediatric population worldwide. Outcomes are typically good with relatively few long term sequelae when recognized and treated in a timely fashion. Pediatric MSKI is traditionally classified based on the type of tissue infected: superficial abscess, septic arthritis, osteomyelitis, deep abscess/pyomyositis and complex MSKI, which involves multiple tissue types (Table 42.1). This chapter will focus primarily on septic arthritis, osteomyelitis, and pyomyositis.

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Children with MSKI typically present with a constellation of symptoms including limp or inability to bear weight and pain. Physical examination will often reveal joint irritability, pain with ROM, and tenderness to palpation. The workup of pediatric MSKI generally includes baseline laboratory work including blood culture, complete blood count (CBC) with differential, c-reactive protein (CRP) and erythrocyte sedimentation rate (ESR). Plain films should also be obtained in order to evaluate for possible trauma. Further work-up is directed by the clinical picture and level of concern by the treating physician and may consist of advanced imaging (ultrasound (US), computed tomography (CT), and/or magnetic resonance imaging (MRI)) to evaluate the location and extent of infection. Some pediatric MSKI can be treated with antibiotics alone, while others require surgical debridement. Early recognition and appropriate treatment leads to favorable outcomes in most cases. Infection is deemed to be disseminated when there are multiple positive blood cultures, multiple foci of infection, evidence of thrombus formation (deep vein thrombosis (DVT) or pulmonary

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Table 42.1 Classification of pediatric MSKI based on tissue type

Diagnosis	Definition	Example
Tissue injury of unknown origin	Diagnosis of exclusion Negative tissue and/or blood culture Joint effusion	Transient synovitis of the hip
Superficial abscess	Superficial to deep fascia of limb or located in hand or foot	Septic pre-patellar bursitis Superficial forearm abscess
Septic joint	Limited to joint space only with no extension to the surrounding muscles or bone Synovial aspirate is grossly purulent, >50,000 cells, positive gram stain and/or positive bacterial culture	Septic knee
Osteomyelitis	Isolated to bone only, no extension to sub-periosteal space or surrounding muscle/joint	Proximal femur osteomyelitis
Deep abscess/Pyomyositis	Deep to fascia of limb Isolated to muscle only, no extension into nearby bone or joint May include multiple muscle groups May be mild (edema only), moderate (Phlegmon) or severe (abscess)	Obturator internus myositis Adductor and rectus femoris myositis
Complex	Involving a combination of bone, muscle and joints	Subperiosteal abscess, pericapsular myositis with ischial osteomyelitis, clavicular osteomyelitis with supraclavicular abscess

embolism (PE)), septic pulmonary emboli or endocarditis. These children have the potential to become very sick and may require intensive care. While long-term morbidity from pediatric MSKI is low, there are recognized complications from delayed diagnosis and treatment. The two most common sequelae are joint destruction and physeal arrest, which can lead to limb length discrepancy or angular deformity.

Septic Arthritis

Diagnosis

Most children with septic arthritis will demonstrate joint irritability with limited ROM, refusal to bear weight, fever, elevated inflammatory markers such as CRP, ESR, and white blood count (WBC) and joint effusion on ultrasound [1–14]. Other pediatric MSKI can mimic septic arthritis, especially when involving the hip joint. While "Kocher Criteria" has long been used to differentiate between children with septic hip arthritis and transient synovitis, recent studies have shown that this diagnostic criteria is unable to distinguish been septic arthritis and other types of infection (osteomyelitis and pyomyositis) around the hip [15]. For this reason, it has been advocated that in cases where there is concern for septic arthritis of the hip based on clinical exam and elevated inflammatory markers that an MRI be obtained as quickly as possible to evaluate the hip and surrounding structures for contiguous infection. One study of 53 patients evaluated in the emergency room for "rule out septic hip arthritis" demonstrated that pelvic pyomyositis was twice as common as isolated septic hip arthritis based on MRI [15] and a different study found that 70/103 patients with septic arthritis had contiguous osteomyelitis on MRI [16]. Rosenfeld et al. found five variables (age above 3.6 years, CRP > 13.8 mg/L, duration of symptoms >3 d, platelets <314 × 10 cells/μL, and ANC > 8.6 × 10 cells/μL) to be predictive of adjacent infection. Patients with ≥3 risk factors were classified as high risk for septic arthritis with adjacent infection (sensitivity: 90 %, specificity: 67 %, positive predictive value: 80 %, negative predictive value: 83 %). The authors of this study recommend preoperative MRI in patients in who meet ≥3 of these criteria [17]. In cases where there is infection adjacent to the joint in question, MRI also has the added benefit of directing the approach for joint aspiration (Fig. 42.1). Hip joint aspiration is commonly performed through a medial approach, just posterior to the adductor longus tendon. It is known, however, that a large percentage of cases of pelvic pyomyositis affect the adductor musculature. Therefore, there is a theoretical risk of contaminating the hip joint if joint arthrocentesis is unknowingly performed through the standard medial approach [15]. In many institutions, however, MRI is not readily available and in younger patients requires sedation. Therefore in a patient for whom the exam is clearly consistent with septic arthritis or who is becoming septic, treatment can be based on the presence of effusion on US and should not be delayed in order to obtain advanced imaging.

The gold standard for diagnosis of a joint infection is the acquisition of a joint fluid sample for gram stain, culture, cell count and occasionally, molecular testing [1, 2, 18]. This can

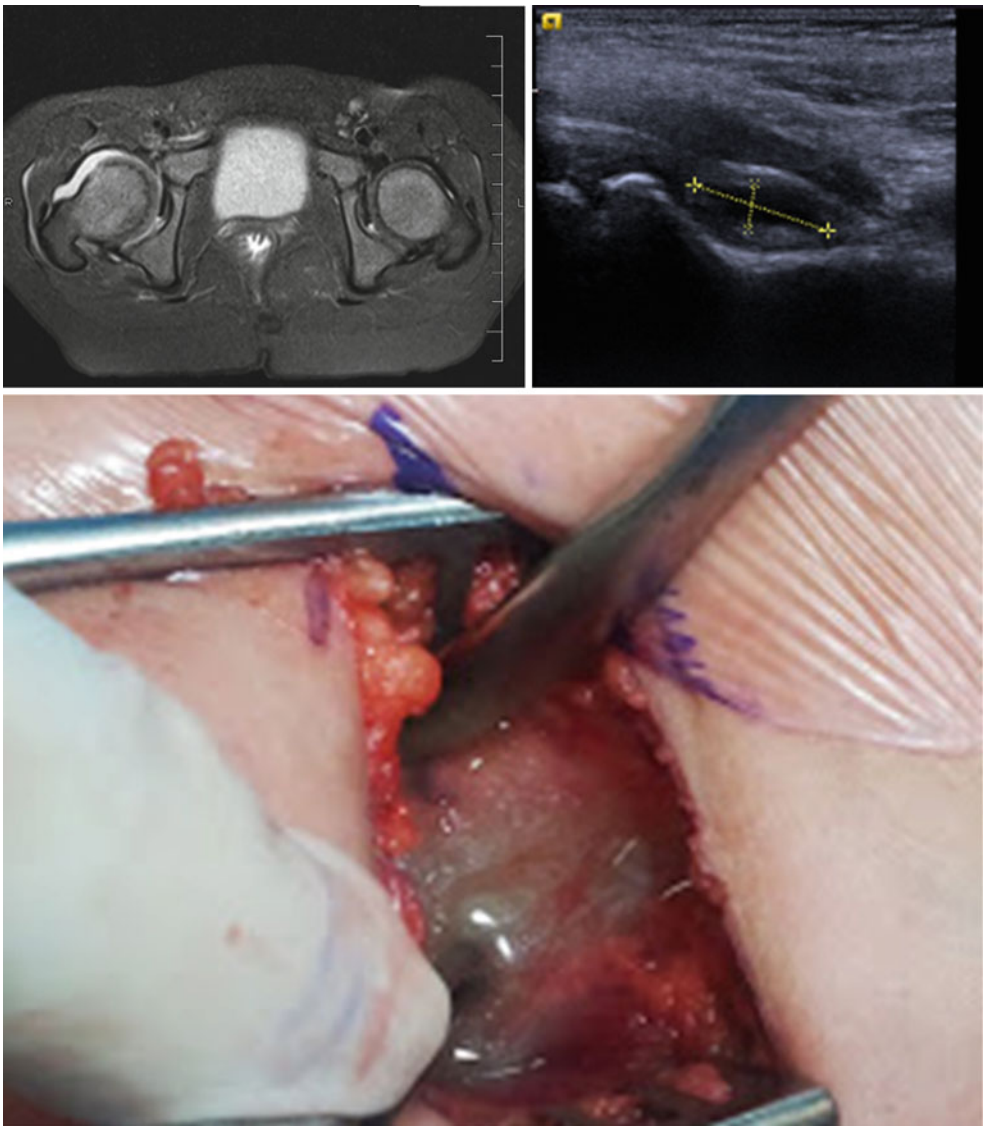

Fig. 42.1 Septic arthritis of the hip joint

be done under conscious sedation in the emergency department or general anesthesia in the operating room. In most children with bacterial arthritis, the nucleated cell count of the joint fluid aspirate is greater than 50,000 cells/mL with >75 % of the cells identified as segmented neutrophils. Occasionally arthrocentesis can lead to diagnostic uncertainty due to intermediate joint fluid cell counts (10,000–75,000 cells/mL) [19]. In this case, the clinician may consider arthrotomy or arthroscopy as a surgical intervention to treat the possibility of septic arthritis due to incomplete evidence. The most common cause of septic arthritis in children is Staphylococcus aureus and Streptococcus.

In endemic communities, Lyme arthritis due to Borrelia burgdorferi, may demonstrate an overall clinical impression of possible septic arthritis and should be differentiated by supplemental tests [20–23]. This is commonly performed using a sensitive enzyme immunoassay or immunofluorescent assay followed by a Western immunoblot. Joint aspiration is recommended in order to rule out bacterial septic arthritis. Joint fluid cell counts in cases of Lyme arthritis may demonstrate a spectrum of findings [19, 23]. For example, the average cell count for a patient with Lyme arthritis is 60,200 cells/mm^3, however, for septic arthritis the average cell count is 123,000 cells/mm^3 [23]. In a study of 506 patients with joint effusions, 49 % of the children who were ultimately diagnosed with Lyme arthritis had a joint fluid cell count >50,000 cells/mm^3, while 13 % had counts >100,000 cells/mm^3. Based on this study, patients with synovial fluid cell counts >100,000 cells/mm^3 were more 3.63× more likely to have septic arthritis than Lyme arthritis [23].

Kingella kingae is a gram-negative organism that is becoming increasingly recognized as a common cause of MSKI in

children age 6 months to 4 years [24–27]. The clinical presentation of MSKI caused by K kingae is more subtle, with normal or only mildly elevated inflammatory markers. The hip joint is a common site of K kingae infection and therefore a high index of suspicion in needed to make the diagnosis, as these cases are often mistaken for transient synovitis of the hip and blood/synovial fluid cultures are never obtained [24, 25, 28–31]. The benign clinical presentation along with the observation that several patients with culture proven K kingae septic arthritis improve without antibiotic treatment [32] calls into question the importance of missing a septic arthritis caused by K kingae [24]. There have been a few case reports of long term adverse sequelae from K kingae MSKI, which indicates that appropriate antibiotics should be given to all children from whom this organism is recovered [24, 25, 33–35]. K kingae is notoriously difficult to culture from synovial fluid using normal culture techniques. Inoculation of synovial fluid into blood culture vials can significantly improve the recovery of this organism and more recently, real time polymerase chain reaction (PCR) assays have become available to further enhance the detection of K kingae in synovial fluid [24, 36–43]. While PCR shows improved rates of detection of K kingae over traditional joint fluid culture, the turnaround time for results is quite delayed, with an average of 14.6 days at one center [43]. PCR also does not provide any information regarding antibiotic sensitivity [43].

Treatment

Adverse sequelae among children with septic arthritis are well described and are attributed to delayed or inadequate treatment. Clinicians should therefore treat bacterial arthritis by urgent irrigation and drainage of the infected joint [44–55]. Favorable outcomes are generally achieved as long as surgical and antibiotic therapy are initiated for bacterial arthritis within 5 days of the clinical onset of infection [48, 49, 54, 56]. Septic arthritis of the shoulder and elbow in children are also associated with delay in recognition and intervention and therefore require a higher index of suspicion [57, 58]. Successful treatment of bacterial arthritis can be accomplished by either arthrotomy or arthroscopy depending on the surgeon's level of comfort with these techniques [59–63]. Following irrigation and debridement of the joint, a drain may be placed to allow continued evacuation of the joint space and is typically removed at bedside 2–3 days later. Primary closure of the wound is associated with shorter hospital stays with no adverse affect on outcomes [64]. While aspiration and lavage of the infected joint may act as a temporizing measure, it is recommended that formal irrigation and debridement of the involved joint be performed after bacterial arthritis has been confirmed, unless the clinical and laboratory improvement are substantial [56, 65]. Serial aspirations of bacterial arthritis are rarely performed due to the logistical challenge of repeatedly aspirating joints in children [65]. Children who do not demonstrate appropriate clinical and laboratory improvement within the first 96 h after joint irrigation and debridement may require repeat surgical intervention. The possibility of contiguous osteomyelitis or pyomyositis should be considered and can be evaluated using MRI prior to repeat surgery.

Levels of Evidence

The majority of literature on pediatric septic arthritis are Level III retrospective reviews, with few well designed prospective studies available (Table 42.2). There are also a significant number of case series with Level IV evidence. Based on the best available literature, septic arthritis is best diagnosed based on clinical exam (fever, refusal to bear weight and joint irritability) in combination with several inflammatory markers, including WBC, CRP, and ESR. CRP has the highest sensitivity and specificity. While children with septic arthritis have joint effusions on US, the mere presence of an effusion alone, in the absence of other clinical signs and symptoms of infection, can be a false positive (as in transient synovitis or Lyme arthritis). MRI is slowly replacing US as a better diagnostic test due to the ability of MRI to demonstrate the presence of contiguous infection. The gold standard for diagnosis of septic arthritis remains a culture positive joint aspirate. While synovial fluid cell counts >50,000 cells/mm^3 are often considered diagnostic of septic arthritis, there is no good evidence to support this number. There is a wide range of synovial fluid cell counts reported in cases of culture positive septic arthritis, as well as in cases of transient synovitis and Lyme arthritis, making this test difficult to interpret. Clinicians should have a high index of suspicion for K kingae and Lyme arthritis in the appropriate patient populations. K kingae is difficult to culture via standard culture methods and detection of this bacteria is enhanced with the addition of real-time PCR. All available literature recommends early arthrotomy and irrigation of septic joints in children to avoid devastating long term sequelae. There are no studies looking at the optimal timing of such intervention. Several studies, including the lone prospective randomized control trial (Level 1B evidence) suggests that arthroscopic debridement is as effective as open arthrotomy and drainage for the management of septic arthritis in children.

Osteomyelitis

Diagnosis

Children with acute hematogenous osteomyelitis (AHO) may present similarly to those with other osteoarticular

Table 42.2 Levels of evidence for pediatric acute hematogenous osteomyelitis

Author	Type of study	Level of evidence	Grade
Kan et al. [67]	RR	III	B
Ju et al. [102]	RR	III	B
Shrader et al. [103]	RR	III	B
Section et al. [104]	RR	III	B
Gwynne-Jones et al. [68]	CS	IV	C
Saavedra-Lozano et al. [69]	RR	III	B
Hawkshead et al. [70]	RR	III	B
Vander Have et al. [71]	CS	IV	C
Gafur et al. [72]	RR	III	B
Creel et al. [105]	CS	IV	C
Yamagishi et al. [106]	CS	IV	C
Hulten et al. [73]	PS	IIB	B
Kaplan et al. [81]	PS	IIB	B
Fergie et al. (2007)	RR	III	B
Naimi et al. [75]	PS	IIB	B
Herold et al. [76]	RR	III	B
Jungk et al. [77]	RR	III	B
Frederiksen et al. [107]	CS	IV	C
Ish-Horowicz et al. [108]	CS	IV	C
Wong et al. [109]	CS	IV	C
Goergens et al. [110]	CS	IV	C
Buckingham et al. [78]	RR	III	B
Martinez-Aguilar et al. [79]	RR	III	B
Gonzalez et al. [80]	CS	IV	C
Kaplan et al. [81]	PS	IIB	B
Zaoutis et al. [111]	RR	III	B
Arnold et al. [112]	RR	III	B
McCaskill et al. [83]	RR	III	B
Browne et al. [84]	RR	III	B
Zimbelman et al. [85]	RR	III	B
Frank et al. [88]	RR	III	B
Sattler et al. [89]	PS	IIB	B
Martinez-Aguilar et al. [90]	RR	III	B
Mishaan et al. [91]	PS	IIB	B
Chang et al. [92]	RR	III	B
Seal et al. [93]	CS	IV	C
Tanir et al. [94]	RR	III	B
Pannaraj et al. [95]	RR	III	B
Hasty et al. [96]	RR	III	B
Olesevich et al. [97]	RR	III	B
Tsuji et al. [98]	RR	III	B
Elliott et al. [99]	RR	III	B
Carrillo-Marquez et al. [100]	RR	III	B
Young et al. [101]	CS	IV	C
Hidayat et al. [113]	PS	IIA	B
Lundy et al. [114]	CS	IV	C
Trobs et al. [115]	CS	IV	C
Danielsson et al. [116]	CS	IV	C
Perlman et al. [117]	CS	IV	C
Aigner et al. [118]	RR	III	B

(continued)

Table 42.2 (continued)

Author	Type of study	Level of evidence	Grade
Unkila-Kallio et al. [119]	PS	IIB	B
Unkila-Kallio et al. [120]	PS	IIB	B
Roine et al. [121]	RR	III	B
Khachatourians et al. [122]	RR	III	B
Arnold et al. [82]	RR	III	B
Roine et al. [123]	RR	III	B
Paakkonen et al. [124]	PS	IIB	B
Copley et al. [125]	RR	III	B
Courtney et al. [126]	RR	III	B
Kan et al. [127]	RR	III	B
Tuason et al. [128]	RR	III	B
Chou et al. [129]	CS	IV	C
Paakkonen et al. [130]	CS	IV	C
Peltola et al. [131]	PSRC	IB	B
Peltola et al. [132]	PSRC	IB	B
Peltola et al. [133]	PSRC	IB	B
Peters et al. [135]	CS	IV	C

infections such as septic arthritis and pyomyositis. On clinical exam, they may have fever and exhibit refusal to bear weight, but typically lack the joint irritability of children with bacterial arthritis. Instead, they may have point tenderness to palpation of the affected bone. Inflammatory markers, including CRP, ESR, and WBC, will be elevated as in other musculoskeletal infections. The best imaging modality to diagnose AHO is MRI, which will demonstrate increased signal intensity within bone on T2 images, but decreased signal intensity on T1 sequences. MRI also has the advantage of also demonstrating contiguous sources of infection, such as sub-periosteal abscess, septic arthritis and pyomyositis [66]. While gadolinium should not be routinely used to evaluate for MSKI in children, contrasted MRI can increased the detection of small abscesses. In a study of 90 children who had MRI with and without contrast performed for evaluation of MSKI, 8 had abscesses requiring surgical intervention that were only identified on post-contrast images. No child with a normal pre-contrast study ended up with a diagnosis of MSKI [67]. Due to the increase in the prevalence of MRSA MSKI in some regions over the last two decades [68–101], one institution created a predictive algorithm to distinguish MRSA vs. MSSA AHO in children. Risk factors for MRSA AHO included hematocrit <34, WBC >12, CRP >13 mg/L and temperature >38. If all 4 factors are present, the risk of MRSA AHO is 92 % [102]. This algorithm, however, has been tested at other institutions with a higher incidence of MRSA infections and was found to have poor diagnostic performance, calling into question the utility of the predictive algorithm outside the original authors' institution [103].

Treatment

Antibiotics are considered the first line of therapy in children with AHO who have no evidence of abscess formation (intra-osseus, sub-periosteal, or extra-periosteal) on imaging and in whom there is no concern for developing sepsis. The timing of appropriate empiric antibiotic administration should balance the priorities of identifying the causative organism and avoiding the unnecessary delay of antibiotic administration. In a study of 860 children with pediatric MSKI, antibiotic exposure, either pre-hospital or within the authors' own institution, was not associated with a lower rate of culture positivity of material from the site of infection [104]. Antibiotics should therefore be held until blood cultures have been obtained and given any time thereafter depending on the clinical situation. Whenever there is clinical concern about the potential for disease dissemination or evolving sepsis (high fever, ill appearing, hemodynamic instability, severely elevated inflammatory markers), antibiotics may be given regardless of the timing of advanced imaging or surgery. If urgent aspiration or surgical debridement is planned after initial evaluation or advanced imaging, antibiotics may be held until local tissue culture is obtained or can be started once the decision is made to pursue conservative treatment without surgical intervention or obtainment of local tissue culture. Anaerobic, fungal and

AFB cultures should not be routinely performed during the initial evaluation of children with MSKI unless there is a history of immunocompromise, penetrating injury or failed primary treatment [104].

Empiric antibiotic therapy for children with MSKI is dependent upon the local microbiology of the community and surveillance monitoring must be done to ensure appropriate empiric antibiotic selection [68–77, 81, 105, 106]. Neonatal deep infection deserves special consideration as these infections are often acquired in the NICU while the newborn is exposed to invasive lines and catheters [107, 108]. Thus, neonates are at risk of exposure to hospital acquired pathogens, including multi-drug resistant methicillin resistant staphylococcus aureus (MRSA) and Candida Albicans [107, 108]. Neonates who are otherwise healthy and leave the hospital after birth may develop deep infection within a few weeks of birth [45, 109]. These neonates are at risk for infection due to Enterobacteriaceae or group B Streptococcus (GBS) to which they were exposed during delivery. Empiric antibiotic selection in the neonate should therefore cover gram negative organisms, GBS and S aureus [37, 45, 109, 110]. Outside of the neonatal group, the most common causative organism is S aureus [68–77, 81, 105, 106]. MRSA is now thought to be responsible for a sufficient percentage of culture positive cases of AHO in some regions to warrant clindamycin or vancomycin as the empiric antibiotic of choice in children [68, 69, 72–81, 83–101, 111, 112]. In some communities, there is increasing evidence of clindamycin resistance, which typically invokes the use of vancomycin [73]. Monitoring of local antibiotic resistance patterns is therefore essential to ensure that empiric antibiotic therapy is rational based on the local microbiologic epidemiology of the community. When vancomycin is used, there is evidence that using a higher dose protocol leads to better efficacy in cases of MRSA infection [113]. In the 6 month to 4 year old age group, ceftriaxone should be added to clindamycin for empiric treatment when K kingae infection is suspected [31, 36, 101, 114, 115]. When a patient appears to have developed sepsis (ICU, fever, bacteremia, multi-focal disease, and/or pulmonary involvement) empiric treatment should include high-dose vancomycin and ceftriaxone.

Surgery is indicated for children with AHO who demonstrate failure to respond to antibiotic therapy, evolving sepsis, or who have imaging findings consistent with abscess formation (intra-osseus, sub-periosteal, or extra-periosteal) (Fig. 42.2). Surgical treatment involves irrigation and debridement of infected or necrotic bone. This is done by making a cortical window in the region of affected bone with care taken to avoid injury to the growth plate or perichondral physeal ring. The bone window should be large enough to perform an adequate debridement but not compromise the overall architecture of the bone. Surgical decompression reduces intra-osseus pressure, which may restore perfusion to the affected area and enhance antibiotic delivery. One study demonstrated antegrade drilling of the femoral neck in children with AHO of the proximal femur to decrease the rate of contiguous septic arthritis and osteonecrosis [116]. Similar to septic arthritis, drains should be left in place to allow for evacuation of the space in the post-operative period. Pre-operative imaging must be studied carefully to identify all foci of infection. Any adjacent or contiguous abscesses (both sub-periosteal or extra-periosteal) should be drained at the time of bone decompression. Involvement of adjacent joints may occur in as many as 42 % of children with AHO [117]. If there is an effusion present in an adjacent joint, this should be aspirated at a minimum and evaluated for the presence of bacterial arthritis and/or irrigated and debrided when appropriate. Post-operative limitations on weight bearing and activity should be considered, depending on the location of infection and extent of debridement, to avoid pathologic fracture.

CRP and ESR levels should be monitored during the course of treatment of AHO to ensure appropriate response to treatment, including both antibiotics and surgery. The acute phase of AHO is accompanied by a rapid rise in CRP which declines at a moderate pace when the infection is being adequately treated. The ESR rises and declines more gradually, and may continue to rise even after treatment of the infection has been initiated [82, 118–124]. Monitoring of inflammatory markers should be accompanied by careful physical exam. Children who fail to demonstrate appropriate clinical or laboratory improvement after treatment has been initiated should be carefully evaluated to determine the need for repeat imaging with MRI. Repeat imaging in the early post-operative setting is advised only if the clinician feels there is a previously unaddressed focus (either local or remote) of infection. Otherwise, clinical and laboratory examinations should be used to guide the decision for repeat surgical intervention of known, or previously addressed foci [125–127]. In a series of 59 children with AHO, 104 repeat MRI studies were assessed regarding the indications for the imaging study and the impact on treatment. Twenty-eight of the MRIs were obtained within 2 weeks of the index imaging study because of a worsening clinical course. In this group of children a change in treatment occurred after 8 of the MRIs (29 %) compared with management changes in only 3 of the 76 MRIs (3.9 %) performed >14 days after the index study. Of the 11 children in whom repeat MRI changed the ultimate treatment plan, CRP levels were increasing in 7 and were elevated or

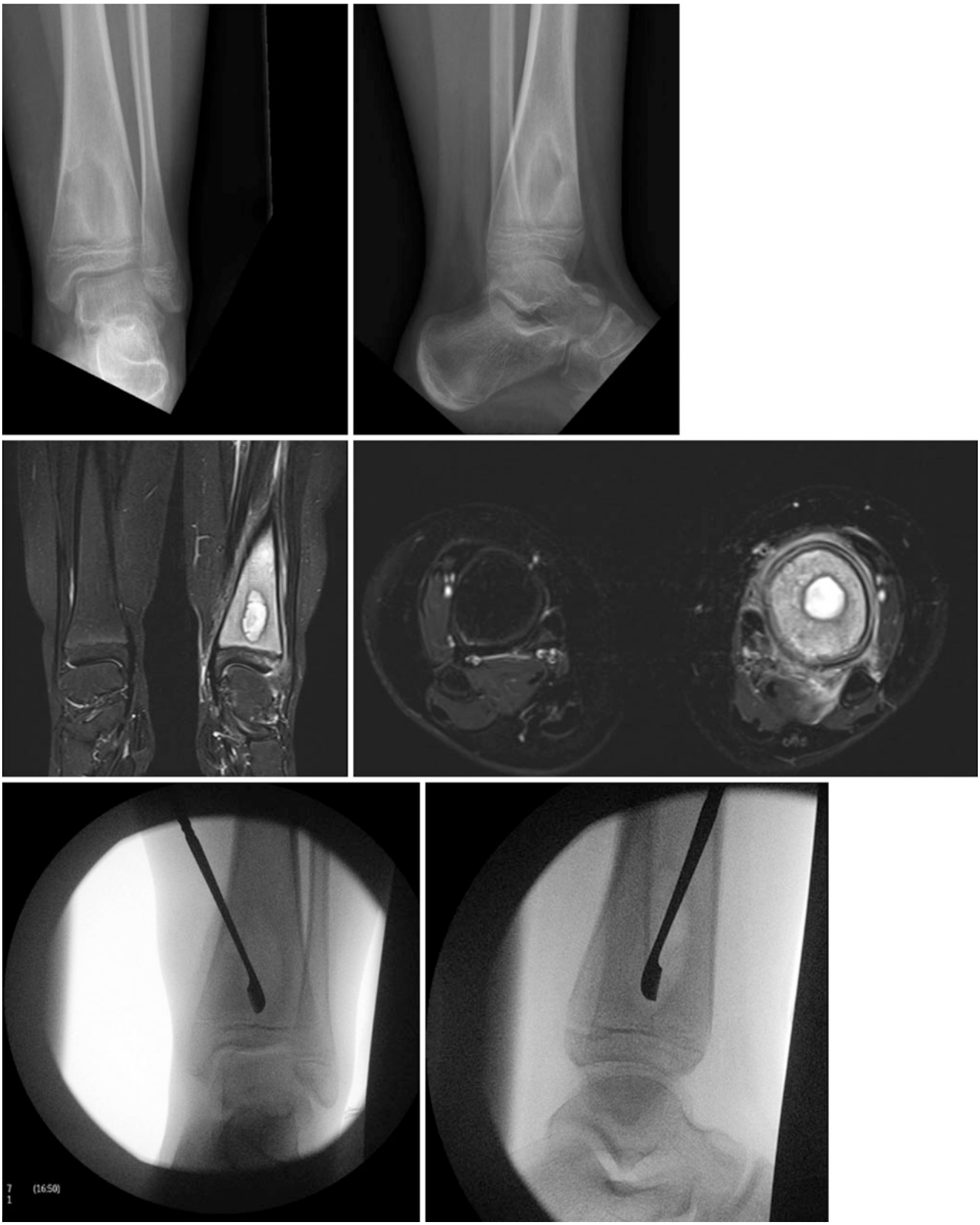

Fig. 42.2 Brodie's abscess treated with incision and drainage

unchanged in 4 [126]. Based on this is it not recommended that MRI be used in the routine monitoring of infection resolution as MRI in the aftermath of infection and surgical intervention has a prolonged appearance that is difficult to interpret. One study of 57 children with AHO showed that children with sustained elevation of CRP after 96 h after their initial surgery and who remain febrile while on antibiotic treatment have an increased likelihood of repeat surgical intervention and should be evaluated carefully for additional surgical treatment [128].

The convalescent phase of infection is accompanied by rapid decline and normalization of CRP (1–2 weeks) and more gradual decline of ESR (3–6 weeks). The patient can be considered for discharge and outpatient management with transition from parental to oral antibiotic therapy when there is sufficient clinical and laboratory improvement to ensure that the acute phase of the infection is over. In a study of 194 children with acute bacterial osteoarticular infections, a CRP of 3 mg/dL (in addition to improving clinical exam) was used as a guide for transitioning from parental to oral antibiotic therapy with 99.5 % success rate. Long term outcomes were similar to those expected with the more traditional extended parental therapy course [82]. Another study employed a CRP of 2 mg/dL as part of the criteria for discharge of children with osteomyelitis within a clinical practice guideline. They reported a shorter length of stay and lower hospital readmission rate among children in whom these criteria was applied [125]. Other institutions consider transition to outpatient care and oral antibiotic therapy when the CRP has reached 50 % of its peak value in addition to improvement in the clinical exam. In one study, 92 % of children with MSKI experienced a 50 % decline in CRP over 4 days [129]. While the clinical exam will not have *normalized*, there should be significant improvement in the following clinical parameters: temperature trend, inspection/palpation of site of infection for erythema, tenderness, swelling, range of motion, surgical site drainage and general appearance/demeanor of the child. The decision to terminate antibiotic therapy altogether is based on the normalization of laboratory markers and is typically dependent upon the normalization of ESR. The clinical exam should normalize over 6–12 weeks, depending on the location and extent of infection. If clinical and laboratory normalization does not occur as anticipated within a 3–6 week time frame, the clinician should continue antibiotic therapy and re-assess laboratory markers every 1–2 weeks. Failure to improve clinical or to normalize markers of infection should be considered a treatment failure and a repeat MRI should be considered to look for residual focus of infection or involucrum that is driving the ongoing inflammatory process. If identified, the clinical should perform repeat irrigation and debridement with new culture acquisition. Complications that can occur from osteomyelitis include: pathologic fracture, growth disturbance leading to limb length difference or deformity, joint contracture, osteonecrosis, spinal degeneration with loss of disc space and arthritis [130–135].

Levels of Evidence

Most studies on pediatric AHO are retrospective reviews or case series (Level III and IV evidence). There are however, a few well designed prospective studies (Level II) and even a few prospective randomize control studies available (Level 1). Based on the best available literature, pediatric AHO is best diagnosed based on clinical exam and elevated inflammatory markers (mainly ESR and CRP) in combination with MRI. MRI with contrast is not necessary to diagnose AHO in children. Although based mostly on Level III and IV evidence, there is evidence to suggest an increase in the number of MSKI due to MRSA (in some areas), as well as the severity of disease. Surgery is required when there is evidence of abscess formation (whether intra-osseous, extra-osseus or subperiosteal). Otherwise, AHO can be successfully treated with antibiotic therapy. Choice of antibiotics should be tailored to the local epidemiology and resistance profile of the institution. Level II evidence suggests that ESR and CRP can be used to monitor the response to treatment, with CRP decreasing to 50 % of peak values in 4 days. This is combination with improving clinical exam can be used as discharge criteria. Level I evidence suggests that most cases of AHO can be treated with a short duration of IV antibiotics, followed by 3–4 weeks of oral antibiotics.

Pyomyositis

Diagnosis

Pyomyositis in children occurs most commonly around the hip joint (hip pericapsular pyomyositis) (Figs. 42.3 and 42.4) and as such, children present with symptoms similar to a septic hip joint – joint irritability with limited ROM, refusal to bear weight, fever, and elevated inflammatory markers such as CRP, ESR, and WBC [15, 86, 95, 136–138]. Clinical exam and laboratory markers frequently fail to distinguish pyomyositis from other infectious etiologies (septic arthritis and AHO) and the diagnosis can be delayed while workup for these other conditions is performed. Pyomyositis is not visualized on plain radiographs and when involving the deep pericapsular musculature around a large joint, pyomyositis is difficult to visualize using US. Therefore, MRI is the most useful imaging modality for diagnosing pyomyositis and has

the advantage of detecting other foci of infection. In a study of 53 children presenting to the ED with an acutely irritable hip, pelvic pyomyositis was found to be twice as common as septic arthritis of the hip based on MRI [15]. In a similar study of 130 children with concern for septic arthritis of the hip, only 39 % were found to have an isolated septic joint while the rest had other areas of infection around the hip. Patients who had joint aspiration before advanced imaging were 2.8 times more likely to require a reoperation than those who had an MRI prior to surgical intervention [139].

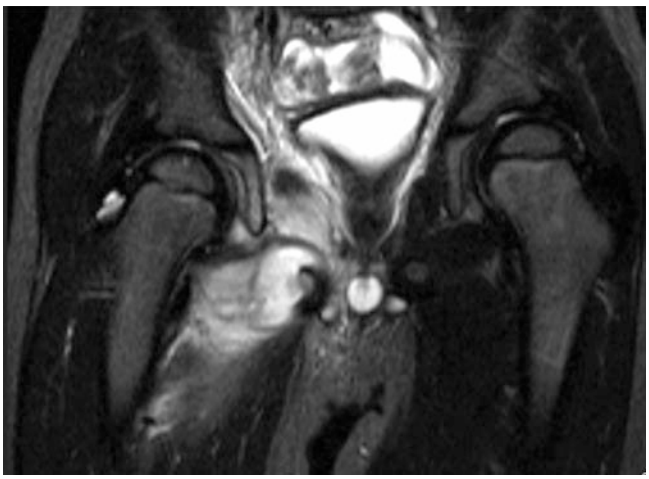

Fig. 42.3 Pyomyositis of psoas, iliacus, piriformis, and obturator internus

Scanner availability and the need for sedation are the two biggest barriers to the use of MRI in the acute setting. The use of limited MRI has been studied and found to be effective at diagnosing cases of pelvis pyomyositis [109]. In one study, a FAST-sequence MRI protocol was utilized to evaluate children for pelvic pyomyositis. The protocol is a non-contrasted, non-sedated study that can be done in less than 30 min and provides a coronal short T1 inversion recovery (STIR) sequence and an axial T2 sequence [15]. For patients with a clinical exam concerning for septic arthritis and elevated inflammatory markers, ideally an MRI should be obtained as quickly as possible to evaluate the hip and surrounding structures for infection. However, in a patient for whom the exam is clearly consistent with septic arthritis or who is becoming septic, treatment should not be delayed in order to obtain advanced imaging.

Treatment

In the early stages of pyomyositis with only inflammation or phlegmon formation on MRI and no evidence of frank abscess formation, the first line of treatment is empiric antibiotic therapy. As in other pediatric MSKI, Staphylococcus and Streptococcus are the most common causative organisms and empiric antibiotic therapy should cover these organisms [15, 86, 95, 136–138]. Once treatment is initiated, clinical exam and inflammatory markers should be monitored for signs of

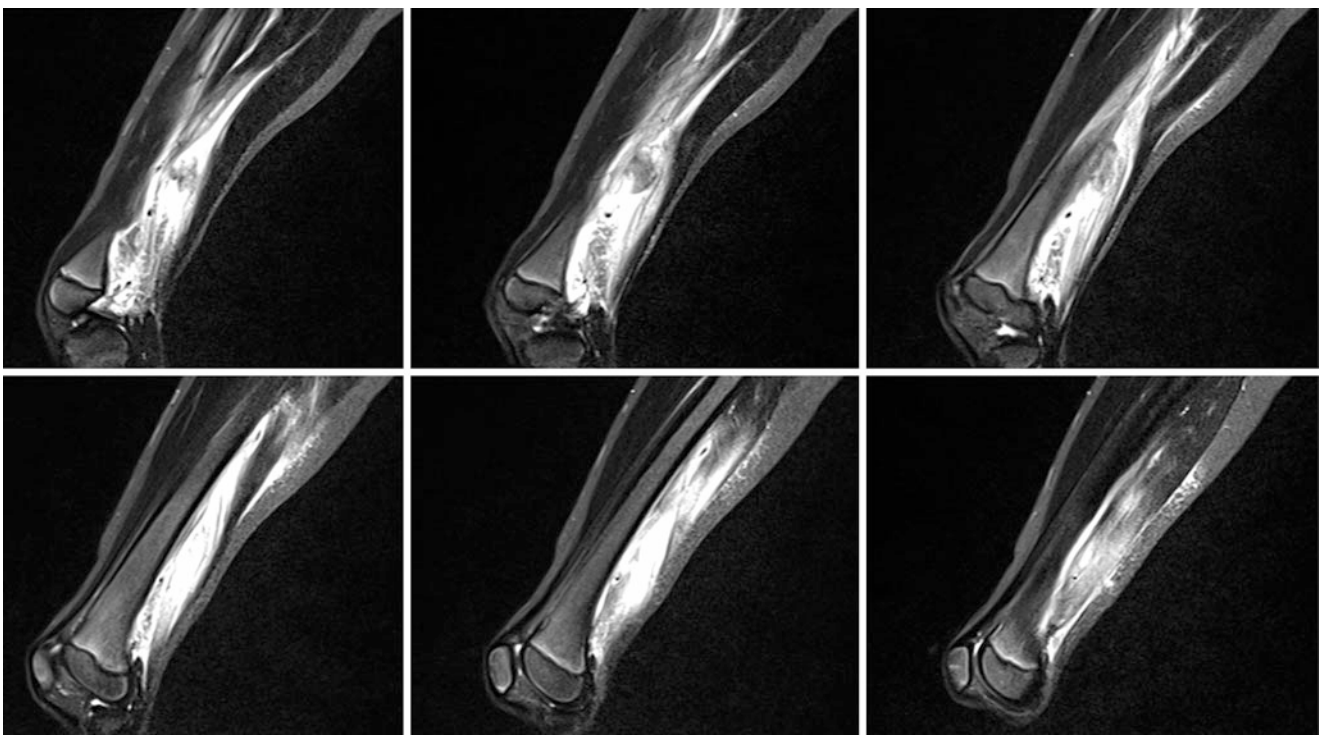

Fig. 42.4 Pyomyositis of the hamstring muscles

improvement. If there is no improvement after 2–3 days, repeat imaging should be considered to evaluate for progression of disease. When advanced imaging shows abscess formation, the mainstay of treatment is evacuation of the abscess [15, 86, 136]. This can be done in the operating room under general anesthesia with surgical debridement of the abscess, as well as any other foci of infection seen on MRI. A drain should be placed in the abscess cavity for 2–3 days postoperatively to allow for continued egress of fluid postoperatively. If the abscess is accessible by interventional radiology, this is an appropriate alternative to formal surgical debridement [138]. One caveat to this is pyomyositis affecting the obturator musculature, which occurs in nearly half of all cases of pelvic pyomyositis [15]. Because of the deep location of these muscles within the pelvis, abscesses in the obturator musculature are often not amenable to drainage by interventional radiology. One institution recently published their experience with a medial approach to safely access the obturator musculature for abscess decompression [140]. In one series, 70 % of pyomyositis cases required debridement despite the early initiation of antibiotic therapy. When MRI reveals a joint effusion in addition to pyomyositis, this finding should not be ignored. Joint aspiration should be performed to evaluate for septic arthritis.

Levels of Evidence

There is very poor evidence in the literature for pyomyositis in children (Table 42.3). Most of the evidence is Level IV (small case series). There are a few retrospective series (Level III) and only 1 prospective study, which provides only grade B evidence. One reason for the poor quality literature on pyomyositis in children is that this was previously thought to be a disease isolated only to the tropics, however, with increasing availability of MRI, we know realize that pyomyositis more common in temperate climates than previous thought. MRI is the best test to diagnose pyomyositis. The majority of cases occur in the pelvis, around the hip joint. Many children have contiguous infection, either septic arthritis or osteomyelitis. Surgical treatment is necessary when there is frank abscess formation, however, there is some level IV evidence that these abscesses can be drained percutaneously by interventional radiology.

Conclusion

Pediatric MSKI represents a wide spectrum of disease, from non-bacterial inflammatory conditions including transient synovitis, to more complex infection involving bone, muscle and/or joint tissue. MRI is becoming increasingly recognized as an important tool in the evaluation of children with MSKI due to its ability to quickly quantify the amount and location of infection. Care of children with MSKI can be complex, requiring the use of precious hospital resources (MRI, operating room time, etc.) as well as the coordination of multiple different services – pediatrics, infectious disease and pediatric orthopaedic surgery. Despite this, early recognition and prompt treatment lead to favorable outcomes in the majority of cases.

A summary of evidence for pediatric MSKI is given in Table 42.4.

Future Questions

Pediatric MSKI leads to significant hospital resource utilization worldwide. Effective clinical practice guidelines are necessary to guide care and maximize the use of hospital resources. Based on the literature, it is clear that there is significant variation in regional epidemiology and clinical practice guidelines for children with MSKI. For example, there is a significant burden of pediatric MSKI caused by S aureus in the southeast, while, in the northeast, less invasive organisms, such as B burgdorferi, are more common. In order to improve care, reduce costs and streamline the use of hospital resources for children with MSKI, prospective multicenter studies need to be performed to determine the regional variations in epidemiology and practice patterns in order to come up with the most efficient and accurate clinical practice guidelines to treat children with MSKI.

Table 42.3 Levels of evidence for pediatric pyomyositis

Author	Type of study	Level of evidence	Grade
Mignemi et al. [15]	PS	IIB	B
Gubbay et al. [86]	CS	IV	C
Pannaraj et al. [95]	RR	III	B
Karmazyn et al. [136]	CS	IV	C
Mitsionis et al. [137]	CS	IV	C
Spiegel et al. [138]	CS	IV	C
Gottschalk et al. [139]	RR	III	B
Menge et al. [140]	RR	III	B

Table 42.4 Summary evidence for pediatric MSKI

Author	Grade	Recommendation
Mignemi et al. [15]	B	MRI is the modality of choice to distinguish pericapsular pyomyositis from septic arthritis of the hip
Heyworth et al. [19]	B	Septic arthritis of the hip is the most common diagnosis with synovial fluid WBC counts between 25,000 and 75,000 cells/mm^3
Chometon et al. [27]	B	K kingae is a common cause of osteoarticular infections and is best identified using real time PCR
El-sayed et al. [60]	B	Septic arthritis can be adequately treated by both arthroscopy and traditional arthrotomy
Kan et al. [67]	B	MRI with contrast should not be routinely used for the workup of non-spine MSI
Unkila-Kallio et al. (1994)	B	CRP and ESR are best tests to diagnosis MSKI and to follow the patient's response to treatment
Copley et al. [125]	B	Evidence-based treatment guidelines applied by a multidisciplinary team resulted in better care of children with MSKI
Peltola et al. [131]	B	AHO can be treated with short period of IV antibiotic, followed by oral antibiotic for 3–4 weeks
Peltola et al. [132]	B	Clindamycin or first-generation cephalosporin can be used with equal efficacy for to treat culture positive MSKI
Gafur et al. [72]	B	Prevalence of MRSA-AHO has increased over the last two decades, empiric antibiotic choice should reflect local epidemiology and antibiotic resistance patterns

References

1. Luhmann SJ, Jones A, Schootman M, Gordon JE, Schoenecker PL, Luhmann JD. Differentiation between septic arthritis and transient synovitis of the hip in children with clinical prediction algorithms. J Bone Joint Surg. 2004;86-A(5):956–62.
2. Li SF, Cassidy C, Chang C, Gharib S, Torres J. Diagnostic utility of laboratory tests in septic arthritis. Emerg Med J. 2007;24(2):75–7. doi:10.1136/emj.2006.037929.
3. Kocher MS, Mandiga R, Murphy JM, et al. A clinical practice guideline for treatment of septic arthritis in children: efficacy in improving process of care and effect on outcome of septic arthritis of the hip. J Bone Joint Surg. 2003;85-A(6):994–9.
4. Eich GF, Superti-Furga A, Umbricht FS, Willi UV. The painful hip: evaluation of criteria for clinical decision-making. Eur J Pediatr. 1999;158(11):923–8.
5. Klein DM, Barbera C, Gray ST, Spero CR, Perrier G, Teicher JL. Sensitivity of objective parameters in the diagnosis of pediatric septic hips. Clin Orthop Relat Res. 1997;338:153–9.
6. Zamzam MM. The role of ultrasound in differentiating septic arthritis from transient synovitis of the hip in children. J Pediatr Orthop B. 2006;15(6):418–22. doi:10.1097/01.bpb.0000228388.32184.7f.
7. Caird MS, Flynn JM, Leung YL, Millman JE, D'Italia JG, Dormans JP. Factors distinguishing septic arthritis from transient synovitis of the hip in children. A prospective study. J Bone Joint Surg. 2006;88(6):1251–7. doi:10.2106/JBJS.E.00216.
8. Levine MJ, McGuire KJ, McGowan KL, Flynn JM. Assessment of the test characteristics of C-reactive protein for septic arthritis in children. J Pediatr Orthop. 2003;23(3):373–7.
9. Jung ST, Rowe SM, Moon ES, Song EK, Yoon TR, Seo HY. Significance of laboratory and radiologic findings for differentiating between septic arthritis and transient synovitis of the hip. J Pediatr Orthop. 2003;23(3):368–72.
10. Sultan J, Hughes PJ. Septic arthritis or transient synovitis of the hip in children: the value of clinical prediction algorithms. J Bone Joint Surg Br. 2010;92(9):1289–93. doi:10.1302/0301-620X.92B9.24286.
11. Gordon JE, Huang M, Dobbs M, Luhmann SJ, Szymanski DA, Schoenecker PL. Causes of false-negative ultrasound scans in the diagnosis of septic arthritis of the hip in children. J Pediatr Orthop. 2002;22(3):312–6.
12. Kang S-N, Sanghera T, Mangwani J, Paterson JMH, Ramachandran M. The management of septic arthritis in children: systematic review of the English language literature. J Bone Joint Surg Br. 2009;91(9):1127–33. doi:10.1302/0301-620X.91B9.22530.
13. Mathews CJ, Kingsley G, Field M, et al. Management of septic arthritis: a systematic review. Ann Rheum Dis. 2007;66(4):440–5. doi:10.1136/ard.2006.058909.
14. Kocher MS, Zurakowski D, Kasser JR. Differentiating between septic arthritis and transient synovitis of the hip in children: an evidence-based clinical prediction algorithm. J Bone Joint Surg. 1999;81(12):1662–70.
15. Mignemi ME, Menge TJ, Cole HA, et al. Epidemiology, diagnosis, and treatment of pericapsular pyomyositis of the hip in children. J Pediatr Orthop. 2014;34(3):316–25. doi:10.1097/BPO.0000000000000106.
16. Monsalve J, Kan JH, Schallert EK, Bisset GS, Zhang W, Rosenfeld SB. Septic arthritis in children: frequency of coexisting unsuspected osteomyelitis and implications on imaging work-up and management. AJR Am J Roentgenol. 2015;204(6):1289–95. doi:10.2214/AJR.14.12891.
17. Rosenfeld S, Bernstein D, Daram S, Dawson J, Zhang W. Predicting the presence of adjacent infections in septic arthritis in children. J Pediatr Orthop. 2015;1. doi:10.1097/BPO.0000000000000389.
18. Bombaci H, Canbora K, Onur G, Görgeç M, Döşoğlu N. Diagnosis of septic arthritis and initial antibiotic treatment. Acta Orthop Traumatol Turc. 2005;39(4):307–15.
19. Heyworth BE, Shore BJ, Donohue KS, Miller PE, Kocher MS, Glotzbecker MP. Management of pediatric patients with synovial fluid white blood-cell counts of 25,000 to 75,000 cells/mm^3 after aspiration of the hip. J Bone Joint Surg Am. 2015;97(5):389–95. doi:10.2106/JBJS.N.00443.

20. Bachman DT, Srivastava G. Emergency department presentations of Lyme disease in children. Pediatr Emerg Care. 1998; 14(5):356–61.
21. Rose CD, Fawcett PT, Eppes SC, Klein JD, Gibney K, Doughty RA. Pediatric Lyme arthritis: clinical spectrum and outcome. J Pediatr Orthop. 1994;14(2):238–41.
22. Willis AA, Widmann RF, Flynn JM, Green DW, Onel KB. Lyme arthritis presenting as acute septic arthritis in children. J Pediatr Orthop. 2003;23(1):114–8.
23. Milewski MD, Cruz AI, Miller CP, Peterson AT, Smith BG. Lyme arthritis in children presenting with joint effusions. J Bone Joint Surg Am. 2011;93(3):252–60. doi:10.2106/JBJS.I.01776.
24. Yagupsky P, Dubnov-Raz G, Gené A, Ephros M, Israeli-Spanish Kingella kingae Research Group. Differentiating Kingella kingae septic arthritis of the hip from transient synovitis in young children. J Pediatr. 2014;165(5):985–989.e1. doi:10.1016/j.jpeds.2014.07.060.
25. Yagupsky P, Porsch E, St Geme JW. Kingella kingae: an emerging pathogen in young children. Pediatrics. 2011;127(3):557–65. doi:10.1542/peds.2010-1867.
26. Cherkaoui A, Ceroni D, Emonet S, Lefevre Y, Schrenzel J. Molecular diagnosis of Kingella kingae osteoarticular infections by specific real-time PCR assay. J Med Microbiol. 2009;58(Pt 1):65–8. doi:10.1099/jmm.0.47707-0.
27. Chometon S, Benito Y, Chaker M, et al. Specific real-time polymerase chain reaction places Kingella kingae as the most common cause of osteoarticular infections in young children. Pediatr Infect Dis J. 2007;26(5):377–81. doi:10.1097/01.inf.0000259954.88139.f4.
28. Basmaci R, Lorrot M, Bidet P, et al. Comparison of clinical and biologic features of Kingella kingae and Staphylococcus aureus arthritis at initial evaluation. Pediatr Infect Dis J. 2011;30(10):902–4. doi:10.1097/INF.0b013e31821fe0f7.
29. Basmaci R, Ilharreborde B, Lorrot M, Bidet P, Bingen E, Bonacorsi S. Predictive score to discriminate Kingella kingae from Staphylococcus aureus arthritis in France. Pediatr Infect Dis J. 2011;30(12):1120–1–authorreply1121–2. doi:10.1097/INF.0b013e31822ce97e.
30. Ceroni D, Cherkaoui A, Combescure C, François P, Kaelin A, Schrenzel J. Differentiating osteoarticular infections caused by Kingella kingae from those due to typical pathogens in young children. Pediatr Infect Dis J. 2011;30(10):906–9. doi:10.1097/INF.0b013e31821c3aee.
31. Dubnov-Raz G, Scheuerman O, Chodick G, Finkelstein Y, Samra Z, Garty B-Z. Invasive Kingella kingae infections in children: clinical and laboratory characteristics. Pediatrics. 2008;122(6):1305–9. doi:10.1542/peds.2007-3070.
32. Yagupsky P, Press J. Unsuspected Kingella kingae infections in afebrile children with mild skeletal symptoms: the importance of blood cultures. Eur J Pediatr. 2004;163(9):563–4. doi:10.1007/s00431-004-1496-8.
33. Lacour M, Duarte M, Beutler A, Auckenthaler R, Suter S. Osteoarticular infections due to Kingella kingae in children. Eur J Pediatr. 1991;150(9):612–8.
34. Mallet C, Ceroni D, Litzelmann E, et al. Unusually severe cases of Kingella kingae osteoarticular infections in children. Pediatr Infect Dis J. 2014;33(1):1–4. doi:10.1097/INF.0b013e3182a22cc6.
35. Dubnov-Raz G, Ephros M, Garty B-Z, et al. Invasive pediatric Kingella kingae Infections: a nationwide collaborative study. Pediatr Infect Dis J. 2010;29(7):639–43. doi:10.1097/INF.0b013e3181d57a6c.
36. Yagupsky P, Bar-Ziv Y, Howard CB, Dagan R. Epidemiology, etiology, and clinical features of septic arthritis in children younger than 24 months. Arch Pediatr Adolesc Med. 1995;149(5): 537–40.
37. Moumile K, Merckx J, Glorion C, Pouliquen JC, Berche P, Ferroni A. Bacterial aetiology of acute osteoarticular infections in children. Acta Paediatr. 2005;94(4):419–22.
38. Fenollar F, Lévy P-Y, Raoult D. Usefulness of broad-range PCR for the diagnosis of osteoarticular infections. Curr Opin Rheumatol. 2008;20(4):463–70. doi:10.1097/BOR.0b013e3283032030.
39. Moumile K, Merckx J, Glorion C, Berche P, Ferroni A. Osteoarticular infections caused by Kingella kingae in children: contribution of polymerase chain reaction to the microbiologic diagnosis. Pediatr Infect Dis J. 2003;22(9):837–9.
40. Ilharreborde B, Bidet P, Lorrot M, et al. New real-time PCR-based method for Kingella kingae DNA detection: application to samples collected from 89 children with acute arthritis. J Clin Microbiol. 2009;47(6):1837–41. doi:10.1128/JCM.00144-09.
41. Lehours P, Freydiere AM, Richer O, et al. The rtxA toxin gene of Kingella kingae: a pertinent target for molecular diagnosis of osteoarticular infections. J Clin Microbiol. 2011;49(4):1245–50. doi:10.1128/JCM.01657-10.
42. Rosey A-L, Abachin E, Quesnes G, et al. Development of a broad-range 16S rDNA real-time PCR for the diagnosis of septic arthritis in children. J Microbiol Methods. 2007;68(1):88–93. doi:10.1016/j.mimet.2006.06.010.
43. Carter K, Doern C, Jo C-H, Copley LAB. The clinical usefulness of polymerase chain reaction as a supplemental diagnostic tool in the evaluation and the treatment of children with septic arthritis. J Pediatr Orthop. 2015;1. doi:10.1097/BPO.0000000000000411.
44. Bos CF, Mol LJ, Obermann WR. Tjin a Ton ER. Late sequelae of neonatal septic arthritis of the shoulder. J Bone Joint Surg Br. 1998;80(4):645–50.
45. Narang A, Mukhopadhyay K, Kumar P, Bhakoo ON. Bone and joint infection in neonates. Indian J Pediatr. 1998;65(3): 461–4.
46. Chang CH, Huang SC. Reconstruction for sequelae of septic hip in children. J Formos Med Assoc. 1997;96(5):353–8.
47. Saisu T, Kawashima A, Kamegaya M, Mikasa M, Moriishi J, Moriya H. Humeral shortening and inferior subluxation as sequelae of septic arthritis of the shoulder in neonates and infants. J Bone Joint Surg. 2007;89(8):1784–93. doi:10.2106/JBJS.E.01021.
48. Nunn TR, Cheung WY, Rollinson PD. A prospective study of pyogenic sepsis of the hip in childhood. J Bone Joint Surg Br. 2007;89(1):100–6. doi:10.1302/0301-620X.89B1.17940.
49. Yuan H-C, Wu K-G, Chen C-J, Tang R-B, Hwang B-T. Characteristics and outcome of septic arthritis in children. J Microbiol Immunol Infect. 2006;39(4):342–7.
50. Cheng JC, Lam TP. Femoral lengthening after type IVB septic arthritis of the hip in children. J Pediatr Orthop. 1996;16(4): 533–9.
51. Lejman T, Strong M, Michno P, Hayman M. Septic arthritis of the shoulder during the first 18 months of life. J Pediatr Orthop. 1995;15(2):172–5.
52. Strong M, Lejman T, Michno P, Hayman M. Sequelae from septic arthritis of the knee during the first two years of life. J Pediatr Orthop. 1994;14(6):745–51.
53. Umer M, Hashmi P, Ahmad T, Ahmed M, Umar M. Septic arthritis of the hip in children–Aga Khan University Hospital experience in Pakistan. J Pak Med Assoc. 2003;53(10):472–8.
54. Kabak S, Halici M, Akcakus M, Cetin N, Narin N. Septic arthritis in patients followed-up in neonatal intensive care unit. Pediatr Int. 2002;44(6):652–7.
55. Forlin E, Milani C. Sequelae of septic arthritis of the hip in children: a new classification and a review of 41 hips. J Pediatr Orthop. 2008;28(5):524–8. doi:10.1097/BPO.0b013e31817bb079.
56. Chen CE, Ko JY, Li CC, Wang CJ. Acute septic arthritis of the hip in children. Arch Orthop Trauma Surg. 2001;121(9):521–6.

57. Belthur MV, Palazzi DL, Miller JA, Phillips WA, Weinberg J. A clinical analysis of shoulder and hip joint infections in children. J Pediatr Orthop. 2009;29(7):828–33. doi:10.1097/BPO.0b013e3181b76a91.
58. Bowakim J, Marti R, Curto A. Elbow septic arthritis in children: clinical presentation and management. J Pediatr Orthop B. 2010;19(3):281–4. doi:10.1097/BPB.0b013e3283387d2d.
59. Jeon I-H, Choi C-H, Seo J-S, Seo K-J, Ko S-H, Park J-Y. Arthroscopic management of septic arthritis of the shoulder joint. J Bone Joint Surg. 2006;88(8):1802–6. doi:10.2106/JBJS.E.00917.
60. El-Sayed AMM. Treatment of early septic arthritis of the hip in children: comparison of results of open arthrotomy versus arthroscopic drainage. J Child Orthop. 2008;2(3):229–37. doi:10.1007/s11832-008-0094-0.
61. Stutz G, Kuster MS, Kleinstück F, Gächter A. Arthroscopic management of septic arthritis: stages of infection and results. Knee Surg Sports Traumatol Arthrosc. 2000;8(5):270–4.
62. Kim S-J, Choi N-H, Ko S-H, Linton JA, Park H-W. Arthroscopic treatment of septic arthritis of the hip. Clin Orthop Relat Res. 2003;407:211–4.
63. Vispo Seara JL, Barthel T, Schmitz H, Eulert J. Arthroscopic treatment of septic joints: prognostic factors. Arch Orthop Trauma Surg. 2002;122(4):204–11. doi:10.1007/s00402-001-0386-z.
64. Gordon JE, Wolff A, Luhmann SJ, Ortman MR, Dobbs MB, Schoenecker PL. Primary and delayed closure after open irrigation and debridement of septic arthritis in children. J Pediatr Orthop B. 2005;14(2):101–4.
65. Griffet J, Oborocianu I, Rubio A, Leroux J, Lauron J, Hayek T. Percutaneous aspiration irrigation drainage technique in the management of septic arthritis in children. J Trauma. 2011;70(2):377–83. doi:10.1097/TA.0b013e31820318f0.
66. Pugmire BS, Shailam R, Gee MS. Role of MRI in the diagnosis and treatment of osteomyelitis in pediatric patients. WJR. 2014;6(8):530–7. doi:10.4329/wjr.v6.i8.530.
67. Kan JH, Young RS, Yu C, Hernanz-Schulman M. Clinical impact of gadolinium in the MRI diagnosis of musculoskeletal infection in children. Pediatr Radiol. 2010;40(7):1197–205. doi:10.1007/s00247-010-1557-2.
68. Gwynne-Jones DP, Stott NS. Community-acquired methicillin-resistant Staphylococcus aureus: a cause of musculoskeletal sepsis in children. J Pediatr Orthop. 1999;19(3):413–6.
69. Saavedra-Lozano J, Mejías A, Ahmad N, et al. Changing trends in acute osteomyelitis in children: impact of methicillin-resistant Staphylococcus aureus infections. J Pediatr Orthop. 2008;28(5):569–75. doi:10.1097/BPO.0b013e31817bb816.
70. Hawkshead JJ, Patel NB, Steele RW, Heinrich SD. Comparative severity of pediatric osteomyelitis attributable to methicillin-resistant versus methicillin-sensitive Staphylococcus aureus. J Pediatr Orthop. 2009;29(1):85–90. doi:10.1097/BPO.0b013e3181901c3a.
71. Vander Have KL, Karmazyn B, Verma M, et al. Community-associated methicillin-resistant Staphylococcus aureus in acute musculoskeletal infection in children: a game changer. J Pediatr Orthop.2009;29(8):927–31.doi:10.1097/BPO.0b013e3181bd1e0c.
72. Gafur OA, Copley LAB, Hollmig ST, Browne RH, Thornton LA, Crawford SE. The impact of the current epidemiology of pediatric musculoskeletal infection on evaluation and treatment guidelines. J Pediatr Orthop. 2008;28(7):777–85. doi:10.1097/BPO.0b013e318186eb4b.
73. Hulten KG, Kaplan SL, Gonzalez BE, et al. Three-year surveillance of community onset health care-associated Staphylococcus aureus infections in children. Pediatr Infect Dis J. 2006;25(4):349–53. doi:10.1097/01.inf.0000207404.50143.1e.
74. Fergie J, Purcell K. The epidemic of methicillin-resistant Staphylococcus aureus colonization and infection in children: effects on the community, health systems, and physician practices. Pediatr Ann. 2007;36(7):404–12.
75. Naimi TS, LeDell KH, Como-Sabetti K, et al. Comparison of community - and health care-associated methicillin-resistant Staphylococcus aureus infection. JAMA. 2003;290(22):2976–84. doi:10.1001/jama.290.22.2976.
76. Herold BC, Immergluck LC, Maranan MC, et al. Community-acquired methicillin-resistant Staphylococcus aureus in children with no identified predisposing risk. JAMA. 1998;279(8):593–8.
77. Jungk J, Como-Sabetti K, Stinchfield P, Ackerman P, Harriman K. Epidemiology of methicillin-resistant Staphylococcus aureus at a pediatric healthcare system, 1991–2003. Pediatr Infect Dis J. 2007;26(4):339–44. doi:10.1097/01.inf.0000257452.58182.e1.
78. Buckingham SC, McDougal LK, Cathey LD, et al. Emergence of community-associated methicillin-resistant Staphylococcus aureus at a Memphis, Tennessee Children's Hospital. Pediatr Infect Dis J. 2004;23(7):619–24.
79. Martínez-Aguilar G, Avalos-Mishaan A, Hulten K, Hammerman W, Mason EO, Kaplan SL. Community-acquired, methicillin-resistant and methicillin-susceptible Staphylococcus aureus musculoskeletal infections in children. Pediatr Infect Dis J. 2004;23(8):701–6.
80. Gonzalez BE, Martínez-Aguilar G, Hulten KG, et al. Severe Staphylococcal sepsis in adolescents in the era of community-acquired methicillin-resistant Staphylococcus aureus. Pediatrics. 2005;115(3):642–8. doi:10.1542/peds.2004-2300.
81. Kaplan SL, Hulten KG, Gonzalez BE, et al. Three-year surveillance of community-acquired Staphylococcus aureus infections in children. Clin Infect Dis. 2005;40(12):1785–91. doi:10.1086/430312.
82. Arnold JC, Cannavino CR, Ross MK, et al. Acute bacterial osteoarticular infections: eight-year analysis of C-reactive protein for oral step-down therapy. Pediatrics. 2012;130(4):e821–8. doi:10.1542/peds.2012-0220.
83. McCaskill ML, Mason EO, Kaplan SL, Hammerman W, Lamberth LB, Hulten KG. Increase of the USA300 clone among community-acquired methicillin-susceptible Staphylococcus aureus causing invasive infections. Pediatr Infect Dis J. 2007;26(12):1122–7. doi:10.1097/INF.0b013e31814536e0.
84. Browne LP, Mason EO, Kaplan SL, Cassady CI, Krishnamurthy R, Guillerman RP. Optimal imaging strategy for community-acquired Staphylococcus aureus musculoskeletal infections in children. Pediatr Radiol. 2008;38(8):841–7. doi:10.1007/s00247-008-0888-8.
85. Zimbelman J, Palmer A, Todd J. Improved outcome of clindamycin compared with beta-lactam antibiotic treatment for invasive Streptococcus pyogenes infection. Pediatr Infect Dis J. 1999;18(12):1096–100.
86. Gubbay AJ, Isaacs D. Pyomyositis in children. Pediatr Infect Dis J. 2000;19(10):1009–12–quiz1013.
87. Garron E, Viehweger E, Launay F, Guillaume JM, Jouve JL, Bollini G. Nontuberculous spondylodiscitis in children. J Pediatr Orthop. 2002;22(3):321–8.
88. Frank AL, Marcinak JF, Mangat PD, et al. Clindamycin treatment of methicillin-resistant Staphylococcus aureus infections in children. Pediatr Infect Dis J. 2002;21(6):530–4.
89. Sattler CA, Mason EO, Kaplan SL. Prospective comparison of risk factors and demographic and clinical characteristics of community-acquired, methicillin-resistant versus methicillin-susceptible Staphylococcus aureus infection in children. Pediatr Infect Dis J. 2002;21(10):910–7. doi:10.1097/01.inf.0000034268.59915.6f.
90. Martínez-Aguilar G, Hammerman WA, Mason EO, Kaplan SL. Clindamycin treatment of invasive infections caused by community-acquired, methicillin-resistant and methicillin-susceptible Staphylococcus aureus in children. Pediatr Infect Dis J. 2003;22(7):593–8. doi:10.1097/01.inf.0000073163.37519.ee.

91. Mishaan AMA, Mason EO, Martínez-Aguilar G, et al. Emergence of a predominant clone of community-acquired Staphylococcus aureus among children in Houston. Texas Pediatr Infect Dis J. 2005;24(3):201–6.
92. Chang W-S, Chiu N-C, Chi H, Li W-C, Huang F-Y. Comparison of the characteristics of culture-negative versus culture-positive septic arthritis in children. J Microbiol Immunol Infect. 2005; 38(3):189–93.
93. Seal J, Glynn L, Statter M, Liu D. A high prevalence of methicillin-resistant Staphylococcus aureus among surgically drained soft-tissue infections in pediatric patients. Pediatr Surg Int. 2006;22(8):683–7. doi:10.1007/s00383-006-1684-x.
94. Tanir G, Tonbul A, Tuygun N, Aydemir C, Ertan U. Soft tissue infections in children: a retrospective analysis of 242 hospitalized patients. Jpn J Infect Dis. 2006;59(4):258–60.
95. Pannaraj PS, Hulten KG, Gonzalez BE, Mason EO, Kaplan SL. Infective pyomyositis and myositis in children in the era of community-acquired, methicillin-resistant Staphylococcus aureus infection. Clin Infect Dis. 2006;43(8):953–60. doi:10.1086/507637.
96. Hasty MB, Klasner A, Kness S, et al. Cutaneous community-associated methicillin-resistant Staphylococcus aureus among all skin and soft-tissue infections in two geographically distant pediatric emergency departments. Acad Emerg Med. 2007;14(1):35–40. doi:10.1197/j.aem.2006.08.008.
97. Olesevich M, Kennedy A. Emergence of community-acquired methicillin-resistant Staphylococcus aureus soft tissue infections. J Pediatr Surg. 2007;42(5):765–8. doi:10.1016/j.jpedsurg.2006.12.027.
98. Tsuji BT, Rybak MJ, Cheung CM, Amjad M, Kaatz GW. Community- and health care-associated methicillin-resistant Staphylococcus aureus: a comparison of molecular epidemiology and antimicrobial activities of various agents. Diagn Microbiol Infect Dis. 2007;58(1):41–7. doi:10.1016/j.diagmicrobio.2006.10.021.
99. Elliott DJ, Zaoutis TE, Troxel AB, Loh A, Keren R. Empiric antimicrobial therapy for pediatric skin and soft-tissue infections in the era of methicillin-resistant Staphylococcus aureus. Pediatrics. 2009;123(6):e959–66. doi:10.1542/peds.2008-2428.
100. Carrillo-Marquez MA, Hulten KG, Hammerman W, Mason EO, Kaplan SL. USA300 is the predominant genotype causing Staphylococcus aureus septic arthritis in children. Pediatr Infect Dis J. 2009;28(12):1076–80. doi:10.1097/INF.0b013e3181adbcfe.
101. Young TP, Maas L, Thorp AW, Brown L. Etiology of septic arthritis in children: an update for the new millennium. Am J Emerg Med. 2011;29(8):899–902. doi:10.1016/j.ajem.2010.04.008.
102. Ju KL, Zurakowski D, Kocher MS. Differentiating between methicillin-resistant and methicillin-sensitive Staphylococcus aureus osteomyelitis in children: an evidence-based clinical prediction algorithm. J Bone Joint Surg Am. 2011;93(18):1693–701. doi:10.2106/JBJS.J.01154.
103. Wade Shrader M, Nowlin M, SEGAL LS. Independent analysis of a clinical predictive algorithm to identify methicillin-resistant Staphylococcus aureus osteomyelitis in children. J Pediatr Orthop. 2013;33(7):759–62. doi:10.1097/BPO.0b013e3182a11cf7.
104. Section J, Gibbons SD, Barton T, Greenberg DE, Jo C-H, Copley LAB. Microbiological culture methods for pediatric musculoskeletal infection: a guideline for optimal use. J Bone Joint Surg Am. 2015;97(6):441–9. doi:10.2106/JBJS.N.00477.
105. Creel AM, Durham SH, Benner KW, Alten JA, Winkler MK. Severe invasive community-associated methicillin-resistant Staphylococcus aureus infections in previously healthy children. Pediatr Crit Care Med. 2009;10(3):323–7. doi:10.1097/PCC.0b013e3181988798.
106. Yamagishi Y, Togawa M, Shiomi M. Septic arthritis and acute hematogenous osteomyelitis in childhood at a tertiary hospital in Japan. Pediatr Int. 2009;51(3):371–6. doi:10.1111/j.1442-200X.2008.02740.x.
107. Frederiksen B, Christiansen P, Knudsen FU. Acute osteomyelitis and septic arthritis in the neonate, risk factors and outcome. Eur J Pediatr. 1993;152(7):577–80.
108. Ish-Horowicz MR, McIntyre P, Nade S. Bone and joint infections caused by multiply resistant Staphylococcus aureus in a neonatal intensive care unit. Pediatr Infect Dis J. 1992;11(2):82–7.
109. Wong M, Isaacs D, Howman-Giles R, Uren R. Clinical and diagnostic features of osteomyelitis occurring in the first three months of life. Pediatr Infect Dis J. 1995;14(12):1047–53.
110. Goergens ED, McEvoy A, Watson M, Barrett IR. Acute osteomyelitis and septic arthritis in children. J Paediatr Child Health. 2005;41(1–2):59–62. doi:10.1111/j.1440-1754.2005.00538.x.
111. Zaoutis TE, Toltzis P, Chu J, et al. Clinical and molecular epidemiology of community-acquired methicillin-resistant Staphylococcus aureus infections among children with risk factors for health care-associated infection: 2001–2003. Pediatr Infect Dis J. 2006; 25(4):343–8. doi:10.1097/01.inf.0000207403.67197.cc.
112. Arnold SR, Elias D, Buckingham SC, et al. Changing patterns of acute hematogenous osteomyelitis and septic arthritis: emergence of community-associated methicillin-resistant Staphylococcus aureus. J Pediatr Orthop. 2006;26(6):703–8. doi:10.1097/01.bpo.0000242431.91489.b4.
113. Hidayat LK, Hsu DI, Quist R, Shriner KA, Wong-Beringer A. High-dose vancomycin therapy for methicillin-resistant Staphylococcus aureus infections: efficacy and toxicity. Arch Intern Med. 2006;166(19):2138–44. doi:10.1001/archinte.166.19.2138.
114. Lundy DW, Kehl DK. Increasing prevalence of Kingella kingae in osteoarticular infections in young children. J Pediatr Orthop. 1998;18(2):262–7.
115. Tröbs R, Möritz R, Bühligen U, et al. Changing pattern of osteomyelitis in infants and children. Pediatr Surg Int. 1999;15(5–6): 363–72.
116. Danielsson LG, Düppe H. Acute hematogenous osteomyelitis of the neck of the femur in children treated with drilling. Acta Orthop Scand. 2002;73(3):311–6. doi:10.1080/000164702320155310.
117. Perlman MH, Patzakis MJ, Kumar PJ, Holtom P. The incidence of joint involvement with adjacent osteomyelitis in pediatric patients. J Pediatr Orthop. 2000;20(1):40–3.
118. Aigner RM, Fueger GF, Vejda M. Follow-up of osteomyelitis of infants with systemic serum parameters and bone scintigraphy. Nuklearmedizin. 1996;35(4):116–21.
119. Unkila-Kallio L, Kallio MJ, Eskola J, Peltola H. Serum C-reactive protein, erythrocyte sedimentation rate, and white blood cell count in acute hematogenous osteomyelitis of children. Pediatrics. 1994;93(1):59–62.
120. Unkila-Kallio L, Kallio MJ, Peltola H. The usefulness of C-reactive protein levels in the identification of concurrent septic arthritis in children who have acute hematogenous osteomyelitis. A comparison with the usefulness of the erythrocyte sedimentation rate and the white blood-cell count. J Bone Joint Surg. 1994;76(6):848–53.
121. Roine I, Faingezicht I, Arguedas A, Herrera JF, Rodríguez F. Serial serum C-reactive protein to monitor recovery from acute hematogenous osteomyelitis in children. Pediatr Infect Dis J. 1995;14(1):40–4.
122. Khachatourians AG, Patzakis MJ, Roidis N, Holtom PD. Laboratory monitoring in pediatric acute osteomyelitis and septic arthritis. Clin Orthop Relat Res. 2003;409(409):186–94. doi:10.1097/01.blo.0000057991.41099.47.
123. Roine I, Arguedas A, Faingezicht I, Rodríguez F. Early detection of sequela-prone osteomyelitis in children with use of simple clinical and laboratory criteria. Clin Infect Dis. 1997;24(5):849–53.
124. Pääkkönen M, Kallio MJT, Kallio PE, Peltola H. Sensitivity of erythrocyte sedimentation rate and C-reactive protein in childhood bone and joint infections. Clin Orthop Relat Res. 2010;468(3):861–6. doi:10.1007/s11999-009-0936-1.

125. Copley LAB, Kinsler MA, Gheen T, Shar A, Sun D, Browne R. The impact of evidence-based clinical practice guidelines applied by a multidisciplinary team for the care of children with osteomyelitis. J Bone Joint Surg Am. 2013;95(8):686–93. doi:10.2106/JBJS.L.00037.
126. Courtney PM, Flynn JM, Jaramillo D, Horn BD, Calabro K, Spiegel DA. Clinical indications for repeat MRI in children with acute hematogenous osteomyelitis. J Pediatr Orthop. 2010;30(8):883–7. doi:10.1097/BPO.0b013e3181fcbae5.
127. Kan JH, Hilmes MA, Martus JE, Yu C, Hernanz-Schulman M. Value of MRI after recent diagnostic or surgical intervention in children with suspected osteomyelitis. AJR Am J Roentgenol. 2008;191(5):1595–600. doi:10.2214/AJR.08.1115.
128. Tuason DA, Gheen T, Sun D, Huang R, Copley L. Clinical and laboratory parameters associated with multiple surgeries in children with acute hematogenous osteomyelitis. J Pediatr Orthop. 2014;34(5):565–70. doi:10.1097/BPO.0000000000000136.
129. Chou ACC, Mahadev A. The Use of C-reactive Protein as a Guide for Transitioning to Oral Antibiotics in Pediatric Osteoarticular Infections. J Pediatr Orthop. 2015;1. doi:10.1097/BPO.0000000000000427.
130. Pääkkönen M, Kallio PE, Kallio MJT, Peltola H. Management of osteoarticular infections caused by Staphylococcus aureus is similar to that of other etiologies: analysis of 199 staphylococcal bone and joint infections. Pediatr Infect Dis J. 2012;31(5):436–8. doi:10.1097/INF.0b013e31824657dd.
131. Peltola H, Pääkkönen M, Kallio P, Kallio MJT. Short – versus long-term antimicrobial treatment for acute hematogenous osteomyelitis of childhood. Pediatr Infect Dis J. 2010;29(12):1123–8. doi:10.1097/INF.0b013e3181f55a89.
132. Peltola H, Pääkkönen M, Kallio P, Kallio MJT. OM-SA study group. Clindamycin vs. first-generation cephalosporins for acute osteoarticular infections of childhood–a prospective quasi-randomized controlled trial. Clin Microbiol Infect. 2012;18(6):582–9. doi:10.1111/j.1469-0691.2011.03643.x.
133. Peltola H, Unkila-Kallio L, Kallio MJ. Simplified treatment of acute staphylococcal osteomyelitis of childhood. Finnish Study Group Pediatr. 1997;99(6):846–50.
134. Shen C-J, Wu M-S, Lin K-H, et al. The use of procalcitonin in the diagnosis of bone and joint infection: a systemic review and meta-analysis. Eur J Clin Microbiol Infect Dis. 2013;32(6):807–14. doi:10.1007/s10096-012-1812-6.
135. Peters W, Irving J, Letts M. Long-term effects of neonatal bone and joint infection on adjacent growth plates. J Pediatr Orthop. 1992;12(6):806–10.
136. Karmazyn B, Kleiman MB, Buckwalter K, Loder RT, Siddiqui A, Applegate KE. Acute pyomyositis of the pelvis: the spectrum of clinical presentations and MR findings. Pediatr Radiol. 2006;36(4):338–43. doi:10.1007/s00247-005-0082-1.
137. Mitsionis GI, Manoudis GN, Lykissas MG, et al. Pyomyositis in children: early diagnosis and treatment. J Pediatr Surg. 2009;44(11):2173–8. doi:10.1016/j.jpedsurg.2009.02.053.
138. Spiegel DA, Meyer JS, Dormans JP, Flynn JM, Drummond DS. Pyomyositis in children and adolescents: report of 12 cases and review of the literature. J Pediatr Orthop. 1999;19(2):143–50.
139. Gottschalk HP, Moor MA, Muhamad AR, Wenger DR, Yaszay B. Improving diagnostic efficiency: analysis of pelvic MRI versus emergency hip aspiration for suspected hip sepsis. J Pediatr Orthop. 2014;34(3):300–6. doi:10.1097/BPO.0000000000000097.
140. Menge TJ, Cole HA, Mignemi ME, et al. Medial approach for drainage of the obturator musculature in children. J Pediatr Orthop. 2014;34(3):307–15. doi:10.1097/BPO.0000000000000118.

Evidence-Based Treatment of Simple Bone Cyst

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Abstract

Simple bone cyst is a common benign lesion of bone. Although it is considered to be benign (non cancerous), it can cause fractures, deformities and psychosocial problems. Several treatments have been advocated with variable success, and the search for better treatments is ongoing. In this chapter, we have summarised the evidence that underpin current treatments.

Keywords

Simple bone cyst • Unicameral bone cyst • Solitary bone cyst • Benign bone tumours • Steroid injection • Bone marrow injection • Pathological fractures • Children tumours

Background

A simple bone cyst (SBC) is a common, benign, fluid-filled lesion of bone. The true prevalence is unknown as many are asymptomatic. It constitutes approximately 3 % of all bone tumors. It is often described as a unicameral (single chamber) cyst; however multiple septations may be present. There is a male: female preponderance of 3:1 [1]. The most common sites for SBC are the proximal humerus and proximal femur which account for nearly 90 % of all SBC. However, any bone may be affected [2–4].

The aetiology remains largely unknown. There are several aetiological theories. The vascular theory was one of the earliest; it postulated that a localized blockage of the drainage of interstitial fluid from cancellous bone is the cause for SBC [5]. A study in seven patients found a higher pressure inside the SBC compared to the contralateral normal bone marrow. Also, the oxygen tension in the cyst fluid was lower than that in venous or arterial samples taken at the same time. These authors suggested that venous obstruction within the bone was the likely cause of such SBC [6].

The cells and biochemistry contents of the cyst fluid have also been the subject of research with increased prostaglandin E_2 levels demonstrated in the cyst fluid [7–9]. Steroids have been reported to inhibit prostaglandin synthesis [10, 11] and this may help explain the beneficial effect of steroid injection in treatments. High levels of oxygen free radicals, IL-1 and an increased lysosomal enzyme activity have been found in SBC and this may play a pathological role in bone erosion, expanding the cyst and increasing the pressure inside the cyst [12–14].

Another theory suggested that SBC may be an intraosseous synovial cyst or entrapped synovial tissue in the bone. Both synovial type A and type B cells were found in the lining of SBC [15, 16]. More recently genetic abnormalities have been described in children with SBC [17].

Cysts can be considered to be in a latent or active phase based on their proximity to the growth plate. A cyst that is juxtaphyseal (<0.5 cm from the physis) is considered active. Epiphyseal involvement is rare, but if present may increase risk of growth disturbance. A cyst that has grown away from the plate is considered latent. Latent cysts continue to have

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growth potential, as proved by their recurrence after treatment [18].

Patients with a SBC usually present with a pathologic fracture or pain. Some patients with SBCs are asymptomatic and discovered incidentally. A plain radiograph is usually diagnostic when the typical features of cyst are present. SBCs are well defined geographic lucent lesions with narrow zone of transition, centrally located and show sclerotic margin in majority of cases with no periosteal reaction or soft tissue component. They sometimes expand the bone with thinning of the endosteum without any breach of the cortex unless there is a pathologic fracture. If present, the fallen-fragment, or "fallen leaf" (a bony fragment that breaks free and falls to the bottom of the cyst), can aid the radiographic diagnosis of a SBC. The fallen leaf sign is found in approximately 20 % of patients who present with a pathologic fracture [19, 20]. MRI is indicated when atypical features such as periosteal reaction, eccentric site, extending to the articular surface or soft tissue shadow. Cases of pseudocystic osteosarcoma or low-grade central osteosarcoma that were mistaken for SBC have been reported [21].

Several treatments variations have been reported in the literature, these can be simply classified into:

1. Non interventional treatment.
2. Intralesional injection such as (corticosteroids [9, 22–24], autologous bone marrow [2, 25], demineralized bone matrix [26], calcium sulfate pellets [27–29] and fibrin sealant [30]).
3. Disruption/curettage of cyst lining or cyst wall with or without grafting [31–33].
4. Decompression of the cyst such as multiple drill holes [13, 34] and cannulated screws [35, 36]. In addition to decompressing and opening the cyst to the intramedullary canal [37], flexible intramedullary nailing provides structural support [36, 38, 39].
5. Subtotal resection with or without bone grafting and total resection [40–43]. These are regarded as historical treatments and are associated with surgical complications and are rarely, if ever, used today [18].
6. Various combinations of the above interventions.

Healing is not often well defined, particularly in old studies. The success rate of healing varies dramatically according to series and intervention.

To better define "healing" of SBC, Neer et al. [44] introduced a grading system which has been modified by several authors [45–47]. Neer's grading consists of:

- Excellent: complete obliteration of the cyst
- Residual defect: one or more static, cyst like residual with good bone strength on x-ray
- Reoperation: subsequent operation required by recurrence

Neer emphasised that incomplete obliteration of the cyst after operation appears to be of little clinical significance, provided there is good bone strength.

Wright et al. [48] conducted a randomised controlled trial comparing intralesional bone marrow and steroid injections for simple bone cysts [49], introducing a new version of the grading system (reverse of Neer's grading):

- Grade 1: cyst clearly visible
- Grade 2: cyst visible, but multilocular and opaque
- Grade 3: sclerosis around or within a partially visible cyst
- Grade 4: complete healing with obliteration of cyst

In the following sections we answer commonly asked clinical questions about SBC substantiating the answers with published evidence.

What Is the Natural History of Simple Bone Cyst?

The common belief is that SBC is benign and tend to improve or resolve in late adolescence. In a study of the natural history of SBC in 57 patients (58 cyst) [50], Kaelin showed that these cysts caused significant number of recurrent fractures. In their series, 31 children with humeral cysts sustained a total of 50 fractures (average 1.6 fractures a child; range 0–5). Twenty-one children with femoral cysts sustained a total of 14 fractures (average 0.7 fractures a child; range 0–2). They observed the natural history of SBC in 11 untreated humeral cysts that were followed for more than 1 year. Spontaneous healing occurred in five cases only (<50 %), who had sustained a total of nine fractures.

Donaldson and Wright [51] evaluated 24 subjects with SBC who participated in a prior randomized clinical trial [49] but had not healed at trial conclusion. Eighteen (75 %) cysts were located in the humerus and 4 (25 %) in the femur. Patients were followed for 7.0 ± 1.0 years following initial treatment with a mean age at follow-up of 17.2 ± 3.2 years and 14 (87 %) of growth plates were closed. Of the 24 subjects, none were graded as completely healed at time of follow-up.

Several studies showed that SBCs resulted in growth disturbance in up to 10 % of patients [52–57] leading to angular deformity or limb length discrepancy (Figs. 43.1 and 43.2). Although simple bone cyst is oncologically not malignant, its course is far from being innocent.

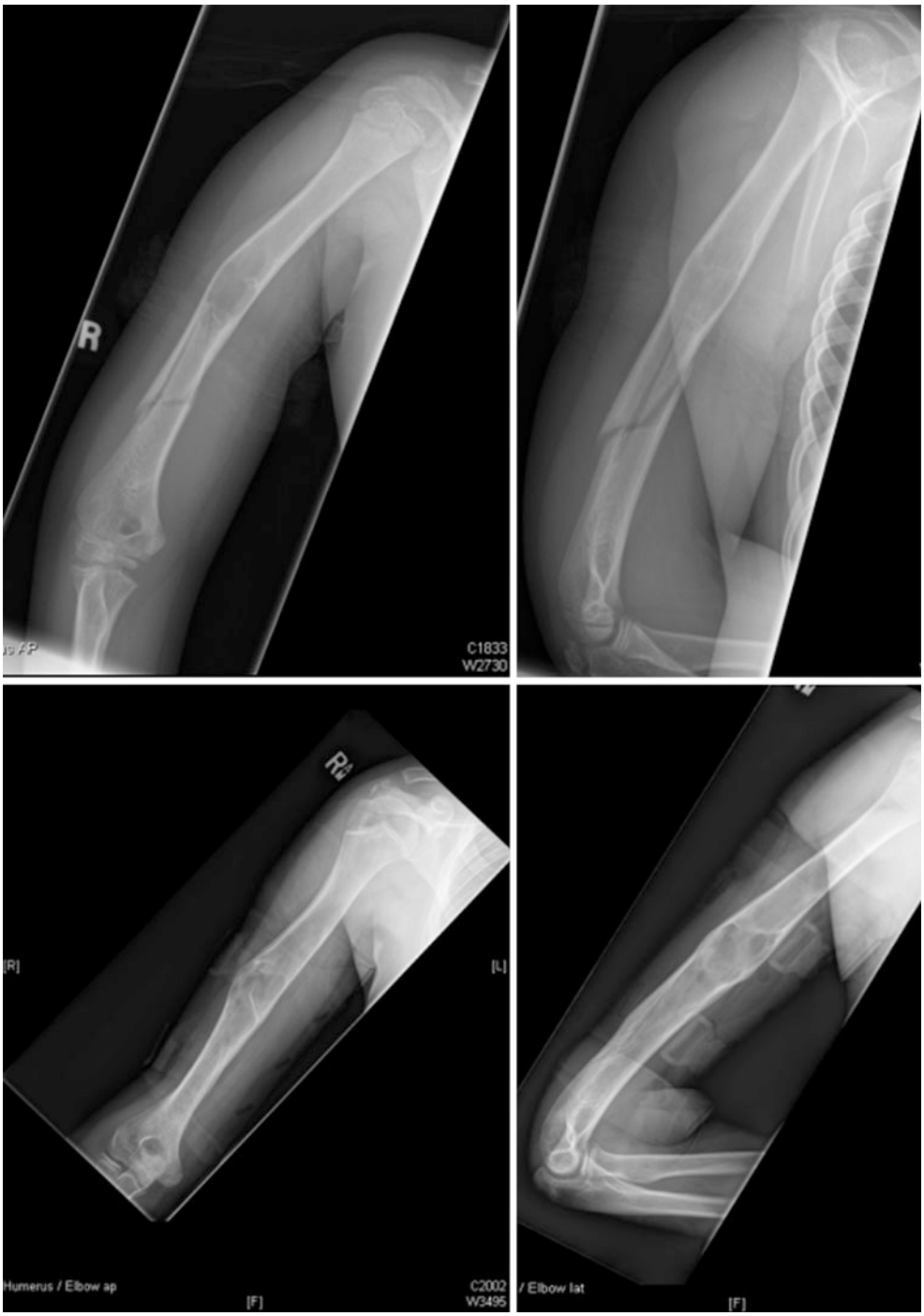

Fig. 43.1 This 13 old boy with a simple bone cyst of the humerus had 5 pathological fractures over 5 years and the cyst did not heal

Do Simple Bone Cysts Heal Spontaneously After a Pathological Fracture?

A simple bone cyst seldom heals after a pathological fracture. While abundant callous may initially form, it tends to resorb after 6 months and there is a low likelihood of the cyst healing following a fracture (Fig. 43.3). Garceau and Gregory [58] showed a 15 % healing rate after a fracture. Others found less than 5 % [59–61]. Moreover, Neer [44] found 70 % of the cysts developed another fracture within 2 years.

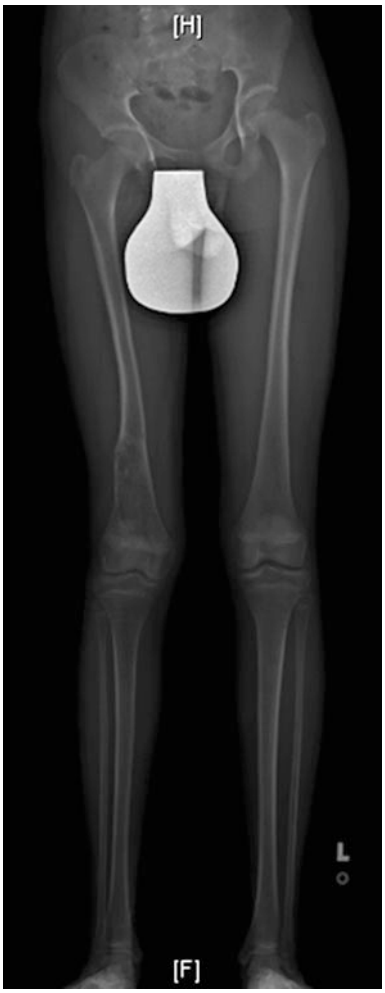

Fig. 43.2 A simple bone cyst caused pathological fracture and subsequent growth arrest at the right distal femur physis

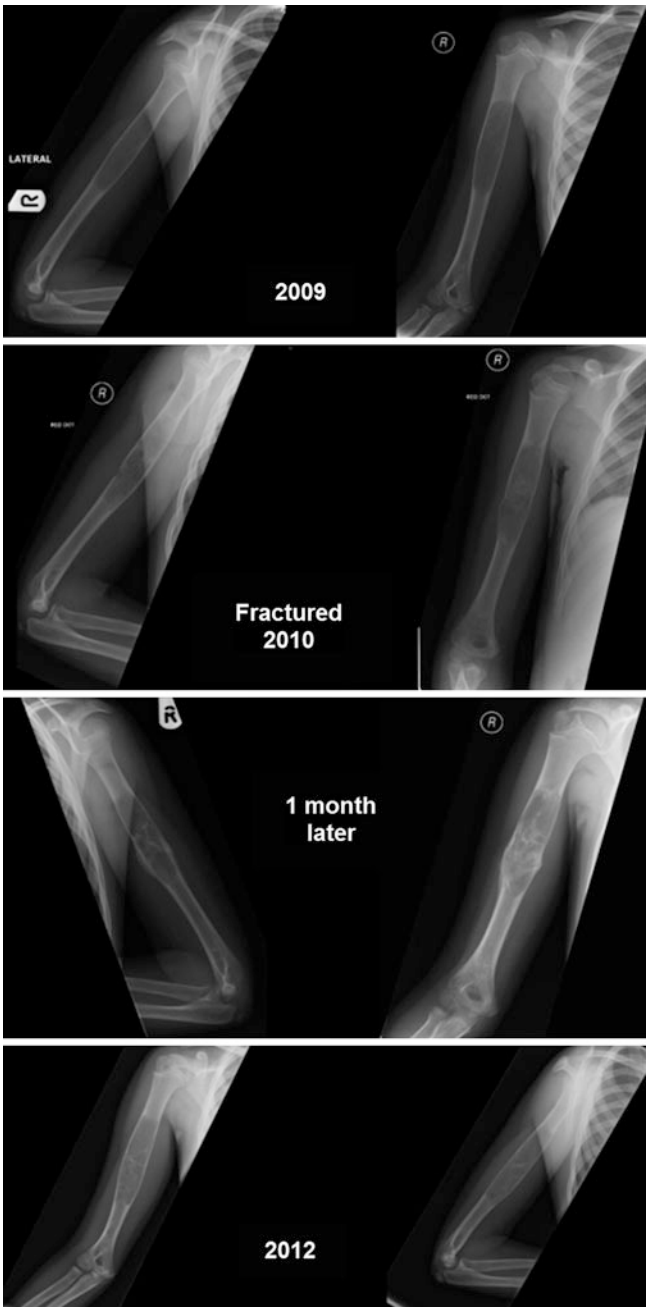

Fig. 43.3 This young boy was treated non-operatively for simple bone cyst. He had a pathological fracture in January 2010. After a month, plain x-ray showed the cyst was filling with bone; however, the cyst was larger after a year (see also Fig. 43.7)

Can We Predict Simple Bone Cysts that Could Cause Problems Such as Fractures or Deformities?

Researchers have been trying to identify patients and/or cyst features that predict outcomes in SBC. Several factors were investigated including but not limited to the size and site of the cyst, symptoms, previous fractures, patients' age, sex, types of interventions and length of follow-up. The literature is contradictory on virtually all these prognostic factors. Issues included definition of prognostic factors, duration of follow-up (cyst often recurs after signs of initial healing), definition of healing, lack of multivariate analysis and lack of blinding.

Kaelin and MacEwen [50] found that the larger the cyst, the more cortex was destroyed and the bone weakened. To quantify the strength of the remaining cortex, which is related to the size of the cyst and the size of the involved bone, they devised the cyst index. This gives the proportion between the radiographic area of the cyst and the size of the involved bone, measured as the diameter of the diaphysis squared:

Cyst Index = Area of the cyst/Diaphysis diameter^2

A low cyst index indicates a small cyst area in relation to the bone; conversely, a high index indicates a large cyst. The

smallest cyst in their series was measured 0.1, the largest 12.78. Recent pathological fractures were found in 53 radiographs and the cyst index was measured in each. The average index in humeral fractures was 6.12 (SD = 1.8) and in femoral fractures 4.74 (SD = 1.19). The humeral index is higher because these fractures occurred later than those in the lower limb.

Before the first fracture, the children are normally active, but once the diagnosis is made activity is often reduced, and for a subsequent fracture the trauma is likely to be less than in the first instance. The index would then be larger, and the results confirmed this hypothesis. However, as discussed below the change in cyst index could be due to the natural history of the cyst and/or reaction to the fracture.

In 21 initial fractures through humeral cysts, the average index was 5.38 (SD = 1.02), and in 14 recurrent fractures the index was 7.38 (SD = 2.4) which was a significant difference ($P < 0.005$). Interestingly it was shown that the cyst index does not show any tendency towards regression after a fracture, in fact the cyst often continues to grow in parallel to the healing process. In these cases the slope of the increase remained in the same range as before fracture, showing a rise of about two points a year. Thus a fracture did not change the natural course of the development of a cyst. Kaelin reported that there were no fractures in 40 patients who had an index lower than 3.5 and who did not curtail their activities.

In all 57 included patients, Kaelin found no further increase in the index occurred after its spontaneous. Cysts with at least two consecutive decreased indices of <3 and a cortical wall thicker >2 mm are considered to be healed. When these conditions are present, fractures never occur and the result is always favourable.

A recent study questioned the reliability of the cyst index as a predictor of fracture [62]. The cyst index of 32 femoral and humeral SBCs was determined by 8 reviewers. The risk group cyst index was compared with whether a fracture took place. Sensitivities, specificities, and positive and negative predictive values were calculated across varying cutoff levels. Intra-observer and inter-observer reliability testing for 10 cysts was also made. The mean value for the cyst index was significantly different for different observers ($P < 0.05$). The sensitivities, specificities, and positive and negative predictive values were small; particularly for the femoral cysts (0.5, 0.33, 0.33 and 0.50 respectively). The study conclusion did not validate the cyst index to be an accurate predictor of fracture. One significant weakness of the study was the short follow-up of 1.4 years.

Ahn and Park [59] carried out a retrospective review of 75 children with SBC to determine which cysts were likely to be at risk and whether healing was accelerated after a fracture. They found the percentage of bone occupied by the cyst in the transverse plane was more than 85 % in both AP and lateral radiographs in every case of pathological fracture. In most cases, the cyst recurred and sometimes became large without any acceleration of healing after fracture.

In another study, Leong et al. [63] investigated the use of quantitative computed tomography-based structural analysis to predict fractures in children with a benign appendicular skeletal lesion between 2002 and 2007. The resistance of the affected bone to compressive, bending, and torsional loads was calculated with rigidity analysis performed with the use of serial transaxial quantitative computed tomography data obtained along the length of the bone containing the lesion and from homologous cross sections through the contralateral, normal bone. At each cross section, the ratio of the structural rigidity of the affected bone to that of the normal, contralateral bone was determined.

Thirty-four of 41 of included individuals completed activity questionnaire at least 2 years after the quantitative computed tomography-based rigidity analysis. None of the patients for whom no increased fracture risk had been predicted by the rigidity analysis sustained a fracture, even though they had not received surgical treatment.

In conclusion, quantitative CT may be the most accurate method of predicting fracture but even this method has false positives and false negatives and does involve risks associated with ionising radiation.

What Are the Indications to Treat Simple Bone Cyst?

Parent and child psycho-social attitude to treatments or activity restriction usually guide indication for surgery. Families may choose observation for an asymptomatic humeral SBC with satisfactory cortical thickness. For cysts that are gradually increasing in size, causing expansions and progressive thinning of the cortex with imminent threat of pathological fracture, families may want surgical treatment.

Some authors have suggested the use of a cyst index aimed at predicting the future risk of a pathologic fracture as shown above. Kaelin et al. recommended observation for humeral cysts with an index of less than 4 and for femoral cysts with an index of less than 3.5. As noted above the value of these is uncertain [62].

What Is the Best Evidence We Have to Support the Current Treatment Options for Simple Bone Cyst?

Treatments of SBC have evolved over time; we present treatments and the underlying evidence, from a historical perspective.

Curettage and Grafting

The lining of SBC is believed to play a role in its formation. Hence removing the lining is a logical action for treatment. Aggressive curettage of the lining was practiced and often this was combined with using phenol or zinc chloride to further destroy the cyst lining. Curettage was often followed by bone grafting. Graft materials were initially autograft. Allograft and other non-biologic materials were also used (no donor site morbidity, less operating time and potentially unlimited quantity).

Neer et al. [44] reported on 175 SBC treated between 1930 and 1960 with a minimum follow-up of 2 years. One hundred and twenty nine cysts were treated by curettage and bone grafting within 1 year of the diagnosis. Twenty-seven underwent chemical cauterisation using phenol or zinc chloride and 44 did not. The results were excellent in 15 (55 %), residual defect in 4 (15 %) and re-operation in 8 (30 %). Forty-four cysts were not cauterised with phenol or zinc chloride and the results were excellent in 21(48 %), residual defect in 17 (39 %) and re-operation in 6 (13 %). He concluded that chemical cauterisation was not of value.

In 93 cysts of the proximal humerus or femur, comparison between bone autograft and allograft was possible. In 35 treated with autograft the results were excellent in 21 (60 %), residual defect in 6 (17 %) and reoperations in 8 (23 %). In 58 cysts that treated with allograft the results were excellent in 28 (48 %), residual defect in 12 (21 %), and reoperation in 8 (36 %) favouring autograft over allograft.

Neer also demonstrated that the results of treatment differed depending upon the specific bone involved; cysts present in infrequent locations heal far better than those present in common locations such as humerus and femur. The incidence of re-operations was 30 % in the upper ends of the humerus, 17 % in the proximal femur, 11 % in the upper end of the tibia and none in the lower end of the tibia, whereas in the infrequent sites or unusual location, the incidence of recurrence was mostly 12 %.

Neer also found that true recurrence following surgical treatment is significantly more frequent in patients over 10 years old and age was a better prognostic criterion than the proximity of these cysts to the epiphyseal plate.

In another large study of 144 SBCs treated by curettage and grafting with freeze-dried, crushed, cortical-bone allograft, all had been followed from 12 to 48 months after surgery, Spence et al. [47] showed that 98 healed primarily and completely and 10 healed with a small, static, non- progressive residual defect, for an over-all rate of healing of 75 %. However there was a recurrence in 36 patients (25 %) which was quite apparent in most cases by 6 months. In none of the recurrent cysts did spontaneous healing occur after the recurrence had been established.

Spence also concluded that age, sex, size and site of the SBC, cyst activity and extent of cyst packing were important factors in cyst healing. In his series, in contrast to that of Neer, above, the rate of recurrence was higher in patients below the age of ten than in patients above that age. Of the 82 cysts in patients under 10 years old, 57 (70 %) healed and 25 (30 %) recurred, while of the 62 cysts in patients over ten, 51 (82 %) healed and 11 (18 %) failed to do so. The correlation between age and healing was consistent regardless of the activity (active or latent) of the cyst and the sex of the patient.

Eighty-one percent of the 103 cysts in male patients healed after the first allograft, while only 61 % of the 41 in female patients healed primarily. The recurrence rate in males was 19.4 % (20 of 103) in comparison to 39 % (16 of 41) in females. This recurrence rate was not related to the location, size, or type of cyst.

The size of the cyst was classified as small if its longest diameter was less than one-eighth of the length of the involved bone; medium, if the length was from one-eighth to one third of the bone length; and large, if its maximum dimension was longer than one-third of the bone length. The incidence of postoperative healing was 73 % for the small, 64 % for the medium and 82 % for the large cysts. Authors stated that low incidence of healing in the medium-sized cysts could not be explained by differences in distribution with respect to sex, age, cyst type, or cyst site. However, testing these findings revealed the difference was not statistically significant (χ^2: P = 0.26).

All cysts in non tubular bones (6 in number) were fully healed. While 73 % of the 78 humeral cysts, 77 % of the 43 femoral cysts healed primarily and 67 % of the 12 tibial cysts healed.

Thirty-one (53 %) of the 59 active cysts and 75 (91 %) of the 85 latent cysts were healed after allografting. The degree of cyst packing was also correlated directly with the healing results since 88 % of 110 cysts which were completely packed healed primarily while only 32 % of the 34 incompletely packed ones healed, giving an over-all recurrence rate of 68 % in cysts not completely packed at the time of surgery.

Other contemporary studies demonstrated that curettage and grafting as a primary treatment was associated with cyst recurrence and/or persistence [40, 64].

Subtotal Resection with and Without Grafting

A more aggressive surgical approach was used to overcome the high recurrence rate of curettage and grafting. Subtotal resection with bone grafting was first presented in a scientific exhibit at the Annual Meeting of The American Academy of Orthopaedic Surgeons in 1962. It involved sub-periosteal

excision of the cyst and the adjacent normal bone, followed by bone autograft harvested from the iliac crest or tibia. Freeze-dried allograft was also used in grafting large defects in younger children.

Fahey [40] published his findings in 20 patients who were treated with subtotal resection and bone grafting. There was only one recurrence (5 %). One patient developed growth arrest after surgery.

McKay and Nason [41] published their series of 21 patients who underwent subtotal resection but without bone grafting. They reported a 90 % (19/21) success rate. No infectious or neurovascular complications were reported, however six patients with humeral lesions and one with a femoral lesion, a fracture occurred at the time of operation as a result of the extensive decortications. The authors reported three patients with humeral growth disturbances before the surgery. To avoid the high rate of fractures, authors used internal fixation [42, 43].

In 1973, Scaglietti et al. [23, 24], an Italian surgeon introduced the approach of using steroids to treat SBC. He found that the chemical composition of fluid obtained from SBC was indistinguishable from that of a tissue transudate seen in joint arthritis [23]. On the basis of these observations, Scaglietti injected corticosteroids into these cysts. Depo-Medrol was chosen because it is a microcrystalline suspension of acetate of methylprednisolone that is relatively insoluble and, therefore, believed to have a prolonged pharmacological effect. Dr Scaglietti reported a 96 % healing success rate which started a new era of percutaneous treatment of simple bone cyst.

Percutaneous Techniques/Corticosteroid Injection

In 1979, Scaglietti published his series of 72 cysts treated with up to 6 injections of methylprednisolone acetate. Follow-up period was a minimum of 18 months; 60 % of patients healed completely, and 36 % healed "almost complete and only a 4 % failure to heal rate." In 1982, he published a larger series (163 cysts) with a longer follow. Complete obliteration of the cyst was noted in 55 % of the cases and recurrence of the lesion was rare. However, in 45 % of the cases in which there was some improvement following injection, e.g., thickening of the cortex, some areas of new bone formation within the cyst interspersed in areas with little evidence of repair, there was a higher "recurrence" rate, i.e., the lesion subsequently enlarged or a lesion of a significant size persisted. He also stated that complete repair of the cystic cavity was observed mainly in patients younger than 11 years who have lesions localised in metaphyses near the epiphyseal line whereas older patients who were nearer to growth termination had arrest of the osteoblastic process and the persistence of some part of the cyst.

Capanna [22, 45, 65, 66] reported similar success rate with steroid injection and recurrence rate of 7 % only. Capanna et al. [67] promoted using contrast examination to identify non-contiguous septated areas to ensure the steroid was distributed through the whole cyst (Fig. 43.4).

Various authors have reported comparable results and suggested refinements of the percutaneous steroid techniques such as using different types of steroids, different doses and combination with bone graft [68–72].

Two studies have compared steroid injection with curettage and grafting (Level III). Oppenheim and Galleno [73] compared 37 patients with SBC treated operatively to 20 patients treated with steroid injection with a minimum follow-up of 2 years. In the operative group the average operative time was 100 min, with a mean estimated blood loss of 300 ml. The recurrence rate was 40 %, rising to 88 % in patients under the age of 10 years with active cysts (less than 1 cm from the physis). Major complications occurred in 15 % and included infection, refracture, coxa vara, extremity shortening, and physeal damage. In contrast, the steroid-injected group had a recurrence rate of 5 %, although 50 % required more than one injection for maximum obliteration. The average operative time was 30 min, with negligible blood loss and a minimum hospital stay and rehabilitation. The only complications were a mild steroid flush in one patient and extremity shortening due to preexisting fracture in another. The end point of healing for this study was reconstitution of cortical thickness, rather than total obliteration at the cyst. No secondary fractures were encountered. They concluded that both operative treatment and percutaneous steroid injection exhibited a high rate of recurrence or persistence. The greater simplicity and lesser morbidity associated with the steroid technique favored it as their method of choice.

Bovill and Skinner [74] reviewed 32 patients with SBC treated in 3 different ways; 15 patients were treated surgically, 12 were given steroid injections, and five were treated nonoperatively. Thirteen of the 32 cysts were latent and 19 were active, judged by their proximity to the growth plate. The average follow-up was 5.6 years. They showed that steroid injections were as effective as surgical intervention while having lower morbidity.

Percutaneous Techniques/Bone Marrow Injection

The cellular contents of bone marrow particularly stem and undifferentiated cells have been the subject of extensive research in various medical fields. Bone marrow transplant

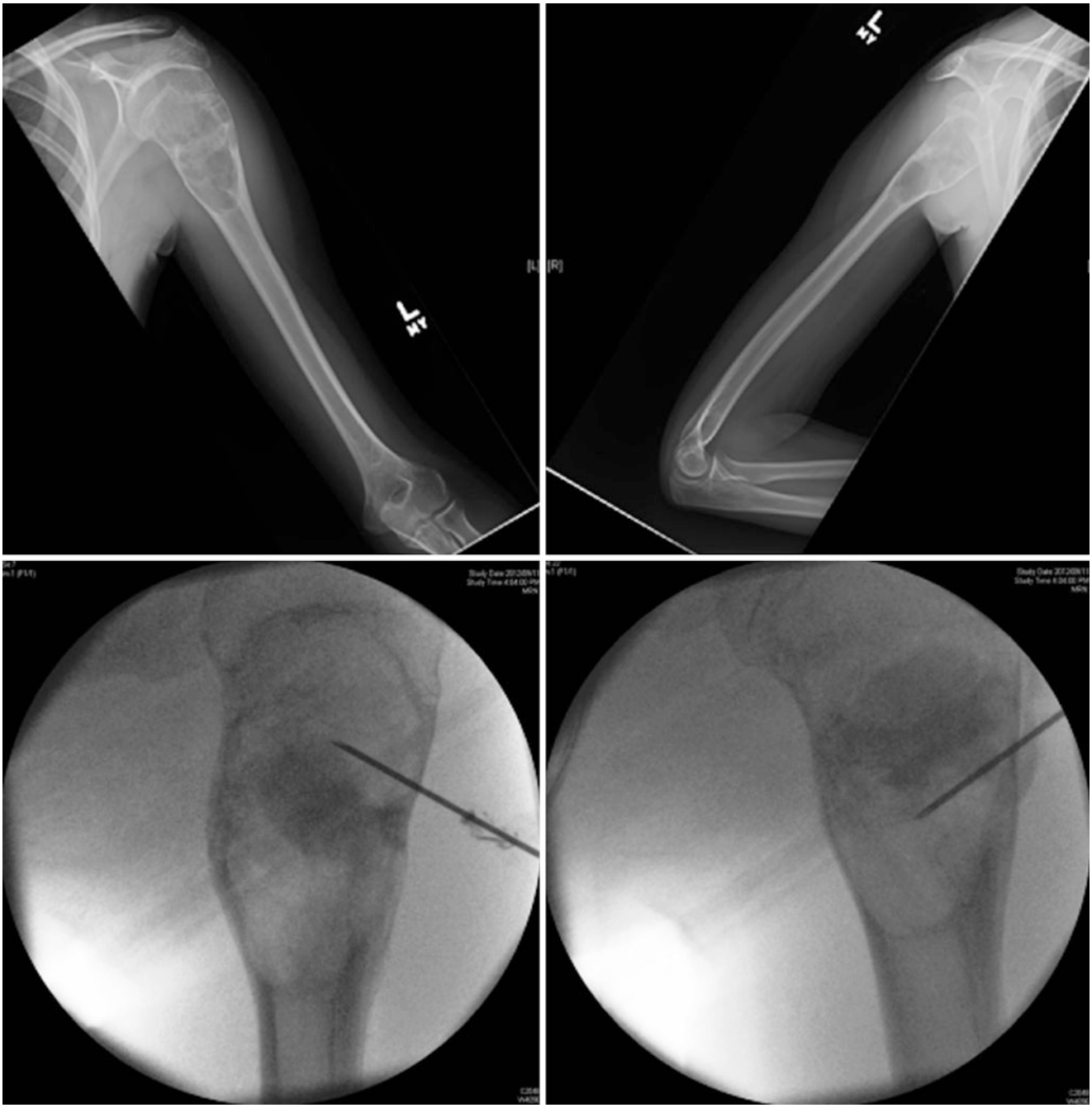

Fig. 43.4 Proximal humerus cyst underwent a cystogram before steroid injection. Bottom images show a non-contiguous septated areas

has been successfully used in haematological malignancy, severe types of haemoglobinopathies and mucopolysaccharoidosis. It has also been used to promote fracture healing.

Lokiec [25] published a preliminary case series of 10 patients with SBC who were treated with bone marrow injection. Of note these patients also had curetting of the cyst, raising the possibility that mechanical disruption rather than the injection was responsible for cyst healing. All the patients became pain free after a mean of 2 weeks and resumed full activities within 6 weeks. All ten cysts consolidated radiologically and showed remodeling within 4 months. Review at 12–48 months showed satisfactory healing without complications.

Several authors reported positive results (but not 100 % healing) [75–77]. Yandow [75] reported 83 % (10/12) suc-

cess rate. Delloye et al. [78] reported good results in approximately 88 % (7/8) of their patients.

Kose et al. [77] prospectively evaluated the outcome of the autologous bone marrow procedure in 12 patients. The mean volume of the lesions was 51.2 cc (27–74 cc). The mean follow-up time was 23 months (range: 18–38 months). Complete healing occurred in two of the patients. Three cysts had residual cystic defect in which two required curettage and bone chip grafts. Six cases recurred. One patient failed to show any signs healing. He found factors such as the size, multi-loculation, and completeness of the filling of the cyst with bone marrow grafting might influence the results.

Chang et al. [79] published a study comparing the results of aspiration and injection of bone marrow with those of aspiration and injection of steroid (level III). All were treated by the same protocol. The only difference was the substance injected into the cysts. The mean radiological follow-up to detect activity in the cyst was 44 months (12–108). Of the 79 patients included, 14 received a total of 27 injections of bone marrow and 65 a total of 99 injections of steroid. Repeated injections were required in 57 % of patients after bone marrow and in 49 % after steroid. No complications were noted in either group. In this series no advantage could be shown for the use of autogenous injection of bone marrow compared with injection of steroid in the management of SBC.

We identified a single randomised controlled trail [48] (level I) and a Cochrane systematic review and meta-analysis [80, 81] that compared bone marrow injection with steroid (methylprednisolone acetate) injection for treating simple bone cysts. The Cochrane systematic review and meta-analysis was published in 2014 and based on the above RCT only.

In their RCT [48] of 90 patients (72 were available after 2 years follow-up) Wright et al. randomly allocated to treatment with injection of either bone-marrow or methylprednisolone. The primary outcome was radiographic evidence of healing. The cyst was judged by independent radiologist to be either:

1. Not healed (grade 1 = a clearly visible cyst or grade 2 = a cyst that was visible but multilocular and opaque).
2. Healed (grade 3 = sclerosis around or within a partially visible cyst or complete healing (obliteration of the cyst)).

Patient function was assessed using the Activity Scale for Kids, and pain was assessed with the Oucher Scale. Sixteen (42 %) of the 38 cysts treated with methylprednisolone acetate healed, and 9 (23 %) of the 39 cysts treated with bone marrow healed ($p = 0.01$). There was no significant difference between the treatment groups ($p > 0.09$) with respect to function, pain, number of injections, additional fractures, or complications. Although the rate of healing of simple bone cysts was low following injection of either bone marrow or methylprednisolone, the latter provided superior healing rates. This trial suggested that even with steroid, the healing rate was low (42 %). Hence the search for better treatments modalities must continue.

Mechanical Disruption of the Cyst with or Without Fixation

Simple mechanical disruption of the cyst wall in a controlled fashion may produce a facture healing like state and potentially continuous drainage of the cyst and open cyst to marrow contents. Kirschner wires (K-wires), elastic nails and cannulated screws have been used to allow such continuous drainage [38, 82–85]. Elastic nails may also provide mechanical support for the bone while cyst is healing (Fig. 43.5).

Komiya et al. [34] introduced the term "trepanation" which consists of drainage of cyst fluid, lavage of the cystic cavity with saline, and the making of multiple drill holes through the cortical and the medullary bone of the cyst wall. Injection of corticosteroid was omitted. He reported good outcome in 91 % (10/11) based on clinical judgment, but no validated clinical or radiological outcome was used. One patient had a pathological fracture and another two had partial recurrence.

Chigira et al. [6] reported similar success with multiple drilling in 86 % (6/7) of their patients. Their technique included leaving 2 mm K-wires in place to allow drainage of the fluid through the cyst wall. They also reported on the long-term results with ten patients with SBC. They stated that cortical thickening was initially observed within 3 months, and minimal new bone formation around K-wires left in situ was observed within 4 months. New bone formation gradually progressed from the cyst wall and around the wire. Consolidation of a cyst cavity was uniformly observed 6–7 months after drilling. In several patients residual cysts were seen between the wire and cortical shell. In younger patients, a cystic cavity reappeared after extraction of the wire, although complete radiographic healing had been observed. These findings suggest that a K-wire may play a role in preventing relapse of SBC after hole drilling [86].

In a retrospective comparative study (level III) of 46 patients with bone cyst treated with curettage involved lining and cyst wall (10 patients), methylprednisolone injection (17 patients) or autologous bone marrow injection (19 patients), the healing rates were 70 % , 41 % and 21 %

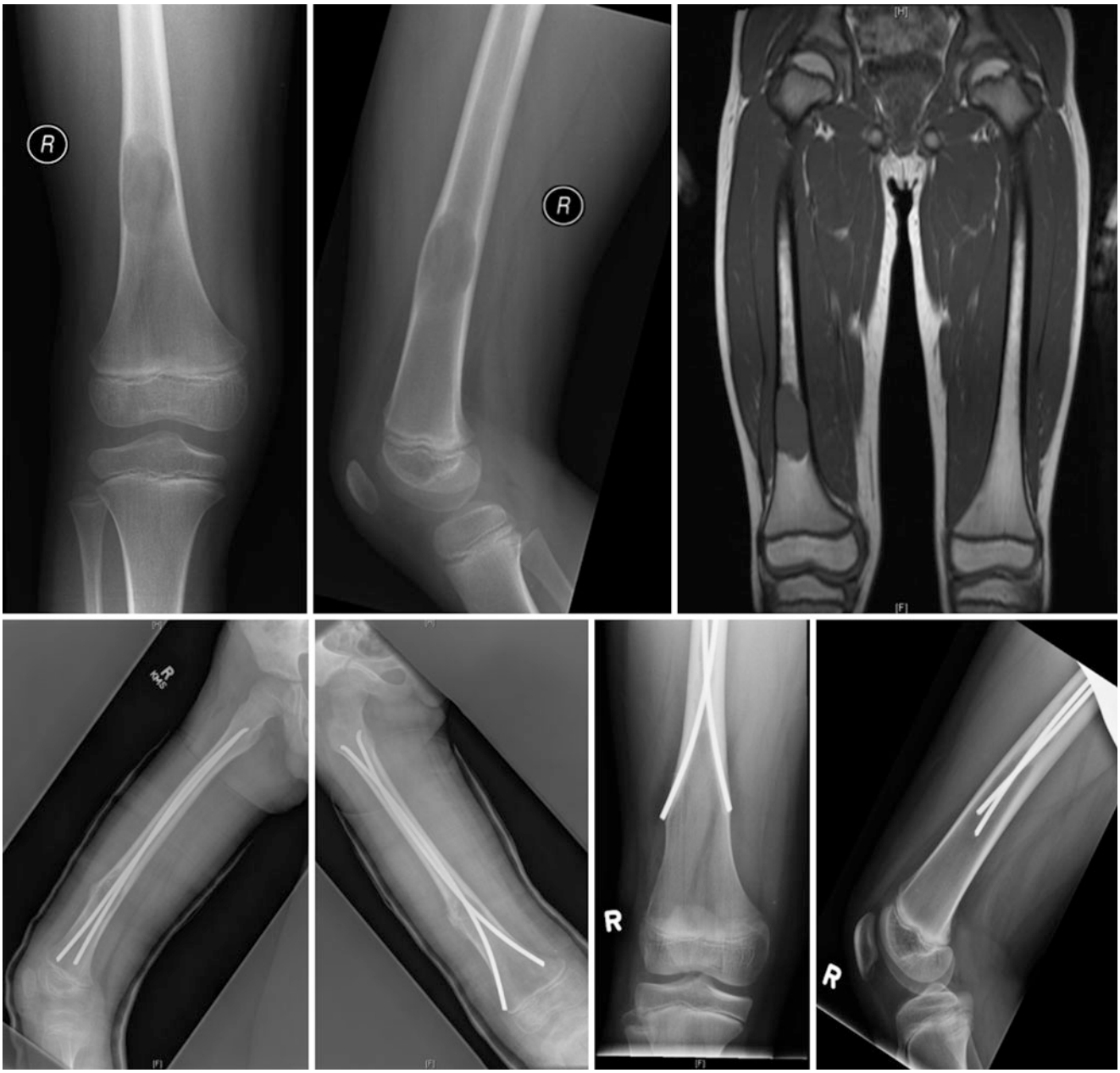

Fig. 43.5 A distal femoral simple bone cyst treated with isolated curettage and stabilisation using flexible intramedullary nailing

respectively (P = 0.08) [32]. In another study [36], 40 patients were treated with one of four methods: serial percutaneous steroid and autologous bone-marrow injection (Group 1, 9 patients); open curettage and grafting with a calcium sulfate bone substitute either without instrumentation (Group 2, 12) or with internal instrumentation (Group 3, 7 patients); or minimally invasive curettage, ethanol cauterization, disruption of the cystic boundary, insertion of a synthetic calcium sulfate bone-graft substitute, and placement of a cannulated screw to provide drainage (Group 4, 12 patients). Group-4 patients had the highest healing rate (11/12) compared with 3/9 in Group 1; 8/12 in Group 2 and 6/7 in Group 3. Group-4 patients also had the shortest mean time to union (mean±SD): 3.7 ± 2.3 months compared with 23.4 ± 14.9, 12.2 ± 8.5, and 6.6 ± 4.3months in Groups 1, 2, and 3, respectively (Figs. 43.6 and 43.7).

In conclusion, the literature reports different healing rates with different treatments. While superior to simple injection of bone marrow, methylprednisolone has low rates of complete cyst healing at 2 years.

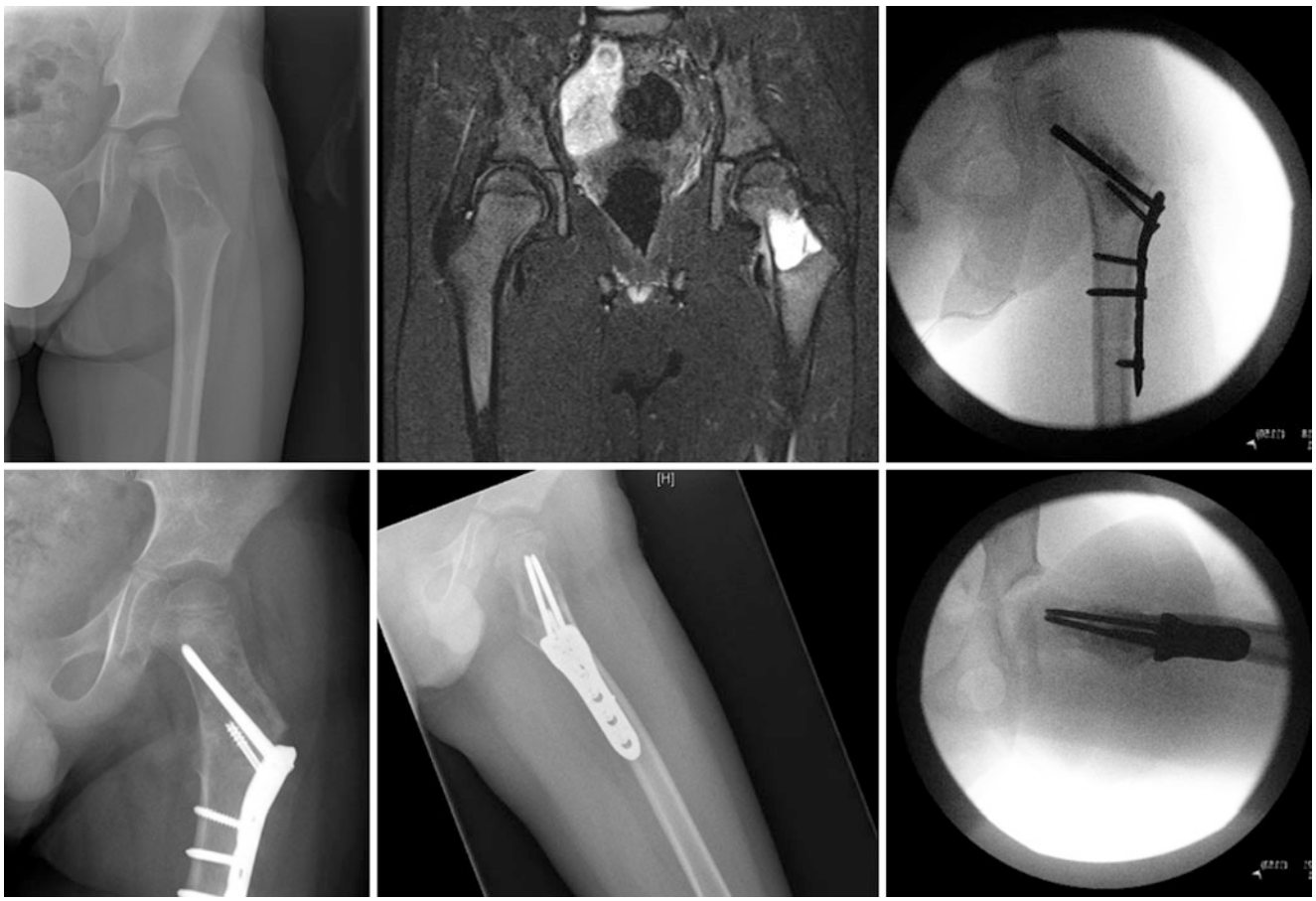

Fig. 43.6 A boy with proximal femoral simple bone cyst who was treated with curettage, osteoset pellets, cannulated screw and stabilization

What Alternative Graft Materials Can Be Used in Treating Simple Bone Cysts?

Studies comparing steroid to bone marrow injection convincingly showed that the nature of the injected material influences the outcome. Wright et al. [48] showed that at 2 years with blinded assessment that steroid provides only 42 % radiographic cyst healing. In a recent review by Donaldson and Wright [87], various materials have been used in treating SBC. All published works have been case series (level IV). Most also included some sort of variation of the above intervention making untangling the effect of material *per se* difficult if not impossible.

Our search identified two ongoing randomised controlled trials comparing various material injections:

i. Simple Bone Cysts in Kids (SBoCK) study [88] comparing curettage with puncture and curettage with puncture and injection of Vitoss morsels.
ii. A phase-2, randomised, open-label, multicentre, ascending dose study of the efficacy, safety and tolerability of Osteogenic Gel I-040302 versus control injection (bone marrow aspirate or steroids) in children and young adults with solitary bone cysts [89].

What Is the Prognosis of Simple Bone Cysts?

As stated earlier the SBCs are noncancerous and malignant transformation is extremely unusual. Six cases of malignant conversion of SBC to sarcoma have been reported, but 5 did not have a biopsy to confirm that they were truly SBC before transformation [90]. There was a single report of malignant transformation to chondrosarcoma from a biopsy-proved SBC [91].

As stated in the natural history section, SBCs do not always run a benign course. In up to 10 % of patients, SBCs in a limb may result in growth disturbance [53–55, 92] leading to angular deformity or limb length discrepancy [52, 57] Thus, children with SBC are at risk for continued pain, activity restriction, anxiety, or recurrent fracture [93]. Recommendations for treatment are outlined in Table 43.1.

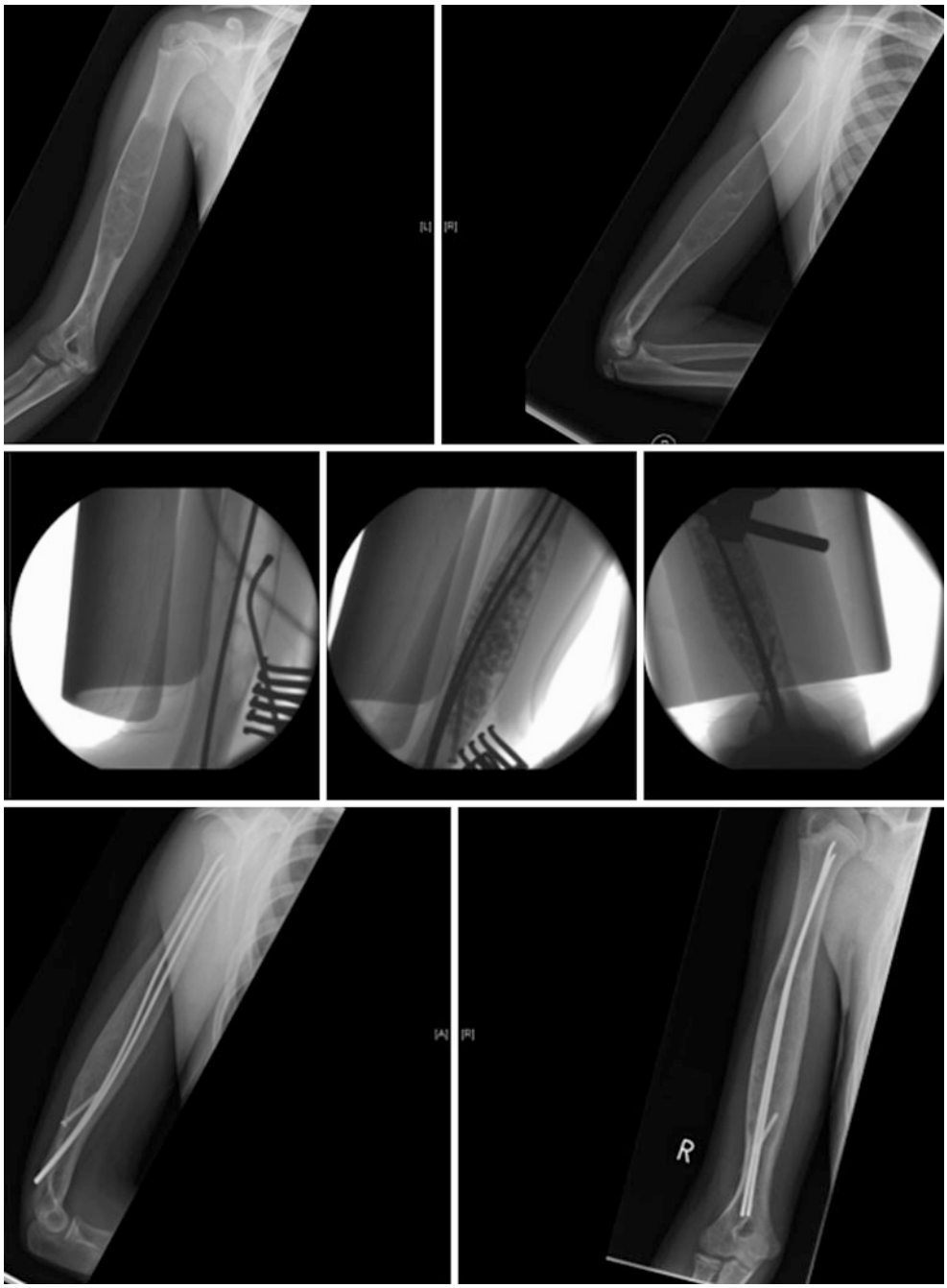

Fig. 43.7 Humeral simple bone cyst treated curettage, osteoset pellets, cannulated screw and stabilisation (same patient in Fig. 43.3)

Table 43.1 Recommendations

Statement	Grade of evidence
Fracture does not heal a simple bone cyst	B
Intervention is superior to observation in healing a simple bone cyst	B
Percutaneous injection of steroid is superior to bone marrow	B
Curettage with or without grafting is superior to percutaneous injection of steroid or bone marrow	C
Curettage, synthetic calcium sulfate bone-graft substitute, and placement of a cannulated screw to provide drainage is superior to all other interventions	I

References

1. Alshryda S, Jones S, Banaszkiewicz P. Postgraduate paediatric orthopaedics: the candidate's guide to the FRCS (Tr and Orth) examination. Cambridge: Cambridge University Press; 2014.
2. Lokiec F et al. Simple bone cysts treated by percutaneous autologous marrow grafting. J Bone Joint Surg. 1996;78(6):934–7.
3. Lokiec F, Wientroub S. Simple bone cyst. J Pediatr Orthop B. 1998;7(4):262–73.
4. Zenmyo M et al. A solitary bone cyst in the spinous process of the cervical spine. Spine (Phila Pa 1976). 2000;25(5):641.
5. Cohen J. Simple bone cysts. Studies of cyst fluid in six cases with a theory of pathogenesis. J Bone Joint Surg Am. 1960;42-A:609–16.
6. Chigira M et al. The aetiology and treatment of simple bone cysts. J Bone Joint Surg Br. 1983;65(5):633–7.
7. Shindell R, Connolly JF, Lippiello L. Prostaglandin levels in a unicameral bone cyst treated by corticosteroid injection. J Pediatr Orthop. 1987;7(2):210–2.
8. Shindell R et al. Prostaglandin levels in unicameral bone cysts treated by intralesional steroid injection. J Pediat Orthop. 1989;9(5):516–9.
9. Shindell R et al. Prostaglandin levels in unicameral bone cysts treated by intralesional steroid injection. J Pediatr Orthop. 1989;9(5):516–9.
10. Kobza Black A, Greaves MW, Hensby CN. The effect of systemic prednisolone on arachidonic acid, and prostaglandin E2 and F2 alpha levels in human cutaneous inflammation. Br J Clin Pharmacol. 1982;14(3):391–4.
11. Hart PH et al. Interleukin-4 stimulates human monocytes to produce tissue-type plasminogen activator. Blood. 1989;74(4):1222–5.
12. Gerasimov AM et al. The role of lysosomes in the pathogenesis of unicameral bone cysts. Clin Orthop Relat Res. 1991;NA(266):53–63.
13. Komiya S, Inoue A. Development of a solitary bone cyst – a report of a case suggesting its pathogenesis. Arch Orthop Trauma Surg. 2000;120(7-8):455–7.
14. Komiya S et al. Oxygen scavengers in simple bone cysts. Clin Orthop Relat Res. 1994;NA(308):199–206.
15. Mirra JM et al. Cementum-like bone production in solitary bone cysts (so-called ???Cementoma??? of long bones) report of three cases. Electron microscopic observations supporting a synovial origin to the simple bone cyst. Clin Orthop Relat Res. 1978;NA(135):295–307.
16. Yu CL, D'Astous J, Finnegan M. Simple bone cysts. The effects of methylprednisolone on synovial cells in culture. Clin Orthop Relat Res. 1991;262:34–41.
17. Richkind KE et al. Translocation (16;20)(p11.2;q13). Sole cytogenetic abnormality in a unicameral bone cyst. Cancer Genet Cytogenet. 2002;137(2):153–5.
18. Tachdjians. In: Herring JA, editor. Pediatric orthopaedics, vol. 1. Philadelphia: Saunders Elsevier; 2013.
19. Struhl S et al. Solitary (unicameral) bone cyst. Skeletal Radiol. 1989;18(4):261–5.
20. Struhl S et al. Solitary (unicameral) bone cyst. The fallen fragment sign revisited. Skeletal Radiol. 1989;18(4):261–5.
21. Steinberg GG. Ewing's sarcoma arising in a unicameral bone cyst. J Pediat Orthop. 1985;5(1):97–100.
22. Campanacci M, Sessa L, Trentani C. Scaglietti's method for conservative treatment of simple bone cysts with local injections of methylprednisolone acetate. Ital J Orthop Traumatol. 1977;3(1):27–36.
23. Scaglietti O, Marchetti PG, Bartolozzi P. Final results obtained in the treatment of bone cysts with methylprednisolone acetate (depomedrol) and a discussion of results achieved in other bone lesions. Clin Orthop Relat Res. 1982;165:33–42.
24. Scaglietti O, Marchetti PG, Bartolozzi P. The effects of methylprednisolone acetate in the treatment of bone cysts. Results of three years follow-up. J Bone Joint Surg Br. 1979;61-B(2):200–4.
25. Lokiec F et al. Simple bone cysts treated by percutaneous autologous marrow grafting. A preliminary report. J Bone Joint Surg Br. 1996;78(6):934–7.
26. Rougraff BT, Kling TJ. Treatment of active unicameral bone cysts with percutaneous injection of demineralized bone matrix and autogenous bone marrow. J Bone Joint Surg Am. 2002;84-A(6):921–9.
27. Dormans JP, Dormans NJ. Use of percutaneous intramedullary decompression and medical-grade calcium sulfate pellets for treatment of unicameral bone cysts of the calcaneus in children. Orthopedics. 2004;27(1 Suppl):s137–9.
28. Dormans JP et al. Percutaneous intramedullary decompression, curettage, and grafting with medical-grade calcium sulfate pellets for unicameral bone cysts in children: a new minimally invasive technique. J Pediatr Orthop. 2005;25(6):804–11.
29. Mik G et al. Results of a minimally invasive technique for treatment of unicameral bone cysts. Clin Orthop Relat Res®. 2009;467(11):2949–54.
30. Tang XY et al. Simple bone cysts in children treated with intracystic fibrin sealant injection. Chin Med J (Engl). 2006;119(6):523–5.
31. Takechi R et al. Solid variant of aneurysmal bone cyst in the tibia treated with simple curettage without bone graft: a case report. World J Surg Oncol. 2012;10:45.
32. Canavese F et al. Unicameral bone cysts: comparison of percutaneous curettage, steroid, and autologous bone marrow injections. J Pediatr Orthop. 2010;31(1):50–5.
33. Hirn M et al. Bone defects following curettage do not necessarily need augmentation. Acta Orthop. 2009;80(1):4–8.
34. Komiya S et al. Simple bone cyst. Treatment by trepanation and studies on bone resorptive factors in cyst fluid with a theory of its pathogenesis. Clin Orthop Relat Res. 1993;287:204–11.
35. Brecelj J, Suhodolcan L. Continuous decompression of unicameral bone cyst with cannulated screws: a comparative study. J Pediatr Orthop B. 2007;16(5):367–72.
36. Hou HY et al. Treatment of unicameral bone cyst: a comparative study of selected techniques. J Bone Joint Surg Am. 2010;92(4):855–62.
37. Bumci I, Vlahovic T. Significance of opening the medullar canal in surgical treatment of simple bone cyst. J Pediatr Orthop. 2002;22(1):125–9.
38. Roposch A, Saraph V, Linhart WE. Flexible intramedullary nailing for the treatment of unicameral bone cysts in long bones. J Bone Joint Surg Am. 2000;82-A(10):1447–53.
39. de Sanctis N, Andreacchio A. Elastic stable intramedullary nailing is the best treatment of unicameral bone cysts of the long bones in children?: prospective long-term follow-up study. J Pediatr Orthop. 2006;26(4):520–5.
40. Fahey JJ, O'Brien ET. Subtotal resection and grafting in selected cases of solitary unicameral bone cyst. J Bone Joint Surg Am. 1973;55(1):59–68.
41. McKay DW, Nason SS. Treatment of unicameral bone cysts by subtotal resection without grafts. J Bone Joint Surg Am. 1977;59(4):515–9.
42. Sturz H, Zenker H, Buckl H. Total subperiosteal resection treatment of solitary bone cysts of the humerus. Arch Orthop Trauma Surg. 1979;93(3):231–9.
43. Chaves D. Treatment of solitary cysts of the humerus. Treated by diaphyseal resection and bone grafting. Int Orthop. 1980;3(4):253–6.
44. Neer 2nd CS et al. Treatment of unicameral bone cyst. A follow-up study of one hundred seventy-five cases. J Bone Joint Surg Am. 1966;48(4):731–45.
45. Capanna R et al. The natural history of unicameral bone cyst after steroid injection. Clin Orthop Relat Res. 1982;NA(166)204–11.

46. Oppenheim WL, Galleno H. Operative treatment versus steroid injection in the management of unicameral bone cysts. J Pediat Orthop. 1984;4(1):1–7.
47. Spence Jr KF et al. Solitary unicameral bone cyst: treatment with freeze-dried crushed cortical-bone allograft. A review of one hundred and forty-four cases. J Bone Joint Surg Am. 1976;58(5):636–41.
48. Wright JG et al. A randomized clinical trial comparing intralesional bone marrow and steroid injections for simple bone cysts. J Bone Joint Surg Am. 2008;90(4):722–30.
49. Wright JG. A randomized clinical trial comparing intralesional bone marrow and steroid injections for simple bone cysts. J Bone Joint Surg Am. 2008;90(4):722.
50. Kaelin AJ, MacEwen GD. Unicameral bone cysts. Natural history and the risk of fracture. Int Orthop. 1989;13(4):275–82.
51. Donaldson S, Wright JG. Simple bone cysts: better with age? J Pediatr Orthop. 2015;35(1):108–14.
52. Haims AH et al. Epiphyseal extension of a unicameral bone cyst. Skeletal Radiol. 1997;26(1):51–4.
53. Stanton RP, Abdel-Mota'al MM. Growth arrest resulting from unicameral bone cyst. J Pediat Orthop. 1998;18(2):198–201.
54. Cohen J. Unicameral bone cysts. A current synthesis of reported cases. Orthop Clin North Am. 1977;8(4):715–36.
55. Violas P et al. Simple bone cysts of the proximal humerus complicated with growth arrest. Acta Orthop Belg. 2004;70(2):166–70.
56. Hashemi-Nejad A, Cole WG. Incomplete healing of simple bone cysts after steroid injections. J Bone Joint Surg. 1997;79(5):727–30.
57. Tey IK et al. Active unicameral bone cysts in the upper limb are at greater risk of fracture. J Orthop Surg (Hong Kong). 2009;17(2):157–60.
58. Garceau GJ, Gregory CF. Solitary unicameral bone cyst. J Bone Joint Surg Am. 1954;36(A:2):267–80.
59. Ahn JI, Park JS. Pathological fractures secondary to unicameral bone cysts. Int Orthop. 1994;18(1):20–2.
60. Galasko CS. Letter: the fate of simple bone cysts which fracture. Clin Orthop Relat Res. 1974;101:302–4.
61. Neer CS et al. Current concepts on the treatment of solitary unicameral bone cyst. Clin Orthop Relat Res. 1973;97:40–51.
62. Vasconcellos DA et al. Cyst index: a nonpredictor of simple bone cyst fracture. J Pediatr Orthop. 2007;27(3):307–10.
63. Leong NL et al. Computed tomography-based structural analysis for predicting fracture risk in children with benign skeletal neoplasms: comparison of specificity with that of plain radiographs. J Bone Joint Surg Am. 2010;92(9):1827–33.
64. Jaffe HL. Solitary unicameral bone cyst. Arch Surg. 1942;44(6):1004.
65. Campanacci M, Capanna R, Picci P. Unicameral and aneurysmal bone cysts. Clin Orthop Relat Res. 1986;204:25–36.
66. Capanna R, Campanacci DA, Manfrini M. Unicameral and aneurysmal bone cysts. Orthop Clin North Am. 1996;27(3):605–614.
67. Capanna R et al. Contrast examination as a prognostic factor in the treatment of solitary bone cyst by cortisone injection. Skeletal Radiol. 1984;12(2):97–102.
68. Carrata A et al. The treatment of simple bone cysts by topical infiltrations of methylprednisolone acetate: technique and results. Eur J Radiol. 1983;3(1):3–8.
69. Fernbach SK et al. Radiographic changes in unicameral bone cysts following direct injection of steroids: a report on 14 cases. Radiology. 1981;140(3):689–95.
70. Thawrani D et al. Successful treatment of unicameral bone cyst by single percutaneous injection of α-BSM. J Pediat Orthop. 2009;29(5):511–7.
71. Pavone V et al. Steroid injections in the treatment of humeral unicameral bone cysts: long-term follow-up and review of the literature. Eur J Orthop Surg Traumatol. 2013;24(4):497–503.
72. Goel AR et al. Unicameral bone cysts: treatment with methylprednisone acetate injections. J Foot Ankle Surg. 1994;33(1):6–15.
73. Oppenheim WL, Galleno H. Operative treatment versus steroid injection in the management of unicameral bone cysts. J Pediatr Orthop. 1984;4(1):1–7.
74. Bovill DF, Skinner HB. Unicameral bone cysts. A comparison of treatment options. Orthop Rev. 1989;18(4):420–7.
75. Yandow SM et al. Autogenic bone marrow injections as a treatment for simple bone cyst. J Pediat Orthop. 1998;18(5):616–20.
76. Arazi M et al. Minimally invasive treatment of simple bone cysts with percutaneous autogenous bone marrow injection. Orthopedics. 2005;28(2):108–12.
77. Kose N et al. Percutaneous autologous bone marrow grafting for simple bone cysts. Bull Hosp Jt Dis. 1999;58(2):105–10.
78. Delloye C et al. Simple bone cysts treated with aspiration and a single bone marrow injection. International Orthopaedics. 1998;22(2):134–8.
79. Chang CH, Stanton RP, Glutting J. Unicameral bone cysts treated by injection of bone marrow or methylprednisolone. J Bone Joint Surg. 2002;84(3):407–12.
80. Zhao JG, Ding N, Huang WJ, Wang J, Shang J, Zhang P. Interventions for treating simple bone cysts in the long bones of children. Cochrane Database Syst Rev. 2014;(9): Art. No.: CD010847. doi:10.1002/14651858.CD010847.pub2. 2014.
81. Zhao J-G et al. Interventions for treating simple bone cysts in the long bones of children. Cochrane Database Syst Rev. 1996, Wiley-Blackwell.
82. Catier P et al. The treatment of upper femoral unicameral bone cysts in children by Ender's nailing technique. Rev Chir Orthop Reparatrice Appar Mot. 1981;67(2):147–9.
83. Tsuchiya H et al. Cannulation of simple bone cysts. J Bone Joint Surg Br. 2002;84(2):245–8.
84. Shirai T et al. Treatment of a simple bone cyst using a cannulated hydroxyapatite pin. Medicine (Baltimore). 2015;94(25):e1027.
85. Roposch A, Saraph V, Linhart WE. Treatment of femoral neck and trochanteric simple bone cysts. Arch Orthop Trauma Surg. 2004;124(7):437–42.
86. Chigira M et al. Radiological evidence of healing of a simple bone cyst after hole drilling. Arch Orthop Trauma Surg. 1986;105(3):150–3.
87. Donaldson S, Wright JG. Recent developments in treatment for simple bone cysts. Curr Opin Pediatr. 2011;23(1):73–7.
88. Wright J. The SBoCK study. https://clinicaltrials.gov/ct2/show/study/NCT02193841?term=simple+bone+cyst&rank=1&show_locs=Y#locn
89. EUCTR2006-001294-18-GB, EUCTR2006-001294-18-GB. A phase 2 randomised, open-label, multi-centre ascending dose study of the efficacy, safety and tolerability of I-040302 versus control injection (bone marrow aspirate or steroids) in children and young adults with solitary bone cysts. http://apps.who.int/%20trialsearch/Trial.aspx?TrialID=EUCTR2006-001294-18-GB. Accessed 10 Feb 2014.
90. Johnson LC, Vetter H, Putschar WG. Sarcomas arising in bone cysts. Virchows Arch Pathol Anat Physiol Klin Med. 1962;335:428–51.
91. Grabias S, Mankin HJ. Chondrosarcoma arising in histologically proved unicameral bone cyst. A case report. J Bone Joint Surg Am. 1974;56(7):1501–9.
92. Tachdjians. In: Herring JA, editor. Pediatric orthopaedics, vol. 1. Philadelphia: Saunders Elsevier; 2008.
93. Norman-Taylor FH et al. Risk of refracture through unicameral bone cysts of the proximal femur. J Pediat Orthop. 2002;22(2):249–54.

Evidence-Based Treatment for Malignant Paediatric Bone Tumours

44

Andrew J. Graydon, Michael M. Hanlon, and Caitlin E. Bodian

Abstract

In Children, the vast majority of bone tumours are benign or benign aggressive entities. For the purposes of this discussion however, we are focusing on the management of primary and secondary bone malignancies.

Keywords

Osteosarcoma • Bone • Limb salvage • Paediatric

In Children, the vast majority of bone tumours are benign or benign aggressive entities. For the purposes of this discussion however, we are focusing on the management of primary and secondary bone malignancies.

Primary Tumours of Bone

Osteosarcoma

Osteosarcoma (OSA) is the most common malignant primary bone tumour and the third most common tumour of adolescence, with highest incidence in the second decade of life [1]. Yet, it is still considered an orphan disease with between 4.4 and 7.08/1,000,000 cases per year [1–3]. It is an osteoid producing tumour with multiple histological subdivisions, osteogenic OSA being the most common. However, these subdivisions have little influence on treatment or prognosis. Due to loss of tumour suppressor genes, OSA exhibits genetic instability, tumour heterogeneity, and rapid metastatic potential. Genetic conditions characterized by genomic instability, such as retinoblastoma, Rothmund Thomson Syndrome, Werner syndrome and Li-Fraumeni syndrome are all associated with a high risk of OSA [4–6]. Children with previous cancer and radiation exposure are also at higher risk [7].

OSA lesions are most frequently located about the knee, particularly the distal femur, followed by proximal tibia. Those who present with singular appendicular lesions can expect a 5-year survival rate of 60–70 % [8, 9]. Approximately 10 % of OSA occur in the axial skeleton, of which have a significantly poorer outcome, with 3-year survival rates between 11 % and 56 % [1, 8]. Joint involvement is rare as the metaphysial plate and articular cartilage serve as barriers to disease spread.

Radiologically, it is aggressive with sclerotic and lucent features, poorly defined margins and wide transitional zone, prominent periosteal reaction and destruction of surrounding cortical bone [4]. 10–20 % of patients present with visible metastatic disease, most commonly located in the lungs and occasionally in other bones or lymph nodes. However, it is speculated that up to 80 % of patients have micro-metastatic disease not detectable on current imaging modalities [4].

Ewing's Sarcoma

Ewing's sarcoma family tumours (ESFT) are small round-cell tumours and the second most common bone tumours in children. Overall, ESFT patients tend to present at a younger age than OSA [10]. Eighty percent occur in patients younger than 20 years of age and slight tendency towards males (1.5:1) [11] ESFT includes classic Ewing's sarcoma (EWS),

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primitive neuroectodermal tumours (PNET) and Askin tumours [12]. These tumours are exceptionally rare (2.93/1,000,000), accounting for less than 5 % of all childhood malignancies [13]. Ninety-five percent of these tumours are derived from a translocation of the EWS gene on chromosome 22 and an ETS gene on chromosome 11, resulting in a chimeric ESW-ETS protein [11]. Like OSA, EWS is also frequently found about the knee, but has a higher rate of axial involvement. EWS is highly aggressive and can quickly metastasise to bone marrow, lungs, and other tissue [12]. Lymph node involvement is rare, but can occur [14]. The 5-year survival rate for EWS is lower than that of OSA, roughly 50 % [15].

Radiologically, lesions demonstrate a mottled mixed lytic and sclerotic appearance. A speculated or layered 'onion skin' periosteal reaction is a classic finding with EWS [14].

Adamantinoma

Adamantinomas (ADA) are rare low-grade osteolytic tumours of the bone, closely associated with osteofibrous dysplasia (OFD) [16, 17]. Extensive investigation involving immunohistochemistry, molecular studies and cell cultures have not been able to conclude whether OFD represents a reparative process related to pre-existing ADA or if OFD, differentiated ADA and classic ADA represent a continuum of tumours [18]. It is most commonly found in adolescents and are almost exclusively localized to the tibia (83–90 % of cases) and are considered slow growing, low grade malignant tumours [17]. Nonetheless, they are capable of distant metastases, occurring in about 10 % of cases. Metastases usually occur in lung and lymph nodes and can occur many years after initial presentation of the primary lesion [19]. Chemotherapy and radiotherapy has been shown to be largely ineffective [20]. Surgical resection is the mainstay of treatment.

Langerhans Cell Histiocytosis

Langerhans Cell Histiocytosis (LCH) is a spectrum of disease associated with the proliferation and accumulation of monoclonal Langerhans' cells. Disease can be single or multisystem, with the skeletal system most commonly involved in either a monostotic or polyostotic process [21]. Incidence is 2–9 per million children under 15 years of age [22]. The aetiology of LCH has been widely debated, with no unanimous agreement to whether it is a disorder of immune regulation or a true malignant condition [22]. Cytogenic abnormalities have been associated with LCH suggest that it is a low grade malignancy [23]. A small number of patients, usually those with multisystem involvement, have rapidly progressive disease courses that can be fatal [24].

The nomenclature regarding LCH is controversial as the disease has historically been categorized according to clinical manifestations. Previously the term Eosinophilic Granuloma (EG) has been used if a solitary lesion presents, but this does not distinguish the varying clinical entities that LCH represents. Other eponymous associations include Hand-Schuller-Christian disease and Letterer-Seiwe disease.

Patients with solitary disease typically present with localised pain, but those with disseminated disease may present with lymphadenopathy, skin lesions, or diabetes insipidus [25]. X-ray appearances of LCH typically show an osteolytic lesion, often with benign features such as circular appearance, sclerotic rim and well-defined margins. Occasionally, LCHs will have a more aggressive characteristic such as moth-eaten appearance and wide zone of transition, similar to that of a Ewing's sarcoma [26].

There is limited information regarding medical treatment for LCH. For local lesions, general observation, injections with steroids, local excision and curettage, chemotherapy and irradiation have been trialled. In younger patients, there is a high rate of spontaneous resolution, or resolution after biopsy [27]. In systemic disease the role of standard therapy (Steroid +/− vincristine) is the subject of ongoing Children's Oncology Group trials (LCH-III and LCH-IV).

Others

Chondrosarcoma is considered an adult tumour of bone and is exceedingly rare in children, comprising approximately 2 % of all paediatric sarcomas [28]. It is a malignant hyaline cartilage tumour. CSA in a patient under 20 is usually the result of genetic disorders such as Ollier's or Maffucci syndrome and very occasionally seen in Li-Fraumeni and multiple hereditary exostoses [29–31]. Currently, surgical excision is the mainstay of treatment. The use of chemotherapy and radiotherapy is controversial, as it is typically resistant to these modalities [32–34]. Recent advances in understanding the molecular variations of CSA have led to potential therapies being piloted in clinical trials [35]. There is considerable diagnostic difficulty in differentiating CSA from chondroblastic OSA, with obvious implications in treatment modalities used. From a practical point of view, the response of Chondroblastic OSA to chemotherapy is usually suboptimal however [36].

Secondary Tumours of Bone

Lymphoma

In the Paediatric age group Lymphoma is almost exclusively seen as a secondary bone tumour. The adult variant of pri-

mary lymphoma of bone is exceedingly rare in the paediatric population.

In disseminated systemic lymphoma with bone involvement, bone lesions often respond well to systemic therapy and it is unusual for treatment of the bone lesions to be required. However in the situation that they are, surgical stabilisation can be considered, or Radiation therapy as either local control or palliative treatment can be used.

Neuroblastoma

Neuroblastoma is a solid tumour of primitive sympathetic nervous cells. It is the most common cancer to be diagnosed in the first year of life. In the United States it affects 10.2 per 1,000,000 under the age of 15 [37, 38]. 70 % of patients present with metastatic disease, with bone marrow, bone, and liver being the most likely metastatic sites [39]. Neuroblastoma cells express proteins leading to maturation and activation of osteoclasts, leading to an osteolytic response. This occurs via the receptor activator RANK-L or activation of mesenchymal bone marrow stem cells and overexpression of IL-6, a strong factor in osteoclast activation [40].

These osteolytic lesions can present with severe pain, pathological fractures, spinal cord and nerve compression and severe hypercalcaemia [39]. Treatment is a combination of chemotherapy, radiotherapy, and surgical excision.

Leukaemia and Other Myeloproliferative Disorders

Haematologic malignancies and their variants often present with vague musculoskeletal pain, but rarely progress to the stage of fracture. A simple blood film is mandatory in any patient presenting with unexplained pain for greater than 6 weeks, but these malignancies rarely require orthopaedic intervention following adequate chemotherapy or systemic therapy.

The long-term effects of chemotherapeutic treatment of Leukaemia can be significant. The high doses of steroids used can be associated with avascular necrosis of various joints such as the hip, shoulder and knee. This complication can cause significant morbidity and treatment can be difficult.

Diagnosis and Staging

Clinical History

Typical presentation for almost all types of malignant bone tumours is weeks to months of dull, aching pain and localised swelling [41]. As patients are young and may be very active, pain is initially attributed to injury or growth [3]. Pain is constant and increasing over time, will be present at night and does not usually respond to simple analgesics. In most cases, patients are otherwise well, with systemic symptoms being very rare [41]. Pathological fractures are present in 10–20 % of cases and usually carry a poorer prognostic outcome.

Imaging

X-ray

Plain x-ray is the required first step in the diagnosis of any bone lesion. X-ray provides information regarding the site of the tumour as well as strong clues to morphology. Lesions that appear benign on X-ray (and are accompanied by a clinical history and examination that supports a benign process) rarely require further imaging. However, radiographic evidence of lysis is not seen until bone mineralization loss reaches 30–50 %, so normal x-rays in the context of abnormal history may warrant further investigation [42]. Ultimately, if there is any suspicion from either clinical or radiological findings, further investigation and imaging is the most appropriate next step [5].

Magnetic Resonance Imaging

MRI accurately analyses the location and extension of the tumour, soft tissue component and neurovascular involvement, and the extent of peritumour oedema. MRI studies should contain at least one coronal or sagittal sequence of the entire involved bone to detect any skip metastases. MRI also has a critical role in pre-operative planning, and assessing planned marginal status.

Computed Tomography

For paediatric populations, the radiation exposure associated with CT limits its use and acceptance. However, in certain circumstances CT provides excellent information. For example, CT more accurately assesses the extent of bone involvement such as pathological fracture and cortical destruction. It is also helpful to obtain CT when investigating tumours in difficult locations such as the scapula, sacrum and pelvis [43, 44]. The use of multi-detector CT in children may result in a reduced Radiation dose, but availability limits widespread use.

Systemic Staging

Laboratory Investigations

Any patient with a suspected malignant lesion requires a full blood workup. These should include CBC, blood film, Electrolytes, CRP/ESR, LDH and LFTS. In Paediatric Sarcoma routine laboratory values are typically normal,

although Alkaline phosphatase (ALP) and inflammatory markers such as ESR/CRP may be raised, particularly in OSA [3]. Lactate Dehydrogenase (LDH) may be of prognostic significance in OSA and EWS. The primary utility in laboratory investigations is providing a baseline before chemotherapy or radiotherapy initiation, particularly as chemotherapy can cause renal, cardiac, and auditory damage. All patients undergoing chemotherapy should have assessment of baseline renal and cardiac function, and audiogram [5].

Genetic Testing

Many bone tumours occur sporadically, however there are multiple conditions that predispose a patient to bone tumours. If there is suspicion of a genetic cause to the tumour, genetic testing should be sought [45]. Identification of a genetic predisposition is important, as the condition can affect management strategies. These patients are also at high risk for subsequent tumour development. Familial testing and counselling may also be required.

CT Imaging

CT modality is an integral part of staging, as the most common location of metastasis is the lungs. Certain subgroups of bone tumours also exhibit lymphatogenous spread and imaging of the Abdomen and Pelvis in addition to the usually imaged chest may be indicated. Post-treatment Surveillance CT scans are also indicated at regular intervals.

Functional Imaging

Bone Scan

A whole body technetium bone scan is also indicated in order to detect distant metastases or skip lesions.

Positron Emission Technology

Positron Emission Technology (PET) combines CT detection with a radiolabelled useless substrate for glycolysis (18-Flurodeoxyglucose 18-FDG) as a measure for tissue biologic activity, and this is then used as a surrogate for assessment of tumour viability. PET-CT is not currently widely used for OSA or EWS initial staging, but is utilized in some facilities for assessing treatment response. The prognostic significance of PET-CT is not yet well established or accepted despite increasing evidence. Issues such as radiation dose, cost and accessibility limit usage currently. The separate issues of unique substrate use by an previously un-imaged tumour is also controversial and poorly understood. Much variability is also seen in tumour detection with PET-CT. For example, whilst in EWS PET-CT has been shown to be more sensitive than bone scans at detecting bone metastases, in OSA bone scans are actually still superior to PET scans at metastatic detection [46, 47].

Biopsy

Obtain representative tissue without compromising further management

Before biopsy can be considered, a full history, examination and appropriate radiographic analysis is required. While core needle biopsy (CNB) and fine needle aspiration (FNA) are utilized in some institutions, incisional biopsy is the gold standard for musculoskeletal tumour diagnosis [48]. The biopsy of any lesion should strictly incorporate the biopsy principles to ensure adequate sampling and to minimize cell spread. Poorly planned biopsies can lead to significantly poorer outcomes. Ideally, the surgeon performing the initial biopsy should also be the surgeon who will perform the wide excision or should be in close communication with the surgeon performing the definitive surgical treatment, in order to position the biopsy tract appropriately. There are a number of basic principles which should be followed;

- Incision should be longitudinal and small as possible, so it may be easily incorporated into the final resection.
- It should be directly perpendicular to the lesion and should not cross multiple planes or compartments to avoid contamination.
- It is critical that meticulous haemostasis be maintained throughout the biopsy to prevent microcellular tumour spread.
- Any biopsy of bone should be oblong in shape running parallel to the axis of the bone to prevent further weakening and pathological fracture [49].
- Three samples should be obtained that give a good representation of the lesions cytology, and microbiology should be obtained.
- Communication with a histologist prior to biopsy is helpful to guarantee a quality sample is obtained prior to closure.
- Samples should be quickly submitted for assessment, preferably within half an hour. Before formalin fixation, tumour imprints can be taken (for tumour-specific translocation by FISH), and freezing of tissue/cell suspensions should be done in cryomolds [5].
- Drains can be utilized in closure, but must exit in line with the biopsy tract.

Failure to adhere to these principles has been associated with misdiagnosis of disease, compromise of definitive surgical options, and negative impact on patient survival (see [50]).

Cultures

The saying ‘culture what you biopsy and biopsy what you culture’ should always be in the mind of the operating surgeon in order to rule out differential diagnoses. Occasionally bone tumours and infection can be difficult

Table 44.1 Enneking staging of malignant sarcoma

Stage	Grade	Location	Distant metastasis
IB	Low grade	Intracompartmental	No
IB	Low grade	Extracompartmental	No
IIA	High Grade	Intracompartmental	No
IIB	High Grade	Extracompartmental	No
III	Low or High	Intra or extra-compartmental	Yes

to distinguish by radiography alone [51]. There are many reported cases demonstrating misdiagnosis of tumour for infection and vice versa [51–53]. This is particularly true for EWS, which can have striking similarities to osteomyelitis on imaging [54].

Staging

The most widely used staging method and the one implemented at our institution is the Enneking/Musculoskeletal Tumour Rating Score (MSTS). The goal of staging is to provide a consistent standard for communicating a patient's disease extent and estimated prognosis and to assist in management decisions, both medical and surgical [55]. The Enneking system for malignant tumours is categorised into three stages (Table 44.1). The most typical grade at presentation for both OSA and EWS is Grade IIb [42].

Other Prognostic Factors

More proximal site of primary disease, tumour volume >100 mL, and presence of metastatic disease indicate poorer prognostic outcome. In OSA and EWS, a high LDH is also considered to be a poor prognostic indicator [56].

Treatment

The cornerstone to all malignant bone tumours is a combination of chemotherapy and wide-margin excision. This requires a multidisciplinary approach in order to create the best management plan. The medical and surgical team should include a musculoskeletal oncologist, a bone pathologist, paediatric oncologist, paediatric medical specialist, radiation oncologist and radiologist.

The major concept of modern treatment is neoadjuvant therapy. This means that systemic treatment is initiated rapidly after diagnosis (chemotherapy), local control treatment can be planned and tailored for specific circumstances (radiation therapy and/or limb salvage/ablation surgery) and then ongoing treatment can further tailored depending on tumour factors such as necrosis rates and margins.

Chemotherapy

Chemotherapy Principles

Chemotherapy has drastically improved the survival rates of cancer patients, when combined with complete tumour resection. Chemotherapy response is the most significant factor in local recurrence. A successful chemotherapeutic response is considered to be 90 % tumour necrosis. Those with less than 90 % necrosis have a higher rate of relapse in the first two years [44]. Protocols for chemotherapy are based on those of the Children's Oncology Group (COG). These are implemented in over 200 centres in the USA, Canada, Europe, Australia and New Zealand that treat children with cancer. Chemotherapy is not routinely required for low-grade intramedullary or surface OSAs and is variable for periosteal for low-grade periosteal OSAs [56]. All high-grade or metastatic OSA, Ewing's and neuroblastomas require chemotherapy according to local protocol.

Complications from chemotherapy are a significant issue for patients. Mucositis and ototoxicity are the most common, but renal damage and cardiac damage can occur and function of these organs should be monitored. Ototoxicity is especially challenging for neuroblastoma patients. The damage is not only bilateral and irreversible, but affects detection of high-frequency sounds required to hear human speech. As nearly 90 % are under the age of 5 and receiving ototoxic agents while language skills are developing, this can have severe implications on social and academic development [57].

Fertility should also be addressed before initiating chemotherapy. Some chemotherapy agents can have irreversible effects on fertility and tends to affect males more than females [58]. Patients may need to be referred for sperm banking/embryo freezing.

OSA

Therapy is a multi-agent regime including various combinations of doxorubicin, cisplatin, methotrexate, cyclophosphamide, ifosfamide, carboplatin, etoposide and vincristine [2]. The COG protocol is pre-operative high-dose methotrexate with doxorubicin and cisplatin. Patients undergo several cycles before surgical excision. Post-operatively, patients with more than 90 % necrosis are divided into two groups. Half receive the same post-operative medications and the other half receives the same post-operative medications with

an added pegylated form of interferon alpha 2b. If tumour necrosis is less than 90 %, post-operative chemotherapy is the same as pre-operative as well as randomly assigned ifosfamide or etoposide [59].

Ewing's Sarcoma

Multiagent therapy for Ewing's sarcoma is for at least 12 weeks in duration and those with metastatic disease should receive treatment for 6-months to a year. Combinations may include doxorubicin, ifosfamide, vincristine and etoposide. Dactinomycin and cyclophosphamide may also be incorporated. VAC/IE is the preferred regimen for local disease and VAdriaC for those with metastases [56]. Restaging with MRI or PET should be done following initial treatment, to determine if local control with surgery is appropriate.

Neuroblastoma

Platinum-based therapy with cisplatin, doxorubicin, cyclophosphamide, teniposide and etoposide is the basis of neuroblastoma treatment [57]. Chemotherapy may be used with the intent of myeloablation followed by autologous bone marrow transplant.

Radiation Therapy

The role of radiation in sarcoma management has diminished as chemotherapy and surgical technique has improved. Currently, it is indicated for patients with very large tumours, or for tumours that cannot be fully resected [12, 44]. It may also be used as an adjunct for marginal control.

There is an increasing evidence and acceptance for the use of Radiation therapy as sole local control therapy for Pelvic Ewings Sarcoma. This is, at least in part, due to the significant morbidity associated with pelvic resection surgery. Several studies have demonstrated equivalence in terms of local recurrence and disease free survival.

The risk of radiation in children is subsequent development of secondary tumours. Radiation is the most significant risk factor for secondary development. The excess relative risk of secondary OS is 1.4 per Gy [7]. Patients who have radiation treatment for a primary tumour require thorough follow-up to facilitate early detection of further tumours.

Surgical Treatment

Surgery should be considered for all patients, even those with metastatic disease. However, paediatric patients pose unique challenges for the operating surgeon. As chemotherapy has increased the survival rate for most childhood cancers, most children who develop OSA and EWS or secondary tumours can expect have a long life expectancy. The fundamental difference in a paediatric population is the potential for the immature skeleton and patient to heal, grow and adapt to the surgical manipulations available. The corollary of this is that the surgical reconstruction needs to be durable potentially able to withstand a 'lifetime' of use.

To decide the best surgical option for a patient, there are multiple factors that must be considered. Complete tumour resection is the priority and any reconstruction must accommodate for this. Other crucial considerations are the patient's age and future growth, the location of the tumour, and the consequences the procedure will have with regards to functional outcome and quality of life.

Finally, quality of life (QOL) is a major influence on treatment choice. This is a broad, ill-defined concept, but encompasses a number of facets that need consideration. QOL includes social, emotional, and physical components. Measuring these can especially be difficult when managing young children. Expectations around quality of life come primarily from parents and may not be congruent with the best surgical option. For example, while amputation carries a very negative preconception, amputees have faster acceptance and better quality of life based on patient surveys. In fact, there is minimal data to support that limb salvage is in any way better than amputation with regards to QOL [60].

Planning

Successful surgical management requires scrupulous preoperative planning and multi-disciplinary input. Communicate with oncology and medical teams to coordinate care, as surgery cannot interfere with chemotherapy. Discussion with surgeons from other specialties such as general, plastics, vascular or neurosurgery may be necessary depending on tumour location and tissue involvement.

MRI studies will be the guide surgical approach and precise excision. T1-weighted images are preferred. They correspond more closely to pathologic findings than STIR images, as STIR images can overestimate tumour extent [61]. MRI is highly accurate at determining neurovascular involvement, which will influence surgical management. Nerve involvement is rare but devastating. In these cases, the only likely option is amputation, as there is little value in sparing a limb if the nerve is resected [62]. The most frequent cause of nerve contamination is from pathological fracture. Vascular involvement is variable as diseased sections can be resected and vessels anastomosed.

If the growth plate has tumour involvement, the age and future growth of the patient are also important factors. A full workup of bone age and estimated adult height is useful when planning surgical measurements.

Ultimately, there is no standard algorithm for planning the correct procedure. It is a complex decision and must be tailored

to the situation of the patient. Furthermore, even with extensive pre-operative planning, there are always unforeseen factors. The definitive surgical decision will ultimately be made intra-operatively and alternative plans should be in place.

Resection

En-block resection of all diseased tissue, previous biopsy tracts and adjacent soft tissue is required to consider treatment as curative. Adequate margins are a fundamental component of surgical treatment for sarcomas, and the second most important factor influencing local recurrence, after chemotherapeutic response [63]. It is universally accepted that positive margins are associated with increased risk of recurrence; however, exactly what is a 'safe' margin is not well established. It is usually defined in quantitative measurements, with recommendations varying between 2 mm and 7 cm of normal tissue surrounding a tumour [64]. Some consider the measurement to not be relevant; the margin is adequate as long as the quality of surrounding tissue is good. Regardless of margin width, there are uncontrollable factors such as tumour location, size, grade and metastasis that will have influence overall prognosis [65]. The final excision biopsy should be assessed for margins, overall tumour necrosis, as well as the size and dimensions of the tumour to ensure complete removal and to guide post-operative chemotherapy [56].

Reconstruction

There are fundamental questions that must be asked when considering limb salvage. First, is limb salvage possible? This is primarily based on location of the tumour and neurovascular involvement. Second, is limb salvage functionally desirable? This will be based on age and future growth, any future reconstruction that may be required and what the functional outcome of reconstruction will be. Currently, over 90 % of patients diagnosed with OSA can be treated with some form of limb salvage. However, it is important to be aware that this is accompanied by a 60 % complication rate and 75 % revision rate.

Following the resection of some bone tumours, reconstruction offers no functional benefit. Examples of this include the proximal fibula (Fig. 44.1), individual ribs, the clavicle and parts of the scapula.

Biologic Reconstruction

Autograft

Autograft can either be involved tumour bone that has been sterilised or unaffected host bone. Unaffected host bone can either be vascularised or non-vascularised.

Irradiated or pasteurized autografts of the patients affected bone have a variable success rate. Experimentation using cryosurgery with liquid nitrogen of the autograft has also been trialled [66]. In order to consider using autograft tumour bone, there must be less than one-third cortical bone destruction, tumour confinement to one compartment and osteoblastic pattern [66, 67]. Positive features include the fact that no foreign material is used and a perfect fit is achieved at reconstruction [68]. Mid-term survivorship is comparable to other methods; however, non-union and infection rates are still very high.

Other biologic autografts may be used. The most common donor site for vascularised grafts is the fibula (Fig. 44.2),

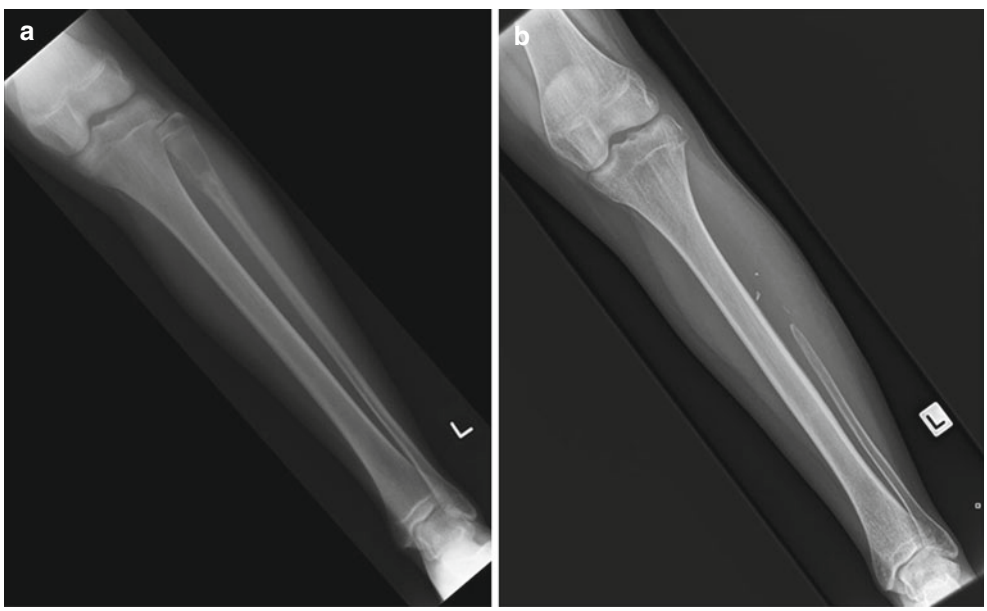

Fig. 44.1 Osteosarcoma of the proximal fibula. Thirteen year female presenting with osteosarcoma of the left proximal fibula; (**a**) presentation film and (**b**) resection of the proximal half of the fibula without reconstruction

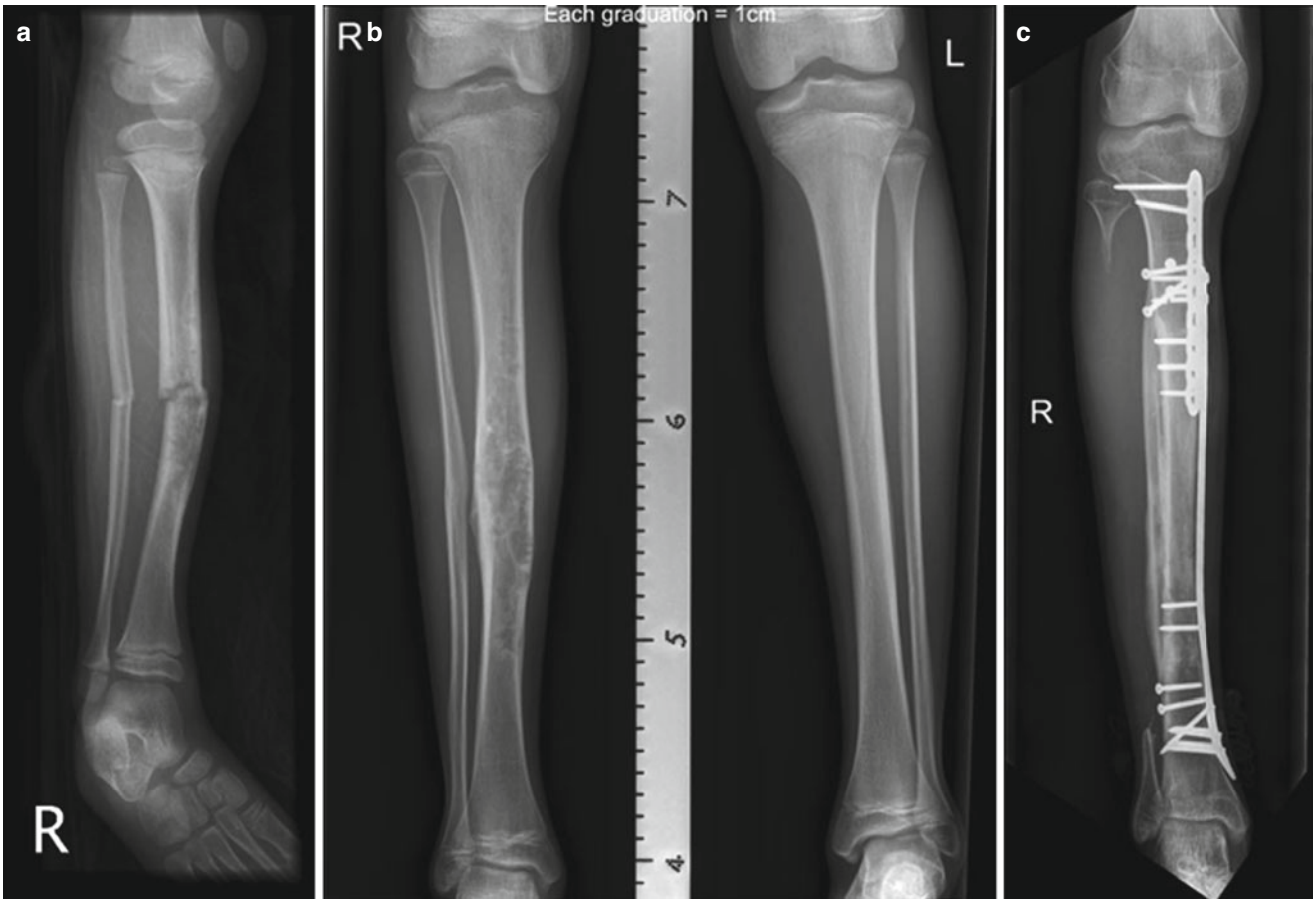

Fig. 44.2 Osteofibrous dysplasia right fibula. Osteofibrous dysplasia right tibia; (**a**) presentation age 6 with pathologic fracture. (**b**) following union and observation, deformity and further pain developed age 12. (**c**) following resection and intercalary allograft reconstruction augmented with an ipsilateral vascularised fibula transport

followed by the tibia and iliac crest. These can be combined with fascial, muscle and cutaneous components. If the bone defect is greater than 6 cm, vascularised autograft is preferable. As it is the patient's own vascular bone, excellent integration and healing is typically seen, and for these reasons they are often combined with allograft usage.

Non-vascularised autograft is typically particulate graft harvested from the iliac crest. This provides good osteoinductive properties but limited osteoconductive properties.

Distraction Osteogenesis

Following tumour resection, a temporary spacer placement can be utilised to encourage vascularised membrane formation followed by a delayed secondary grafting procedure. This technique was popularised by Masquelet and colleagues. They describe excellent longterm results with this technique [69, 93].

Distraction Osteogenesis utilises a similar principle to achieve biologic filling of a bony defect [94].

Intercalary Allograft

Allografts can be composed of either cancellous bone or cortical bone. The use of intercalary allografts for metaphyseal and diaphyseal lesions (with or without vascularized fibula) can provide excellent outcomes and sparing of the growth plate eliminates the issues of limb length discrepancy [4, 70] (Fig. 44.3). Consolidation time is long, averaging 6.5 months at the metaphyseal and 16 months in the diaphyseal osteotomy sites and overall survival rate of the graft is approximately 76 % at 10-years. The most significant risk is non-union, thus adequate fixation of cortical grafts is vital, achieved with plates. Allograft collapse is also a possibility; therefore, the number of screw holes placed in the graft should be minimal to prevent structural compromise. Intramedullary nails have also been used for fixation but non-union of the diaphyseal junctions are higher with nail fixation, likely due to inadequate compression between graft and host bone [71]. Some studies using compressible intramedullary devices have shown improved rates of union [72].

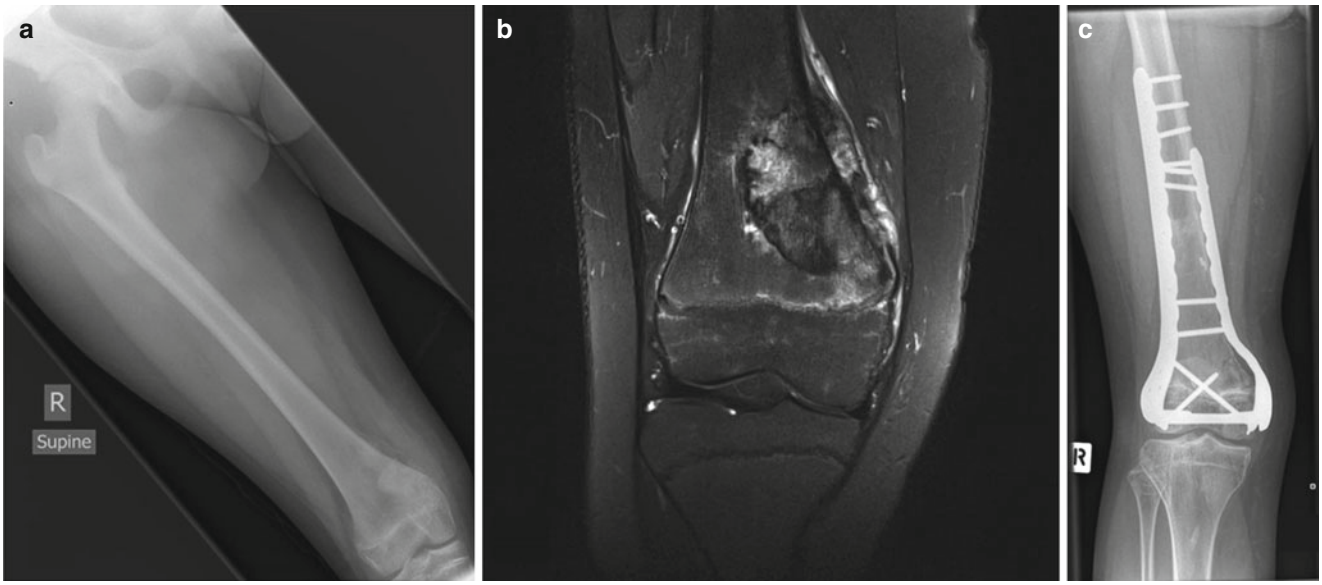

Fig. 44.3 Right distal femoral osteosarcoma. A 13 year. female with right distal femoral osteosarcoma; (**a**) Presentation film, (**b**) Coronal MRI demonstrating tumour extent and oedema at the level of the physis, (**c**) Following transepiphyseal resection and intercalary allograft reconstruction

Osteoarticular Allograft

Osteo-articular allografts are an option when the joint cannot be preserved. Fracture, non-union, joint instability, articular degeneration, and infection are common, thus it is utilised as a temporary solution for patients who still have significant growth [73]. Graft survival is 37–71 % at 5–10 years [73]. It carries a very high re-operation rate. Once skeletal maturity is reached, a conversion to a composite endoprosthetic joint can be performed.

Transepiphyseal Resection

Transepiphyseal resection may be considered a safe method of limb salvage for patients with juxta-articular lesion. If there is no indication of tumour involvement within the joint on MRI and a safe margin can be obtained, an osteotomy can be made through the epiphysis to preserve the articular portions. Reconstruction has been achieved with epiphyseal fragments as little as 0.5–2 cm, with no reported incidences of AVN or epiphyseal non-union. Reconstruction is with an intercalary graft with or without vascularised fibula [74]. Fixation is achieved with a lateral plate and screws. Diaphyseal fixation is with a plate or with rod.

Complications include deep infection, joint stiffness, diaphyseal non-union and stress fractures. In follow-up studies of patients receiving a transepiphyseal resection (82 patients in 5 studies over an average follow-up time of 54.2 months), there were no cases of local recurrence. Functional outcome is very good, with no restrictions of physical activity [74–76].

Arthrodesis

Arthrodesis is utilised primarily for the salvage of a failed allograft or failed ER. It usually is not desirable as it removes the joint function. However in certain circumstances it is a useful treatment option. Joints which can be arthrodesed with minimal functional loss include the wrist, shoulder, hip and ankle. Arthrodesis of the knee greatly decreases gait efficiency, but it is durable and functional with 86 % of patients achieving independent ambulation [60, 77]. In general, patients with arthrodesis have reasonable functional outcomes with average MSTS scores of 77 % [77] (Fig. 44.4).

Prosthetic and Alloprosthetic Reconstruction

Endoprosthetic Reconstruction

At first glance, endoprosthetic reconstruction appears to be the silver bullet for limb salvage surgery (LSS). They can be modular or custom made while the patient is undergoing pre-operative chemotherapy. It enables early mobility and is aesthetically desirable to patients (and parents) as it recreates the entire limb. It creates a mechanical joint for patients who have joint involvement and for many has very good functional outcomes in the short term. MSTS scores are very high with ER in the first post-operative years.

Its downfall is in longevity and durability and for growing patients, a lack of dynamicity. Paediatric patients have many decades of active life ahead of them, and it is unlikely that an endoprosthetic reconstruction will withstand a lifetime of wear and tear. Implant survival is variable and can be

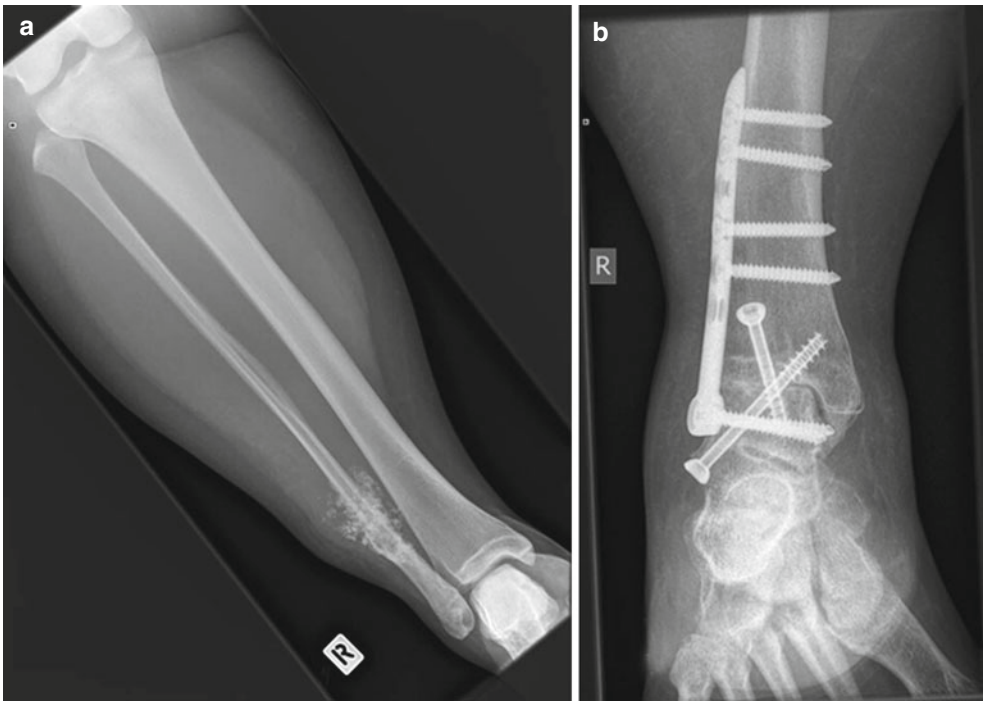

Fig. 44.4 Chondroblastic osteosarcoma right distal fibula. A 17 year. female with chondroblastic OSA right distal fibula; (**a**) presentation films and (**b**) following fibula resection and tibiotalar arthrodesis

anywhere from 50 % to 90 % at 5–10 years [78, 79]. ER has a high complication rate including aseptic loosening, mechanical failure, periprothetic fracture and infection [79]. Thus, when performing an ER on a young patient, one should anticipate and plan for future revisions. Surprisingly LSS does not correlate with significantly improved levels of quality of life when compared to other options such as rotationplasty or amputation.

Children with more than 2–3 cm of growth should not be considered for ER, as there is much anticipated growth and the limb length discrepancy will be too significant [44]. This has been challenged with the advent of telescopic devices that "grow" and extend with the child. Newer versions use an electromagnetically driven gearbox that turns a drive screw to extend the prosthetic rod. Lengthening is a non-invasive outpatient procedure and avoids subsequent surgeries. Short term outcomes are attractive but are still fraught with risk [80]. Infection rates between 25 % and 40 % are the most common complication, but implant breakage, gearbox failure and loosening also can occur, leading to high rates of revision [60, 81, 82].

For all endoprosthetic devices, infection risk is high. A significant contributing factor to infection is poor soft-tissue coverage and sufficient coverage of implants is essential. This may be through the use of muscle flaps. For example, in tibial reconstruction, introducing a gastrocnemius flap reduced the risk of infection from 36 % to 12 % [83] (Fig. 44.5).

Allograft Prosthetic Composite

APC techniques replace the necessary bone stock with frozen allograft combined with a metallic implant. The allograft provides soft tissue attachments and the metallic joint implant provides better stability and function than osteoarticular allografts. Common sites for APC are the distal femur and proximal tibia, proximal femur and proximal humerus. APCs pose the same complication risks as ER and allograft, but can provide excellent functional results in the reconstruction of difficult soft tissue organs such as the extensor mechanism (Fig. 44.6).

Rotationplasty

Rotationplasty is a modified amputation reserved for patients for which limb salvage is not feasible. It involves the resection of the tumour site and rotation of the distal limb portion, creating a biological recreation of a joint. Pre-operative planning is imperative in this operation as children undergoing rotationplasty are young. The remaining growth of the child needs to be taken into consideration to minimize limb length discrepancies. Contraindications include compromise of the sciatic nerve, poor ankle or foot function, or limb infection.

The most common variation is the Vann-Nes [84], where tumours about the knee are resected en block and the neurovascular bundle is spared with the distal portion of the

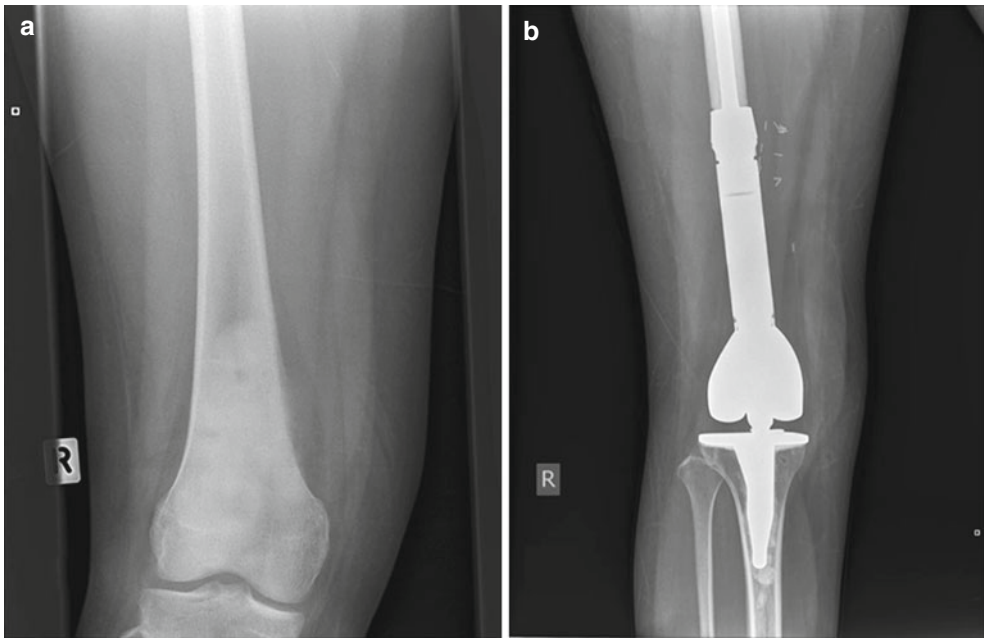

Fig. 44.5 Right distal femoral OSA. A 14 year. female with right distal femoral OSA, metastatic at presentation; (**a**) presentation film and (**b**) following resection and distal femoral endoprosthetic reconstruction

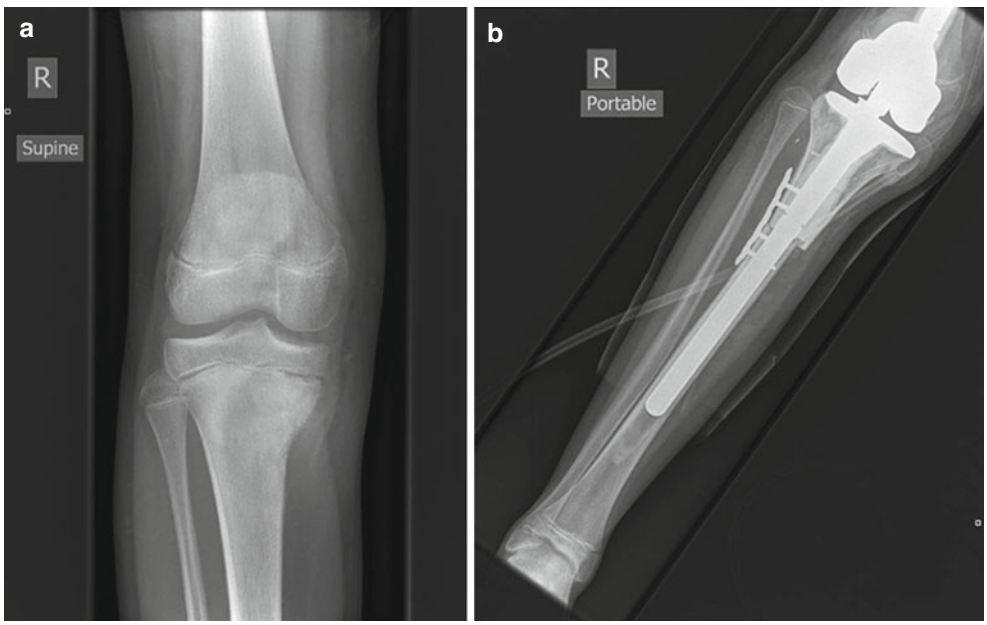

Fig. 44.6 Osteosarcoma of right proximal tibia. A 13 year. male with OSA right proximal tibia; (**a**) presentation film and (**b**) following resection and allograft-prosthetic reconstruction

leg. The remaining leg is rotated 180 degrees and reattached with the foot at the level of the contralateral knee when accounting for future growth. To provide plantar flexion of the ankle, the femoral origins of the gastrocnemius muscle are augmented to the quadriceps muscles at their origins [84]. The anterior and posterior tibial and peroneal muscles usually stay fixed to the tibia and fibula providing dorsal flexion of the ankle joint or can be fixed to the hamstrings fascia.

This procedure is a valuable option for patients not suitable for limb salvage. It eliminates the issue of growth in the skeletally immature, avoids risks of endoprostheses such as infection or device failure, has better functional results over amputation, maintains foot proprioception and overcomes phantom limb pain by sparing the sciatic nerve. Patients who receive a rotationplasty have excellent functional outcomes compared to amputation [85]. In fact, even when compared to endoprosthetic reconstruction, rotationplasty is not

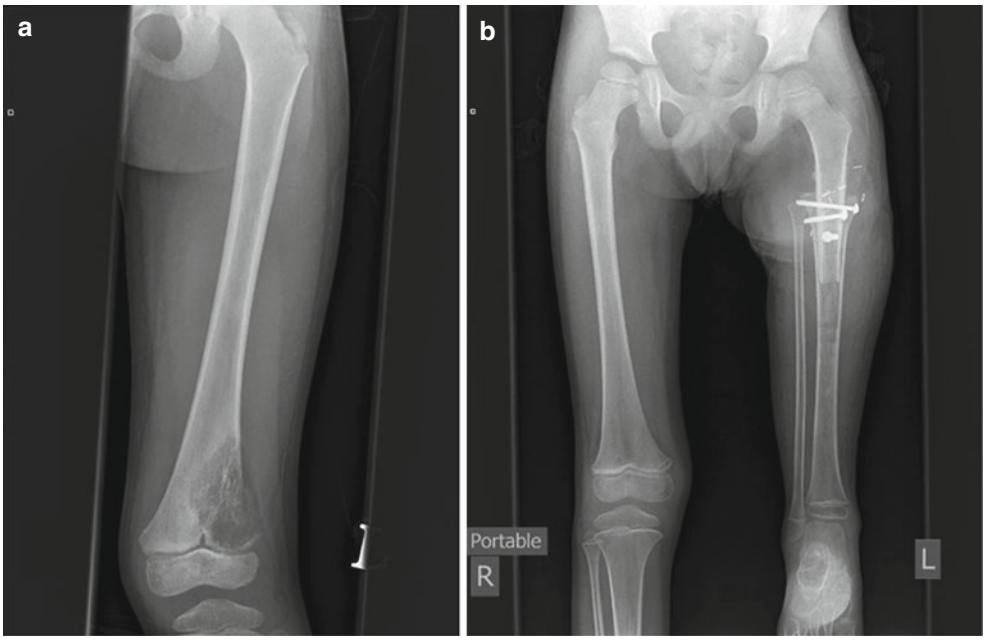

Fig. 44.7 Left distal femoral Ewing sarcoma. Four year male with left distal femoral EWS; (**a**) Presentation film and (**b**) following rotationplasty of the right lower limb. Note the telescoped femur inside the proximal tibia and limited fixation required

associated with any functional disadvantages or differences in quality of life [86]. Patients receiving a rotationplasty can participate in high intensity activities more frequently than children receiving endoprosthesis as there is no risk of reconstruction failure [87, 95].

The only apparent drawback to rotationplasty is cosmetic appearance. There has been much literature exploring the risk of psychological difficulties due to the appearance of a rotationplasty. This has not observed this at our institution nor in other published evaluations [88, 89] (Fig. 44.7).

Amputation

For nearly all patients, amputation should be the last resort. Unless the tumour cannot be fully resected or the function of the limb will be useless, there is no advantage in amputation with regards to disease survival or local recurrence [90]. If there is a close margin and or poor chemotherapy response, limb salvage should still be considered first. While amputation achieves better local control, there is no significant increase in survival when compared to patients receiving an limb salvage [65]. In some circumstances, it may be reasonable to consider amputation first if there is the likelihood of multiple operations or factors that will prevent good functional results.

Other than poorer functional outcomes, amputation in young children can be problematic. As a child grows, the remaining bone can either too short or may outgrow the surrounding soft tissue at the distal end, requiring surgical revision [60].

Post-operative Considerations

Accurate evaluation of post-operative complications and true implant/reconstruction failure rates is difficult to assess. The competing risks of local recurrence and death combine to eliminate 30–40 % of patients from long-term follow-up.

Early Post-operative Care

Infection rates are generally high in the immediate post-operative phase and are influenced by the type of reconstruction implemented. It is the most common complication and can occur in the short or long term, and is also influenced by the adjuvant therapies of chemotherapy and radiation therapy.

Skin care and wound healing is key to prevent delays in post-operative chemotherapy. Rehabilitation begins directly after surgery. Active mobilization that does not compromise the operated limb promotes blood flow, preserves strength and motor function in other limbs.

Infection

Infection is the leading cause of implant revision over the long term. Time to infection development can range from 1 to 50 months, and is usually Staphylococcus aureus and Staphylococcus epidermis [83].

Table 44.2 Evidence-based recommendations

Statements	LoE
Paediatric orthopaedic oncology should be managed in a specialised multidisciplinary way	2C
Biopsy of paediatric orthopaedic oncologic lesions should be performed in a methodical way under the direction of an orthopaedic oncologist	2a
Chemotherapy should be provided to paediatric patients with malignant musculoskeletal tumours	1b
Local control of the primary malignancy in osteosarcoma should involve negative margin resection surgery	3a
Local control of the primary malignancy in Ewing's sarcoma can involve either surgery and/or radiation therapy	3b
Reconstructive options should be tailored to the individual needs and demands of the Paediatric patient and their family	5

Aseptic Loosing

Rates of aseptic loosening in ER are approximately 6 % at 10–15 years and 14 % at 20 years [91]. Bone:stem ratio is the major factor predicating aseptic failure. Patients with small stem sizes and higher bone:stem ratios are more likely to develop aseptic loosening.

Non-union

Non-union is particular high in allograft reconstruction, but is a risk for any procedure. Those who receive extensive adjuvant chemotherapy have an increased risk of non-union due to its adverse effects on bone quality.

Fracture

Fracture can be associated with the bone around the implant or in other areas secondary to decreased bone mineral density (BMD) from chemotherapy. BMD is significantly lower in survivors who received chemotherapy when compared to age-matched reference populations, leading to a high rate of fractures in the long-term.

Long-Term Follow Up

After completing post-operative chemotherapy patients should undergo regular radiographic follow-up occurring every 3 months for the first year, every 6 months in the second year and annually thereafter. Five years after final therapy is considered ling-term survival. Follow-up should also include functional measures and prosthetic function evaluation. The MSTS score is a useful subjective and objective assessment of functional outcome. It incorporates pain levels, functional levels, emotional acceptance, support needs, waling ability and gait [92].

Another consideration in follow-up is screening depending on adjuvant treatment, as chemotherapy and radiotherapy have long-term consequences. For example, abdominal or pelvic radiation requires colorectal cancer screening, gonadal function and renal screening. Patients who received cisplatin and carboplatin may need ongoing cardiovascular renal and audiological assessment. This list is not exhaustive and it is the role of the managing team to ensure appropriate screening is arranged based on the patient's needs. There are detailed guidelines developed by the Children's Oncology Group for recommended long-term follow up. Table 44.2 outlines the evidence-based recommendations.

References

1. Duong LA, Richardson LC. Descriptive epidemiology of malignant primary OSA using population-based registries, United States, 1999–2008. J Registry Manag. 2013;40(2):59–64.
2. Luetke A, Meyers PA, Lewis I, Juergens H. OSA treatment–where do we stand? A state of the art review. Cancer Treat Rev. 2014;40(4):523–32.
3. Geller DS, Gorlick R. OSA: a review of diagnosis, management, and treatment strategies. Clin Adv Hematol Oncol. 2010;8(10):705–18.
4. Moore DD, Luu HH. OSA. In: Orthopaedic oncology. Springer; Chicago. 2014. pp. 65–92.
5. Group ESESNW. Bone sarcomas: ESMO clinical practice guidelines for diagnosis, treatment and follow-up. Ann Oncol. 2012;23(suppl 7):vii100–9.
6. Larizza L, Roversi G, Volpi L. Rothmund-thomson syndrome. Orphanet J Rare Dis. 2010;5(2).
7. Henderson TO, Rajaraman P, Stovall M, Constine LS, Olive A, Smith SA, et al. Risk factors associated with secondary sarcomas in childhood cancer survivors: a report from the childhood cancer survivor study. Int J Radiat Oncol Biol Phys. 2012;84(1):224–30.
8. Meazza C, Luksch R, Daolio P, Podda M, Luzzati A, Gronchi A, et al. Axial skeletal OSA: a 25-year monoinstitutional experience in children and adolescents. Med Oncol. 2014;31(4):1–6.
9. Bacci G, Ferrari S, Bertoni F, Ruggieri P, Picci P, Longhi A, et al. Long-term outcome for patients with nonmetastatic OSA of the extremity treated at the istituto ortopedico rizzoli according to the istituto ortopedico rizzoli/OSA-2 protocol: an updated report. J Clin Oncol. 2000;18(24):4016–27.
10. Willeumier J, Fiocco M, Nout R, Dijkstra S, Aston W, Pollock R, et al. High-grade soft tissue sarcomas of the extremities: surgical margins influence only local recurrence not overall survival. Int Orthop. 2015:1–7.
11. Iwamoto Y. Diagnosis and treatment of Ewing's sarcoma. Jpn J Clin Oncol. 2007;37(2):79–89.
12. Ross KA, Smyth NA, Murawski CD, Kennedy JG. The biology of ewing sarcoma. ISRN Oncol. 2013;2013.

13. Balamuth NJ, Womer RB. Ewing's sarcoma. Lancet Oncol. 2010;11(2):184–92.
14. Bernstein M, Kovar H, Paulussen M, Randall RL, Schuck A, Teot LA, et al. Ewing's sarcoma family of tumors: current management. Oncologist. 2006;11(5):503–19.
15. Dehal A, Copeland H, Kheradpour A, Martin M, Wallen J, Zaheer S. Ewing's Sarcoma of the sternum: a case report and literature review. J Surg [Jurnalul de Chirurgie]. 2014;10(15):67–9.
16. Springfield DS, Rosenberg AE, Mankin HJ, Mindell ER. Relationship between osteofibrous dysplasia and ADA. Clin Orthop Relat Res. 1994;309:234–44.
17. Keeney GL, Unni KK, Beabout JW, Pritchard DJ. ADA of long bones. A clinicopathologic study of 85 cases. Cancer. 1989;64(3):730–7.
18. Taylor RM, Kashima TG, Ferguson DJ, Szuhai K, Hogendoorn PC, Athanasou NA. Analysis of stromal cells in osteofibrous dysplasia and ADA of long bones. Mod Pathol. 2012;25(1):56–64.
19. Gudaganatti SB, Jadhav MN, Patil RK, Kittur SK. Recurrent ADA of the tibia and lymph node metastasis. Medica. 2014;3(2):127.
20. Duan P-G, Li R-Y, Jiang Y-Q, Wang H-r, Zhou X-G, Li X-L, et al. Recurrent ADA in the thoracolumbar spine successfully treated by three-level total en bloc spondylectomy by a single posterior approach. Eur Spine J. 2014:1–8.
21. Haupt R, Minkov M, Astigarraga I, Schäfer E, Nanduri V, Jubran R, et al. Langerhans cell histiocytosis (LCH): guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years. Pediatr Blood Cancer. 2013;60(2):175–84.
22. Degar BA, Rollins BJ. Langerhans cell histiocytosis: malignancy or inflammatory disorder doing a great job of imitating one? Dis Model Mech. 2009;2(9–10):436–9.
23. Betts DR, Leibundgut KE, Feldges A. PlÃ¼ss HJ, Niggli FK. Cytogenetic abnormalities in Langerhans cell histiocytosis. Br J Cancer. 1998;77(4):552.
24. Howarth DM, Gilchrist GS, Mullan BP, Wiseman GA, Edmonson JH, Schomberg PJ. Langerhans cell histiocytosis. Cancer. 1999;85(10):2278–90.
25. Nishal AJ, Modi JP, Balar HR. Langerhans cell histiocytosis. Indian J Otol. 2013;19(4):194.
26. Picci P, Manfrini M, Fabbri N, Gambarotti M, Vanel D. Langerhans cell hystocytosis in: atlas of musculoskeletal tumors and tumorlike lesions: the rizzoli case archive. Springer Science & Business Media; Switzerland. 2014.
27. Langerhans L. Eosinophilic granuloma. J Bone Joint Surg Br. 2002;2002(84-B):870–2.
28. Eyre R, Feltbower RG, Mubwandarikwa E, Jenkinson HC, Parkes S, Birch JM, et al. Incidence and survival of childhood bone cancer in northern England and the West Midlands, 1981–2002. Br J Cancer. 2009;100(1):188–93.
29. Kivioja A, Ervasti H, Kinnunen J, Kaitila I, Wolf M, Böhling T. Chondrosarcoma in a family with multiple hereditary exostoses. J Bone Joint Surg Br. 2000;82(2):261–6.
30. Madan S, Robinson K, Kasliwal P, Bell M, Saleh M, Fernandes J. Limb reconstruction in Ollier's disease. Strateg Trauma Limb Reconstr. 2015;10(1):49–54.
31. Verdegaal SH, Bovée JV, Pansuriya TC, Grimer RJ, Ozger H, Jutte PC, et al. Incidence, predictive factors, and prognosis of chondrosarcoma in patients with Ollier disease and Maffucci syndrome: an international multicenter study of 161 patients. Oncologist. 2011;16(12):1771–9.
32. Reddy DSK, Kumar RK, Gali R, Kannubaddy SR, Rao M, Akheel M. Central chondrosarcoma of a pediatric mandibular condyle: a case report and review. Ann Maxillofac Surg. 2014;4(1):85.
33. Bovée JV, Cleton-Jansen A-M, Taminiau AH, Hogendoorn PC. Emerging pathways in the development of chondrosarcoma of bone and implications for targeted treatment. Lancet Oncol. 2005;6(8):599–607.
34. Fabbri N, Donati D. Central chondrosarcoma. In: Atlas of musculoskeletal tumors and tumorlike lesions. Springer; Switzerland. 2014. pp. 113–7.
35. van Maldegem AM, Bovée JV, Gelderblom H. Comprehensive analysis of published studies involving systemic treatment for chondrosarcoma of bone between 2000 and 2013. Clinical Sarcoma Research. 2014;4(1):11.
36. Akpolat N, Yildirim H, Poyraz K. Sacral chondroblastic OSA misdiagnosed as chondrosarcoma and chordoma. Turkish J Med Sci. 2007;37(4):243–9.
37. Cheung N-KV, Dyer MA. Neuroblastoma: developmental biology, cancer genomics and immunotherapy. Nat Rev Cancer. 2013;13(6):397–411.
38. Brodeur GM. Neuroblastoma: biological insights into a clinical enigma. Nat Rev Cancer. 2003;3(3):203–16.
39. Zhao H, Cai W, Li S, Da Z, Sun H, Ma L, et al. Characterization of neuroblastoma bone invasion/metastasis in established bone metastatic model of SY5Y and KCNR cell lines. Childs Nerv Syst. 2013;29(7):1097–105.
40. Sohara Y, Shimada H, DeClerck YA. Mechanisms of bone invasion and metastasis in human neuroblastoma. Cancer Lett. 2005;228(1):203–9.
41. Picci P. OSA (osteogenic sarcoma). Orphanet J Rare Dis. 2007;2(6).
42. Picci P. Classic OSA. In: Atlas of musculoskeletal tumors and tumorlike lesions. Springer; Switzerland. 2014. pp. 147–52.
43. Stacy GS, Mahal RS, Peabody TD. Staging of bone tumors: a review with illustrative examples. Am J Roentgenol. 2006;186(4):967–76.
44. San-Julian M, Vázquez-García BL, Sierrasesúmaga L. Limb salvage in children. Eur Surg Orthop Traumatol EFORT Textbook. 2014:4251–80.
45. Heymann D. Bone cancer: Primary bone cancers and bone metastases. Elsevier; San Diego, CA. 2014.
46. Moore DD, Haydon RC. Ewing's sarcoma of bone. In: Orthopaedic oncology. Springer; Chicago, IL. 2014. pp. 93–115.
47. Franzius C, Sciuk J, Daldrup-Link HE, Jürgens H, Schober O. FDG-PET for detection of osseous metastases from malignant primary bone tumours: comparison with bone scintigraphy. Eur J Nucl Med. 2000;27(9):1305–11.
48. Avedian RS. Principles of musculoskeletal biopsy. In: Orthopaedic oncology. Springer; Chicago, IL. 2014. pp. 1–7.
49. Clark CR, Morgan C, Sonstegard DA, Matthews LS. The effect of biopsy-hole shape and size on bone strength. J Bone Joint Surg. 1977;59(2):213–7.
50. Bickles J, Jelinek JS, Shmookler BM, Neff RS, Malawer MM. Biopsy of musculoskeletal tumors: current concepts. Clin Orthop Relat Res. 1999;368:212–9.
51. Huang P-Y, Wu P-K, Chen C-F, Lee F-T, Wu H-T, Liu C-L, et al. Osteomyelitis of the femur mimicking bone tumors: a review of 10 cases. World J Surg Oncol. 2013;11:283.
52. Davido N, Rigolet A, Kerner S, Gruffaz F, Boucher Y. Case of Ewing's sarcoma misdiagnosed as a periapical lesion of maxillary incisor. J Endod. 2011;37(2):259–64.
53. Gould CF, Ly JQ, Lattin GE, Beall DP, Sutcliffe JB. Bone tumor mimics: avoiding misdiagnosis. Curr Probl Diagn Radiol. 2007;36(3):124–41.
54. Tow BP, Tan MH. Delayed diagnosis of Ewing's sarcoma of the right humerus initially treated as chronic osteomyelitis: a case report. Journal of Orthopaedic Surgery. 2005;13(1).
55. Enneking WF, Spanier SS, Goodman MA. A system for the surgical staging of musculoskeletal sarcoma. Clin Orthop Relat Res. 1980;153:106–20.
56. National Comprehensive Cancer N. NCCN Clinical Practice Guidelines in Oncology-Bone Cancer, 2012. 2013. https://www.nccn.org/professionals/physician_gls/f_guidelines.asp.
57. Landier W, Knight K, Wong FL, Lee J, Thomas O, Kim H, et al. Ototoxicity in children with high-risk neuroblastoma: prevalence,

risk factors, and concordance of grading scales –a report from the children's oncology group. Journal of Clinical Oncology. 2014:JCO. 2013.51. 38.
58. Longhi A, Macchiagodena M, Vitali G, Bacci G. Fertility in male patients treated with neoadjuvant chemotherapy for OSA. J Pediatr Hematol Oncol. 2003;25(4):292–6.
59. Isakoff MS, Stefan S, Bielack BM. Osteosarcoma: current treatment and a collaborative pathway to success. J Clin Oncol. 2015;33:3029–35.
60. Nagarajan R, Neglia JP, Clohisy DR, Robison LL. Limb salvage and amputation in survivors of pediatric lower-extremity bone tumors: what are the long-term implications? J Clin Oncol. 2002;20(22):4493–501.
61. Onikul E, Fletcher BD, Parham DM, Chen G. Accuracy of MR imaging for estimating intraosseous extent of OSA. AJR Am J Roentgenol. 1996;167(5):1211–5.
62. Van Trommel MF, Kroon HM, Bloem JL, Hogendoorn PCW, Taminiau AHM. MR imaging based strategies in limb salvage surgery for OSA of the distal femur. Skeletal Radiol. 1997;26(11):636–41.
63. Picci P, Sangiorgi L, Rougraff BT, Neff JR, Casadei R, Campanacci M. Relationship of chemotherapy-induced necrosis and surgical margins to local recurrence in OSA. J Clin Oncol. 1994;12(12):2699–705.
64. Gouin F, Heymann M-F. Margins and bone tumors–what are we talking about? Bone Cancer: Prim Bone Cancers Bone Metastases. 2014;287.
65. Reddy KA, Wafa H, Gaston CL, Grimer RJ, Abudu AT, Jeys LM, et al. Does amputation offer any survival benefit over limb salvage in OSA patients with poor chemonecrosis and close margins? Bone Joint J. 2015;97(1):115–20.
66. Tsuchiya H, Wan SL, Sakayama K, Yamamoto N, Nishida H, Tomita K. Reconstruction using an autograft containing tumour treated by liquid nitrogen. J Bone Joint Surg. 2005;87(2):218–25.
67. Jeon D-G, Kim MS, Cho WH, Song WS, Lee S-Y. Pasteurized autograft–prosthesis composite for distal femoral OSA. J Orthop Sci. 2007;12(6):542–9.
68. Sys G, Uyttendaele D, Poffyn B, Verdonk R, Verstraete K. Extracorporeally irradiated autografts in pelvic reconstruction after malignant tumour resection. Int Orthop. 2002;26(3):174–8.
69. Masquelet AC, Fitoussi F, Begue T, Muller GP. Reconstruction of the long bones by the induced membrane and spongy autograft. Ann Chir Plast Esthet. 2000;45(3):346–53.
70. Virkus WV, Henshaw RM, Miller B, Girelis S. Use of allografts and segmental prostheses for reconstruction of segmental bone defects. In: Operative techniques in orthopaedic surgery. Lippincott Williams and Wilkins; Philadelphia, PA. 2010.
71. Aponte-Tinao L, Farfalli GL, Ritacco LE, Ayerza MA, Muscolo DL. Intercalary femur allografts are an acceptable alternative after tumor resection. Clin Orthop Relat Res. 2012;470(3):728–34.
72. Miller BJ, Virkus WW. Intercalary allograft reconstructions using a compressible intramedullary nail: a preliminary report. Clin Orthop Relat Res. 2010;468(9):2507–13.
73. Ramseier LE, Malinin TI, Temple HT, Mnaymneh WA, Exner GU. Allograft reconstruction for bone sarcoma of the tibia in the growing child. J Bone Joint Surg. 2006;88(1):95–9.
74. Manfrini M, Gasbarrini A, Malaguti C, Ceruso M, Innocenti M, Bini S, et al. Intraepiphyseal resection of the proximal tibia and its impact on lower limb growth. Clin Orthop Relat Res. 1999;358:111–9.
75. Muscolo DL, Ayerza MA, Aponte-Tinao LA, Ranalletta M. Partial epiphyseal preservation and intercalary allograft reconstruction in high-grade metaphyseal OSA of the knee. J Bone Joint Surg. 2004;86(12):2686–93.
76. Tsuchiya H, Abdel-Wanis ME, Sakurakichi K, Yamashiro T, Tomita K. OSA around the knee Intraepiphyseal excision and biological reconstruction with distraction osteogenesis. J Bone Joint Surg. 2002;84(8):1162–6.
77. Wolf RE, Scarborough MT, Enneking WF. Long term followup of patients with autogenous resection arthrodesis of the knee. Clin Orthop Relat Res. 1999;358:36–40.
78. Chandrasekar CR et al. Modular endoprosthetic replacement for tumours of the proximal femur. J Bone Joint Surg. 2009;91:108–12.
79. Kinkel S, Lehner B, Kleinhans JA, Jakubowitz E, Ewerbeck V, Heisel C. Medium to long-term results after reconstruction of bone defects at the knee with tumour endoprostheses. J Surg Oncol. 2010;101:166–9.
80. Nystrom LM, Morcuende JA. Expanding endoprosthesis for pediatric musculoskeletal malignancy: current concepts and results. Iowa Orthop J. 2010;30:141.
81. Picardo NE, Blunn GW, Shekkeris AS, Meswania J, Aston WJ, Pollock RC, et al. The medium-term results of the Stanmore non-invasive extendible endoprosthesis in the treatment of paediatric bone tumours. J Bone Joint Surg. 2012;94(3):425–30.
82. Mavrogenis AF, Papagelopoulos PJ. Expandable prostheses for the leg in children. Orthopedics. 2012;35(3):173–5.
83. Grimer RJ, Carter SR, Tillman RM, Sneath RS, Walker PS, Unwin PS, et al. Endoprosthetic replacement of the proximal tibia. J Bone Joint Surg. 1999;81(3):488–94.
84. van de Sande MAJ, Vochteloo AJH, Dijkstra PDS, Taminiau AHM. Van Nes-Borggreve rotationplasty of the knee. Eur Surg Orthop Traumatol EFORT Textbook. 2014;(1):4135–47.
85. So NF, Andrews KL, Anderson K, Gozola MA, Shives TC, Rose PS, et al. Prosthetic fitting after rotationplasty of the knee. Am J Phys Med Rehabil. 2014;93(4):328–34.
86. Hillmann A, Hoffmann C, Gosheger G, Krakau H, Winkelmann W. Malignant tumor of the distal part of the femur or the proximal part of the Tibia: endoprosthetic replacement or rotationplasty. Functional outcome and quality-of-life measurements*. J Bone Joint Surg. 1999;81(4):462–8.
87. Heare T, Hensley MA, Dell'Orfano S. Bone tumors: OSA and Ewing's sarcoma. Curr Opin Pediatr. 2009;21(3):365–72.
88. Bernthal NM, Monument MJ, Randall RL, Jones KB. Rotationplasty: beauty is in the eye of the beholder. Oper Tech Orthop. 2014;24(2):103–10.
89. Petri M, Omar M, Horstmann H, Brand S, Krettek C. Eighteen-year follow-up after rotationplasty for a grade IIIC open fracture of the distal femur. Arch Orthop Trauma Surg. 2013;133(3):351–5.
90. Grimer RJ, Carter SR, Tillman RM, Abudu S, Jeys L. Osa with poor necrosis and close margins–amputation or limb salvage? J Bone Joint Surg Br. 2012;94(SUPP XIV):45.
91. Bergin PF, Noveau JB, Jelinek JS, Henshaw RM. Aseptic loosening rates in distal femoral endoprostheses: does stem size matter? Clin Orthop Relat Res®. 2012;470(3):743–50.
92. Enneking WF, Dunham W, Gebhardt MC, Malawar M, Pritchard DJ. A system for the functional evaluation of reconstructive procedures after surgical treatment of tumors of the musculoskeletal system. Clin Orthop Relat Res. 1993;286:241–6.
93. Malevez C, Dujardin T, Glorieux V, Swennen G, Schutyser F, Van Cleynenbreugel J. Bone distraction osteogenesis. Revue belge de medecine dentaire. 2001;57(2):137–49.
94. Tsuchiya H, Tomita K, Minematsu K, Mori Y, Asada N, Kitano S. Limb salvage using distraction osteogenesis a classification of the technique. J Bone Joint Surg Br. 1997;79(3):403–11.
95. Hanlon MJ, Krajbick IJ. Rotationplasty in skeletally immature patients: long term follow-up results. Clin Orthop Relat Res. 1999; 385.

Part XI

Miscellaneous Paediatric Conditions

Physeal Injury, Epiphysiodesis and Guided Growth

45

Laura Deriu and Deborah M. Eastwood

Abstract

The evidence for the best treatment of acute physeal injuries is based mainly on epidemiologic studies, retrospective reviews, and expert opinion (LoE IV/V).

Epiphysiodesis is indicated to treat mild to moderate leg length discrepancy (<5 cm). Percutaneous epiphysiodesis produces a growth arrest as effectively as open procedures but with a significantly lower complication rate. A percutaneous (drill) epiphysiodesis is more effective than staples, PETS technique (percutaneous ephiphysiodesis with transphyseal screws) and guided growth with tension band devices and has fewer complications (LoE III). Staples work significantly better than PETS to correct leg length discrepancy but show a significantly higher complication rate. 8-plates show significantly less peri-operative complications than staples and PETS but do not significantly reduce growth.

Hemiepiphysiodesis with staples and transphyseal screw is a safe and reliable procedure to correct angular deformity around the knee. The latter is less invasive, more cosmetic and does not produce permanent physeal arrest. Angular deformity correction with staples and 8-plate is equally effective even in younger patients. Correction of angular deformity with 8-plate in the presence of a pathological physis shows a slower correction rate. Treatment should be considered at younger age to improve the overall appearance of the affected and non-affected physes.

Keywords

Physis • Physeal injury • Growth arrest • Physeal bar • Langenskiold procedure • Epiphysiodesis • Guided growth

Physeal injury

Introduction

The physis is the primary growth centre of the immature skeleton. It appears as a cartilaginous disc organised in five layers or zones (germinal, proliferative, columnar, hypertrophic, and provisional calcification) [1]. Discoid physes occur at the ends of long bones whilst spherical physes are the growth centres of the epiphyses and the carpal and tarsal bones. Apophyses have a similar structure but are usually subjected to tension forces rather than compression forces.

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Any injury to the growth plate, particularly if it involves the germinal cell layer, may produce irreversible damage resulting in growth disturbance [2].

Physeal injuries are most commonly caused by trauma and they may account for approximately 30 % of long bone fractures in children [3]. Less frequent causes include [2, 4]:

- Infection
- Physical agents such as laser, radiation, and heat
- Child abuse
- Tumor (benign or malignant)
- Disordered growth
  - Metabolic bone disease
  - Neuromuscular conditions

The most obvious and catastrophic consequence of a physeal injury is complete cessation of growth; this is, however, infrequent and its significance depends on patient age and the site of the injury. Complete growth arrest may result in significant limb length inequality with functional impairment in younger children and if it occurs in one of the paired bones (in the lower leg or forearm) there may also be significant deformity and restriction of joint movement. Partial growth arrest occurs when a central or peripheral physeal bone bridge links the epiphysis to the metaphysis tethering growth [4]. The size and location of the bar determine the type of deformity and dictate treatment [2].

How Are Physeal Injuries and Their Late Effects Recognised?

If the x-ray beam is tangential to the radiolucent physis, a displaced physeal fracture can be recognised indirectly on standard plain radiographs by observing the relative positions of the epiphysis to the metaphysis. Interpretation becomes difficult when the physis is oriented obliquely to the x-ray beam [5], in such cases comparison with the contralateral extremity may be helpful (LoE V) although it is essential to limit radiation exposure in the immature skeleton [6] (Fig. 45.1).

Although rarely required, Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) have proven to be reliable tools for recognizing physeal injuries and for identifying fracture fragments [7–11]. Carey et al. looked at the plain radiographs and MRI of 14 patients: 9 injuries were noted on plain radiographs and one occult injury was identified on MRI. Additional MRI information changed the Salter–Harris classification in two of the nine patients and management changed in 5/14 cases [11]. Similarly, Seifert et al. investigated

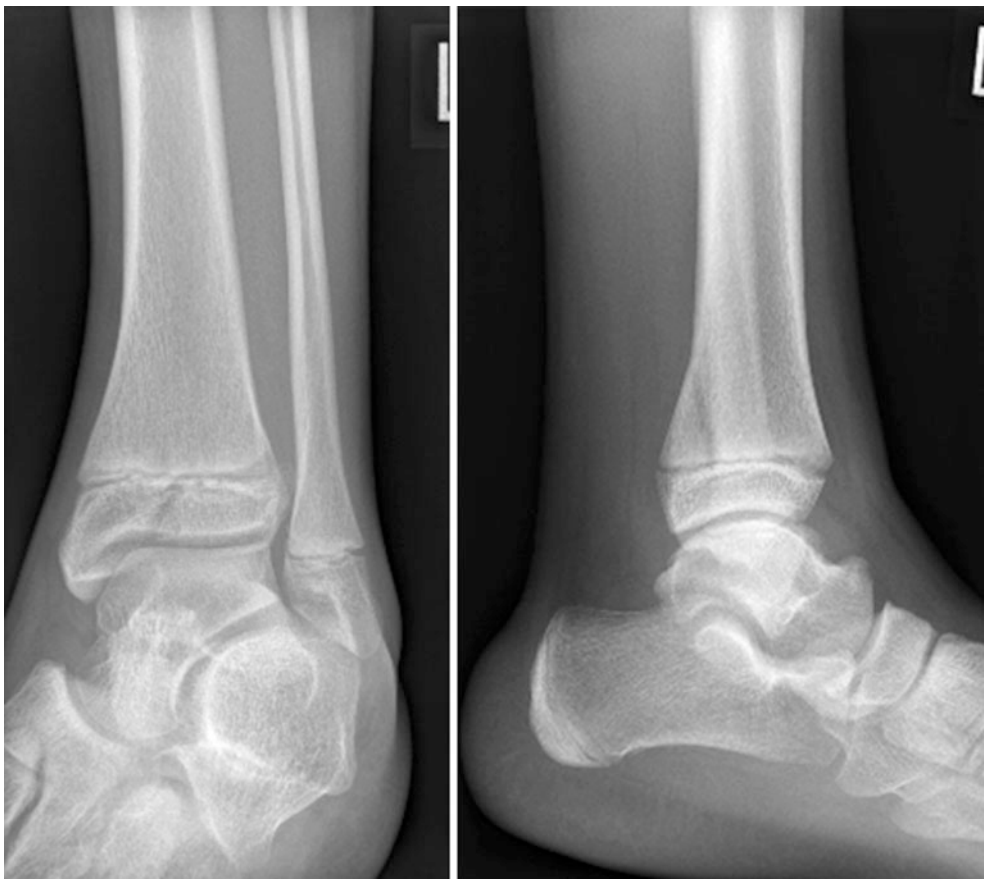

Fig. 45.1 Subtle distal tibial injury involving the physis and extending into the joint: notice the relative position of the epiphysis to the metaphysis on the lateral view

the role of MRI in the diagnosis of distal tibia fractures in 22 adolescents comparing standard radiographs and MRI images for each patient. They found that fracture type was misclassified and displacement underestimated on plain radiographs. MRI provided better anatomic details, moreover, it helped to diagnose a transitional fracture in two patients [11]. Hence, MRI was recommended for complex physeal injuries or when a growth plate injury was suspected (LoE III). [11, 12] MRI can also be useful for distinguishing between radiograph negative Salter Harris Type I fractures of the distal fibula and ligament sprains allowing management to be tailored appropriately [13] (Fig. 45.2).

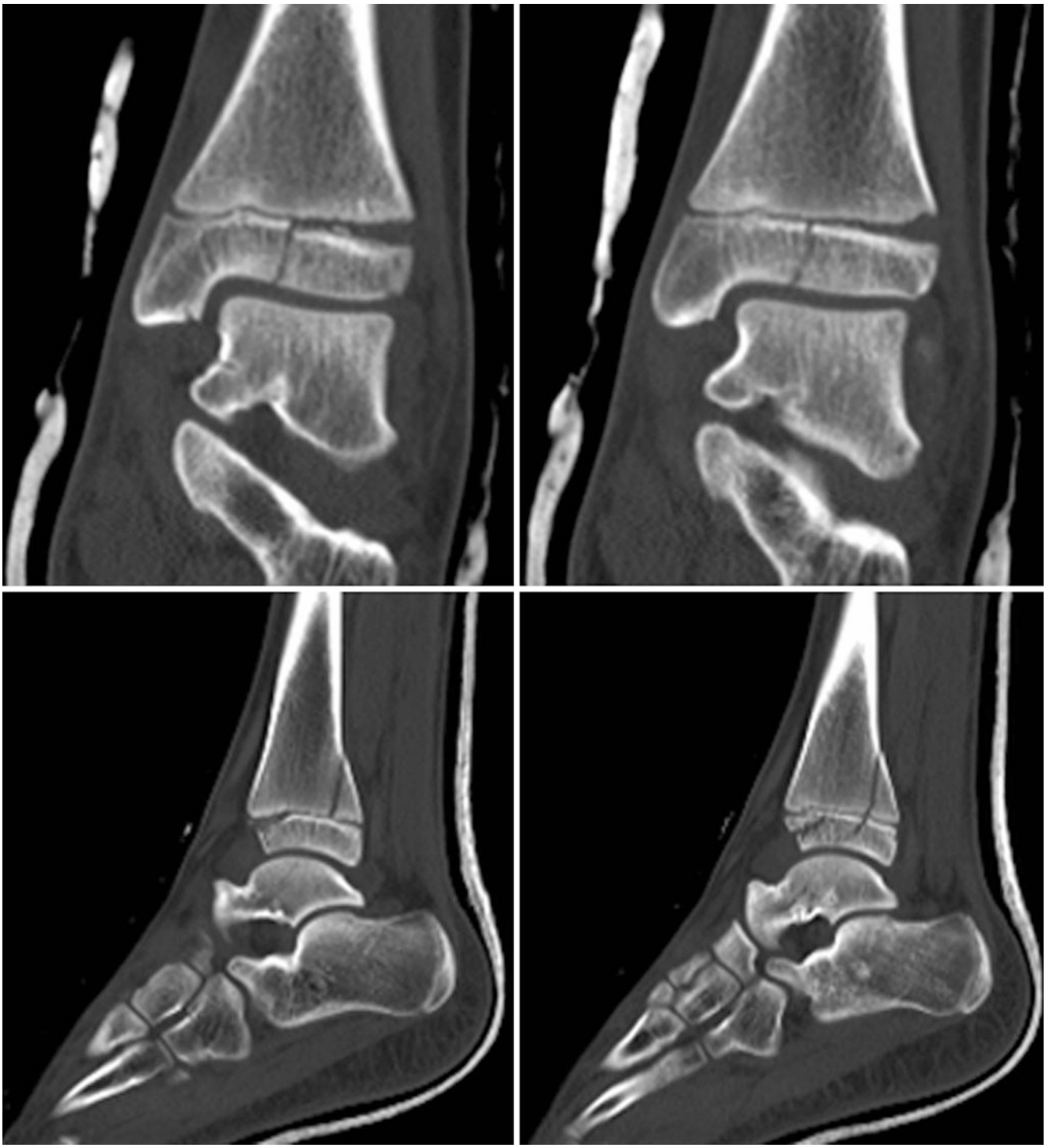

Fig. 45.2 CT scan of physeal ankle fracture. Four images of a CT scan of the same patient as in Fig. 45.1 which revealed that the fracture line on the lateral view extends into the epiphysis (i.e Salter Harris type IV rather than type II) which may affect the choice of the fixation method

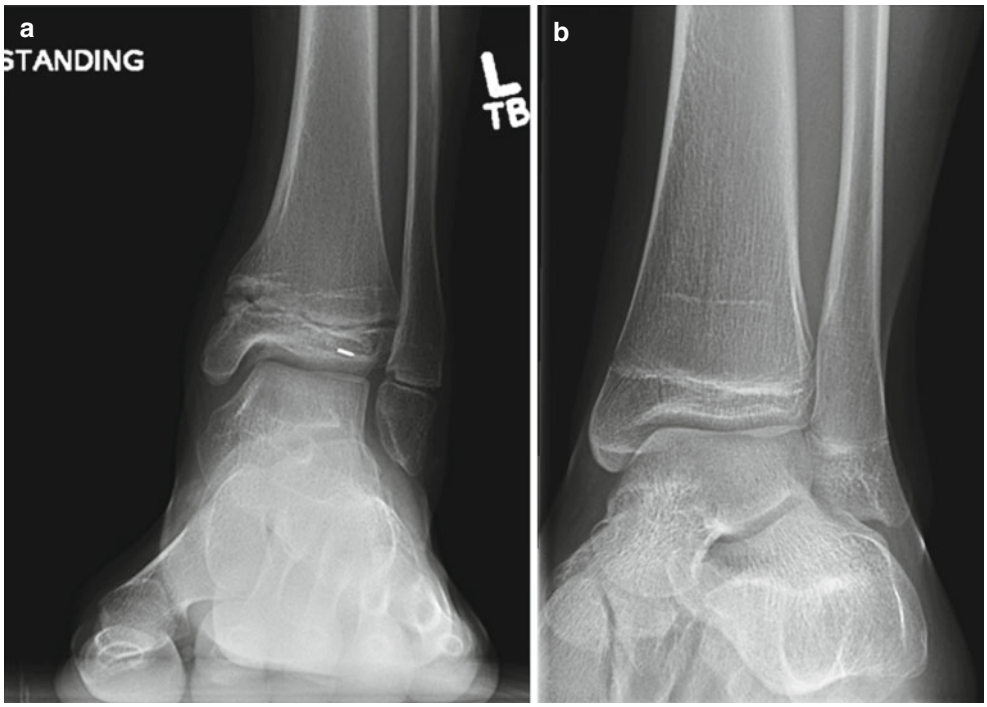

Fig. 45.3 Harris growth arrest lines; parallel (**b**) and angled (**a**): the angled line 'points' to the site of the arrest

Physeal fractures are followed-up with radiographs in the short term to ensure reduction has been maintained, and in the longer term to ensure that growth arrest has not occurred. There is no evidence that guides the length of follow-up. It is commonly believed (LoE V) that a Harris line [14] parallel to and some distance from the growth plate at 6–12 months post injury is a sign that normal growth has resumed.

An angled Harris line [14], a sclerotic bridge of bone, blurring and/or narrowing of the physis (especially in comparison to the non-injured and/or contralateral physis) all suggest permanent physeal damage (Fig. 45.3).

CT defines the extent of the physeal bridge and enables measurement of the area affected [7]. Loder reported that helical CT mapped the location and size of bony bars accurately in patients where bar excision was planned. The use of helical CT significantly reduced the radiation dose and scanning time (such that sedation was not required) and gave better differentiation between bone and physeal cartilage. (LoE IV) [15]. Despite this, there seems to be common agreement that MRI is the method of choice for evaluation of physeal bridges. It maps the bridge accurately in terms of size and distance from known landmarks while demonstrating the injured and uninjured areas of the growth cartilage [9, 16–20], and can be used to confirm complete bar resection and/or detect recurrence (LoE IV/V) [21]. 3D MRI reconstruction is useful to visualize very small physeal bars such as in the phalanges [20, 22]. Semi-automated segmentation techniques allow for reliable 3D modelling of the physeal bar [23]. As any movement reduces the quality of the scan, the main disadvantage of MRI is the time required and thus the need for sedation/general anaesthesia particularly in the younger child.

Can You Predict Which Physeal Injuries Will Have a Poor Outcome?

The most important factors influencing both treatment and prognosis of physeal fractures are: the age of the child, the fracture pattern and the specific physis affected.

Age of the Child

The child's age correlates to the amount of growth remaining and, hence, the potential growth disturbance if a complete or partial growth arrest occurs.

Fracture Pattern

Many classification systems have been proposed but the most widely used one is that of Salter and Harris [24] based on joint congruity and physeal involvement and alignment.

Treatment varies with fracture type and site, but in general, best practice guidelines are based on case series (LoE IV) [2, 24–27]. Care must be taken with manipulations, surgical approaches and the use of fixation devices to ensure that no additional, iatrogenic damage takes place.

In *Salter-Harris type I and II injuries* permanent growth disturbance is rare and healing is usually uneventful unless damage to the blood supply co-exists e.g. in intra-articular physeal injury (proximal femur, humerus and radius) (LoE IV) [24, 28]. Treatment varies with the specific site of

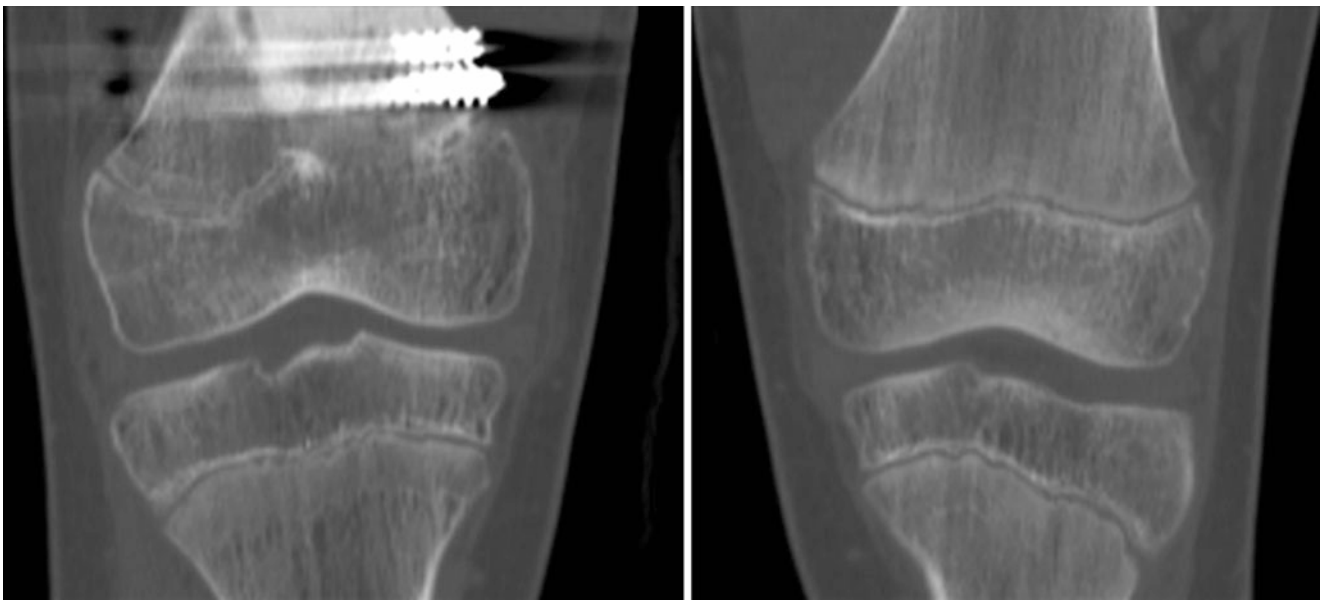

Fig. 45.4 Distal femoral physeal growth arrest affecting the medial portion of the physis

the fracture. In general, reduction should be gentle and anatomical: closed reduction and cast immobilisation is considered adequate treatment but, if anatomic closed reduction cannot be achieved then open reduction should be considered. If the reduction is unstable, fixation is required (LoE IV) [24, 26, 29].

In *Salter-Harris type III and IV injuries* both the physis and the articular cartilage are involved; both must be reduced anatomically and open reduction and fracture stabilisation is common [29].

In *Salter-Harris fracture types I-IV*, fixation, when necessary, is usually achieved with non-threaded wires or cannulated screws in the metaphysis or epiphysis parallel to the physis, or non-threaded wires across the growth plate supplemented by cast immobilisation [26, 29]. Fixation methods have not been compared. Experimental and clinical studies reported the use of biodegradable rods or screws across the growth plate with no evidence of growth arrest at 2 years (LoE IV) [30, 31]. However Bostman described a high incidence of discharging inflammatory foreign-body reaction and osteolytic foci in patients with displaced malleolar fractures treated by open reduction and internal fixation using absorbable polyglycolide rods [32]. Moreover, pin fracture did occur and the technique was not recommended for supracondylar fractures [33]. The technique has fallen out of favour over recent years.

Salter-Harris type V injuries are compression injuries to the growth plate. They usually lead to a complete growth arrest.

The Salter-Harris type VI injury was not included in the original Salter-Harris classification. It was first described by Rang in 1969 as a compression injury to the periosteum or perichondrial ring resulting in a bony bridge, and subsequent angular deformity [27].

Salter-Harris type V and VI injuries are often not diagnosed acutely; therefore no specific treatment recommendation is available.

Site of Growth Plate Injury

Growth arrest can affect any injured physis, however it is more likely to affect some physes than others. Growth disturbance has been reported after injuries to the triradiate cartilage [34, 35], the physes of the spine [36], the clavicle, [37] and those of the hands [38], and feet [39], but these are relatively rare events.

Distal Femur. Fractures of the distal femoral growth plate represent approximately 5 % of growth plate injuries and nearly 50 % of them are associated with growth disturbance (LoE IV) [25, 40]. Expert opinion is that closed reduction should be attempted for all mild to moderately displaced *Salter–Harris type I and II fractures*. Re-displacement is common in un-fixed type I fractures suggesting that fixation with one or two smooth heavy pins should be used to stabilize reduction (LoE IV) [41, 42]. Fixation methods have not been compared. In *Salter–Harris type II fractures* percutaneous screws across the metaphyseal fragment are often used but transphyseal fixation may be required for stability (LoE IV) [41, 42]. *Salter–Harris type III and IV fractures* can be fixed with intra-epiphyseal screws (LoE IV) [25, 40] (Fig. 45.4).

Proximal Tibia. Fractures of the proximal tibial growth plate are not common but have a high rate of serious complications, including neurovascular injury [43, 44]. Case series (LoE IV) show that the incidence of growth arrest does not correlate with the fracture pattern, therefore close follow-up should be considered in all cases [43, 44]. Closed reduction

and long leg cast immobilization is usually adequate treatment for minimally displaced fracture. All unstable and/or displaced *Salter-Harris type III and IV fractures* should be reduced and stabilized with internal fixation and cast (LoE IV). [25] Apophyseal growth arrest can lead to significant recurvatum.

Distal Tibia. Salter–Harris I and II fractures are usually treated with closed reduction and cast; acceptable displacement in children with at least 2 years of growth remaining consists of no more than 15° of plantar tilt, 10° of valgus, and no varus. In children with less than 2 years of growth remaining, acceptable angulation is 5° degrees in all planes. Premature physeal closure is unusual (LoE V) [45]. *Salter-Harris III and IV fractures* require surgical intervention (LoE IV) [25, 46]. Major deformity can be avoided with early treatment and close follow-up (LoE IV) [47]. Open or arthroscopic visualization of the joint surface may be required to confirm anatomic reduction. 3.5/4.0 mm cannulated screws stabilize the fracture but the screw placement must be assessed carefully in two planes (LoE IV) [25]. *Tillaux fractures* should be reduced and fixed in the presence of more than 2 mm displacement; best judged on CT (LoE IV/V) [25, 26]. Closed reduction, open reduction, and a percutaneous method have all been described with no strong evidence favouring any of these techniques (LoE IV) [25, 48]. *Triplane fractures* are assessed with standard radiographs and CT. Ertl et al. demonstrated that more than 2 mm intra-articular displacement led to poorer long-term results [49], hence open reduction and stable fixation are indicated to improve the outcome (LoE IV) [25, 49] (Fig. 45.5).

Distal Radius and Ulna. Salter Harris type I and II fractures of the distal radius are common. Seventy-five percent of forearm growth occurs from the distal physis and thus whilst growth arrest is rare (complicating only 1–7 % of injuries) if it affects only one bone, significant deformity can occur (LoE IV) [50, 51]. Waters et al. reported excellent clinical outcomes in a cohort of 30 patients with forearm deformity secondary to physeal arrest who underwent multiple procedures to achieve correction. All patients had good or excellent results, with decreased pain and increased activity level (LoE IV) [52] (Fig. 45.6).

How Do You Manage Growth Arrest?

The key is to anticipate the problem, diagnose it promptly and quantify it accurately. The most important factors influencing the treatment are: the amount of growth remaining at that growth plate, the location of the physis, the type of growth arrest whether complete or partial, and when partial the proportion of the growth plate injured (LoE IV/V) [2].

Complete Growth Arrest

Treatment aims to prevent and/or manage the length inequality. Options consist of no intervention, compensatory orthoses for lower limb discrepancies, epiphyseodesis or shortening of the contralateral or paired bone, ipsilateral bone lengthening, or a combination of these (LoE IV/ V) [2].

Partial Growth Arrest

Bright [4] classified partial growth arrest into three types: peripheral (type I), central (type II), and combined (type III). Partial arrests often result in some loss of length in addition to angular deformity and both problems need to be considered.

Angular deformity can be treated by acute (osteotomy) or gradual correction (distraction osteogenesis) in combination with a formal physeal closure and /or contralateral epiphyseodesis; or by physeal bar excision with interposition of an inert material. In the past, angular deformities were also treated by distraction of the growth plate and bar in children close to skeletal maturity (Hemichondrodiastasis) [2] (Fig. 45.7).

The Langenskiold Procedure for Resection of a Physeal Bar. In 1967, Langenskiöld reported the case of a 15-year-old boy with genu recurvatum secondary to a bone bar in the anterior proximal tibia whose etiology and duration were unknown. The bar was excised and the gap filled with autogenous buttock fat. During the 1.5 year follow-up, the recurvatum improved by 10° [53]. Over the years bone bar resection has become more popular and several interposition materials have been used; but evidence is based mainly on case series and a few comparative studies (LoE III and IV) [4, 53–62].

Physeal bar excision can be considered if less than 30–50 % of growth plate is involved (LoE IV/V) [2]. Younger children tend to have a better prognosis and less than 2 years of remaining growth is a relative contraindication for bone bridge resection (LoE IV) [63, 64]. It has also been noted that central bars are more amenable to resection than peripheral ones: probably related to the periosteal stripping required to resect peripheral lesions. Ischemic or septic-related bone bars have a poorer prognosis (LoE IV) [2]. The forces associated with normal growth can overcome a small physeal bar and although correction up to 30° has been reported, the degree of correction is variable and inconsistent (LoE IV) [4]. Angular deformities greater than 20° are unlikely to correct following bar resection and thus a corrective osteotomy is required. (LoE IV) [2, 62, 65]. Several interposition materials have been used: fat [53, 66, 67], muscle [67], polymeric silicone [68–70], bone wax [71], and bone cement, [63] and none has been shown to be superior. Current literature on the Langenskiold procedure – for the treatment of partial growth arrest is summarized in Table 45.1 [5, 51–58, 60, 69–71]. Recent experimental studies have explored the use of autogenous chondrocytes to fill the defect [72]. The main problem

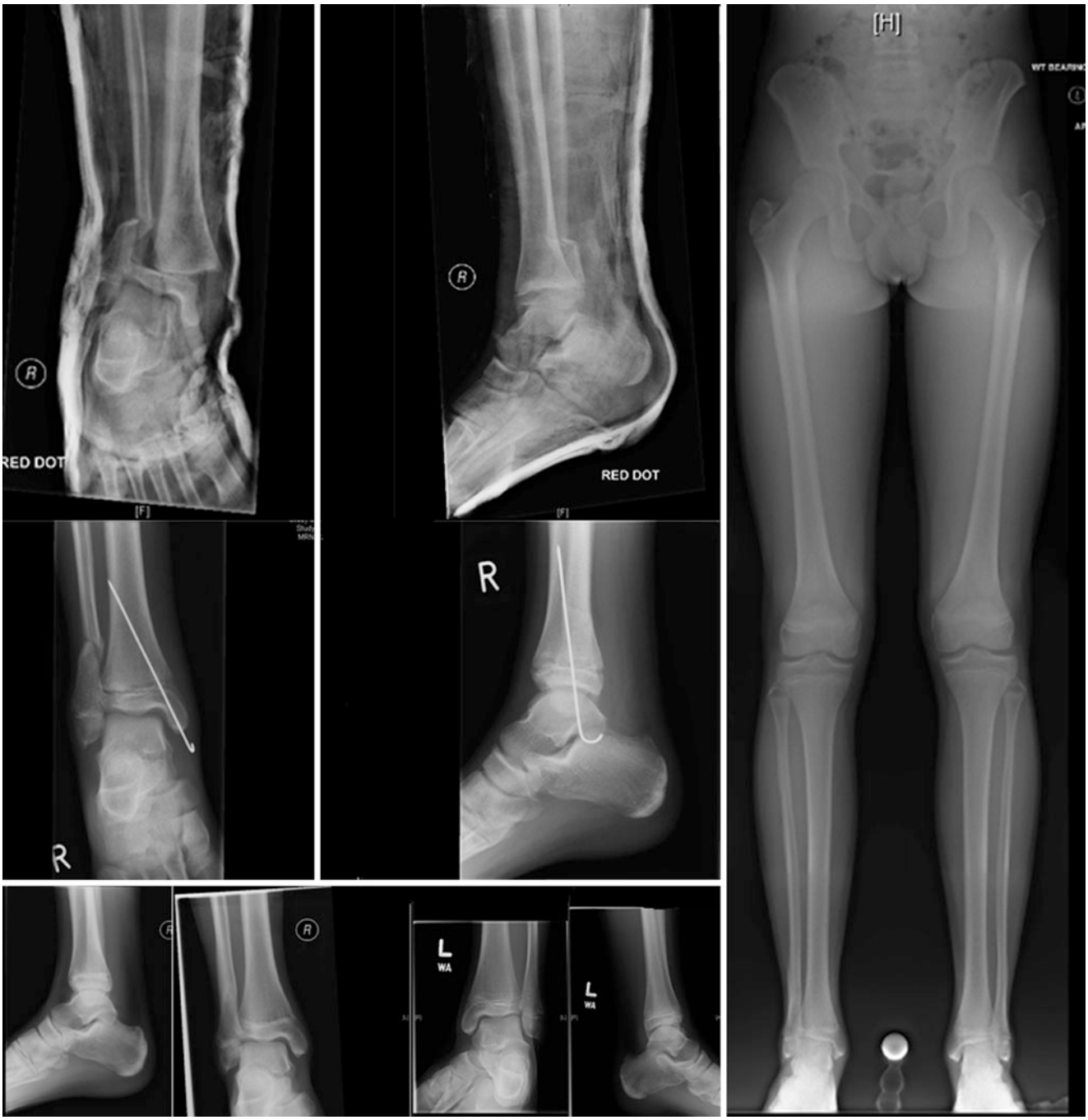

Fig. 45.5 Salter Harris Type II fracture of the distal tibial physis that caused a growth arrest note the slight asymmetry of the buttock creases and pelvic obliquity suggesting a short right leg

is to find a suitable donor site with cells retaining a certain growth potential. These experimental studies are still in the preclinical evaluation phase.

Hemichondrodiastasis Closed gradual distraction of the growth plate to correct angular deformity has been described (LoE IV) [73, 74]. Authors observed that the best results were achieved in posttraumatic deformities when the bone bridge occupied less than 20–30 % of the physeal plate. Moreover, the procedure was best performed toward the end of growth, or earlier if a progressive deformity exceeded 15–20^0 (LoE IV) [73].

Epiphysiodesis

Introduction

The purpose of epiphysiodesis is to produce either a permanent or temporary growth arrest. Its primary indication is a

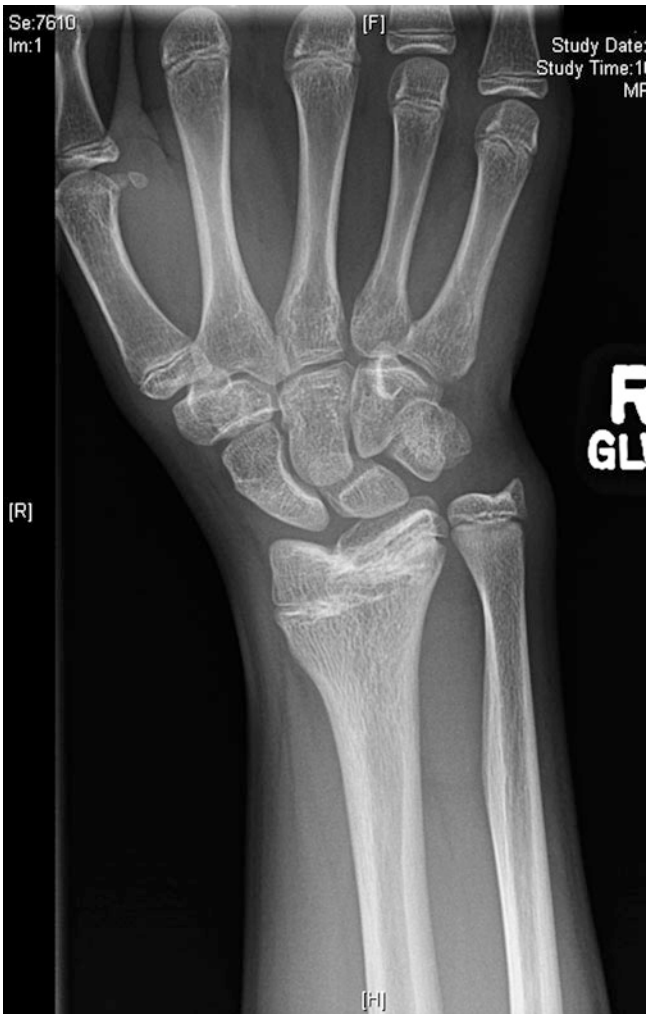

Fig. 45.6 Growth arrest of the distal radius leading to a 'long' ulna

mild to moderate, (2–5 cm), actual or predicted leg length discrepancy in a skeletally immature child with adequate growth remaining and an acceptable final height at skeletal maturity [75–79].

Epiphysiodesis must be timed accurately so that leg lengths becomes equal at skeletal maturity: the timing of surgery is the most challenging aspect of this procedure. [75–80]. Different methods have been proposed to determine the amount of growth remaining at any individual time point: each has advantages and disadvantages [75, 80–83].

Several surgical techniques for epiphysiodesis are available. Some destroy the physis permanently [84, 85] while others, implant-mediated, are reversible and produce a temporary growth arrest [86–91]. Current literature on epiphysiodesis consists mainly of retrospective reviews, case series and expert opinion with few comparative studies (LoE III, IV and V).

Does Epiphysiodesis Restrict Limb Growth and Correct Leg Length Difference?

Permanent Epiphysiodesis Phemister is credited with the first description of the technique of permanent epiphysiodesis [85]. In 1933, he described his "epiphyseodiaphyseal fusion", and reported the case of a girl with Ollier's disease and deformity with shortening of the left upper and lower limbs. Prior to treatment, at the age of 8.5 years, she had 6 cm leg length inequality. He excised the proximal physis of the left radius to correct the forearm deformity, and the proximal femoral physis of the unaffected lower limb to arrest longitudinal growth. At age 18, the residual discrepancy was 2.5 cm. He later reported good results on 20 additional patients treated by epiphysiodesis for leg length inequality (LoE IV) [85]. White and Stubbing modified the shape and size of the Phemister cancellous bone plug [84] and reported their epiphysiodesis results on 202 physes (149 patients, differing aetiologies) showing a growth retardation of 3/8 inch per year at the distal femur and ¼ inch per year at the proximal tibia and fibula. The complication rate was lower than the original Phemister technique (LoE IV) [84]. Stephens et al. reported their results of the White and Stubbins technique. Only 4 patients out of 56 had equal limb lengths at follow-up. Overcorrection and undercorrection were observed in 52 patients but only 4 of them required additional surgery (7 % of the group). Patients with residual discrepancy did not notice the difference and treatment was considered successful. Finally, authors noticed a relative overgrowth of 5 mm in the fibula in those cases where proximal tibia epiphysiodesis was not accompanied by a proximal fibula epiphysiodesis (LoE IV) [92].

Green and Anderson reported preliminary results of 77 procedures in 50 patients using several epiphysiodesis techniques, including their modified Phemister technique [93]. Later on they evaluated the results on 237 patients, 173 had permanent epiphysiodeses and 83 staplings [94]. With few exceptions, results were good or excellent in the 173 epiphysiodesis patients. Two had overcorrection greater than ½ inch, and 5 others underwent a contralateral epiphysiodesis to prevent overcorrection. Five patients developed angular deformity necessitating a corrective osteotomy in 4 and a repeat epiphysiodesis in 1. One patient developed osteomyelitis. Their overall rate of complications (including slow fusion) was 9.3 % (LoE III) [94].

Concerns about the cosmetic appearance of the scars and joint stiffness with the open technique, prompted interest in percutaneous modifications.

In 1984, Bowen and Johnson published the results of their percutaneous technique of epiphysiodesis which consisted of curetting the physis from either side and inserting an osteotome to a depth of 5 mm. They observed complete

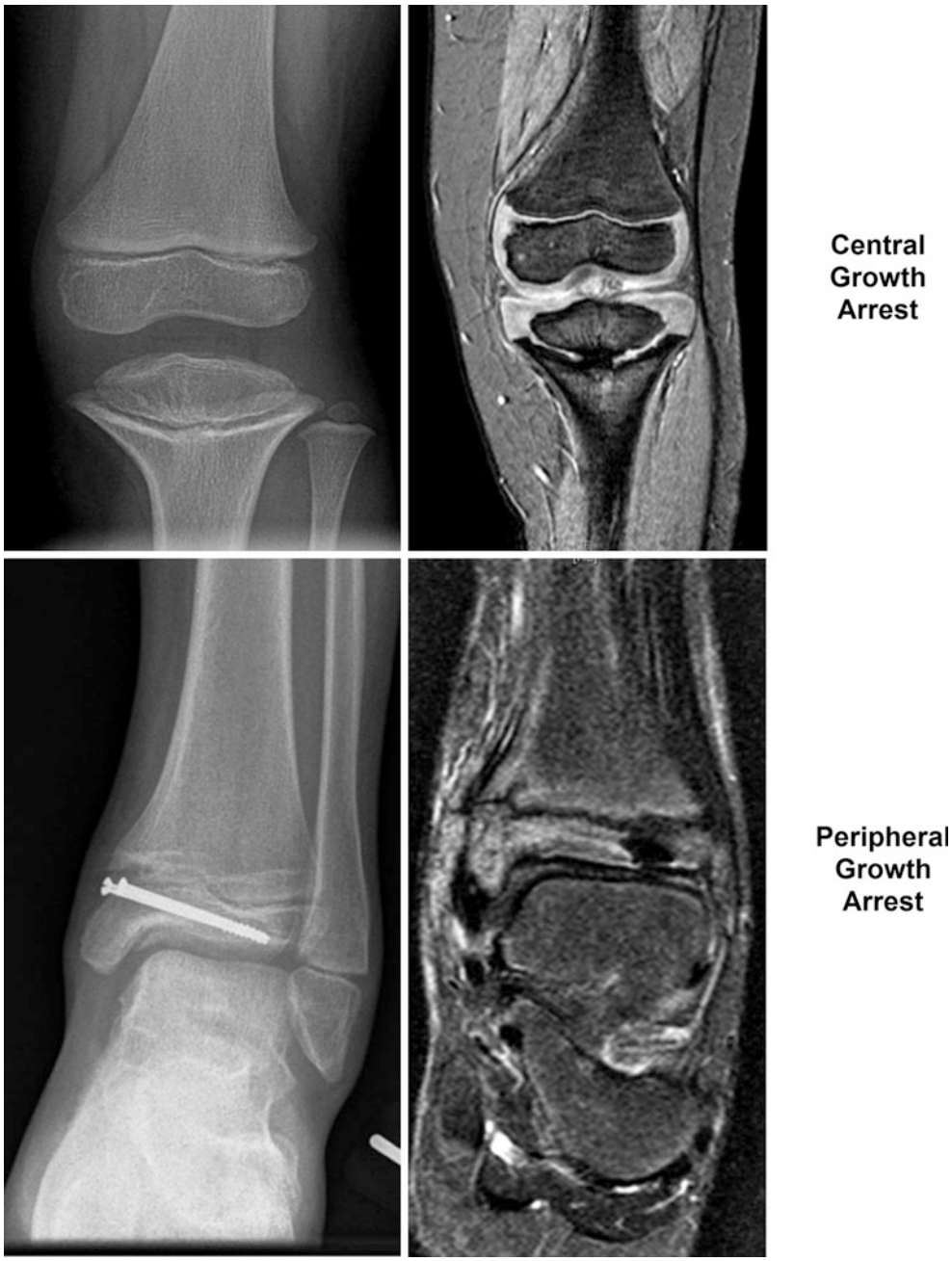

Fig. 45.7 Growth arrest: *top* is central in the proximal tibia and *bottom* is peripheral affecting the medial distal tibial physis

physeal arrest within 4 months in all 12 patients treated: the only complication was a keloid scar (LoE IV) [95]. In 1986, both Canale and Ogilvie, independently, reported that percutaneous techniques using a combination of drills and high-speed burrs effectively produced the desired epiphysiodesis effect in an animal model [96, 97]. These experimental findings have since been confirmed by many authors in clinical series using a variety of instruments (alone or in combination) such as cannulated tube saws or reamers, drills, burrs and curettes. Complete growth arrest was observed within 3–6 months from surgery in all series (LoE III and IV) [96, 98–107].

Temporary Epiphysiodesis Guiding growth to correct a deformity is perhaps one of the oldest concepts in orthopaedics. Implant-mediated guided growth is used for the correction of angular deformities (*Guided growth* pg 464) and the technique has recently been applied for the correction of leg length discrepancy.

Haas described the first method of temporary, reversible epiphysiodesis, observing the retardation of bone growth when using a wire loop around the physis in an animal model. He used the same technique in five patients: in two the wire loop broke and had to be replaced but he noted growth retardation while the wires were in place, and

Table 45.1 Current literature on physeal bar resection for the treatment of partial growth arrest

Year	Author	Interposition material	# Patients (Age)	# Bars	F-U (years)	Results	Positive prognostic factor	Conclusions (comments)	Level of evidence
1982	Bright [4]	Silastic	100 (9.4)	NK	NK	Resumed growth in 81 % of patients	–	Growth not always equivalent to normal side	IV
1983	Langenskiold and Osterman [54]	Fat	29	NK	NK	Resumed growth in most cases	–	–	IV
1986	Botte et al. [55]	Silastic	15 (10.2)	NK	3	Growth resumed in 80 % pts Angulation improved in 50 % pts	–	–	IV
1986	Coleman [56]	Fat	18 (NK)	NK	5	50 % complete correction	–	Common trend of premature closure	IV
1987	Talbert and Wilkins [57]	Silastic	29 (NK)	33	1	10 excellent 6 good 5 fair 8 poor	Young age Short timing injury/ surgery Small peripheral bars	–	IV
1988	Hume and Burstein [59]	Silastic	NK (9)	29	4.5	17 excellent 4 good 2 fair 6 failures	–	90 % excellent/good Corrective osteotomy should be in case of failure	IV
1989	Macksound and Bright [58]	Silastic	NK (10.8)	21	NK	78 % pts resumed growth ≥5 mm 8 pts ≥ 50 % of expected growth	–	Better results in younger patients	IV
1989	Vickers [60]	Fat	80 (NK)	NK	17	Growth re-established in 90 % Recurrence rare Early physeal closure but close to maturity	–	(Distal radius physis only)	IV
1990	Williamson and Staheli [61]	Fat	22 (NK)	NK	2	11 excellent 5 good 2 fair 4 poor	–	Mean growth 83 % of expected 96 % excellent/good	IV
1993	Mayo Clinic experience [63, 156, 157, 158]	153 cranioplast 23 fat 1 PMMA 1 Silastic + Gelfoam	178 (10.8)	NK	98 pt to maturity	Average resumed growth expressed as percentage of expected growth: 78 % distal femur 88 % Prox Tibia 93 % Distal Tibia	Distal physis Small bar Short timing injury/ surgery Bar area <45 %	50 % around the knee Result *not* correlated with gender or physeal location of the bar	III
1999	Dunn [159]	5 fat 5 cranioplast	NK (9.8)	9	Only 4 pts to maturity	All physes resumed growth	–	–	III

subsequent resumption of growth when wires broke (LoE IV) [86]. Subsequently, he used staples in an animal model and obtained similar effects on growth inhibition but growth resumption after staple removal was not normal and not universal [87]. Blount and Clarke published the first clinical results of the use staples in 13 patients, 7 treated for leg length inequality and 6 for angular deformity. They noted that inserting three staples on either side of the physis, stopped growth immediately and almost completely (LoE IV) [88]. Green and Anderson evaluated 83 stapling procedures in 61 patients at skeletal maturity and found stapling to be effective, although the distal femur grew an average of 6 mm after stapling (LoE III) [94]. Similar experience was reported by other authors (LoE IV) [89, 108–110]. Bylander observed a gradual cessation of growth across the stapled physis over 6 months; moreover the incidence of staple extrusion was lower if staples had been bent prior to insertion (LoE IV) [108, 109]. Blount reported that 426 operations were necessary in 185 patients, but only 2 patients required osteotomy for final correction (LoE IV) [89]. Sengupta et al. found that 71 % of 503 patients treated with stapling had less than 1 cm of discrepancy at skeletal maturity and concluded that stapling is a simple, effective procedure in developing countries (LoE IV) [110].

The main complications reported for epiphyseal stapling were slow arrest, asymmetric growth, overgrowth or reduced growth after staple removal, staple extrusion and a need for surgery to correct residual deformities [89, 94, 109, 111–113].

In 1998, Metaizeau described his technique of temporary percutaneous epiphysiodesis using transphyseal screws (PETS). He reported results on 32 patients with limb length inequality concluding that the screws began to cause detectable growth inhibition within 6 months of insertion, slowing down the distal femoral and proximal tibial physes by 68% and 56 %, respectively. Maximum growth retardation was achieved at 12 months, when the distal femoral physis was inhibited by 89 % and the proximal tibial physis by 95 %. At skeletal maturity, total femoral growth was 45 % and total tibial growth was 52 % of the normal side (LoE IV) [90].

Several reports have documented the success of PETS in the management of limb length difference (LLD). Nouth reported on nine patients, showing an average reduction in LLD from 3.33 to 1.36 cm over an average 2.2 year follow-up: 56 % reached an LLD of less than 1 cm (LoE IV) [114]. Khoury followed 30 patients to maturity: PETS was successful in all cases and the average final femoral and tibial lengths were 0.15 cm and 0.05 cm, respectively, from predicted length (LoE IV). Ilharreborde observed a mean efficacy of the femoral epiphysiodesis of 35 % at 6 months, and 66 % at maturity and of the tibial epiphysiodesis, 46 % at 6 months, and 66 % at maturity when looking at 45 patients. The revision rate was 18 % (8 patients): 7 of 8 revisions (87.5 %) involved the tibia and the authors advised caution using the technique at this site. (LoE IV) [115]. In a retrospective series of 59 patients by Song, PETS showed an average LLD correction of 75.5 % in the distal femur and 78.9 % at the proximal tibia: the authors recommended that PETS should be performed at least 1 year earlier than the estimated epiphysiodesis timing in order to achieve equal leg length (LoE IV) [116]. Monier reported similar results in 16 PETS patients and concluded that results would have been better if the procedure had been performed at an earlier skeletal age [117].

The most recent implant introduced on the marked for growth modulation is the 8-plate (LoE IV) [91, 118]. Early experience was on the correction of angular deformities and only recently has the implant been used to correct leg length discrepancy. Pendleton reported his results on 34 patients with either congenital or developmental LLD followed until screw removal or skeletal maturity. The LLD change in patients who had guided growth of the tibia was minimal, but in those patients who had femoral or combined femoral and tibial guided growth the change was an average 10 mm. Twenty of 33 patients had a LLD of less than 1 cm at maturity or screw removal. One patient required treatment for angular deformity after guided growth for LLD (LoE IV) [91]. Stewart reported 11 patients who had dual 8-plate for LLD and observed a mean 4 mm improvement after 18 months, compared toa mean 15.5 mm improvement in those patients who had physeal ablation (LoE) [119]. Gaumetou reported similar results with a mean efficacy for femoral epiphysiodesis of 23 % at 6 months and 68 % at 18 months. The procedure was even less effective on the proximal tibia, with only 42 % of the expected growth arrest at 18 months (LoE IV) [120].

Which Technique Is the Most Effective?

Current literature [94, 98, 102, 119, 121–125] is summarized in Table 45.2.

Based on the evidence available:

- Percutaneous epiphysiodesis produces a growth arrest within 6 months similar to an open epiphysiodesis (Phemister's technique) but shows a significantly lower complication rate. (LoE III) [98, 102, 121, 122].
- Percutaneous epiphysiodesis is significantly more effective and shows significantly less complications than staples, PETS and 8-plate (LoE III) [94, 119, 123, 125].
- Staples work significantly better than PETS to correct leg length discrepancy but show a significantly higher complication rate. Staples and 8-plates are equally effective but the latter show significantly less complications. 8-plates show significantly less peri-operative complications than staples and PETS but do not significantly reduce growth (LoE III) [120, 124, 126].

Table 45.2 Current literature on physeal bar resection for the treatment of partial growth arrest

Author	# Patients	Skeletal age	Prediction LLD @ maturity	Timing for epiphysiodesis	Age (mos)	Etiology	Pre-op LLD (cm)	F-U (mos)	Post-op LLD (cm)	Correction (cm/year)/growth arrest	Complications	Level of evidence
Phemister (Phem) vs Percutaneous Epiphysiodesis (P.E.)												
Liotta [102]	70: Phem: 44 P.E.: 26	–	Green & Anderson	Moseley	Phem: 12.9 P.E.: 13.2	–	–	–	–	100 % growth arrest @ 4–6 mos	Phem: 4.5 % P.E.: 4 %	III
Little [121]	110: Phem: 49 P.E.: 61	Gruelich & Pyle	Green & Anderson Moseley Menelaus	Green & Anderson Moseley Menelaus	Male: 13.5 Female: 11.6	Idiopathic, congenital, trauma, LCPD, DDH, CTEV, poliomyelitis, other	3.12 (1.4–7.4)	48	1.05 (−2.0–4.4)	No significant difference	14 further lenghtenings, 1 infection, 1stiffness, 1 scar pain (Phem)	III
Scott [98]	24: Phem: 12 physis P.E.: 20 physis	Gruelich & Pyle	Moseley	Moseley	Phem: 12.5 P.E.: 12.2	Idiopathic, congenital, trauma, infection, poliomyelitis, AVN, hemiplegia	Phem: 5.8 P.E.: 5.6 (Predicted LLD)	Phem: 2.2 P.E.: 2.4	Phem: 1.7 P.E.: 3	Phem: 12 % further growth P.E.: 15 % further growth	Phem: 1 stiffness P.E.: 2 wound infection	III
Surdam [123]	96: Phem: 40 P.E.: 56	–	–	–	12.9	Idiopathic, trauma, DDH, SUFE, CTEV, congenital, LCPD, infection, poliomyelitis	–	–	–	Phem: 100 % growth arrest P.E.: 3 pt delayed closure	Phem: 1 deep infection P.E.: 3 lenghtenings, 2 infections	III
Percutaneous Epiphysiodesis (P.E.) vs Staples												
Green [89]	256: P.E.: 173 Staples: 83	Gruelich & Pyle	Green & Anderson	Moseley	13 (10.5–14.5)	Idiopathic, Trauma, LCPD, Fibula hemimelia, CSF, Hemiparesis, Ollier's disease, Klippel–Trenaunay sdr, Hemihypertrophy, McCune–Albright sdr	–	>24	–	P.E.: 68 % femur, 45 % tibia @ 1 yr Staples:58 % femur, 38 % tibia @1 yr	P.E. 9.3 % (overcorrecion, angular deformity, reoperation) Staples: 25.3 % (extrusion, undercorrection, reoperation)	III
Phemister (Phem) vs Percutaneous Epiphysiodesis (P.E.) vs PETS												
Campens [124]	82: Phem:33 P.E.: 34 PETS: 15	Gruelich & Pyle	Green & Anderson	–	13.4 (9.3–16.3)	Idiopathic, congenital, trauma, LCPD, DDH, infection, CTEV, NF, hemiparesis, tumor, Ollier, syndromic, other	Phem:33 P.E.: 2.8 PETS: 3.0	Phem:38 P.E.: 3.6 PETS: 3.1	Phem:38 P.E.: 3.6 PETS: 3.1	LLD < 1.5 cm Phem:82 % P.E.: 89 % PETS: 70 %	Phem:6 % P.E.: 9 % PETS: 7 %	III

Percutaneous Epiphysiodesis (P.E.) vs PETS												
Babu [126]	40: P.E: 26 PETS: 14	TW3	P.E: Paley PETS: Moseley	P.E: Paley PETS: Moseley	Male: 13.3 Female: 11.8	DDH, PFFD, trauma, LCPD, SUFE, other	P.E: 3.7 PETS: 3.2	26 (12–72)	P.E: 26 PETS: 28	P.E: 1.2 PETS: 1.4	Complication half in P.E	III
Percutaneous Epiphysiodesis (P.E.) vs Guided Growth (8-plates)												
Stewart [119]	27: P.E.: 16 8-plate: 11	–	–	–	P.E.: 12.5 8-plate: 13	–	–	P.E.: 18 8-plate: 26	–	P.E.: 1.5 8-plates: 0.4 *P < 05*	P.E.: 18.75 % 8-plates: 0 % Further surgery: 1 correction of a fistula & 2 lengthenings	III
Staples vs PETS vs Guided Growth (8-plates)												
Lykissas [124]	39: Staples: 8 PETS: 22 8-plate: 9	Gruelich & Pyle	Green & Anderson	Moseley	13 (10.5–14.5)	Idiopathic, trauma, LCPD, fibula hemimelia, CSF, hemiparesis, Ollier's disease, Klippel–Trenaunay sdr, hemihypertrophy, McCune–Albright sdr	Staples: 3.65 PETS: 3.15 8-plates: 4.1	46 (24–88)	Staples: 1.95 PETS: 1.45 8-plates: 1.11	Staples: 1.22 PETS: 0.59* 8-plates: 1.11 *P < 05 Staples/PETS*	Staples: 50 % PETS: 36 % 8-plates: 44 % Further surgery: 1 correction angular deformity in PETS; 1 lengthening in 8-plate	III

*Refers to significance level

Does It Matter if the Limb Has a Pathological Growth Rate or Not?

The generic term "sick physes" is used to describe the radiographic appearance of structurally deficient or pathological physes in patients with generalized conditions affecting skeletal growth, including rickets, endocrinopathies, and skeletal dysplasias [127]. Correction of angular deformity in patients with pathological physes has showed a slower correction rate, however the evidence is limited [128–131]. Similarly, experts have hypothesized a slow correction for leg length discrepancy, however, to date, no literature has been published to substantiate this.

Guided Growth

Introduction

The first experiences on "guided growth" have been outlined in the previous section ("*Temporary Epiphysiodesis*" pg 459) [86–88]. This technique has been used for the treatment of leg length discrepancy (LLD) as well as the correction of angular deformity mostly at the knee. For decades, hemiepiphysiodesis with staples, as described by Blount and Clarke, has been the only technique available to correct deformities in the coronal plane [88, 132–139], but more recently new implants have been introduced to treat, primarily coronal plane, deformities and these are now superseding staples [90, 118, 140].

Does Guided Growth Work in Correcting Angular Deformity?

Hemiepiphysiodesis, employing multiple staples, has received mixed reviews since its introduction more than five decades ago. Blount and Clarke first reported their results on six patients with angular deformity of the knee. They observed full correction of the deformity in all patients (LoE IV) [88] and multiple further studies confirmed this (LoE IV) [132–139]. Hemiepiphyseal stapling was particularly effective in the treatment of knee deformities in patients ≤10 years of age. [132, 136] Metalwork fatigue, failure or migration, and under-correction with the need for further surgery were the main complications reported (LoE IV) [132–139]. Furthermore, a certain degree of rebound growth was observed after staple removal (LoE IV) [134] and other authors expressed concerns about the risk of permanent physeal closure if the implants were retained for too long (LoE IV) [141–143]. In many centres, hemiepiphyseal stapling has been abandoned.

Percutaneous hemiepiphysiodesis using transphyseal screws (PETS) became popular in Europe following Metaizeau et al's description of the technique. They reported deformity correction to within 3° of the mechanical axis in 9 patients with mild genu valgum (mean 7°, range 4–12°): only one patient required screw removal to prevent overcorrection (LoE IV) [90]. Subsequent studies have confirmed the validity of this technique in patients with greater deformity. Nouh and Kuo reported an average 12.5° correction by 2.6 years in 9 patients whose mean initial deformity was 18°. The only failure was a patient with hypophosphatemic rickets (LoE IV) [114]. Khoury et al. recorded differential corrective rates of 0.8° per month in 4 patients with Blount disease and 0.2° per month in those with idiopathic genu valgum, suggesting that both site and pathology influenced the outcome. Screws were removed in 13 skeletally immature patients, and 6 of them had recurrence of deformity ranging from 2–15° (LoE IV) [144]. Mesa et al. reported satisfactory results in 98 % of their patients with no major complications (LoE IV) [145].

A recent development is the use of the 8-plate (Orthofix, Verona, Italy) (or similar) device which acts as a "tension band" with an extra-physeal fulcrum rather than a compression device at/within the physis like staples and transphyseal screws. This, theoretically, reduces the risk of permanent physeal arrest [118] and may enhance the rate of deformity correction: it was designed for the correction of angular deformity but has been used (with less success) for the management of LLD (*"Temporary Epiphysiodesis"* pg 461) In his prospective series of 34 patients (65 physes), Stevens described neutralization of the mechanical axis with a parallel knee joint line in all but 2 patients. These 2 had recalcitrant Blount's disease and required screw readjustment. No premature physeal arrests were noted. Four patients, aged <11 years, with bilateral idiopathic genu valgum experienced recurrence of deformity after implant removal (LoE IV) [140]. Subsequent studies have confirmed these results (LoE IV). [130, 146–148]

Burghardt et al. in their series of 11 patients observed full correction of the joint orientation angles and the mechanical axis in all cases, with the exception of 1 boy who had a resected osteosarcoma and a compromised growth plate. No hardware failures, extrusion, growth arrest, or other complications including further treatment were observed (LoE IV) [146]. Ten of eleven patients were followed up for an average of 16 months (range, 10–24 months) after plate removal and an average rebound of the MAD of 15.7 mm or 1.0 mm/month, and of the joint orientation angles of 3.7 degrees or 0.23 degrees/month was noted [149]. Guzman et al. and Ballal et al. obtained comparable result and noted faster deformity correction younger patients (LoE IV) [147, 148].

Schroerlucke et al. showed similar results in patients with idiopathic genu varum and genu valgum but reported an overall metalwork failure rate of 26 % in cases where the primary diagnosis was Blount's disease. Excessive weight and a pathologic physis were presumed to place excess stress on the metalwork causing fatigue failure (LoE IV) [130].

These studies have two main weak points: all except one [135] are small retrospective case series and with a short follow-up limited to the period of correction only. Only 2 studies [148, 149] focused on the period after removal of the implant, which is critical to evaluate the rate of deformity recurrence after treatment with guided growth.

Which Technique Is the Most Effective?

Comparative studies available in the literature are summarized in Table 45.3 and Table 45.4 [131, 150–154]. Studies summarized are comparative retrospective reviews (LoE III), except one RCT (LoE I) [154] which has a small sample size and a short follow-up.

Sung et al. performed a decision analysis based on current evidence in the relevant literature to investigate the best treatment modality for coronal angular deformity in growing children. Authors compared temporary hemiepiphysiodesis using staples, percutaneous screws, or a tension band plate and used quality of life to unify the clinical outcomes with metal failure and incomplete correction of deformity. Their decision analysis model favored temporary hemiepiphysiodesis using a tension band plate which showed an overall complication rate lower than 15.7 % and a better QoL over temporary hemiepiphysiodesis using PETS or stapling (LoE II) [155].

Based on the current literature, the evidence is that:

- Hemiepiphysiodesis with a transphyseal screw is as safe and reliable as stapling to correct angular deformity of the knee in skeletally immature patients. Furthermore, this technique is less invasive, more cosmetic and does not produce permanent physeal arrest (LoE III) [146–148].
- The rate of correction of a deformity treated with staples and PETS is lower in older children, but the correction of the deformity at proximal tibia is faster with PETS (LoE III) [148].
- Despite a growing consensus that the 8-plate has superseded the staple, both techniques are equally effective even in younger patients. Complication rate seems higher in staples but it is not significantly different (LoE I-III) [149–151].
- Both techniques show higher complication rates in patients with pathologic physes (LoE III) [149].

Does It Matter if the Limb Has a Pathological Growth Rate or Not?

Historically, it has been considered that pathological physes, such as in rickets, Blount's disease or skeletal dysplasia would not tolerate direct surgical manipulation. The main concern was that 'sick' physes could shut down completely, resulting in additional, iatrogenic deformities. Arbitrarily, the timing of hemiepiphysiodesis to correct angular deformities, has often been deferred until after 8–10 years of age but this can undoubtedly lead to deformity progression, gait disturbance and possibly further impairment of growth not only of the knee but of the hip and ankle as well.

In recent years, there has been a paradigm shift. Stevens and Klatt reported their results on 14 children with rickets who underwent hemepiphysiodesis with staples (10 patients) and 8-plates (4 patients) for a total of 68 hemiepiphysiodeses and 35 osteotomies collectively. In the group of patients treated with staples they noted 45 % migration rate and rebound of the deformity in 41 % of cases. No metalwork failure or rebound of the deformity was seen in the patients treated with 8-plates but the follow-up was too short in this group. The authors observed that whilst correcting the mechanical axis, the overall appearance of the physes improved including the physes at the hip and ankle (LoE IV) [129]. Boero et al. performed guided growth with 8-plates in 58 patients with pathologic and idiopathic knee angular deformities. They noted full correction of all idiopathic deformities but in the group with pathologic physes they only observed complete correction in 78.5 %. This latter group was treated at a younger age. "Sick" physes showed significantly slower correction rates reaching neutral mechanical axis in 18 months with a correction rate of 0.3°/month. Idiopathic deformities demonstrated neutralization of the mechanical axis in 11 months with a correction rate of 0.6°/month. Moreover, rebound of the deformity was observed in 3.8 % of patients with pathologic physes and in none of the patients with idiopathic deformity (LoE IV) [128]. Additional medical management must be optimized when appropriate.

Weimann et al. observed a similar pattern in their small group of patients with "sick" physes and concluded that children with skeletal dysplasia and abnormal physes were significantly more likely to require osteotomy to correct residual angular deformity after hemiepiphysiodesis (LoE IV) [131]. Schroerlucke observed higher failure rates in patients with Blount's disease perhaps related to their obesity (LoE IV). Several studies have shown no difference in the time taken for deformity correction in patients with skeletal dysplasia and Blount's disease who had hemiepiphysiodesis using PETS (LoE IV) [114, 144, 151].

Table 45.3 Current literature comparing stapling vs PETS in the treatment of angular deformities of the knee

Author	# Patients (physes)	Chr. age (Skel. age)	Type of deformity	Etiology	F-U (mos)	Pre-op data	Post-op data	Complications	Conclusions (authors)	Level of evidence
De Brauwer [150] Degreef [134]	Staples: 44 M: 26 F: 18 PETS: 25 M: 9 F: 16	Staples M: 13.2 (13.4) F:12.6 (12.7) PETS M: 14.5 (14.3) F:12.7 (12.8)	Genu Valgum	Idiopathic	26	Staples: IMD: 11 cm HKA: 5° valgus PETS: IMD: 10.5 cm HKA: 4.6° valgus	Staples: IMD: 2 cm HKA (Mean correction): 9°@ removal, 5°@ maturity PETS: IMD: 4.8 cm HKA (Mean correction): 4.35°@removal, 3.85° @ maturity	Staples: avg rebound 4°, (6 pts) sagittal plane deformity, (1 pts) overcorrection PETS: avg rebound 2°, (3 pts) overcorrection, (4 pts) anterior knee pain	PETS is as safe and reliable as stapling but less invasive and more cosmetic treatment	III
Shin [151]	Staples: 19 (43) PETS: 23 (27)	Staples M: 12.1 F: 10.4 PETS M: 12.9 F:11.8	NK	Staples: MED, MPS-IV, metabolic, idiopathic, congenital, trauma PETS: MHE, MED, MCD, SED, SMD, Blount, Infection, Trauma	Staples: 46 PETS: 30 Up to Skel. Maturity	–	Staples: F: Mean correction 3°; Rate of correction 0.68°/mos T: Mean correction 7.8° Rate of correction 0.55°/mos Hospital stay: 3.6 day PETS: F: Mean correction 8.3° Rate of correction 0.92°/mos T: Mean correction 7° Rate of correction 0.52°/mos Hospital stay: 1.6 day	Staples: rebound (6 physes), undercorrection (1 pt, reached skel. maturity), physeal arrest (1 pt), staple extrusion (1 pt) PETS: rebound (6 physes), undercorrection (1 pt, reached skel. maturity)	PETS is as effective as staples, it is minimally invasive and in this series, did not cause permanent physeal arrest	III

Sung [155]	78 (175) M: 107 F: 68 Staple: 44 PETS: 119	15 (10.2)	Genu Valgum	MED, MPS-IV, Genetic, Metabolic Idiopathic, congenital trauma, post-septic	14.6 (3–51)	–	Results not stratified by surgical technique Mean correction: aLDFA: 7.3° aLPTA: 5.2° Rate of correction: Younger children (M < 14, F < 12) F: 0.71°/mos T: 0.4°/mos Older children (M > 14, F > 12) F: 0.39°/mos T:0.29°/mos Multivariate analysis Rate of correction: Staples: F: 0.64°/mos T: 0.26°/mos PETS: F: 0.64°/mos T: 0.42°/mos	–	The rate of correction at the distal femur is lower in older children, and that at the proximal tibia is faster with PETS	III

Table 45.4 Current literature comparing stapling vs 8-plates in the treatment of angular deformities of the knee

Author	# Patients (physes)	Chr. age (Skel. age)	Type of deformity	Etiology	F-U (mos)	Pre-op data	Post-op data	Complications	Conclusions (authors)	Level of evidence
Weimann [149]	63 Staples: 39 M: 24 F: 15 8-plate: 24 M: 11 F: 13	Staples: 12.6 8-plate: 11.1	Genu Valgum Genu varum	Idiopathic, Blounts, gentic, congenital, trauma	Staples: 16.2 (5–34) 8-plate: 14.1 (6–27)	–	Staples: Rate of correction: 9.9°/year 8-plate: Rate of correction: 11.1°/year	Staples: further surgery for deformity correction (5 pts, 12.8 %) 8-plates: further surgery for deformity correction (3 pts, 12.5 %), screw fatigue failure (Blunt's disease)	8-plate is as effective as staple even in younger patients Higher complication rates are observed in patients with pathologic physes	III
Jelinek [153]	Staples: 18 M: 11 F: 7 8-plate: 16 M: 8 F: 9	Staples: 13 8-plate: 11.6	Genu Valgum Genu varum	Staples: MHE, metabolic, idiopathic, congenital, trauma, Ehler-Danlos 8-plate: MHE, idiopathic, achondroplasia, metabolic, infection, trauma	Staples: 11 8-plate: 11.9	Staple: MAD Valgus: 3 cm, Varus: 1.7 cm mLDFA Valgus: 83°, Varus: 93° mMPTA Valgus: 91°, Varus: 83° 8-plate: MAD Valgus: 2.9 cm, Varus: 3.8 cm mLDFA Valgus: 84°, Varus: 99° mMPTA Valgus: 93°, Varus: 83°	Staple: MAD Valgus: 0.7 cm , Varus: 0.4 cm mLDFA Valgus: 89°, Varus: 86° mMPTA Valgus: 87°, Varus: 85° 8-plate: MAD Valgus: 0.5, Varus: 0.2 mLDFA Valgus: 89°, Varus: 90° mMPTA Valgus: 88°, Varus: 91°	Staples: wound problem (1 pt) requiring surgery, metalwork failure (2 pt) 8-plate: reduced knee flexion (1 pt), overcorrection (2 pts) requiring surgery	Both techniques are effective methods for correction of angular deformity of the knee. However, a shorter operating time was noted for the 8-plate	III

Gottliebsen [154]	20 Staple: 10 M: 3 F: 7 8-plates: 10 M: 8 F: 2	Staples: 11.1 (6–13) 8-plate: 10.1 (8–14)	Genu Valgum	MED, MPS-IV, genetic, metabolic idiopathic, congenital trauma, post-septic	14.6 (3–51)	Staples IMD: 92 (82–102) mLDFA: 85.7 (84–87) MPTA: 90 (89–91) MAD: −12.2 (−20 to −5) 8-plate IMD: 95 (78–111) mLDFA: 85.5 (84–87) MPTA: 89.8 (88–92) MAD: −13 (−18 to −8)	Staples IMD: −3 (−8 to 2) mLDFA: 94 (92–96) MPTA: 92 (91–93) MAD: 10 (1–18) 8-plate IMD: −2 (−6 to 3) mLDFA: 92 (90–95) MPTA: 90 (88–96) MAD: 10 (3–15) Similar VAS score	8-plate: rebound in 1 pt 15 mos after plate removal	Treatment time was not significantly different. 8-plate and stapling seemed to have a similar effect regarding correction of genu valgum	I (RCT)

In conclusion, based on the current literature (LoE IV):

- In the presence of a pathological physis, 8-plate correction of angular deformity shows a slower correction rate. Therefore a longer time is required to achieve full correction. Treatment should be commenced at a younger age to improve the overall appearance of the affected and non-affected physes (LoE IV) [128, 129].
- Correction of angular deformity with PETS in patients with skeletal dysplasia and Blount's shows similar correction rate to patients with idiopathic deformities (LoE IV) [114, 144, 151].
- Obese patients with Blount's disease show a higher rate of 8-plate failure rate compared to patients with idiopathic deformities (LoE IV) [130].

References

1. Brighton CT. Structure and function of the growth plate. Clin Orthop Relat Res. 1978;136:22–32.
2. Peterson HA. Physeal injuries and growth arrest. In: Beaty JH, Kasser JR, editors. Rockwood and Wilkins' fractures in children. 5th ed. Philadelphia: Lippincott Williams & Wilkins; 2001. p. 91–138.
3. Mann DC, Rajmaira S. Distribution of physeal and nonphyseal fractures in 2,650 long-bone fractures in children aged 0–16 years. J Pediatr Orthop. 1990;10(6):713–6.
4. Bright RW. Partial growth arrest: identification, classification and results of treatment. Orthop Trans. [Abstract]. 1982;6:65–6.
5. Ogden JA, Ganey T, Light TR, Southwick WO. The pathology of acute chondro-osseous injury in the child. Yale J Biol Med. 1993;66(3):219–33.
6. Gautier E, Ziran BH, Egger B, Slongo T, Jakob RP. Growth disturbances after injuries of the proximal tibial epiphysis. Arch Orthop Trauma Surg. 1998;118(1–2):37–41.
7. Young JW, Bright RW, Whitley NO. Computed tomography in the evaluation of partial growth plate arrest in children. Skeletal Radiol. 1986;15(7):530–5.
8. White PG, Mah JY, Friedman L. Magnetic resonance imaging in acute physeal injuries. Skeletal Radiol. 1994;23(8):627–31.
9. Smith BG, Rand F, Jaramillo D, Shapiro F. Early MR imaging of lower-extremity physeal fracture-separations: a preliminary report. J Pediatr Orthop. 1994;14(4):526–33.
10. Petit P, Panuel M, Faure F, Jouve JL, Bourliere-Najean B, Bollini G, et al. Acute fracture of the distal tibial physis: role of gradient-echo MR imaging versus plain film examination. AJR Am J Roentgenol. 1996;166(5):1203–6.
11. Seifert J, Matthes G, Hinz P, Paris S, Mutze S, Ekkernkamp A, et al. Role of magnetic resonance imaging in the diagnosis of distal tibia fractures in adolescents. J Pediatr Orthop. 2003;23(6):727–32.
12. Carey J, Spence L, Blickman H, Eustace S. MRI of pediatric growth plate injury: correlation with plain film radiographs and clinical outcome. Skeletal Radiol. 1998;27(5):250–5.
13. Boutis K, Plint A, Stimec J, Miller E, Babyn P, Schuh S, et al. Radiograph-negative lateral ankle injuries in children: occult growth plate fracture or sprain? JAMA Pediatr. 2016;170(1):e154114.
14. Harris HA. Lines of arrested growth in the long bones in childhood: the correlation of histological and radiographic appearances in clinical and experimental conditions. Br J Radiol. 1931;4(47):561–88.
15. Loder RT, Swinford AE, Kuhns LR. The use of helical computed tomographic scan to assess bony physeal bridges. J Pediatr Orthop. 1997;17(3):356–9.
16. Havranek P, Lizler J. Magnetic resonance imaging in the evaluation of partial growth arrest after physeal injuries in children. J Bone Joint Surg Am. 1991;73(8):1234–41.
17. Ecklund K, Jaramillo D. Patterns of premature physeal arrest: MR imaging of 111 children. AJR Am J Roentgenol. 2002;178(4):967–72.
18. Ecklund K, Jaramillo D. Imaging of growth disturbance in children. Radiol Clin North Am. 2001;39(4):823–41.
19. Gabel GT, Peterson HA, Berquist TH. Premature partial physeal arrest. Diagnosis by magnetic resonance imaging in two cases. Clin Orthop Relat Res. 1991;272:242–7.
20. Lohman M, Kivisaari A, Vehmas T, Kallio P, Puntila J, Kivisaari L. MRI in the assessment of growth arrest. Pediatr Radiol. 2002;32(1):41–5.
21. Hasler CC, Foster BK. Secondary tethers after physeal bar resection: a common source of failure? Clin Orthop Relat Res. 2002;405:242–9.
22. Craig JG, Cramer KE, Cody DD, Hearshen DO, Ceulemans RY, van Holsbeeck MT, et al. Premature partial closure and other deformities of the growth plate: MR imaging and three-dimensional modeling. Radiology. 1999;210(3):835–43.
23. Lurie B, Koff MF, Shah P, Feldmann EJ, Amacker N, Downey-Zayas T, et al. Three-dimensional magnetic resonance imaging of physeal injury: reliability and clinical utility. J Pediatr Orthop. 2014;34(3):239–45.
24. Salter RB, Harris WR. Injuries involving the epiphyseal plate. J Bone Joint Surg. 1963;45(3):587–622.
25. Flynn JM, Skaggs DL, Sponseller PD, Ganley TJ, Kay RM, Leitch KK. The surgical management of pediatric fractures of the lower extremity. Instr Course Lect. 2003;52:647–59.
26. Canale ST. Physeal injuries. In: Green NE, Swiontkowski MF, editors. Skeletal trauma in children. 2nd ed. Philadelphia: WB Saunders Company; 1997. p. 17–58.
27. Rang M. The growth plate and its disorders. In: Rang M, editor. The growth plate and its disorders. Edinburgh/London: E&S Livingstone; 1969.
28. Ogden JA. Injury to the growth mechanisms of the immature skeleton. Skeletal Radiol. 1981;6(4):237–53.
29. Bright RW. Physeal injuries. Fractures: fractures in children. 3rd ed. Philadelphia: Lippincott-Raven; 1991. p. 87–186.
30. Hara Y, Tagawa M, Ejima H, Orima H, Fujita M, Yamagami T, et al. Application of oriented poly-L-lactide screws for experimental Salter-Harris type 4 fracture in distal femoral condyle of the dog. J Vet Med Sci. 1994;56(5):817–22.
31. Makela EA, Bostman O, Kekomaki M, Sodergard J, Vainio J, Tormala P, et al. Biodegradable fixation of distal humeral physeal fractures. Clin Orthop Relat Res. 1992;283:237–43.
32. Bostman OM. Osteolytic changes accompanying degradation of absorbable fracture fixation implants. J Bone Joint Surg Br. 1991;73(4):679–82.
33. Bostman O, Makela EA, Sodergard J, Hirvensalo E, Tormala P, Rokkanen P. Absorbable polyglycolide pins in internal fixation of fractures in children. J Pediatr Orthop. 1993;13(2):242–5.
34. Bucholz RW, Ezaki M, Ogden JA. Injury to the acetabular triradiate physeal cartilage. J Bone Joint Surg Am. 1982;64(4):600–9.
35. Heeg M, Visser JD, Oostvogel HJ. Injuries of the acetabular triradiate cartilage and sacroiliac joint. J Bone Joint Surg Br. 1988;70(1):34–7.
36. Lawson JP, Ogden JA, Bucholz RW, Hughes SA. Physeal injuries of the cervical spine. J Pediatr Orthop. 1987;7(4):428–35.
37. Havranek P. Injuries of distal clavicular physis in children. J Pediatr Orthop. 1989;9(2):213–5.

38. Letts M, Esser D. Fractures of the triquetrum in children. J Pediatr Orthop. 1993;13(2):228–31.
39. Noonan KJ, Saltzman CL, Dietz FR. Open physeal fractures of the distal phalanx of the great toe. A case report. J Bone Joint Surg Am. 1994;76(1):122–5.
40. Riseborough EJ, Barrett IR, Shapiro F. Growth disturbances following distal femoral physeal fracture-separations. J Bone Joint Surg Am. 1983;65(7):885–93.
41. Graham JM, Gross RH. Distal femoral physeal problem fractures. Clin Orthop Relat Res. 1990;255:51–3.
42. Thomson JD, Stricker SJ, Williams MM. Fractures of the distal femoral epiphyseal plate. J Pediatr Orthop. 1995;15(4):474–8.
43. Burkhart SS, Peterson HA. Fractures of the proximal tibial epiphysis. J Bone Joint Surg Am. 1979;61(7):996–1002.
44. Shelton WR, Canale ST. Fractures of the tibia through the proximal tibial epiphyseal cartilage. J Bone Joint Surg Am. 1979; 61(2):167–73.
45. Cummings RJ. Distal tibial and fi bular fractures. In: Rockwood Jr CA, Wilkins KE, Beaty JH, editors. Fractures in children. 4th ed. Philadelphia: Lippincott-Raven; 1996. p. 1377–428.
46. de Sanctis N, Della Corte S, Pempinello C. Distal tibial and fibular epiphyseal fractures in children: prognostic criteria and long-term results in 158 patients. J Pediatr Orthop B. 2000;9(1):40–4.
47. Lalonde KA, Letts M. Traumatic growth arrest of the distal tibia: a clinical and radiographic review. Can J Surg. 2005;48(2): 143–7.
48. Schlesinger I, Wedge JH. Percutaneous reduction and fixation of displaced juvenile Tillaux fractures: a new surgical technique. J Pediatr Orthop. 1993;13(3):389–91.
49. Ertl JP, Barrack RL, Alexander AH, VanBuecken K. Triplane fracture of the distal tibial epiphysis. Long-term follow-up. J Bone Joint Surg Am. 1988;70(7):967–76.
50. Buterbaugh GA, Palmer AK. Fractures and dislocations of the distal radioulnar joint. Hand Clin. 1988;4(3):361–75.
51. Lee BS, Esterhai Jr JL, Das M. Fracture of the distal radial epiphysis. Characteristics and surgical treatment of premature, posttraumatic epiphyseal closure. Clin Orthop Relat Res. 1984;185: 90–6.
52. Waters PM, Bae DS, Montgomery KD. Surgical management of posttraumatic distal radial growth arrest in adolescents. J Pediatr Orthop. 2002;22(6):717–24.
53. Langenskiöld A. The possibilities of eliminating premature partial closure of an epiphyseal plate caused by trauma or disease. Acta Orthop. 1967;38(1–4):267–79.
54. Langenskiold A, Osterman K. Surgical elimination of posttraumatic partial fusion of the growth plate. In: Houghton G, Thompson G, editors. Problematic musculoskeletal injuries in children. London: Butterworth's Internat'l Med Reviews Ortho I; 1983. p. 14–31.
55. Botte MJ, Sutherland DH, Mubarak SJ. Treatment of partial epiphyseal arrest by resection of the osseous bridge and interposition with silicone rubber. Orthop Trans. 1986;10:457.
56. Coleman SS. Physolysis: resection of premature incomplete fusion of the physis. Orthop Trans. 1986;10:554–5.
57. Talbert RE, Wilkins KE. Physeal bar resection: factors contributing to success. Orthop Trans. 1987;11:549.
58. Macksound WS, Bright R. Bar resection and silastic interposition in distal radial physeal arrest. Orthop Trans. 1989;12:1–2.
59. Hume MC, Burstein SM. Silastic implantation after epiphyseal bar resection. Orthop Trans. 1988;12:151.
60. Vickers D. Epiphysiolysis. Curr Orthop. 1989;3:41–447.
61. Williamson RV, Staheli L. Partial epiphyseal growth arrest. Treatment by bridge resection and fat interposition. Orthop Trans. 1990;11:443–4.
62. Langenskiold A. Surgical treatment of partial closure of the growth plate. J Pediatr Orthop. 1981;1(1):3–11.
63. Klassen RA, Peterson HA, Peterson LFA. Excision of physeal bars: the Mayo Clinic experience 1968–1978. Orthop Trans. 1982;6:65.
64. Kasser JR. Physeal bar resections after growth arrest about the knee. Clin Orthop Relat Res. 1990;255:68–74.
65. Ogden JA. The evaluation and treatment of partial physeal arrest. J Bone Joint Surg Am. 1987;69(8):1297–302.
66. Osterman K. Operative elimination of partial premature epiphyseal closure. An experimental study. Acta Orthop Scand Suppl. 1972:3–79.
67. Martiana K, Low CK, Tan SK, Pang MW. Comparison of various interpositional materials in the prevention of transphyseal bone bridge formation. Clin Orthop Relat Res. 1996;325:218–24.
68. Campbell CJ, Grisolia A, Zanconato G. The effects produced in the cartilaginous epiphyseal plate of immature dogs by experimental surgical traumata. J Bone Joint Surg Am. 1959;41-A:1221–42.
69. Bright RW. Operative correction of partial epiphyseal plate closure by osseous-bridge resection and silicone-rubber implant. An experimental study in dogs. J Bone Joint Surg Am. 1974;56(4): 655–64.
70. Lee EH, Gao GX, Bose K. Management of partial growth arrest: physis, fat, or silastic? J Pediatr Orthop. 1993;13(3):368–72.
71. Broughton NS, Dickens DR, Cole WG, Menelaus MB. Epiphyseolysis for partial growth plate arrest. Results after four years or at maturity. J Bone Joint Surg Br. 1989;71(1):13–6.
72. Lee EH, Chen F, Chan J, Bose K. Treatment of growth arrest by transfer of cultured chondrocytes into physeal defects. J Pediatr Orthop. 1998;18(2):155–60.
73. Aldegheri R, Trivella G, Lavini F. Epiphyseal distraction. Hemichondrodiatasis. Clin Orthop Relat Res. 1989;241:128–36.
74. Canadell J, de Pablos J. Correction of angular deformities by physeal distraction. Clin Orthop Relat Res. 1992;283:98–105.
75. CF M. Leg length discrepancy and angular deformity of the lower limbs. In: Morrissy RT, Weinstein SL, editors. Lovell and Winter's pediatric orthopaedics. 4th ed. Philadelhia: Lippincott-Raven; 1996. p. 849.
76. Stanitski DF. Limb-length inequality: assessment and treatment options. J Am Acad Orthop Surg. 1999;7(3):143–53.
77. Vitale MA, Choe JC, Sesko AM, Hyman JE, Lee FY, Roye Jr DP, et al. The effect of limb length discrepancy on health-related quality of life: is the '2 cm rule' appropriate? J Pediatr Orthop B. 2006;15(1):1–5.
78. Song KM, Halliday SE, Little DG. The effect of limb-length discrepancy on gait. J Bone Joint Surg Am. 1997;79(11):1690–8.
79. Gross RH. Leg length discrepancy: how much is too much? Orthopedics. 1978;1(4):307–10.
80. Paley D, Bhave A, Herzenberg JE, Bowen JR. Multiplier method for predicting limb-length discrepancy. J Bone Joint Surg Am. 2000;82-A(10):1432–46.
81. Menelaus MB. Correction of leg length discrepancy by epiphysial arrest. J Bone Joint Surg Br. 1966;48(2):336–9.
82. Anderson M, Messner MB, Green WT. Distribution of lengths of the normal femur and tibia in children from one to eighteen years of age. J Bone Joint Surg Am. 1964;46:1197–202.
83. Anderson M, Green WT, Messner MB. Growth and predictions of growth in the lower extremities. J Bone Joint Surg Am. 1963; 45-A:1–14.
84. White J, Stubbins Jr SG. Growth arrest for equalilzing leg lengths. J Am Med Assoc. 1944;126(18):1146–9.
85. Phemister DB. Operative arrestment of longitudinal growth of bones in the treatment of deformities. J Bone Joint Surg. 1933; 15(1):1–15.
86. Haas SL. Retardation of bone growth by a wire loop. J Bone Joint Surg. 1945;27(1):25–36
87. Haas SL. Mechanical retardation of bone growth. J Bone Joint Surg. 1948;30(2):506–12

88. Blount WP, Clarke GR. Control of bone growth by epiphyseal stapling; a preliminary report. J Bone Joint Surg Am. 1949;31A(3):464–78.
89. Blount WP. A mature look at epiphyseal stapling. Clin Orthop Relat Res. 1971;77:158–63.
90. Metaizeau JP, Wong-Chung J, Bertrand H, Pasquier P. Percutaneous epiphysiodesis using transphyseal screws (PETS). J Pediatr Orthop. 1998;18(3):363–9.
91. Pendleton AM, Stevens PM, Hung M. Guided growth for the treatment of moderate leg-length discrepancy. Orthopedics. 2013;36(5):e575–80.
92. Stephens DC, Herrick W, MacEwen GD. Epiphysiodesis for limb length inequality: results and indications. Clin Orthop Relat Res. 1978;136:41–8.
93. Green WT, Anderson M. Experiences with epiphyseal arrest in correcting discrepancies in length of the lower extremities in infantile paralysis: a method of predicting the effect. J Bone Joint Surg Am. 1947;29(3):659–75.
94. Green WT, Anderson M. Epiphyseal arrest for the correction of discrepancies in length of the lower extremities. J Bone Joint Surg Am. 1957;39-A(4):853–72; discussion, 72; passim.
95. Bowen JR, Johnson WJ. Percutaneous epiphysiodesis. Clin Orthop Relat Res. 1984;190:170–3.
96. Canale ST, Russell TA, Holcomb RL. Percutaneous epiphysiodesis: experimental study and preliminary clinical results. J Pediatr Orthop. 1986;6(2):150–6.
97. Ogilvie JW. Epiphysiodesis: evaluation of a new technique. J Pediatr Orthop. 1986;6(2):147–9.
98. Scott AC, Urquhart BA, Cain TE. Percutaneous vs modified phemister epiphysiodesis of the lower extremity. Orthopedics. 1996;19(10):857–61.
99. Porat S, Peyser A, Robin GC. Equalization of lower limbs by epiphysiodesis: results of treatment. J Pediatr Orthop. 1991;11(4):442–8.
100. Ogilvie JW, King K. Epiphysiodesis: two-year clinical results using a new technique. J Pediatr Orthop. 1990;10(6):809–11.
101. Macnicol MF, Gupta MS. Epiphysiodesis using a cannulated tube-saw. J Bone Joint Surg Br. 1997;79(2):307–9.
102. Liotta FJ, Ambrose 2nd TA, Eilert RE. Fluoroscopic technique versus Phemister technique for epiphysiodesis. J Pediatr Orthop. 1992;12(2):248–51.
103. Horton GA, Olney BW. Epiphysiodesis of the lower extremity: results of the percutaneous technique. J Pediatr Orthop. 1996;16(2):180–2.
104. Canale ST, Christian CA. Techniques for epiphysiodesis about the knee. Clin Orthop Relat Res. 1990;255:81–5.
105. Gabriel KR, Crawford AH, Roy DR, True MS, Sauntry S. Percutaneous epiphyseodesis. J Pediatr Orthop. 1994;14(3):358–62.
106. Atar D, Lehman WB, Grant AD, Strongwater A. Percutaneous epiphysiodesis. J Bone Joint Surg Br. 1991;73(1):173.
107. Timperlake RW, Bowen JR, Guille JT, Choi IH. Prospective evaluation of fifty-three consecutive percutaneous epiphysiodeses of the distal femur and proximal tibia and fibula. J Pediatr Orthop. 1991;11(3):350–7.
108. Bylander B, Selvik G, Hansson LI, Aronson S. A roentgen stereophotogrammetric analysis of growth arrest by stapling. J Pediatr Orthop. 1981;1(1):81–90.
109. Bylander B, Hansson LI, Selvik G. Pattern of growth retardation after Blount stapling: a roentgen stereophotogrammetric analysis. J Pediatr Orthop. 1983;3(1):63–72.
110. Sengupta A, Gupta P. Epiphyseal stapling for leg equalization in developing countries. Int Orthop. 1993;17(1):37–42.
111. Frantz CH. Epiphyseal stapling: a comprehensive review. Clin Orthop Relat Res. 1971;77:149–57.
112. May Jr VR, Clements EL. Epiphyseal stapling: with special reference to complications. South Med J. 1965;58(10):1203–7.
113. Trias A, Mueller K, Ray RD. Epiphysial stapling. Surg Gynecol Obstet. 1961;113:315–23.
114. Nouth F, Kuo LA. Percutaneous epiphysiodesis using transphyseal screws (PETS): prospective case study and review. J Pediatr Orthop. 2004;24(6):721–5.
115. Ilharreborde B, Gaumetou E, Souchet P, Fitoussi F, Presedo A, Pennecot GF, et al. Efficacy and late complications of percutaneous epiphysiodesis with transphyseal screws. J Bone Joint Surg Br. 2012;94(2):270–5.
116. Song MH, Choi ES, Park MS, Yoo WJ, Chung CY, Choi IH, et al. Percutaneous epiphysiodesis using transphyseal screws in the management of leg length discrepancy: optimal operation timing and techniques to avoid complications. J Pediatr Orthop. 2015;35(1):89–93.
117. Monier BC, Aronsson DD, Sun M. Percutaneous epiphysiodesis using transphyseal screws for limb-length discrepancies: high variability among growth predictor models. J Child Orthop. 2015;9(5):403–10.
118. Stevens PM, Pease F. Hemiepiphysiodesis for posttraumatic tibial valgus. J Pediatr Orthop. 2006;26(3):385–92.
119. Stewart D, Cheema A, Szalay EA. Dual 8-plate technique is not as effective as ablation for epiphysiodesis about the knee. J Pediatr Orthop. 2013;33(8):843–6.
120. Gaumetou E, Mallet C, Souchet P, Mazda K, Ilharreborde B. Poor efficiency of eight-plates in the treatment of lower limb discrepancy. J Pediatr Orthop 2015;15.
121. Little DG, Nigo L, Aiona MD. Deficiencies of current methods for the timing of epiphysiodesis. J Pediatr Orthop. 1996;16(2):173–9.
122. Surdam JW, Morris CD, DeWeese JD, Drvaric DM. Leg length inequality and epiphysiodesis: review of 96 cases. J Pediatr Orthop. 2003;23(3):381–4.
123. Campens C, Mousny M, Docquier PL. Comparison of three surgical epiphysiodesis techniques for the treatment of lower limb length discrepancy. Acta Orthop Belg. 2010;76(2):226–32.
124. Lykissas MG, Jain VV, Manickam V, Nathan S, Eismann EA, McCarthy JJ. Guided growth for the treatment of limb length discrepancy: a comparative study of the three most commonly used surgical techniques. J Pediatr Orthop B. 2013;22(4):311–7.
125. Babu LV, Evans O, Sankar A, Davies AG, Jones S, Fernandes JA. Epiphysiodesis for limb length discrepancy: a comparison of two methods. Strateg Trauma Limb Reconstr. 2014;9(1):1–3.
126. Lauge-Pedersen H, Hagglund G. Eight plate should not be used for treating leg length discrepancy. J Child Orthop. 2013;7(4):285–8.
127. Mankin HJ. Rickets, osteomalacia, and renal osteodystrophy. Part II. J Bone Joint Surg Am. 1974;56(2):352–86.
128. Boero S, Michelis MB, Riganti S. Use of the eight-plate for angular correction of knee deformities due to idiopathic and pathologic physis: initiating treatment according to etiology. J Child Orthop. 2011;5(3):209–16.
129. Stevens PM, Klatt JB. Guided growth for pathological physes: radiographic improvement during realignment. J Pediatr Orthop. 2008;28(6):632–9.
130. Schroerlucke S, Bertrand S, Clapp J, Bundy J, Gregg FO. Failure of Orthofix eight-plate for the treatment of Blount disease. J Pediatr Orthop. 2009;29(1):57–60.
131. Wiemann JM, Tryon C, Szalay EA. Physeal stapling versus 8-plate hemiepiphysiodesis for guided correction of angular deformity about the knee. J Pediatr Orthop. 2009;29(5):481–5.
132. Mielke CH, Stevens PM. Hemiepiphyseal stapling for knee deformities in children younger than 10 years: a preliminary report. J Pediatr Orthop. 1996;16(4):423–9.
133. Stevens PM, Maguire M, Dales MD, Robins AJ. Physeal stapling for idiopathic genu valgum. J Pediatr Orthop. 1999;19(5):645–9.

134. Degreef I, Moens P, Fabry G. Temporary epiphysiodesis with Blount stapling for treatment of idiopathic genua valga in children. Acta Orthop Belg. 2003;69(5):426–32.
135. Westberry DE, Davids JR, Pugh LI, Blackhurst D. Tibia vara: results of hemiepiphyseodesis. J Pediatr Orthop B. 2004;13(6): 374–8.
136. Park SS, Gordon JE, Luhmann SJ, Dobbs MB, Schoenecker PL. Outcome of hemiepiphyseal stapling for late-onset tibia vara. J Bone Joint Surg Am. 2005;87(10):2259–66.
137. Courvoisier A, Eid A, Merloz P. Epiphyseal stapling of the proximal tibia for idiopathic genu valgum. J Child Orthop. 2009; 3(3):217–21.
138. Zuege RC, Kempken TG, Blount WP. Epiphyseal stapling for angular deformity at the knee. J Bone Joint Surg Am. 1979; 61(3):320–9.
139. Fraser RK, Dickens DR, Cole WG. Medial physeal stapling for primary and secondary genu valgum in late childhood and adolescence. J Bone Joint Surg Br. 1995;77(5):733–5.
140. Stevens PM. Guided growth for angular correction: a preliminary series using a tension band plate. J Pediatr Orthop. 2007;27(3): 253–9.
141. Farnum CE, Nixon A, Lee AO, Kwan DT, Belanger L, Wilsman NJ. Quantitative three-dimensional analysis of chondrocytic kinetic responses to short-term stapling of the rat proximal tibial growth plate. Cells Tissues Organs. 2000;167(4): 247–58.
142. Bylski-Austrow DI, Wall EJ, Rupert MP, Roy DR, Crawford AH. Growth plate forces in the adolescent human knee: a radiographic and mechanical study of epiphyseal staples. J Pediatr Orthop. 2001;21(6):817–23.
143. Aykut US, Yazici M, Kandemir U, Gedikoglu G, Aksoy MC, Cil A, et al. The effect of temporary hemiepiphyseal stapling on the growth plate: a radiologic and immunohistochemical study in rabbits. J Pediatr Orthop. 2005;25(3):336–41.
144. Khoury JG, Tavares JO, McConnell S, Zeiders G, Sanders JO. Results of screw epiphysiodesis for the treatment of limb length discrepancy and angular deformity. J Pediatr Orthop. 2007;27(6):623–8.
145. Mesa PA, Yamhure FH. Percutaneous hemi-epiphysiodesis using transphyseal cannulated screws for genu valgum in adolescents. J Child Orthop. 2009;3(5):397–403.
146. Burghardt RD, Herzenberg JE, Standard SC, Paley D. Temporary hemiepiphyseal arrest using a screw and plate device to treat knee and ankle deformities in children: a preliminary report. J Child Orthop. 2008;2(3):187–97.
147. Guzman H, Yaszay B, Scott VP, Bastrom TP, Mubarak SJ. Early experience with medial femoral tension band plating in idiopathic genu valgum. J Child Orthop. 2011;5(1):11–7.
148. Ballal MS, Bruce CE, Nayagam S. Correcting genu varum and genu valgum in children by guided growth: temporary hemiepiphysiodesis using tension band plates. J Bone Joint Surg Br. 2010;92(2):273–6.
149. Burghardt RD, Herzenberg JE. Temporary hemiepiphysiodesis with the eight-Plate for angular deformities: mid-term results. J Orthop Sci. 2010;15(5):699–704.
150. De Brauwer V, Moens P. Temporary hemiepiphysiodesis for idiopathic genua valga in adolescents: percutaneous transphyseal screws (PETS) versus stapling. J Pediatr Orthop. 2008;28(5):549–54.
151. Shin SJ, Cho TJ, Park MS, Bae JY, Yoo WJ, Chung CY, et al. Angular deformity correction by asymmetrical physeal suppression in growing children: stapling versus percutaneous transphyseal screw. J Pediatr Orthop. 2010;30(6):588–93.
152. Sung KH, Ahn S, Chung CY, Lee KM, Kim TW, Han HS, et al. Rate of correction after asymmetrical physeal suppression in valgus deformity: analysis using a linear mixed model application. J Pediatr Orthop. 2012;32(8):805–14.
153. Jelinek EM, Bittersohl B, Martiny F, Scharfstadt A, Krauspe R, Westhoff B. The 8-plate versus physeal stapling for temporary hemiepiphyseodesis correcting genu valgum and genu varum: a retrospective analysis of thirty five patients. Int Orthop. 2012; 36(3):599–605.
154. Gottliebsen M, Rahbek O, Hvid I, Davidsen M, Hellfritzsch MB, Moller-Madsen B. Hemiepiphysiodesis: similar treatment time for tension-band plating and for stapling: a randomized clinical trial on guided growth for idiopathic genu valgum. Acta Orthop. 2013;84(2):202–6.
155. Sung KH, Chung CY, Lee KM, Lee SY, Choi IH, Cho TJ, et al. Determining the best treatment for coronal angular deformity of the knee joint in growing children: a decision analysis. BioMed Res Int. 2014;2014:603432.
156. Peterson HA. Partial growth plate arrest and its treatment. In: Morrissey RT, editor. Lovell and Winter's Pediatric orthopaedics. 3rd ed. Philadelphia: Lippincott; 1990. p. 1071–89.
157. Peterson HA. Premature partial physeal closure: assessment and treatment. J Bone Joint Surg. 1987;69B:154.
158. Peterson HA. Management of partial physeal arrest. In: Chapman MV, editor. Operative orthopaedics. Philadelphia: J B Lippincott; 1998. p. 3065–75.
159. Dunn RN. Physeal bar resection for partial growth arrest J Bone Joint Surg American volume. 1999;81B(Suppl III):213.

Evidence-Based Management of Limb Length Discrepancy

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Abstract

Limb length discrepancy is a common paediatric orthopaedic problem. Treatment is based on predicting the leg length difference at skeletal maturity and equalizing them if it is deemed necessary. Several methods have been used to achieve the above; predicting the leg length difference at skeletal maturity and matching them. In this chapter, we explored the published evidence behind the available methods.

Keywords

Limb length discrepancy • LLD • Long leg • Short leg • Bone age • Leg lengthening • Bone lengthening • Leg shortening • Leg equalization

Background

The management guidelines of limb length discrepancy LLD in childhood are based on the magnitude of the predicted LLD at skeletal maturity. Predicted LLD < 2 cm requires no treatment, while discrepancies between 2 cm and 5 cm are usually treated with growth arrest procedure of the contralateral normal or long side; discrepancies 5–15 cm need lengthening of the affected short side with or without timely contralateral physeal arrest surgery; and cases of massive discrepancies more than 15 cm might require prosthetic replacement with or without partial amputation [1]. Growth plate arrest techniques or epiphysiodesis of the long or healthy leg would allow the shorter limb to catch up with growth, gradually decreasing and correcting the limb length inequality. Many techniques were described for permanent epiphysiodesis like the original Phemister technique [2], Green-Phemister technique [3], or percutaneous epiphysiodesis by either transphyseal drilling and curettage [4] or transphyseal screws (PETS) described by Metaizeau et al. [5]. The idea of growth modulation in the treatment of LLD, to allow temporary arrest of the physeal growth and decrease the likelihood of overcorrection in case of too early epiphysiodesis, was first introduced by Blount using physeal plate stapling [6] and recently by using bilateral 8-plates designed originally by Stevens [7] for temporary unilateral growth modulation in the treatment angular deformities.

On the other hand, lengthening of the short or affected limb can be appealing because it corrects the diseased side and equalize limbs length without sacrificing physeal growth and losing some height. Current techniques of limb lengthening utilize the distraction osteogenesis principles that were popularized and refined by Gavril Ilizarov in the early 1950th [8–10]. The landmark research done by Ilizarov in that field emphasized the importance of stable fixation, preservation of soft tissues and vascularity of the bony fragments and the influence of distraction rate and rhythm on the quality and success of bone regeneration. Different forms of external fixation were used originally in limb lengthening [9, 11, 12] and the main disadvantages were pin track infections, muscle contractures and joint stiffness, axial deviation, neurovascular injuries, acute or chronic refractures and poor patient acceptance for the bulky external fixator [13, 14]. Complication rates up to 200 % were reported for limb lengthening using external fixation [13, 15]. Another

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achievement in the field of limb lengthening surgery is the development of intramedullary expandable nails which are used for intramedullary lengthening. Different nail designs were developed like the motorized lengthening nail (Fitbone) [16], the mechanically activated lengthening nails as Albizzia nail [17] and the intramedullary skeletal kinetic distractor (ISKD) [18] and more recently the magnetically driven intramedullary lengthening nail (PRECICE nail) [19]. To date, the available intramedullary lengthening nails for clinical use are the Fitbone and the PRECICE nail while the mechanically activated lengthening nails fell out of favour due problems with distraction ratchets and mechanism which led to either failure of distraction (jammed nail) or non-controlled distraction (runaway nail) [20, 21].

When a pediatric orthopedic surgeon is confronted with a child with limb length inequality, his primary interest is to find out the final or the predicted LLD at skeletal maturity without surgery and then treatment decision would be based upon the magnitude of that predicted LLD. In case of choosing growth arrest procedure or the combination of lengthening and growth arrest procedure, the second task would be to determine the optimal timing and location of the growth arrest procedure. In fact, predicting the future limb growth and the LLD at skeletal maturity and the calculation of optimal timing for growth arrest procedures are the most controversial aspects in the management of limb length inequality in a growing child. Predicting the timing of epiphysiodesis is still an imperfect science with relative reported inaccuracies in all different systems used in the determination of skeletal maturation [1]. Therefore, we aimed at this part to try to find the best evidence and to analyze the difficulties encountered in predicting future limb growth.

Predicting Future Limb Growth

Growth is a dynamic process with a great deal of individual variation. Limb equalization surgery for limb length discrepancy in a growing child needs a good judgment for the amount of growth remaining in each limb segment and careful calculation of the predicted limb length discrepancy (LLD) at skeletal maturity [22–24]. To achieve an acceptable accuracy in the prediction of future growth and limb length at skeletal maturity, a good understanding for the growth phases is required [25]. There are three distinct growth phases in childhood:

1. The rapidly decelerating growth phase from birth to 5-years of age at which the standing height reaches 60 % of the adult height,
2. The steady growth phase between the age of 6 to 9-years which is characterized by a stable annual growth rate reaching 80 % of the adult height at the age of 9-years,
3. The final acceleration (pubertal) phase which starts by the age of 10-years when the growth patterns and rates of boys and girls diverge markedly reaching the maximum growth rate between the age of 10 to 12-years in girls and 12 to 14-years in boys by the onset of the pubertal growth spurt. The rate of growth (increase in height) during pubertal growth spurt and the first pubertal skeletal sign is more than 0.5 cm/month or 6 cm/year [25].

It must be remembered that the age at which this maximum annual growth rate would be achieved and the absolute value of this increase are markedly variable between individual children [22]. For example, according to Anderson et al. [22] the rate of increase in standing height at the age interval 8–9 years is in average 5.7 ± 0.77 cm and 5.7 ± 0.88 cm for girls and boys respectively, while the growth rates at the middle of the pubertal growth spurt, 11–12 years of age for girls and 13–14 years of age in boys, are 6.5 ± 1.91 cm and 7.4 ± 2.02 cm respectively. The high standard deviations of growth rate values denote the inherent variability at pubertal phase of growth.

Difficulties encountered with the prediction of final limb length and the final or predicted LLD at maturity fall in one of three main categories:

1. **The reliability of different methods used to assess the maturity status; skeletal and physical maturity.**
   a. Skeletal maturity or the assessment of bone or skeletal age
      i. Greulich & Pyle skeletal atlas of hand and wrist radiographs [26]
      ii. Sauvegrain method using elbow radiographs [27, 28]
      iii. The shorthand bone age assessment method [29]
      iv. Tanner-Whitehouse (TW) methods:
   - TW2 … 20 bones are assessed which include distal radius and ulna, 7 carpal bones (excluding the pisiform), first, third, and fifth metacarpals and their associated phalanges (2, 3, and 3 respectively) [30].
   - TW3 (TW3/RUS) system … refers to R = Radius, U = Ulan and S = Short bones (metacarpals and their phalanges). The carpal bones were no longer included [31].

   b. Physical maturity
      i. Tanner staging for primary and secondary sexual characters [32, 33].
      ii. Voice changes
      iii. Onset of menarche
   c. Rate of annual growth [25].
2. **Difficulties with the determination of the growth percentile**

3. **The calculation of the growth inhibition rate and the developmental growth pattern of the affected short limb and the state of the its growth plates** [34].

Maturity and Skeletal Age Determination

The maturity status is very essential in detecting the start of the pubertal growth spurt which is a very important period for physeal growth arrest procedures. The pubertal growth spurt can be divided into two phases: the acceleration and deceleration phases. In the acceleration or the ascending phase (11–13 years and 13–15 years ***skeletal age*** in girls and boys respectively), the standing height gain is in average 12–14 cm/year for girls and boys respectively and this phase is the most important period for planned epiphysiodesis procedures. The deceleration or the descending phase (13–16 years and 15–18 years skeletal age in girls and boys respectively) during which the rate of growth progressively decreases till complete cessation [27]. To evaluate the maturity status of an individual, both skeletal maturity or skeletal age and physical signs of maturity should be assessed. Skeletal maturity is determined by the skeletal or bone age, while physical maturity takes in account the development of the secondary sexual characters, the onset of menarche, voice change and the annual growth velocity. The most commonly used methods in the determination of the skeletal age are Greulich and Pyle atlas of hand and wrist radiographs [26], Sauvegrain's method using elbow radiographs [28] and the shorthand bone age assessment method [29].

Greulich and Pyle atlas of the hand and wrist [26] uses postero-anterior radiographs of the left hand and wrist and remains the most widely used method for the assessment of skeletal age and maturation. The main disadvantage is that there are no radiographic data corresponding to 14.5 years of age in boys and 11.5 and 12.5 years of age in girls which are very important time periods during the pubertal growth spurt. Furthermore, during the phase of increased growth velocity between 11–13 years in girls and 13–15 years in boys, the morphologic changes in the hand and wrist are neither obvious nor important [27]. The start of puberty is marked by the appearance of the sesamoid bone of the first metacarpal while the end is marked by fusion of the epiphysis of the distal phalanx of the thumb and the first metacarpal. Other radiographic signs in between as the shape of the radial and ulnar epiphyses, the outline of the hook of the hamate, or the wideness of the metacarpal epiphyses are difficult to evaluate [27]. The interobserver reliability is questionable as well. In a study by Cundy et al. [35], 60 hand and wrist radiographs of children with known LLD were evaluated independently by 4 radiologists to determine the skeletal age. Significant variations were found, with 50 % of the children were assigned a skeletal age that differed by >1 year between radiologists and 10 % more than 2 years (Level III evidence).

Dimeglio et al. [27] have studied the accuracy and the reliability (interobserver and intraobserver) of a modified version of the Sauvegrain method [28] or the French method in the determination of the skeletal age by using elbow antero-posterior and lateral radiographs in comparison to the Greulich and Pyle system (level II evidence). The Sauvegrain method uses the anatomical landmarks around the elbow: the lateral condyle and epicondyle, trochlea, olecranon apophysis and proximal radial apophysis. Antero-posterior and lateral radiographs of the left elbow and postero-anterior radiographs of the left hand and wrist were obtained at the same point of time for 60 boys and 60 girls. Three observers independently assessed the skeletal age using the Sauvegrain method in two separate occasions, 4 weeks apart for every observer. Then the data was compared to the skeletal ages of the same 60 boys and 60 girls determined form the hand and wrist radiographs according to the system of Greulich and Pyle [26]. The same review protocol was followed for the Greulich and Pyle method.

The mean interobserver correlation coefficients were r = 0.93 and 0.88 for the Sauvegrain method and Greulich and Pyle system respectively, while the intraobserver correlation coefficients were r = 0.96 and 0.92 respectively. Correlation between methods in the determination of skeletal age was strong with mean correlation coefficient of r = 0.85. The interobserver agreement was analyzed; the observers differed by 2 years or more in 5 % and 8 % of the girls and boys respectively when using the atlas of Greulich and Pyle while disagreement of 2 years or more was not found with the use of the Sauvegrain method. When the two methods were compared, the method of Sauvegrain was found to be more accurate because it allowed a clearer differentiation of skeletal age in 6-month interval.

Advantages of the Sauvegrain method compared to Greulich and Pyle atlas for the determination of the skeletal age are: (1) The ossification centers around the elbow have clear sequence in their development and fusion. At the onset puberty (11 years of age in girls and 13 years in boys), the elbow is still largely cartilaginous. Two years later, fusion of the elbow growth centers is complete marking the critical period of the pubertal growth spurt. (2) Sauvegrain method have the ability of identifying the two phases of pubertal growth spurt; the acceleration and deceleration phases. The beginning of the acceleration phase is marked by the appearance of the second ossific center of the olecranon apophysis. The deceleration or the descending phase is marked by elbow physeal closure and Risser sign starts to appear 6 months after its onset. (3) According to the experience of Dimeglio and his coworkers, the changes of the olecranon apophysis alone are very characteristic (Table 46.1) and give direct clue the skeletal age by itself.

Table 46.1 Characteristic morphologic changes of the Olecranon apophysis according the skeletal age in boys and girls

	Skeletal age (Y)	
Olecranon apophysis morphology	Girls	Boys
Two ossific centers	11	13
Half-moon image	11.5	13.5
Rectangular aspect	12	14
Beginning of fusion	12.5	14.5
Complete fusion	13	15

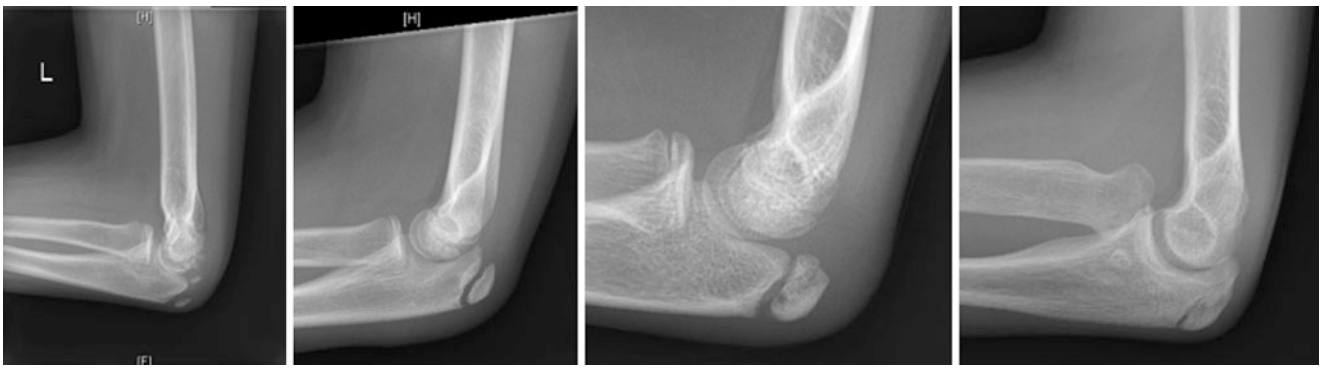

Fig. 46.1 Sauvegrain skeletal bone age

The main disadvantage of the Sauvegrain method is the limited use to the pubertal phase and the year preceding it (10–13 years and 11–14 years in girls and boys respectively). Prior to the pubertal phase, the elbow is largely cartilaginous and changes in the ossific centers cannot be clearly differentiated (Fig. 46.1).

The shorthand bone age assessment method developed by **Heyworth et al.** [29] was to simplify the determination of skeletal age by using a single radiographic criterion from Greulich and Pyle atlas for males between the ages of 12.5 to 16 years and females from 10 to 16 years (Table 46.2). Good intraobserver and interobserver reliability was reported in assessing bone age of 140 males and 120 females. There was substantial agreement and strong correlation when compared with Greulich and Pyle assessments (Level III evidence).

Growth Tables and Charts and the Determination of Growth Percentiles

The first step in the prediction of the future growth remaining in a limb or a limb segment is to determine the growth percentile or the relative size for age (chronologic or skeletal age) within which this limb or limb segment would lie relative to the standard growth records. This is very important because an individual growth tends to be within the same growth percentile throughout different growth phases [23] and in turn would allow us to track and predict the future growth remaining along the records of his peers within the same percentile till skeletal maturity. The growth percentile can be assessed by using different available age and gender specific growth tables and charts which document the average values and percentiles [22, 23, 36, 37].

The most complete set of longitudinal records of height and lower extremity growth in children and the most commonly used growth database in clinical practice is that published by Green and Anderson [22, 23]. Green, Anderson and Messner published two landmark studies. In the first one [23], they used longitudinal data for femoral and tibial lengths of healthy children, recorded by annual lower extremity radiographs from 1 to 18 years of age (67 boys and 67 girls). Those growth data were for North American white males and females of predominately Irish origin and were collected as a part of a longitudinal series of child health and development studies conducted at Harvard School of Public Health, Boston from 1930 to 1956 [38]. They were able to construct gender specific growth tables and charts based on chronologic age from 1 to 18 years defining annual average growth values and standard deviations or growth percentiles (mean ± 1 and 2 SD). In a separate work [22], they published growth remaining charts and tables for the normal femur and tibia relative to chronologic and skeletal age of 50 boys and 50 girls between the age of 8 and 18 years. Forty-nine children (25 girls and 24 boys) had unilateral paralytic poliomyelitis which affected only one lower extremity and therefore data of the healthy side were used. All children had annual orthoroentgenograms for the recording of femoral and tibial lengths as well as hand and wrist radiographic assessment of

Table 46.2 The shorthand bone age assessment method

Radiographic criteria	Skeletal age (Y)	
	Girls	Boys
Appearance of hook of hamate ossific nucleus	10	12.5
Appearance of thumb sesamoid ossific nucleus	11	13
Width of proximal aspect of distal radius epiphysis equals width of distal aspect of distal radius metaphysis, but has not yet begun to cap	–	13.5
Capping of distal radius epiphysis	12	14
Closure of thumb distal phalanx physis	13	15
Closure index finger distal phalanx physis	13.5	15.5
Closure of index finger proximal phalanx physis	14	16
Closure of index finger distal metacarpal physis	15	–
Closure of distal ulna physis	16	–
Closure of distal radius physis	17	–

the skeletal age according to Greulich-Pyle Atlas [22, 26]. They further calculated the annual growth contribution and the growth remaining in the distal femur and proximal tibia (71 % of the total femoral growth and 57 % of the total tibial growth respectively) according to the skeletal age. Growth remaining tables and charts for the normal distal femur and proximal tibia were derived based on the skeletal age between 8 and 17 years with annual average values and four percentile levels (90th, 75th, 25th and 10th) and those tables and charts were the bases of Green and Anderson growth remaining method in predicting the optimal timing of epiphysiodesis. Green and Anderson work became the bases of Moseley straight line graph [39] and Paley's multiplier method [24].

Other racial specific growth charts and curves were developed by Pritchett and Bortel [37] for Scandinavian American children in Denver, Colorado, by Beumer et al. [36] for Dutch children in Rotterdam, Netherlands and by Ha et al. [40] for Korean children. Beumer et al. [36] studied the femoral and tibial growth data in 182 Dutch children by serial orthoroentgenograms from 1979 to 1994. They found significant differences in the length of the femur in girls aged 8–9 years and boys 10–15 years and in the length of the tibia in boys and girls between the ages of 6–16 years. The Dutch children tend to be taller compared to the data of Anderson et al. [23]. They developed the Rotterdam straight line graph RSLG which is similar but not identical to Moseley straight line graph based on their own growth data.

Growth Inhibition Rate and Pattern of the Affected Extremity

Fredric Shapiro published an important article [34] in which he retrospectively reviewed longitudinal data of 803 children with LLD followed by the growth study unit at Boston Children's Hospital over a 40-year period (1940–1980). All patients in this series were assessed at least annually (or more often semi-annually), for a minimum 5 years either to skeletal maturity or to the time of bony surgery. The disease entities which were included were: proximal focal femoral deficiency (PFFD), congenital coxa vara and congenitally short femur, Ollier's disease, physeal destruction, poliomyelitis, septic arthritic of the hip, fractured femoral diaphysis, hemangioma, neurofibromatosis, hemiplegic cerebral palsy, hemiatrophy or hemihypertrophy, juvenile rheumatoid arthritis and Legg-Calvé-Perthe's disease (LCPD). Shapiro was able to describe five main basic developmental patterns for progressive lower extremity length discrepancies (Table 46.3 and Fig. 46.2).

The calculation of the predicted LLD is easiest with **type I** discrepancies which increase at a constant rate during the whole growth. In other words, the percentage of the length achieved by the short limb relative to the healthy side at any age would be the same during the remaining growth and therefore, the first step is to determine the length percentile of the normal bone or limb and calculate its final length at maturity for that percentile. The short limb length is then easily calculated as a percentage of the normal side maturity length. **Type II** is a difficult pattern to project as the information available from the period of constant increase before the deceleration phase has no predictive value. Multiple assessments for the growth inhibition rate are needed to allow for additional calculations. An example of type II discrepancies is poliomyelitis in which there is a tendency for the discrepancy to increase most rapidly in the first four or five years following infection then the rate of increased discrepancy diminishes gradually thereafter. In **type III** patterns, once a plateau has reached, the LLD will be the same throughout the remaining growth. The typical example for type III LLD is the overgrowth following femoral shaft fractures. **Type IV** discrepancies were found to be characteristic for hip diseases in childhood. Premature closure of the proximal capital femoral epiphysis is the cause for the late upward slope following a long time of plateau phase and once detected, it is easy to calculate the remaining growth of the entire femur and to add a 30 % from that value (the contribution of the proximal

Table 46.3 Developmental patterns of lower extremity length discrepancies according to Shapiro

Type	Description	Common disease entities
Type I – upward slope pattern	The discrepancy increases at a constant rate as the growth inhibition rate remains the same throughout growth	Congenital diseases like PFFD, physeal destruction
Type II – upward slope-deceleration pattern	The discrepancy increases at a constant rate for a variable period of time and then shows decremental rate of increase which is variable from patient to patient and from condition to condition	Poliomyelitis, Juvenile rheumatoid arthritis
Type III – upward slope-plateau pattern	The discrepancy increases with time and then a plateau is reached and it will not change throughout the remaining growth	Overgrowth following femoral fractures, Infections like diaphyseal osteomyelitis
Type IV – upward slope-plateau-upward slope pattern	The discrepancy increases then stabilizes for a variable period of time and then increases again towards the end of growth	Hip disease (septic arthritis of the hip, LCPD, AVN with the treatment of DDH)
Type V – upward slope-plateau-downward slope pattern	The discrepancy increases and then stabilizes and then decreases without surgery	Juvenile rheumatoid arthritis

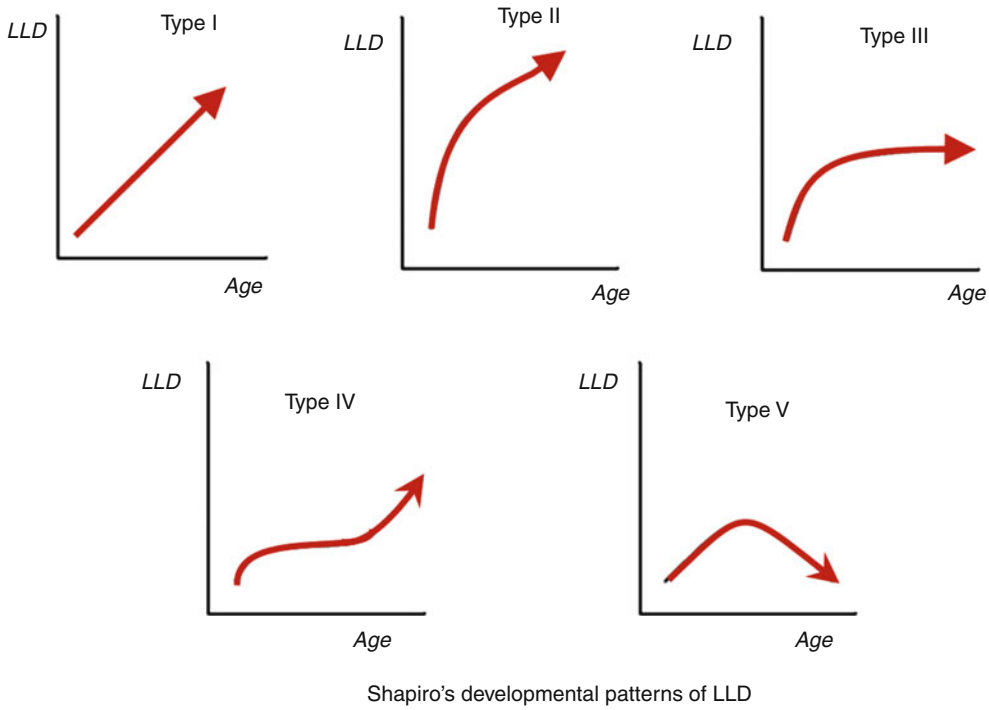

Fig. 46.2 Shapiro's developmental patterns of LLD

femoral physis the normal femoral growth) to pre-existing discrepancy to give the final LLD. In **type V**, the discrepancy starts to correct itself. It is commonly seen in chronic inflammatory diseases which stimulates growth under 10 years of age and then causes premature growth cessation towards the end of growth.

Calculating LLD at Skeletal Maturity and the Timing of Epiphysiodesis

To calculate the predicted LLD at skeletal maturity, one should firstly know the growth percentile of the long or healthy limb. With the help of the standard growth curves, the length of the long limb can then be projected to skeletal maturity to get its final length. When the growth inhibition rate and pattern of the short limb are taken in account, the final length of the affected limb at maturity and therefore the LLD at skeletal maturity can be calculated.

The most commonly used methods for predicting the limb length at skeletal maturity and calculating the timing of epiphysiodesis are the Menelaus rule of thumb [41], Green and Anderson growth remaining method [22, 23], Moseley straight line graph [39] and Paley's multiplier method [24].

Menelaus Rule of Thumb

Menelaus rule of thumb was a simplified approach to project the limb length discrepancy at maturity based on chronological age rather than skeletal age and it didn't take in account the growth percentile of the individual. It was based on the assumptions of White and Stubbins [42] that the lower femoral physis will grow by 3/8 inch/year while the upper tibial physis will grow by 1/4 inch/year. He assumed that growth would stop at the age of 16 years in boys and 14 years in girls [41]. He studied the results of 53 epiphysiodeses in 44 children who had the timing of surgery calculated based on this assumption. At maturity, 52 % of this group had a residual LLD of ¼ inch, 41 % within ¾ inch and 7 % more than ¾ of an inch (level IV evidence).

Green and Anderson Growth Remaining Method

To use Green and Anderson method, firstly the normal leg length (the femoral and tibial length) is compared to the data reported by Anderson et al. [23] for the current age and sex to determine the current growth percentile. The leg length is then projected to skeletal maturity for that percentile group to predict its final maturity length. The short leg length at maturity can be known after calculating the growth inhibition rate from previous assessments [(Growth of the long leg – Growth of the short leg)/growth of the long leg] and applying the same growth inhibition rate to the future growth of the long leg.

The next step is to use the growth remaining charts and tables to estimate the effect of physeal growth arrest procedures on the final length of the limb within the same growth percentile and hence the optimal timing of epiphysiodesis [22].

Moseley Straight Line Graph

Moseley converted Green and Anderson growth curve of the normal limb into a straight line of 45 ° slope by shifting the data points along the x axis and altering the distance between the age scale on the x axis by a comparable amount [39]. Another important concept for the straight line graph is the addition of a nomogram relating leg length to skeletal age for each gender which would provide a mechanism for taking the child's growth percentile into account in predicting at what lengths the growth of the legs will stop. The main advantage of using Moseley straight line graphs is that it can demonstrate and take in account the rate of growth inhibition of the affected limb in predicting LLD and calculating timing of surgery. Furthermore, he added three reference lines which represent the amount of growth inhibition to be caused by epiphysiodesis of the proximal tibial and/or distal femoral growth plates (28 % and 37 % of the total growth of the leg respectively). Moseley compared retrospectively the use of straight line graph with Green and Anderson growth remaining method [22] to predict the final LLD at skeletal maturity in 30 patients (Level III evidence). In a group of 23 children who had been followed up for more than 1 year prior to surgery, the mean error in predicting final LLD was 0.6 cm for the straight line graph compared to 0.9 cm with the growth remaining method and the difference was statistically significant. The accuracy of the straight line graph was even more striking with a mean error of 0.6 cm compared to 1.2 cm with the growth remaining method when the growth inhibition rate of the affected side was taken in account.

Paley's Lower Limb Multiplier

Paley and coworkers [24] used the growth data of Anderson et al. [23] and divided the femoral and tibial lengths at skeletal maturity by the femoral and tibial lengths at different ages during growth for each percentile group to get an age and gender specific multiplier (M = Lm/L, where M is the gender- and age-specific multiplier, Lm is the bone or limb length at maturity, and L is the age-specific bone or limb length). Maresh [43] reported growth data of 175 children using roentgenographic measurements of femoral and tibial lengths, ranging in age from birth to skeletal maturity. Those data were included as well to complete the period from birth to one year of age. They further used growth data from 18 additional growth databases, 9 based on radiographic or clinical length measurements and 9 based on anthropological measurements of femoral and tibial bones. The multiplier derived from all those database were similar. Therefore, the multiplier method should be independent of growth percentile, regional, racial, ethnic and generational differences in growth as it represents the percentage of growth remaining [44, 45]. Whatever the race or eventual limb length will be at maturity, 50 % of growth remains in the lower limb at 4 years of age. Other advantages for Paley's method is that it is very useful in very young children and in cases with no available previous radiographs [46]. The main limitation is that it can be used only in patients with Shapiro type I progression pattern [34]. Paley et al. [24] compared the accuracy of their method to Moseley straight line graph in predicting the final actual LLD in two groups of patients, an epiphysiodesis group of 16 patients and a limb lengthening group of 14 patients. The correlation coefficients of comparing the Moseley predictions and the multiplier predictions were excellent >0.9 in both groups. If a threshold of acceptable accuracy was set to ±1.5 cm in the epiphysiodesis group, 5 out of 16 predictions by Moseley method and one multiplier prediction fell out of this threshold (Level III evidence).

Which Growth Prediction Methods Is the Most Accurate?

Evaluation of the accuracy of the different methods used to predict final LLD at skeletal maturity was studied extensively by several authors [44–49].

Lampe et al. [48] studied a group of children (15 boys and 15 girls) with LLD exceeding 2 cm who had 33 eiphysiodeses and the timing was based on Moseley straight line graph (Level IV evidence). Skeletal age was determined depending on hand and wrist radiographs according to Greulich and Pyle atlas [26]. The mean predicted LLD was 5.2 cm (range 3–11 cm) and the mean final LLD was 1.4 cm (range 0–4.3 cm). Eleven patients had failures, eight children had a final LLI exceeding 1.5 cm, two had to be operated on twice, and one patient ended with a final LLI exceeding 1.5 cm despite two surgeries. Influence of skeletal maturation was analyzed in cases with failures and they found that delayed skeletal maturation was responsible for overcorrection in 8 patients (6 patients had overcorrection <1.5 cm and 2 patients >1.5 cm). Contralateral distal femoral epiphysiodesis was performed in one patient with overcorrection >1.5 cm. Failures due to delay in skeletal maturation don't seem to be preventable as they take place after the surgery. Variations in the determination of skeletal age is another source of error that would affect the appreciation of the stage of skeletal maturation.

Dewaele and Fabry [47] retrospectively compared two groups of patients who had the timing of epiphysiodesis calculated by either Anderson & Green tables (Group A 47 patients) or by Moseley straight line graph (Group B 36 patients) (Level III evidence). Skeletal age was assessed using hand and wrist radiographs with the help of Greulich and Pyle atlas. In group A, 24/47 patients (51 %) had satisfactory results (final LLD <1.5 cm) while 23/36 (64 %) in group B. Six patients in group A and 2 in group B had overcorrection. Fifteen patients had poor results due to mistakes in estimation of the bone age (7/23 and 8/13 for group A and B respectively), while in 9 patients, unpredictable change in the growth rate caused poor results (7/23 and 2/13). Two patients had contralateral epiphysoidesis for overcorrection >1.5 cm. They agreed with Lamp et al. [48] that the assessment of the status of skeletal maturity is a major source of error and the main reason was due to difficulties with the determination of bone age.

In another retrospective comparative study by **Little et al.** [49], eight variations of three methods (Anderson and Green method, Moseley straight line graph and Menelaus rule of thumb) were used for the prediction of post-epiphysiodesis final LLD in 71 patients (42 girls and 29 boys) (Level III evidence). Greulich and Pyle atlas was used for the determination of the skeletal age. The variations used were (Table 46.4):

The computer generated straight line graph was the only method found to be significantly less accurate in predicting final LLD following epiphysidesis with 27 % of the calculations deviated >2 cm and 39 % >1.5 cm. For all other methods used excluding the computer generated straight line graph, the percentage of calculation errors >2 cm in predicting final LLD ranged from 10 % to 20 % of the patients while if errors >1.5 cm were considered, the range was 18–37 %. The accuracy of predicting the length of the untreated limb at maturity using Moseley method was not acceptable. Moseley predictions deviated >2 cm (range −9.1 to 10.9 cm) in 52 % of the patients when hand charts were used and in 61 % of the cases with the computer based graphs. The conclusion was that, all current methods of predicting the timing of epiphysiodesis and the final LLD have similar and limited accuracy and that there was no significant difference between methods relying on chronological age compared to those based on skeletal age. Furthermore, the main shortcomings of the Moseley method in predicting the final limb length at skeletal maturity were: (1) the data used to construct the straight line graph was derived from the second growth study by Anderson et al. [23] in which they reported femoral and tibial lengths in relation to chronological age. Therefore, we are actually plotting skeletal age data on a chronological age database which might be a source of error. (2) the nomogram used for the determination of the standard deviation of the limb growth (the growth percentile) was derived by adding the standard deviations of the femoral and tibial lengths reported by Anderson et al. [23] (as the original Green and Anderson database is not available) which is not statistically valid.

In a retrospective study by **Lee et al.** [46], they compared the accuracy of the original Green and Anderson growth remaining method to Moseley straight line graph and to the multiplier method in predicting the timing and the results of epiphysiodesis (Level III evidence). Forty-four patients were included in this study (27 boys and 17 girls). Preoperatively, a modified variant from Green and Anderson growth remaining method was used to plan the timing of epiphysiodesis in all patients. The pre-operative LLD was assumed to be the expected LLD at skeletal maturity without surgery. The pattern of growth inhibition of the affected limb (Shapiro type) was not taken in account as well as the SD of the height (the growth percentile). Instead, the timing of epiphysiodesis was calculated using the bone age and the Green and Anderson growth remaining chart (with the average value of growth remaining). Then the accuracy in predicting the effect of epiphysiodesis (the final LLD compared to the expected LLD at maturity with surgery) was compared to four other methods assuming that all the five were used preoperatively. In Method 2: Green and Anderson growth remaining method was used as well, however the growth inhibition pattern of the affected side (Shapiro type) and the SD of the height

Table 46.4 The methods that were compared by Little et al.

Anderson and Green growth remaining method	**Method 1:** The growth remaining was read simply as the mean value for skeletal age on the date of surgery (the growth percentile and pattern was not documented)	Mean values (no growth percentile)
	Method 2: as method 1 + calculating the percentage of growth inhibition from previous data and was used for the determination of the final LLD after epiphysiodesis	Mean values + % growth inhibition
	Method 3: The standard deviation of the preoperative limb length for age was determined and used on the Anderson and Green chart to predict outcome (the growth percentile was taken into account)	Standard deviation (SD)
	Method 4: as method 3 + adding the percentage of growth inhibition to the calculations	SD + % growth inhibition
Menelaus calculation	**Method 5:** The Menelaus calculation was applied with the assumption that the distal femoral physis contribute by 1.0 cm/year to growth, and the proximal tibia 0.6 cm/year	Multiple points of time used for the calculation
	Method 6: A simpler version of this method, ignoring all previous data and growth patterns, was also calculated. This was to assess the accuracy of the method if the patients had been seen late	Single direct preoperative point of time
Moseley straight line graph	**Method 7:** The Moseley straight-line graph was generated and the reference slope was moved to the date of surgery, predicting the outcome in terms of the final discrepancy at maturity after epiphysiodesis. The predicted length of the untreated limb at maturity was also recorded	Hand chart
	Method 8: A computer program for plotting the straight-line graph was used	Computer program

Table 46.5 The summary of the methods used by Lee et al.

Method	SD height	Growth remaining of the long leg	Age	Shapiro type
Modified Green and Anderson growth remaining method	Not applied	Not calculated (assumed the preoperative LLD is the LLD at maturity without surgery)	Bone age	Not applied
Original Green and Anderson method	Applied	Calculated	Bone age	Applied
Paley's multiplier	Unnecessary	Unnecessary	Bone age	Applied
Paley's multiplier	Unnecessary	Unnecessary	Chronological age	Applied
Moseley straight line graph	Unnecessary	Unnecessary	Bone age	Applied

(growth percentile) were taken in account. In methods 3 and 4: Paley's multiplier was used based on the bone age or the chronologic age respectively while in method 5, Moseley straight line graph was used (Table 46.5).

Regarding the growth inhibition rate and pattern, the classification described by Shapiro was used [34] and there were 22 type I, 13 type II, 9 type III and no type IV or V. For Shapiro type I, when using method 2, the growth inhibition rate was calculated and then the growth remaining of the long leg was calculated by Green and Anderson growth chart and the expected LLD at maturity without surgery and the timing of epiphysiodesis were calculated. With Paley's multiplier method, patients with Shapiro type I were divided into either congenital or developmental as originally described by Paley et al. [24]. In case of Shapiro types II or III patients, the LLD at the deceleration or plateau phase was assumed to be the expected LLD at maturity without surgery. Interesting results were reported by this study. In 5 patients (11.4 %), the final LLD at maturity with surgery was > 2 m. There was statistically significant difference between the expected LLD at maturity with surgery and the final LLD for all five methods, however, method 2 (the original Green and Anderson

growth remaining method) was the most accurate and method 4 (Paley's multiplier with the use of the chronologic age) was the least accurate. Furthermore, the greatest correlation coefficient between the expected LLD at maturity with surgery and the final LLD was for method 2. The main reported disadvantage for Green and Anderson growth remaining method is that it uses only the most recent assessment of bone age for the determination of the timing of epiphysiodesis (single assessment). Furthermore, concerns about racial and generational differences do exist. For Moseley's straight line graph, it assumes that the growth inhibition pattern is always linear (Shapiro type I) and therefore, other growth inhibition patterns cannot be plotted correctly and would be forced into a straight line introducing an inevitable error.

Aguilar et al. [44, 45] published 2-parts work trying to validate the accuracy of their multiplier method compared to both the original Anderson et al. [23] growth charts and Moseley's straight line graph [39] (Level III evidence). They used in their studies the data reported by Little et al. [49]. In the first part of their work, they studied the accuracy of those different methods in predicting individual non-epiphysiodesed bones (femora and tibiae) and the short limb lengths at skeletal maturity using radiographs of 60 patients treated for LLD. Greulich and Pyle atlas was used for the determination of skeletal age. The average error and standard deviation in predicting the length of the short limb (femur + tibia) at maturity was 2.5 ± 1.6 cm (95 % CI 6.3 cm) and 2.7 ± 1.8 cm (95 % CI 7.1 cm) for the multiplier method using chronologic or skeletal age respectively, 2.8 ± 2.3 cm (95 % CI 9.1 cm) and 2.6 ± 1.6 cm (95 % CI 6.3 cm) for Anderson method and Moseley's graph respectively. The multiplier method using chronologic age over-predicted 58 % of the maturity lengths of the normal non-epiphysiodesed and short bones (mean 1.1 cm; range 0.1–8 cm), under-predicted 39 % (mean 1.1 cm; range 0.1–4.2 cm), and exactly predicted 3 %. The accuracy of the multiplier method varied by chronologic age. The highest prediction error was at 5 years or younger for both boys and girls. The lowest error was at age 9 years for boys and 7 years for girls. Another noticeable increase in error occurred during the adolescent period for both sexes. Therefore, the best time to predict limb lengths is between the ages of 7 and 9 years. This is consistent with the findings of Anderson et al. [22] who observed greater variability of growth, based on chronologic age, after age 9 years for girls and 12 years for boys, with maximum variability occurring at approximately age 11 years in girls and 13 years in boys. Several factors contribute to this variability as the individual differences in in the extent and the chronologic age of the occurrence of the pubertal growth spurt [22], the deviation between chronologic age and skeletal age which starts around the beginning of the pubertal growth spurt [22, 36] and inaccuracies in the determination of skeletal age [35]. Therefore, the use of skeletal age instead of chronologic age would be recommended or girls who are older than 9 years and boys who are older than 12 years.

In the second part, they compared the multiplier method using the chronologic age and the skeletal age to Moseley' method in predicting post-epiphysiodesis LLD and the epiphysiodesed limb length at maturity. The average error in predicting the epiphysiodesed limb length at maturity was exactly the same among the three methods, 1.6 ± 1.2 cm (95 % CI 4.7 cm). The average error in predicting the final LLD at maturity following epiphysiodesis was 0.9 ± 0.69 cm (95 % CI 2.6 cm) and 1 ± 0.72 cm (95 % CI 2.7 cm) for the multiplier method using chronologic and skeletal age respectively and 1.3 ± 0.93 cm (95 % CI 3.6 cm) for Moseley's method.

Recommendations

The evidence for the use of any growth prediction method compared to the other is still very limited and inconsistent. Additional research work with higher levels of evidence and superior methodology is required to compare different methods used to study the future growth of the lower extremities. Furthermore, the determination of the skeletal maturation stage and the skeletal age seems to be the most conflicting and the major source of error. The interobserver reliability of using Greulich and Pyle atlas seems to be questionable [27, 35]. This reliability might be increased by the use of a modified short version, the shorthand bone age assessment method, which determines a single radiographic criterion from Greulich and Pyle atlas for males between the ages of 12.5 to 16 years and females from 10 to 16 years [29]. The interobserver agreement seems to be higher with the use of Sauvegrain's method based on elbow radiographs compared to Greulich and Pyle method and it allows assessment of the skeletal age in 6-months interval during the pubertal growth spurt which is advantageous.

Growth curves are not linear in particular during the pubertal growth spurt period in which physeal growth arrest procedure are usually indicated [22, 25]. Moreover, growth percentile might change during the rapid phase of the pubertal growth spurt. This is mostly due to the inherent inaccuracies and difficulties in the determination of the stage of skeletal maturity and due to the presence of great individual variability of growth rates during this interesting phase [22, 23]. Therefore, it might be more accurate and consistent to assess the individual relative size or growth percentile and to predict the final limb length at maturity during the steady phase of growth between the age of 7–9 years [25, 45].

Finally, racial, ethnic and generational differences do exist in skeletal growth and should be known and taken in consideration [36, 39, 40]. More research is needed

Table 46.6 Summary of recommendations

Statement		LoE/grades of recommendation	References
1.	Significant variations were found using Greulich – Pyle skeletal atlas, with 50 % of the children were assigned a skeletal age that differed by >1 year between radiologists and 10 % more than 2 years	III/C	[35]
2.	The interobserver agreement seems to be higher with Sauvegrain method compared to the use of Greulich and Pyle atlas which might be due to the clearer landmarks	II/C	[27]
3.	The shorthand bone age assessment method has strong correlation to the Greulich and Pyle atlas with good interobserver and intraobserver correlation	III/C	[29]
4.	The accuracy of different methods used for predicting the LLD at maturity are limited mainly due to difficulties with the assessment of skeletal maturation and difficulties and errors in bone age determination	III/C	[47, 48]
5.	There are inconsistent findings regarding the use of skeletal age in predicting the growth of the limbs and if increases the accuracy of the current methods or not	III/I	[44, 45, 49]
6.	It might be more accurate and consistent to determine the growth percentile and predict future limb length during the steady phase of skeletal growth (age 6–9 years)	III/C	[45]

incorporating growth data from different racial and ethnic backgrounds to refine the current growth charts and tables. On the other hand, factors like illness, nutrition, level of activity and socioeconomic level might change the growth percentile and should be studied and taken in account [36] (Table 46.6).

References

1. Shapiro F. Pediatric orthopedic deformities. Cham: Springer; 2016.
2. Phemister DP. Operative arrestment of longitudinal growth of bones in the treatment of deformities. J Bone Joint Surg Am. 1933;15(1):1–15.
3. Green WT, Anderson M. Epiphyseal arrest for the correction of discrepancies in length of the lower extremities. J Bone Joint Surg Am. 1957;39-A(4):853–72 ; discussion, 872; passim.
4. Bowen JR, Johnson WJ. Percutaneous epiphysiodesis. Clin Orthop Relat Res. 1984;190:170–3.
5. Metaizeau JP et al. Percutaneous epiphysiodesis using transphyseal screws (PETS). J Pediatr Orthop. 1998;18(3):363–9.
6. Blount WP, Clarke GR. Control of bone growth by epiphyseal stapling; a preliminary report. J Bone Joint Surg Am. 1949; 31A(3):464–78.
7. Stevens PM. Guided growth for angular correction: a preliminary series using a tension band plate. J Pediatr Orthop. 2007;27(3): 253–9.
8. Ilizarov GA. The tension-stress effect on the genesis and growth of tissues: part II. The influence of the rate and frequency of distraction. Clin Orthop Relat Res. 1989;239:263–85.
9. Ilizarov GA. The tension-stress effect on the genesis and growth of tissues. Part I. The influence of stability of fixation and soft-tissue preservation. Clin Orthop Relat Res. 1989;238:249–81.
10. Ilizarov GA. Clinical application of the tension-stress effect for limb lengthening. Clin Orthop Relat Res. 1990;250:8–26.
11. De Bastiani G et al. Limb lengthening by callus distraction (callotasis). J Pediatr Orthop. 1987;7(2):129–34.
12. Wagner H. Operative lengthening of the femur. Clin Orthop Relat Res. 1978;136:125–42.
13. Paley D. Problems, obstacles, and complications of limb lengthening by the Ilizarov technique. Clin Orthop Relat Res. 1990; 250:81–104.
14. Paley D et al. Femoral lengthening over an intramedullary nail. A matched-case comparison with Ilizarov femoral lengthening. J Bone Joint Surg Am. 1997;79(10):1464–80.
15. Naudie D et al. Complications of limb-lengthening in children who have an underlying bone disorder. J Bone Joint Surg Am. 1998;80(1):18–24.
16. Baumgart R, Betz A, Schweiberer L. A fully implantable motorized intramedullary nail for limb lengthening and bone transport. Clin Orthop Relat Res. 1997;343:135–43.
17. Guichet JM, Casar RS. Mechanical characterization of a totally intramedullary gradual elongation nail. Clin Orthop Relat Res. 1997;337:281–90.
18. Cole JD et al. The intramedullary skeletal kinetic distractor (ISKD): first clinical results of a new intramedullary nail for lengthening of the femur and tibia. Injury. 2001;32(Suppl 4):SD129–39.
19. Paley D. PRECICE intramedullary limb lengthening system. Expert Rev Med Devices. 2015;12(3):231–49.
20. Kenawey M et al. Insufficient bone regenerate after intramedullary femoral lengthening: risk factors and classification system. Clin Orthop Relat Res. 2011;469(1):264–73.

21. Kenawey M et al. Leg lengthening using intramedullay skeletal kinetic distractor: results of 57 consecutive applications. Injury. 2011;42(2):150–5.
22. Anderson M, Green WT, Messner MB. Growth and predictions of growth in the lower extremities. J Bone Joint Surg Am. 1963;45-A:1–14.
23. Anderson M, Messner MB, Green WT. Distribution of lengths of the normal femur and tibia in children from one to eighteen years of age. J Bone Joint Surg Am. 1964;46:1197–202.
24. Paley D et al. Multiplier method for predicting limb-length discrepancy. J Bone Joint Surg Am. 2000;82-A(10):1432–46.
25. Dimeglio A. Growth in pediatric orthopaedics. J Pediatr Orthop. 2001;21(4):549–55.
26. Greulich WW, Pyle SI. Radiographic atlas of skeletal development of the hand and wrist. 2nd ed. Stanford: Stanford University Press; 1999.
27. Dimeglio A et al. Accuracy of the Sauvegrain method in determining skeletal age during puberty. J Bone Joint Surg Am. 2005;87(8):1689–96.
28. Sauvegrain J, Nahum H, Bronstein H. Study of bone maturation of the elbow. Ann Radiol (Paris). 1962;5:542–50.
29. Heyworth BE et al. The shorthand bone age assessment: a simpler alternative to current methods. J Pediatr Orthop. 2013;33(5): 569–74.
30. Tanner JM et al. Assessment of skeletal maturity and prediction of adult height (TW2 method). 3rd ed. London: Academic; 1983.
31. Tanner JM et al. Assessment of skeletal maturity and prediction of adult height (TW3 method). 3rd ed. London: W.B Saunders; 2001.
32. Marshall WA, Tanner JM. Variations in pattern of pubertal changes in girls. Arch Dis Child. 1969;44(235):291–303.
33. Marshall WA, Tanner JM. Variations in the pattern of pubertal changes in boys. Arch Dis Child. 1970;45(239):13–23.
34. Shapiro F. Developmental patterns in lower-extremity length discrepancies. J Bone Joint Surg Am. 1982;64(5):639–51.
35. Cundy P et al. Skeletal age estimation in leg length discrepancy. J Pediatr Orthop. 1988;8(5):513–5.
36. Beumer A et al. The straight line graph in limb length inequality. A new design based on 182 Dutch children. Acta Orthop Scand. 1997;68(4):355–60.
37. Pritchett JW, Bortel DT. Single bone straight line graphs for the lower extremity. Clin Orthop Relat Res. 1997;342:132–40.
38. Stuart HC, Reed RB. Longitudinal studies of child health and development – series II. Description of project. Pediatrics. 1959;24:875–85.
39. Moseley CF. A straight-line graph for leg-length discrepancies. J Bone Joint Surg Am. 1977;59(2):174–9.
40. Ha JH et al. Distribution of lengths of the normal femur and tibia in Korean children from three to sixteen years of age. J Korean Med Sci. 2003;18(5):715–21.
41. Menelaus MB. Correction of leg length discrepancy by epiphysial arrest. J Bone Joint Surg Br. 1966;48(2):336–9.
42. White J, Stubbins S. Growth arrest for equalizing leg lengths. JAMA. 1944;126:1146.
43. Maresh MM. Linear growth of long bones of extremities from infancy through adolescence; continuing studies. AMA Am J Dis Child. 1955;89(6):725–42.
44. Aguilar JA et al. Clinical validation of the multiplier method for predicting limb length discrepancy and outcome of epiphysiodesis, part II. J Pediatr Orthop. 2005;25(2):192–6.
45. Aguilar JA et al. Clinical validation of the multiplier method for predicting limb length at maturity, part I. J Pediatr Orthop. 2005;25(2):186–91.
46. Lee SC et al. The accuracy of current methods in determining the timing of epiphysiodesis. Bone Joint J. 2013;95-B(7):993–1000.
47. Dewaele J, Fabry G. The timing of epiphysiodesis. A comparative study between the use of the method of Anderson and Green and the Moseley chart. Acta Orthop Belg. 1992;58(1):43–7.
48. Lampe HI, Swierstra BA, Diepstraten AF. Timing of physiodesis in limb length inequality. The straight line graph applied in 30 patients. Acta Orthop Scand. 1992;63(6):672–4.
49. Little DG, Nigo L, Aiona MD. Deficiencies of current methods for the timing of epiphysiodesis. J Pediatr Orthop. 1996;16(2):173–9.

Evidence-Based Treatment of Accessory Navicular Bone

47

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Abstract

Accessory navicular bone is a normal variant which can cause symptoms. Various operative and non operative treatments are used to improve symptoms. There is lack of good quality published literature on either non-operative or operative management. It is generally agreed that non-operative management in the form of symptomatic control, orthoses and physiotherapy is the first line of treatment method. Surgical options include excision of the accessory navicular bone, excision with posterior tibialis tendon reconstruction, arthrodesis of the accessory to the anatomical navicular and percutaneous drilling. Flat foot deformity should be assessed because of its potential role in the development of symptoms and need to be managed together with the accessory navicular bone.

Keywords

Accessory navicular • Accessory tarsal navicular • Prehallux • Accessory scaphoid • Os tibiale externum • Os naviculare secundarium • Navicular secundum • Adolescent

Background

Accessory navicular bone is a normal anatomic variant usually located medial and plantar in relation to the anatomical navicular bone. The navicular bone is the last tarsal bone to ossify, occurring between the age 1–3 year in girls and 3–5 year in boys. The accessory navicular bone ossifies even later. A proportion persists through adult life [1].

In the modern English literature, accessory navicular bone is further divided into three types according to location and relationship with the navicular bone. Type 1 is a small round ossicle within the substance of the posterior tibialis tendon, Type 2 is larger triangular shaped and connected to the navicular by a cartilaginous or fibrocartilaginous synchondrosis whereas Type 3 is a cornuate shaped navicular following the fusion between the accessory and the anatomical navicular bones (Fig. 47.1).

The incidence and frequency of types varies according to geographical and age group population studies. Corkun et al. found 11 % of 650 Turkish adult displayed radiographic appearance of accessory navicular bone with similar distribution within the three types (33 %, 31 % and 46 % respectively) [2].

In a study of 148 patients younger than 18 year old with accessory navicular bone in Korea, there were more patients exhibiting Type 2 variant (76 % vs 15 % Type 1, 9 % Type 3) and 87 % of patients had bilateral accessory navicular bone [3].

Why Does It Become a Problem?

There are arguments for a traumatic origin with repetitive chronic stress. Histological examination showed areas of microfracture with acute and chronic inflammation and tissue cellular proliferation around the synchondrosis [4]. In this case, the accessory navicular bone is acting as an irritant. On the other hand, there are also proponents of an inbuilt anatomical anomaly or abnormal posterior tibialis

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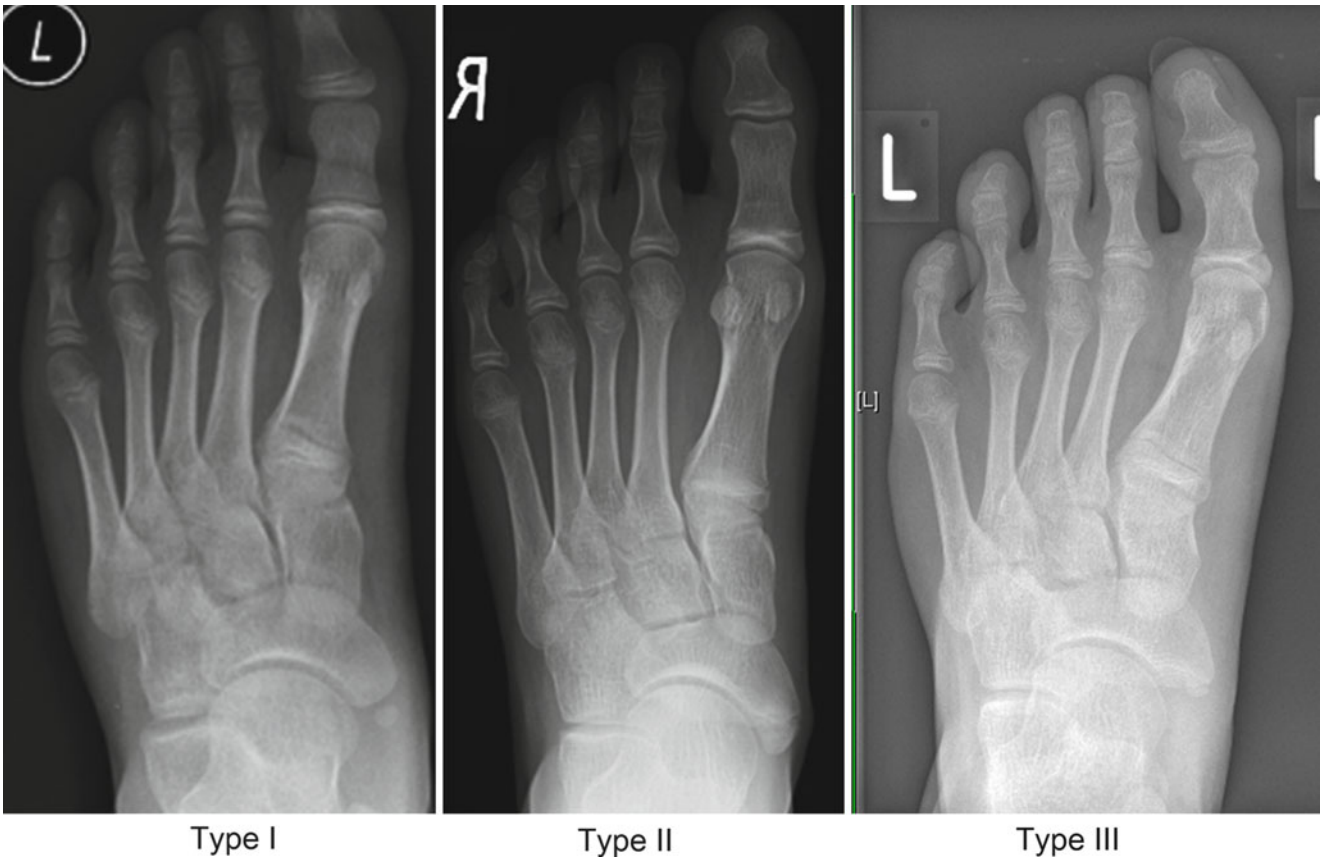

Fig. 47.1 Types of accessory navicular bone

tendon insertion with abnormal tissue between the accessory and navicular bones [5].

Accessory navicular bone can become symptomatic with or without trauma [6, 7]. Pain is usually over the enlarged area of accessory navicular on the medial aspect of foot just at the insertion of posterior tibialis tendon. Tight shoes, walking and exercise exacerbate pain. There is increased pain with resisted inversion of the foot.

External oblique (medial to lateral) plain radiograph complements the dorsoplantar view in diagnosing the accessory navicular bone. Magnetic resonance imaging is sensitive in showing marrow oedema in symptomatic adolescents. The marrow oedema also diminishes following the relief of symptoms after non-operative management [8]. Technetium bone scan is sensitive in showing increased tracer uptake but not specific because half of asymptomatic patients demonstrate the similar features of symptomatic patients [9].

How to Treat Symptomatic Accessory Navicular Bone

Non-operative Management

Non-operative management including symptomatic management in the form of soft pads between the foot and sole of shoe, footwear modification, physiotherapy, orthoses to offload midfoot and oral anti-inflammatory can be effective even for active adolescent [10–12]. Non-operative treatments are usually individualised according to patient and provider factors and there is no known literature on the most effective or widely agreed non-operative protocol or comparison against operative treatment. Most authors tried at least three months of non-operative management before proceeding with surgery [6, 7, 12–17].

Injection

We could not find published English literature using digital search engines on the topic of efficacy of injection in the management of symptomatic accessory navicular bone.

Surgery

Surgery aims to improve pain by removing the accessory bone or stabilising the synchondrosis and protecting the posterior tibialis tendon. Most common accessory navicular bone requiring surgery was Type 2. Table 47.1 summarises the references discussed below.

Excision

Bennett et al. recommended excision surgery with repair of the posterior tibialis tendon without advancement due to its simplicity and low rate of complication [18] (Fig. 47.2).

Table 47.1 Surgical treatment of symptomatic accessory navicular bone

Studies	Study type	No. of patients	Age (Y)	Accessory navicular type	Onset to surgery (month)	Preop mx duration (month)	Surgery	Follow up	Outcomes	Notes
Bennett 1990	n/a	50 (38F:12M) 75 ft (56F:19M)	12.6 (8–18)	2 (77 %) 3	n/a	>6	Excision of accessory bone or prominent cornuate navicular + postop cast 2 w in equinovarus	2–22 y	77 % 'excellent' 16 % 'good' 4 % 'fair' 3 % 'poor' All females able to wear high heels	20 FFF 8 % residual prominence, 1 % neuroma, 5 % superficial inflammation, 7 % scar problems
Kiter 2000	n/a	17 (13F: 4M)	14–36	2	Chronic	n/a	Excision and rasping of remaining bone + postop cast 2 w	3 y (2–5 y)	11 patient no pain, no restriction of activity, no shoewear modification	Inability to single-heel raise persist postop in flatfoot
Jasiewicz 2008	n/a	22 (17F: 5M) 34 ft	14 (9–22)	1, 2 (50 %), 3	n/a	3	Excision +/– PTT reattachment + supinating insoles 4 w	5.6 y (1–13 y) 1 lost to f/u	Improved VAS in all except 1 pt. unchanged in pain and shoewear	Single heel rise test positive pre and post in all cases 2 prolonged wound healing
Prichasuk 1999	n/a	28 (20F: 8M) 31 ft	23 (11–38)	2	n/a	>6	Kidner + cast 6 w + insoles 6 m	3.2 y (1–14 y)	27 pts 'good' results	3/25 FFF improved arch
Lee 2012	Retrospective	41 (8F: 33M) 50 ft	20 (8–48)	2	n/a	6, (7 athletes straight to surgery)	Modified Kidner +cast 6 w	5 y	Improved AOFAS midfoot	
Macnicol 1984	Retrospective	47 62 ft	10–50	n/a	33 (3 m–19 y)	n/a	Kidner (bilateral), n = 26 + postop cast 6 w, **vs.** excision, n = 21 Both groups have FF	Kidner 10 y (3–19 y) Excision 12 y (3–20 y)	Kidner – 19/22 FFF pain relieved, 14 shape improve Excision – in those aged 11–12, 15 with normal feet pain free, 4/5 in FFF painfree	4 recurrent AN in cases without trimming of navicular tuberosity with 1 persistent pain Kidner patient also report protracted medial pain postop
Cha 2013	Prospective, non randomised	50 (29F:21M) 50 ft	9–15	Unilateral Type 2 +FFF	9.3–10.4	>6	Excision, n = 25 + supinating insoles **vs.** Kidner, n = 25 + casting	36–52 m	Improved AOFAS midfoot scale and VAS, both groups not statistically different	Similar restoration of medial longitudinal arch in two groups

(continued)

Table 47.1 (continued)

Studies	Study type	No. of patients	Age (Y)	Accessory navicular type	Onset to surgery (month)	Preop mx duration (month)	Surgery	Follow up	Outcomes	Notes
Pretell-mazzini 2014	Retrospective	27 (22F: 5M) 32 ft	13.2 (9–16)	1, 2 (75 %), 3	10 (2–36)	n/a	Excision, n = 14 **vs**. Kidner, n = 18 (both postop cast 4–6 w) Similar distribution FF both groups	n/a	87.5 % (93 % excision, 83 % Kidner) Good-to-excellent function (no significant difference between 2 groups)	1 tendon Achilles lengthening and lateral column lengthening More complications (painful scar, tendinitis) and reoperation after tendon advancement, but not statistically significant
Scott 2009	Prospective, non-randomised	20 (9F: 11M)	25 (10–52)	2	n/a	n/a	Fusion screw, n = 10 **vs.** modified Kidner, n = 10	35 m (21–71 m) **vs.** 48 m (24–68 m)	Improved final AOFAS midfoot but no significant difference between groups	Corrective surgery performed for significant FF Fusion – 1 removal of screw Worse outcomes in progressive FF
Nakayama 2005	n/a	29 (14F: 15M) 31 ft	10–18	2	4–30	n/a	Percutaneous drillings + postop cast 3 w	32 m (24–37 m)	79 % (23 patient) returned to sports within 3 m, none worse. 18 (feet) union, 13 non-union (all union returned to sports within 3 m)	1 patient with further surgery for severe planovalgus
Garras 2012	Retrospective	20 (14F: 6M) 23 flatfeet	10–27	2	n/a	>6	Subtalar arthroereisis + modified Kidner + cast 6 w	54 m (24–92 m)	Improved AOFAS hindfoot score and VAS	3 removal of implant for impingement
Kim 2014	n/a	13 patients (4F: 9M) 21 ft	10–16	2	n/a	n/a	Calcaneo-cuboid-cuneiform osteotomy + modified Kidner (+ TA lengthening in 4 cases) + postop cast 6 w	12–36 m	ACFAS score from 26.1 to 44.8, improvement in 3 components; pain, appearance, function	

FF flatfeet, *FFF* flexible flatfeet, *PTT* posterior tibialis tendon, *w* weeks, *m* months, *AN* accessory navicular

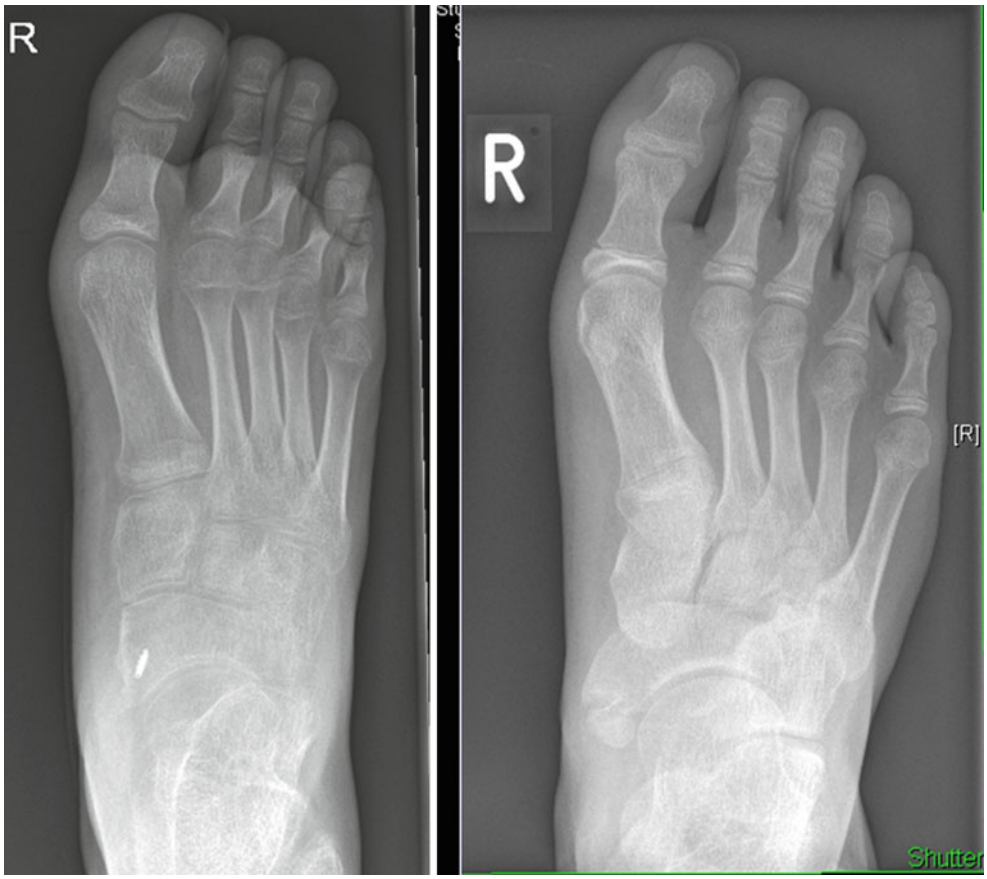

Fig. 47.2 Excision and reattachment of accessory navicular bone

They observed that 77 % of patients reported 'excellent' outcomes after an average 12 years (range from 2 to 22 years). This was subjective patient rating of having 'painless feet and had no shoeware problems'. Seven percent of the patients reported less than good outcome; experiencing 'mild foot pain with activity but not restricting activity plus or minus shoeware modification' or 'moderate foot pain restricting activity plus or minus shoeware modification'.

Kiter et al. reported on the outcome of excision of the accessory bone plus rasping of the remaining bone in patients aged 14–36 year old [12]. After a mean of three years (range 2–5), 11 out of 17 reported no pain, no restriction to activity and no shoewear modification. Excision resulted in improvement of pain and footwear, but it was noticed that patients with flatfoot and not able to perform single-heel rise test before the surgery still could not perform the test after surgery [12, 16]. This may be due to the older population in their studies. Following this observation, Kiter et al. suggested that excision alone is unwise in patient flatfoot [12].

Kidner Procedure

Kidner procedure involved shelling out of the accessory navicular bone and release of posterior tibialis tendon insertion with a thin layer of bone which is then reattached to the undersurface of the navicular body [19]. Modifications of the technique of tendon release and fixation is recognised. Patients were immobilised in below knee cast following this procedure for 4–6 weeks [7, 13–15, 17, 19, 20]. Series of patients undergoing excision of accessory navicular and reattachment of posterior tibialis tendon reported 'good' results and improved AOFAS midfoot scores [15, 17]. Despite reattachment of the tendon, Prichasuk and Sinphurmsukskul only observed that three out of 25 patients with flexible flatfoot had improved arch after the surgery [17]. Similar to some reports, their patients included patients of older aged group [12, 16].

Excision vs. Kidner Procedure

Macnicol and Voutsinas reported positive outcomes in patients with symptomatic accessory navicular undergoing Kidner procedure or simply excision [7]. Both groups of patients experienced improvement in pain. In contrast to other more recent reports [12, 16, 17], 14 of 26 flatfeet improved in shape following Kidner procedure. However, there were more complaints of protracted medial pain postoperatively after Kidner procedure [7].

There were improvements in study methodology in the recent years. In a prospective non-randomised comparison of 25 consecutive excisions with postoperative insoles and 25 consecutive Kidner procedures with postoperative casting,

Cha et al. reported improvement in both AOFAS midfoot scores and Visual Analogue Scale for pain with no statistical significance between both groups [13]. They also reported similar rate of restoration of medial longitudinal arch in both groups.

In another retrospective study, Pretell-Mazzini et al. reported no statistically significant difference in the subjective reported outcomes between patients undergoing excision (93 % good-to-excellent outcome) or Kidner procedure (83 % good-to-excellent outcome) [20]. They also reported more complications in patients undergoing Kidner procedure namely painful scar and tendinitis. There were four reoperations for painful scar, three of which following Kidner procedure.

Table 47.2 Table of recommendation

Statement	Grade of recommendation
First line treatment for symptomatic accessory navicular bone in paediatric patients is non-operative	C
Associated flatfoot deformity is a predictor of less favourable outcome with surgery and may require treatment as well	I
Excision is the preferred surgical option for symptomatic accessory navicular	B
Percutaneous drilling is an effective, least invasive surgery for symptomatic Type 2 accessory navicular	I

Arthrodesis

Scott et al. prospectively evaluated 20 patients undergoing fusion of the accessory navicular using 3.5 mm cannulated screw [19]. The surgical technique was changed to a modified Kidner procedure after 10 patients due to technical difficulty where the large size of the metalwork split the accessory bone. Comparison of the two groups of surgical technique showed improvement in the final AOFAS midfoot scores but not statistically different. They noted three cases of progressive loss of the medial longitudinal arch with recalcitrant medial midfoot pain in the Kidner group.

Percutaneous Drilling

Nakayama et al. experienced non-union and metalware complications after attempted fusion using screw [6]. Hence, they performed percutaneous drilling under radiological guidance. A 1.0 mm K-wire was introduced from posterior prominence on the accessory navicular to the primary navicular through the synchondrosis at five to seven points. The foot was then immobilised a below knee cast for 3 weeks. Their 29 subjects consisted of adolescents aged 10–18 and 79 % reported returning to sports within three months. There were 43 % cases reported to be non-union but all reported improvement in symptoms (92 % good to excellent, 8 % fair). No patients reported a worse outcome or complication. One potential disadvantage of this procedure was there may be residual symptom from the prominent bone [21] but which may also not be solved by excision [18, 22].

Accessory Navicular and Flatfoot

A patient with flatfoot and symptomatic accessory navicular bone can present challenge to treatment, partly due to incomplete understanding of the cause and effect relationship between these two Phenomena. In the adolescence, non-operative management would aim to correct the flatfoot with symptomatic relief of the accessory navicular in parallel with the natural development of the medial longitudinal arch. In cases of protracted symptoms, Garras et al. retrospectively reported improved AOFAS hindfoot and VAS scores at least 2 years after subtalar arthroereisis performed with modified Kidner procedure in patients with flexible flatfoot aged between 10 year old and 27 year old [14]. In the younger patient group aged 10–16 year old with severe flexible flatfoot, modified Kidner procedure was supplemented with calcaneo-cuboid-cuneiform osteotomy [23]. Post-operative outcomes in pain, appearance and functional capacity were significantly improved at one-year follow-up.

Prognosis

Majority of patient satisfaction at one-year following surgery for symptomatic accessory navicular were favourable in case series reporting on surgical outcomes following a period of non-operative management [6, 7, 12–20, 23]. There had been no demonstrable significant difference in the outcomes between excision surgery and Kidner procedure. However, one need to consider there is no good quality study to support or dispute surgery or non-operative management. Most studies were limited in the small number of cases, long duration of patient recruitment, heterogenous patient characteristics and variations of named procedure.

Common complications following excision or Kidner procedure were residual prominence, scar problems such as pain, superficial wound inflammation and recurrence of accessory navicular [7, 18, 20–22].

Table 47.2 provides a list of recommendations for treatment of accessory navicular bone.

References

1. Zadek I, Gold AM. The accessory tarsal scaphoid. J Bone Joint Surg Am. 1948;30A(4):957–68.
2. Corkun NK, Arican RY, Utuk A, Ozcanli H, Sindel T. The incidence of accessory navicular bone types in Turkish subjects. Surg Radiol Anat. 2009;31:675–9.
3. Park H et al. The relationship between accessory navicular and flat foot. J Pediatr Orthop. 2015;35(7):739–45.
4. Grogan DP, Gasser SI, Ogden JA. The painful accessory navicular: a clinical and histopathological study. Foot Ankle Int. 1989;10(3): 164–9.
5. Kiter E et al. Tibialis posterior tendon abnormalities in feet with accessory navicular bone and flatfoot. Acta Orthop Scand. 1999; 70(6):618–21.
6. Nakayama S. Percutaneous drilling of symptomatic accessory navicular in young athletes. Am J Sports Med. 2005;33(4):531–5.
7. Macnicol MF, Voutsinas S. Surgical treatment of the symptomatic accessory navicular. J Bone Joint Surg Br. 1984;66(2):218–26.
8. Takahashi M et al. Magnetic resonance imaging in adolescent symptomatic navicular tuberosity. J Med Invest. 2014;61(1–2):22–7.
9. Chiu NT et al. Symptomatic and asymptomatic accessory navicular bones: findings of Tc-99 m MDP bone scintigraphy. Clin Radiol. 2000;55(5):353–5.
10. Smith TR. Management of dancers with symptomatic accessory navicular: 2 case reports. J Orthop Sports Phys Ther. 2012;42(5):465–73.
11. Requejo SM, Kulig K, Thordarson DB. Management of foot pain associated with accessory bones of the foot: two clinical case reports. J Orthop Sports Phys Ther. 2000;30(10):580–94.
12. Kiter E et al. Evaluation of simple excision in the treatment of symptomatic accessory navicular associated with flat feet. J Orthop Sci. 2000;5(4):333–5.
13. Cha SM et al. Simple excision vs the Kidner procedure for type 2 accessory navicular associated with flatfoot in pediatric population. Foot Ankle Int. 2013;34(2):167–72.
14. Garras DN et al. Outcome of modified Kidner procedure with subtalar arthroereisis for painful accessory navicular associated with planovalgus deformity. Foot Ankle Int. 2012;33(11):934–9.
15. Lee KT et al. Midterm outcome of modified Kidner procedure. Foot Ankle Int. 2012;33(2):122–7.
16. Jasiewicz B et al. Results of simple excision technique in the surgical treatment of symptomatic accessory navicular bones. Foot Ankle Surg. 2008;14(2):57–61.
17. Prichasuk S, Sinphurmsukskul O. Kidner procedure for symptomatic accessory navicular and its relation to pes planus. Foot Ankle Int. 1995;16(8):500–3.
18. Bennett GL, Weiner DS, Leighley B. Surgical treatment of symptomatic accessory tarsal navicular. J Pediatr Orthop. 1990;10(4): 445–9.
19. Scott AT et al. Fusion versus excision of the symptomatic type II accessory navicular: a prospective study. Foot Ankle Int. 2009; 30(01):10–5.
20. Pretell-Mazzini J et al. Surgical treatment of symptomatic accessory navicular in children and adolescents. Am J Orthop (Belle Mead NJ). 2014;43(3):110–3.
21. Vaughan P, Singh D. Ongoing pain and deformity after an excision of the accessory navicular. Foot Ankle Clin. 2014;19(3):541–53.
22. Ricco AI, Richards BS, Herring JA. Disorders of the foot. In: Herring JA, editor. Tachdjian's pediatric orthopaedics. 5th ed. Philadelphia: Elsevier Inc.; 2014.
23. Kim JR et al. Concomitant calcaneo-cuboid-cuneiform osteotomies and the modified Kidner procedure for severe flatfoot associated with symptomatic accessory navicular in children and adolescents. J Orthop Surg Res. 2014;9:131.

Evidence-Based Treatment of Ingrown Toenails

48

Arif Razak and Mubashshar Ahmad

Abstract

Ingrown toenail is a common condition affecting toenails causing inflammation and pain. It is caused by the edge of the nail growing into the surrounding soft tissue. In this chapter we appraised the evidence that underpin the current approaches and we concluded that surgical treatments are superior to non surgical ones in term of recurrence rates (grade B). The use of chemical ablation reduces the recurrences rates regardless of the surgical interventions (grade B). Wedge excision is the first line of surgical treatment (grade C). Phenol chemical ablation has been the most widely used ablating agent and it should be compared to other agents in more robust studies.

Keywords

Ingrowing toenail • Ingrown toenail • Painful toenail • Infected toenail

Background

Ingrown toenails are a common condition in adolescent and young adults. The nail plate penetrates into the skin causing foreign body inflammation which is presented as redness, pain, and swelling (Fig. 48.1) [1]. The big toe is the most commonly affected [2]. The aetiology is not fully identified and often there is no evident cause. Several causes have been implicated including improper nail trimming, tight fitting shoes, trauma, genetic predisposition and familial causes [3]. The incidence is not known as a large proportion does not come to medical attention. There is no racial or gender predilection and teenage seems to be the commonest age to be involved.

Ingrown toenail can be classified into 3 stages. Stage 1 is characterised by pain and inflammation. Stage 2 is essentially stage 1 with infection. Stage 3 is stage 2 with lateral nail fold hypertrophy [2].

Patients seek treatment when become symptomatic. Treatments options vary depending on the severity of the condition, previous treatment, frequency of recurrence and the healthcare provider expertise. Various surgical and non-surgical treatments have been advocated but there is a lack of evidence-based hierarchical indications for these treatments [4]. General hygienic measures are essential for successful treatment whether surgical or non surgical. They are also important in reducing the risk of infection and recurrence following surgery [5].

What Non-operative Methods Have Been Described in the Literature?

The main principle of non-operative treatments is to prevent any physical contact between the nail edge and its nail fold. This will reduce the pressure in the surrounding soft tissue to allow for the inflammation and pain to settle down.

Cotton wool pledgets or wisps can be placed underneath the ingrown nail edge (Fig. 48.2). Senapati described their technique, which involved cutting the nail straight across, placing a small piece of cotton wool under the free corner of the nail and cauterising the granulation tissue with a silver

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nitrate stick. They quoted a success rate of 79 % at mean of 23.7 weeks follow up [6]. Connolly and Fitzgerald used cotton wool pledgets in children with 72 % success rate at mean of 2.5 years follow up [7].

Toe taping is another alternative, where an elastic adhesive tape is used to pull the affected nail fold away from the lateral nail edge. It may take away the pain instantly. However, the symptoms can easily recur if the taping treatment is discontinued early. The usual length of time for this treatment is around 2 months. The success rate of this technique was reported to be less than 50 % [8, 9]. Tsunoda et al. showed that 265 out of their 541 patients who were treated with taping required additional treatment [9].

Splints such as gutter or resin splints have been described in the literature. They are affixed to the nail edge and this allows the nail edge to grow over the nail fold. Hence, the treatment sometimes takes several months to complete [10]. Matsumoto study quoted resin splint recurrence rate as 8.2 % with average duration of splint placement of 9.3 months [11] (Fig. 48.3).

Shape-memory alloy was recently developed in Korea and reviewed by Park et al. [12] in their short-term study. They claimed that its application is simple. The alloy has a hook at each end, which hold onto the nail edge keeping it away from the nail fold. In 24 patients (31 nails) who were treated with these alloys the recurrence rate for stage 1, 2 and 3 was 22.2 %, 33.3 % and 14.2 % respectively. One patient lost his nail. They claimed that they are simple to apply; they cause no deformity and patients have high satisfaction.

What Operative Methods Have Been Described in the Literature?

Several surgical techniques have been described in the literature to treat ingrown toenails. This indicates that none of these techniques is clearly more successful than others. Newer techniques keep coming and rarely bring substantive progress [5]. Surgical approaches are usually fall in one of three categories:

1. Nail folds optimisation (Debulking of the hypertrophic nail fold and granulation tissue)
2. Nail plate optimisation (including nail matrix ablation)
3. A combination of the above two approaches

The nail fold can be excised without disturbing the nail and its matrix. Several techniques have been described with slight modification regarding the size, the direction of the incision and the amount of soft tissue excised [13–17]. The reported short term results are good however the long term results are often missing. A study in 50 children reported no recurrence with very high patient satisfaction [18]. Potential complications in this technique are bleeding, nail deformity, and excessive granulation tissue. In another study, authors reported excellent short term results in 23 patients who were treated with foldplasty with no recurrences or severe complications over 12 months follow up period (Fig. 48.4).

Simple nail removal, either part or all of the nail plate has a recurrence rate up to 70 % [5, 19, 20]. That is why the nail matrix is normally either excised or chemically destroyed

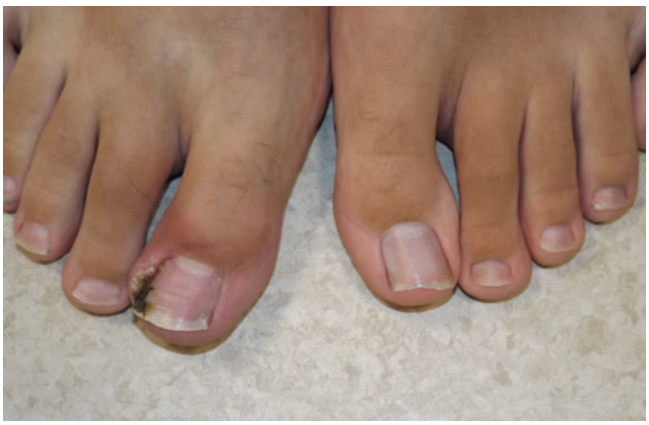

Fig. 48.1 Clinical photograph-ingrown toenail

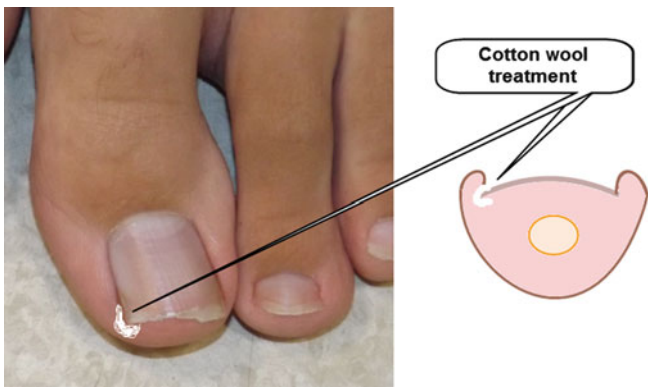

Fig. 48.2 A drawing illustrating the cotton wall treatment

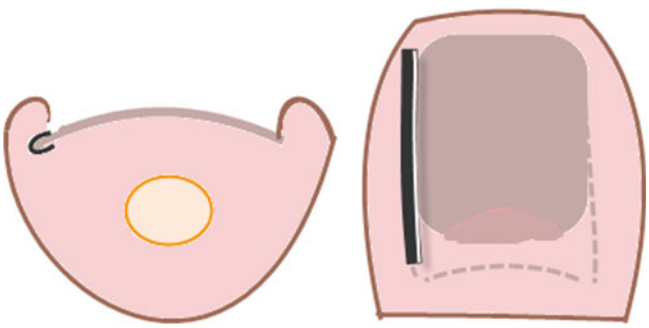

Fig. 48.3 Schematic illustration of gutter splint treatment

with either phenol or sodium hydroxide (corrosive base) as well. This is also known as matricectomy.

Total nail bed and matrix excision (Zadik's operation) [21] has been promoted as the last resort treatment for patients with significant morbidity associated with ingrown toe nail. However, the reported recurrence rates with total nail ablation are high (18–60 %) undermining the reasoning for the indication [22, 23].

Partial excision of the nail and its matrix seems a less invasive intervention. This was first described by Winograd [24] and subsequently underwent several modifications. The most common modification in use is the wedge resection (Fig. 48.5). The nail plate edge is trimmed by about 2 mm and the matrix is destroyed surgically using a curette or blade. The reported recurrence rate varied from 16 % to 30 % [25, 26].

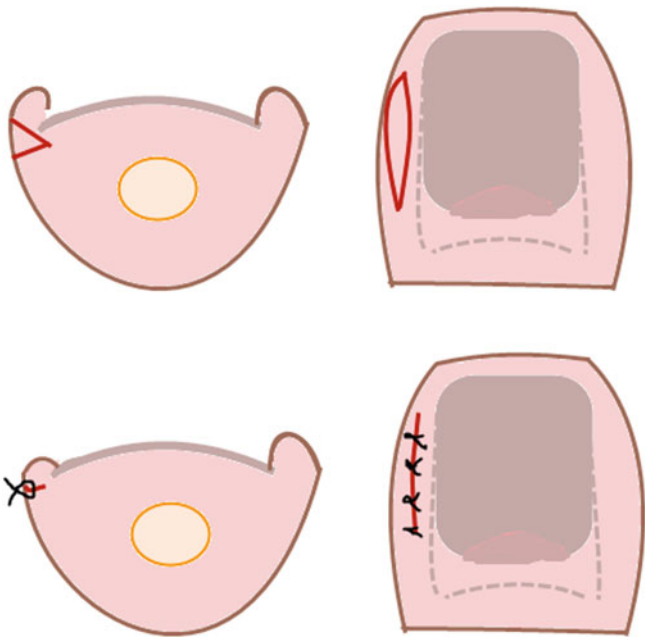

Fig. 48.4 Schematic illustration of a fusiform excision to reduce the hypertrophic nail fold

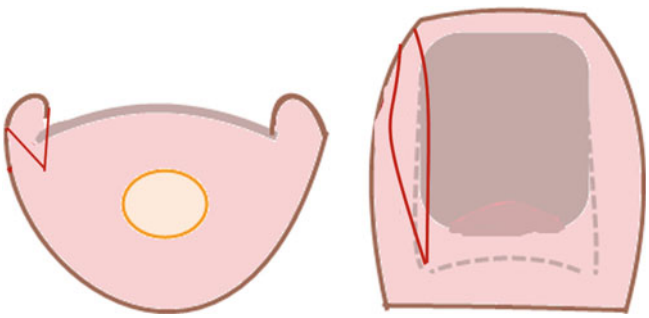

Fig. 48.5 Schematic illustration of wedge excision

Physical and chemical cauterisations have been used to further ensure complete destruction of the matrix and reduce the risk of recurrence. The addition of [27] Phenol has been shown to have better short and long term results compared to matrix surgical excision alone [4, 25, 28–33]. Phenol is widely used in practice compared to sodium hydroxide and both have good comparable results compared to matrix excision only [23, 34]. Bostanci et al. reviewed these two chemical agents in their patients and found that sodium hydroxide causes less postoperative morbidity and provides faster recovery [20]. Cryotherapy and electrocautery are another treatment options for matricectomy with low recurrence rates reported by Küçüktaş [35]. Matrix phenolisation however had reduced healing time compared to matrix electrocautery in a study by Misiak [36].

What Is the Best Treatment for Ingrown Toenail?

In the previous sections we presented the most commonly used treatments (surgical and non surgical) for ingrown toenails with brief description, pros and cons. In a comprehensive systematic review and meta-analysis published by Cochrane library, Eekhof et al. compared the outcomes of these treatments [4]. Twenty-four randomised controlled trials were included with a total of 2826 participants. Five studies investigated various non surgical treatments and 19 studied surgical treatments. Recurrence rate was the commonest outcome used. They found surgical treatments in general were superior to non surgical ones in term of recurrence rates (13 % versus 32 % respectively). The use of chemical ablation seems to reduce the recurrences rates regardless the surgical interventions although the comparatives varied. The crude recurrence rates were 20 % in wedge resection alone. The addition of chemical will reduce the recurrence rate to around 11 %. Moreover, wedge excision achieved similar recurrent rate as total avulsion of the nail [28] but performed better than foldoplasty [37] (Table 48.1).

Table 48.1 Table of recommendations

Statement	Grade of recommendation
Surgical treatments are superior to non surgical ones in term of recurrence rates	B
The use of chemical ablation reduce the recurrences rates regardless the surgical interventions	B
Wedge excision is the first line surgical treatment	C
Phenol chemical ablation is superior to other chemical and physical ablating agents	I

References

1. Aydin N. Partial removal of nail matrix in the treatment of ingrowing toe nail. Acta Orthop Traumatol Turc. 2008;42:174–7.
2. Zuber TJ, Pfenninger JL. Management of ingrown toenails. Am Fam Physician. 1995;52(1):181–90.
3. Reijnen JAM, Goris RJA. Conservative treatment of ingrowing toenails. Br J Surg. 1989;76(9):955–7.
4. Eekhof JAH et al. Interventions for ingrowing toenails. In: Cochrane database of systematic reviews: Wiley-Blackwell; 2012.
5. Haneke E. Controversies in the treatment of ingrown nails. Dermatol Res Pract. 2012;2012:783924.
6. Senapati A. Conservative outpatient management of ingrowing toenails. J R Soc Med. 1986;79(6):339–40.
7. Connolly B, Fitzgerald RJ. Pledgets in ingrowing toenails. Arch Dis Child. 1988;63(1):71–2.
8. Nlshioka K et al. Taping for embedded toenails. Br J Dermatol. 1985;113(2):246–7.
9. Tsunoda M, Tsunoda K. Patient-controlled taping for the treatment of ingrown toenails. Ann Fam Med. 2014;12(6):553–5.
10. Arai H et al. Formable acrylic treatment for ingrowing nail with gutter splint and sculptured nail. Int J Dermatol. 2004;43(10): 759–65.
11. Matsumoto K et al. Resin splint as a new conservative treatment for ingrown toenails. J Med Invest. 2010;57(3, 4):321–5.
12. Park S-W et al. Treatment of ingrown nail with a special device composed of shape-memory alloy. J Dermatol. 2014;41(4):292–5.
13. NoeL B. Surgical treatment of ingrown toenail without matricectomy. Dermatol Surg. 2008;34(1):79–83.
14. NoeL B. Surgical treatment of ingrown toenail without matricectomy. Dermatol Surg. 2007;34(1):79–83.
15. Sarifakioglu E, Yilmaz AE, Gorpelioglu C. Nail alterations in 250 infant patients: a clinical study. J Eur Acad Dermatol Venereol. 2008;22(6):741–4.
16. DuVries HL. Surgery of the Foot, Mosby. St. Louis, Mo, USA: Wiley-Blackwell; 1966.
17. Ney GC. An operation for ingrowing toe nails. JAMA. 1923; 80(6):374.
18. Haricharan RN, Masquijo J, Bettolli M. Nail-fold excision for the treatment of ingrown toenail in children. J Pediatr. 162(2):398–402.
19. Bostanci S, Ekmekçi P, Gürgey E. Chemical matricectomy with phenol for the treatment of ingrowing toenail: a review of the literature and follow-up of 172 treated patients. Acta Derm Venereol. 2001;81(3):181–3.
20. Bostanci S, Kocyigit P, GÃœRgey E. Comparison of phenol and sodium hydroxide chemical matricectomies for the treatment of ingrowing toenails. Dermatol Surg. 2007;33(6):680–5.
21. Zadik FR. Obliteration of the nail bed of the great toe without shortening the terminal phalanx. J Bone Joint Surg. 1950;32(1):66–7.
22. Mitchell S, Jackson CR, Wilson-Storey D. Surgical treatment of ingrown toenails in children: what is best practice? Ann R Coll Surg Engl. 2011;93(2):99–102.
23. Shaath N et al. A prospective randomized comparison of the Zadik procedure and chemical ablation in the treatment of ingrown toenails. Foot Ankle Int. 2005;26(5):401–5.
24. Winograd AM. A modification in the technic of operation for ingrown toenail. JAMA. 1929;92(3):229–30.
25. Ham AC, Hackeng CA, Yo TI. The treatment of ingrowing toenails. A randomised comparison of wedge excision and phenol cauterisation. J Bone Joint Surg Br. 1990;72(3):507–9.
26. Morkane AJ, Robertson RW, Inglis GS. Segmental phenolization of ingrowing toenails: a randomized controlled study. Br J Surg. 1984;71(7):526–7.
27. Küçüktaş M et al. Comparison of effectiveness of electrocautery and cryotherapy in partial matrixectomy after partial nail extraction in the treatment of ingrown nails. Dermatol Surg. 39(2):274–80.
28. Grieg JD et al. The surgical treatment of ingrowing toenails. J Bone Joint Surg Br. 1991;73(1):131–3.
29. Herold N, Houshian S, Riegels-Nielsen P. A prospective comparison of wedge matrix resection with nail matrix phenolization for the treatment of ingrown toenail. J Foot Ankle Surg. 2001;40(6): 390–5.
30. Bos AMC et al. Randomized clinical trial of surgical technique and local antibiotics for ingrowing toenail. Br J Surg. 2007;94(3):292–6.
31. Korkmaz M et al. Teenage patients with ingrown toenails: treatment with partial matrix excision or segmental phenolization. Indian J Dermatol. 2013;58(4):327.
32. Karaca N, Dereli T. Treatment of ingrown toenail with proximolateral matrix partial excision and matrix phenolization. Ann Fam Med. 10(6):556–9.
33. Zaraa I et al. Segmental phenolization for the treatment of ingrown toenails: technique report, follow up of 146 patients, and review of the literature. Dermatol Online J. 2013;19(6):18560.
34. Perez-Rey J, Mediavilla-Saldana L, Martinez-Nova A. Exploring postoperative outcomes for ingrown toenails. NaOH vs wedge resection techniques. Dermatol Surg. 2014;40(3):281–7.
35. Küçüktaş M et al. Comparison of effectiveness of electrocautery and cryotherapy in partial matrixectomy after partial nail extraction in the treatment of ingrown nails. Dermatol Surg. 2013;39(2):274–80.
36. Misiak P et al. Comparison of effectiveness of electrocautery and phenol application in partial matricectomy after partial nail extraction in the treatment of ingrown nails. Pol Przegl Chir. 2014;86(2): 89–93.
37. Perry E et al. A comparison of four nail-conserving procedures for ingrowing toenail. Br J Surg. 1984;71(11):912.

Evidence-Based Treatment of Deformity in Multiple Osteochondromatosis

49

Daniel Porter and Li Fei

Abstract

Multiple Osteochondromatosis is a relatively common genetic orthopaedic condition. Although the molecular basis of inheritance is well established, clinical features are variable. It is known that deformity can occur, however its natural history in relation to the presence of local exostoses is poorly understood. The literature review identifies some features of local deformity that suggest a causal effect however there are no level III studies. The result of exostosis excision has only been studied in the forearm in four case-series and the results are contradictory. In a proportion of patients an improvement may be expected to occur. Optimal timing of surgery in relation to patient age and degree of deformity has not been established. Downsides to excision surgery are chiefly exostosis recurrence and failure to achieve the desired improvement in deformity.

Keywords

Multiple Osteochondromatosis • Deformity • Excision

Introduction

Multiple Osteochondromatosis (MO) (hereditary multiple exostoses, multiple hereditary exostoses, multiple cartilaginous exostoses, diaphyseal aclasis) is an inherited condition affecting 1/50,000 individuals which results in a combination of osteochondromas (characteristic tumours) and growth disturbance (characteristic skeletal dysplasias). In this regard the condition is unique – the most comparable condition; Ollier's disease, is not heritable.

Several theories regarding pathogenesis have been proposed [1]. The genetic and behavioural characteristics of osteochondromas tend to support a neoplastic pathogenesis for the condition. If this were the sole driver of pathology, then growth disturbance would be secondary to osteochondroma development. However growth disturbance can itself is genetically determined (nail-patella syndrome, short stature syndromes, and some congenital limb 'failure of formation' syndromes).

This question is not esoteric, since a purely neoplastic pathogenesis of growth disturbance means that removal of local osteochondromas should remove the stimulus for growth disturbance, hence halting the progress of deformity. On the other hand, a genetic process causing growth disturbance via a wider 'field-change' effect would not be ameliorated by removal of a local osteochondroma (Figs. 49.1 and 49.2).

A field-change effect could cause both an increase in local osteochondroma formation and growth disturbance. Hence local osteochondroma 'burden' might be associated with deformity without invoking 'cause and effect'. A putative 'post-axial' (ulnar and fibular) preference for a 'field-change' effect would result both in greater osteochondroma volume and the genetic effect would then result in more exostoses and a shorter ulna. However this theory becomes more difficult to sustain if some patients are observed to have a 'pre-axial' preference and others a post-axial one. This is especially so of different preferences were to be observed in opposite limbs of the same patient; in this case a neoplastic

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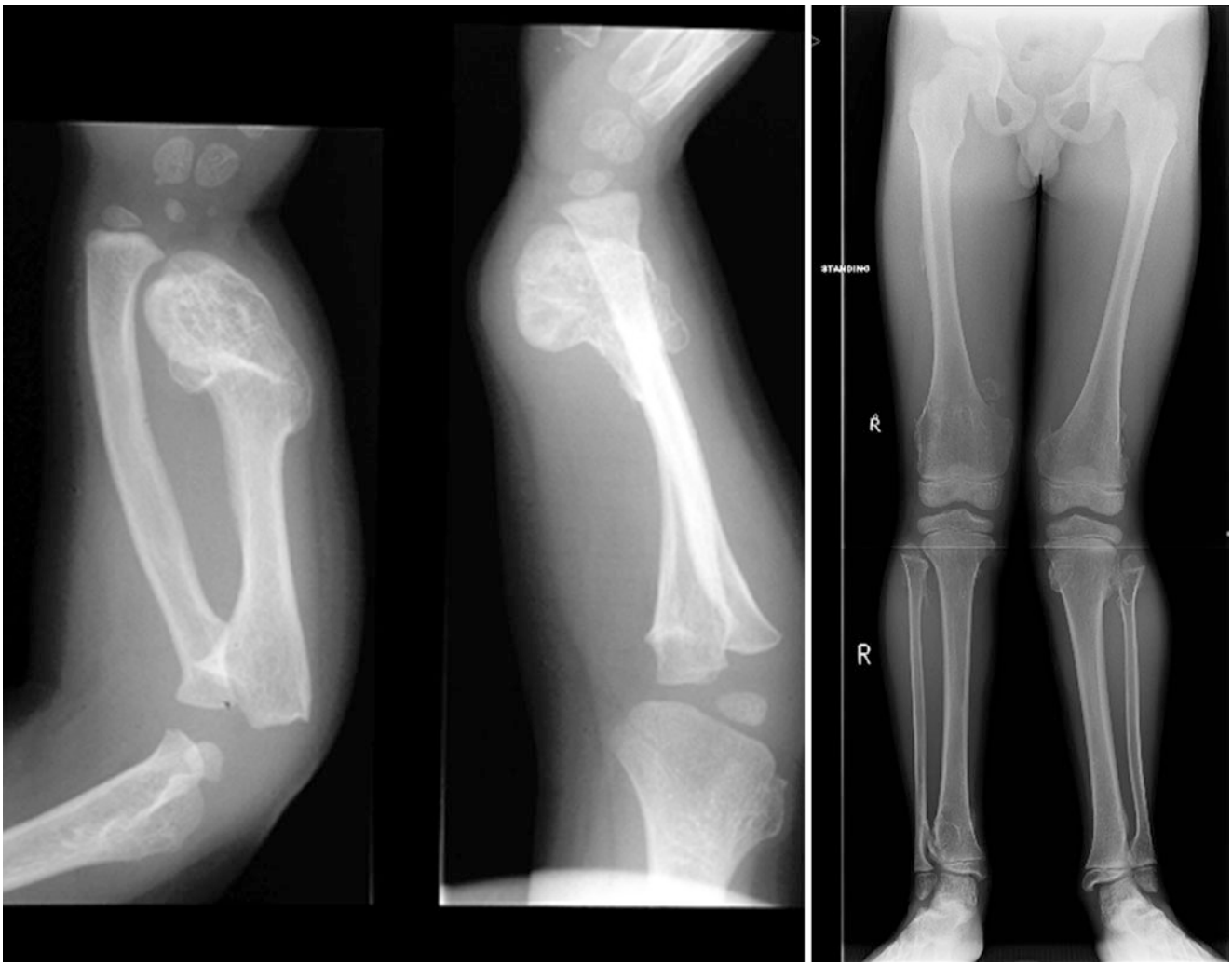

Fig. 49.1 A plain radiograph of a patient with multiple bony exostosis

pathogenesis for local growth disturbance becomes more likely.

Solitary osteochondromas are not hereditary, but they appear identical to osteochondromas arising in the multiple form. Several studies have suggested that bi-allelic mutations can be found in solitary osteochondromas, however this is not ubiquitous and perhaps other non-EXT pathways are involved in the development of these benign tumours [2]. Nevertheless, their clinical behaviour and pathological features are indistinguishable from those in MO. The clonal nature of neoplasia means that the formation of a solitary osteochondroma will initially involve a genetic change in a single cell of the chondral mesenchyme of the peripheral physis. A field change is unlikely to be responsible as there is no germline defect such as occurs in MO. Hence growth disturbance caused by a solitary exostosis cannot easily invoke a field-change effect to explain its occurrence. Consequently, studies which identify growth disturbance in solitary exostoses can shed important light on the same effect in MO.

When a child with MO presents, the parents wish to know what the future is likely to hold. Without an understanding of pathogenesis there can be no certain advice about preventing the growth disturbance which in some children can be severe. If evidence favours solely a neoplastic pathogenesis then the surgeon will choose to remove exostoses in association with deformity as they develop at key sites (eg distal forearm, distal tibia and fibula). He/she may even decide NOT to remove some exostoses if that decision might result in more balanced growth disturbance affecting paired bones. On the other hand, if evidence favoured a field-change effect, then removal of exostoses could not be justified on grounds of amelioration of the disease process, only in order to reduce pain, improve range of movement or cosmesis. Finally it is possible that there is a mixed aetiology for growth disturbance; in which a field-change effect might cause growth restriction at preferred sites in the absence of osteochondromas yet additionally a local effect of a large osteochondroma could modify that effect. In that situation advice to parents

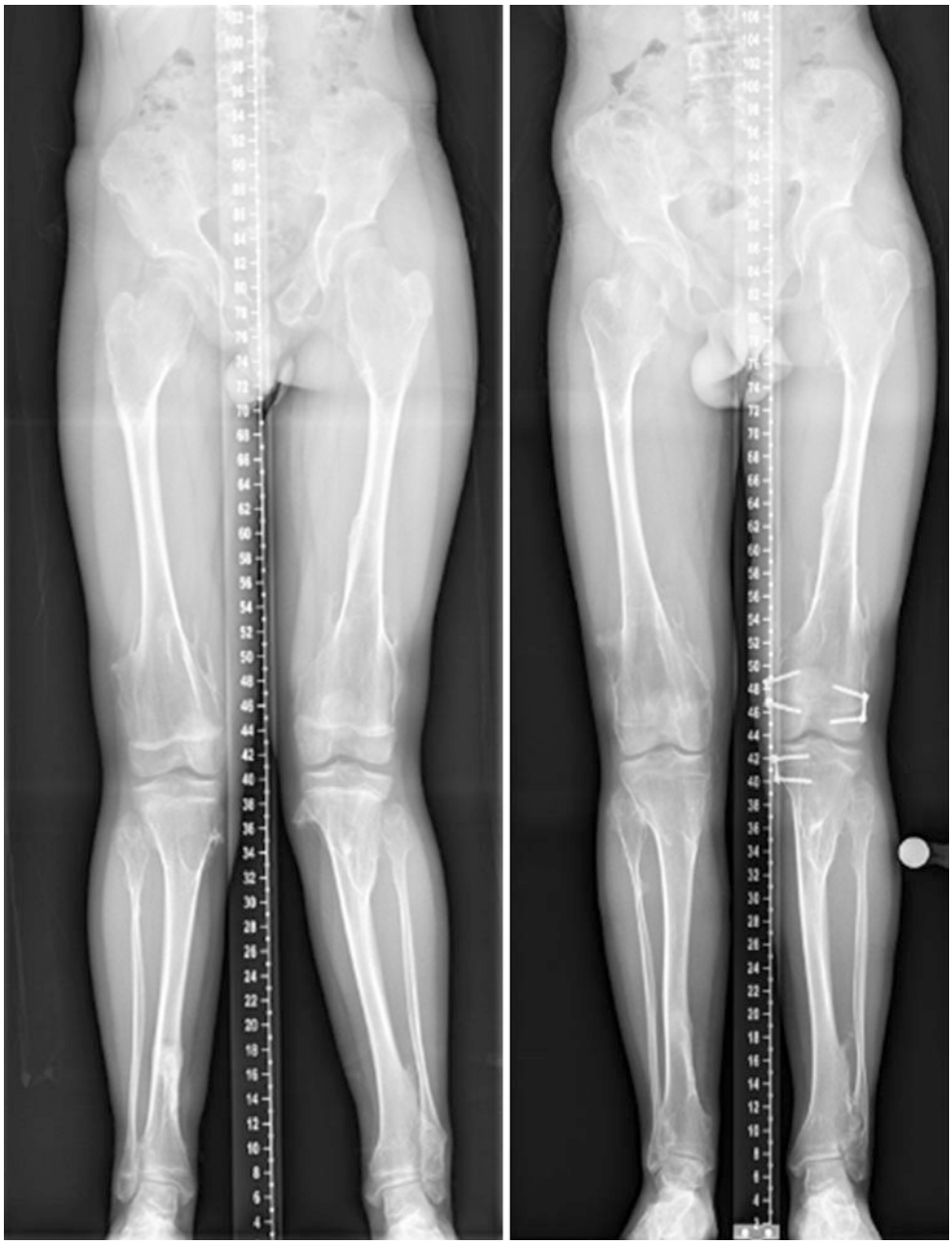

Fig. 49.2 Plain radiograph of a patient with MO and deformity of lower limbs. This patient with multiple bony exostosis developed left tibia knee valgus deformity and leg length discrepancy He was treated with guided growth using 8 plates

about the result of surgery would be more guarded, and allow for both possibilities. If it is believed that the excision of exostoses might reduce growth-disturbance effects, then the question of timing (early or late) becomes important since early exostoses are confluent with the growth plate, but later ones may have already resulted in significant deformity.

Questions

Faced with a child with an established or developing growth disturbance at one of several sites (distal forearm, proximal femur, around the knee, distal tibia/fibula) and one or more local exostoses which are not of themselves painful or causing significant loss of function, the surgeon has the option to either to remove the local exostosis and observe the effect, or to remain vigilant and undertake deformity-correction surgery at a later date. The purpose of this review is to seek evidence to help the surgeon in this decision-making process.

Key questions to answer therefore are:

1. Are local exostoses associated CAUSALLY with local growth disturbance?
2. Does excision of exostoses result in amelioration of growth disturbance?

3. Is early removal better than awaiting the onset of disturbance prior to excision?
4. Even if excision does ameliorate the pace of deformity, are there balancing risks to consider?

Levels of Evidence

MO is a rare condition. Even the largest tertiary medical centres are unlikely to treat enough patients to allow for inter-subject randomisation. However intra-subject randomisation might be achievable, for example with one limb acting as a control. It is recognised, however, that most if not all studies are likely to provide Level IV evidence only.

Most research into MO is based on an investigation at an anatomical location. Hence we filtered our search to identify publications which had both a diagnosis under the headings below AND an anatomical location as shown in Table 49.1.

This review has used Endnote® software (Thomson Scientific Inc) to identify publications found in Medline, PUBMED or EMBase at all time periods. Following deletion of duplications in each set and cross-set, this search generated searchable references and abstracts (Table 49.1). Each abstract was read to identify whether a growth-disturbance effect of MO might be identified within the publication. Single case-reports were included since it was anticipated that the quality of evidence would not exceed level IV. Full texts were sought and read (Table 49.1) in order to populate the data fields in Table 50.3, on which evidence the discussion section is written. In reading these full-texts, where reference was made to a publication of importance not already identified in the search, these were also sought and read and added to the database under the heading 'General' (Table 49.1).

There are no level I, II or III studies which address the question of growth disturbance in MO. Almost all evidence is found in case reports and case series. There are a very few retrospectively acquired longitudinal studies and two retrospective case-control study which do not reach the quality standard for classification at level III (Table 49.2).

Discussion

The Literature review identified 16 publications in which there was no description of deformity [1, 3, 14, 18, 20, 28, 49, 51, 58, 65–67, 73, 79, 83, 94]. These could provide no evidence to assist in answering our key questions

Are Local Exostoses Associated CAUSALLY with Local Growth Disturbance?

Most paediatric surgeons will have seen with their own eyes some evidence of deformity in children with MO. However it is necessary from time to time to review the published data which supports such beliefs. In this regard, a total of 8 papers reported exostoses, without deformity of significance. These included case reports of single exostoses, for example intra-articular acetabular lesions [6, 30], larger metaphyseal solitary exostoses [34, 40, 44, 52] and 'post-traumatic' exostoses [41, 50]. There were no reports in patients with MO in whom deformity could be definitely excluded.

In contrast multiple reports described exostoses in which deformity existed in some form. These included 10 in which deformity may be a misnomer – occurring simply due to physical pressure from an exostosis such as dislocation of the radial head due to the presence of a large proximal radial

Table 49.1 Summary of keywords and search output

Diagnostic keyword	Anatomical keyword	Total number of abstracts	Full texts for Tables 49.2 And 49.3 Below
Cartilaginous exostoses	Forearm	74	32
Diaphyseal aclasia	Ulna	94	9
Diaphyseal aclasis	Radius	32	1
Exostoses	Elbow	101	6
Hereditary multiple exostoses	Wrist	37	1
Hereditary multiple exostosis	Lower limb	21	0
Multiple cartilaginous exostoses	Hip	269	18
Multiple hereditary exostoses	Ankle	211	18
Multiple osteochondroma	Knee	233	6
Multiple osteochondromas	Tibia	109	5
Multiple osteochondromatosis	Fibula	82	15
	Femur	68	7
	Femoral	94	5
	Malleolar	8	3
	General		11

Table 49.2 Study details with regard to questions on effect of exostoses and excision of exostoses on growth disturbance

Reference	LoE	Location	N	Exostosis associated with deformity	Exostosis and deformity linked by causality	Excision exostosis stopped deformity?	Timing of surgery and deformity?	Complications	Summary
[3]	IV case series	Proximal fibula	46	n/a	n/a	n/a	n/a	n/a	No mention deformity
[4]	IV case report	Distal femur	1	(Y)	(Y)	n/a	n/a	n/a	Solitary, Figs. 1 and 2 look like ante-curvatum but not mentioned in report
[5]	V case report	Ulna	1	(Y)	n/a	n/a	n/a	n/a	Solitary exostosis proximal ulna. Dislocation by physical pressure
[6]	V case report	Proximal femur	1	n/a	n/a	n/a	n/a	n/a	No deformity noted
[7]	IV case series	Distal radius & ulna	21(33), but only 10(11) had excision surgery	Y	n/a	N	Age 5–16	None	11 forearms in 10 patients had exostosis excision only at mean age 11. No description of extent of surgery. Average carpal slip, radial articular angle and ulnar variance all deteriorated at final follow-up, but no significant difference (in all groups mean 13 years FU). Unclear when was final follow-up. No control group
[8]	IV case series	Radius and ulna	36 had excisions	Y	n/a	n/a	n/a	One unhappy with scars	No procedure done 'to prevent abnormal growth patterns of the involved bones'. Telephone questionnaire for function only
[9]	IV case series	Distal tibia & fibula	4(6)	n/a	n/a	n/a	n/a	n/a	Use of a screw for hemiepihyseodesis in ankle valgus 4 MO patients. No comment on exostosis site
[10]	IV case series	Distal tib & fib	6	Y	n/a	n/a	n/a	n/a	Permanent epiphysiodesis only
[11]	IV case series	Radius & ulna	7(8)	Y	n/a	n/a	n/a	n/a	8 forearms with ulnar lengthening & excision exostoses without separated procedures in Masada I & IIb(short ulna, ulnar exostoses)
[12]	IV case series	Radius & ulna	16(20)	Y	(Y)	n/a	n/a	n/a	Authors argue degree of forearm deformity associated with degree of metacarpal shortening. Only 1 Taniguchi type IIIA (radius short. This showed only a radial exostosis)

(continued)

Table 49.2 (continued)

Reference	LoE	Location	N	Exostosis associated with deformity	Exostosis and deformity linked by causality	Excision exostosis stopped deformity?	Timing of surgery and deformity?	Complications	Summary
[13]	IV case control	Radius & ulna	35(65)	Y	N	n/a	n/a	n/a	X-sectional study. Relative forearm length compared with controls. Most forearms had both short radius and ulna cf controls. Exostosis presence evaluated (no tabulation) and found not to associate with relative length of the ulna or radius. Only 3 forearms had isolated radial exostosis. These had normal lengths compared with controls
[14]	V case report	Whole body	1	n/a	n/a	n/a	n/a	n/a	Descriptive of multiple sites in one patient
[15]	IV case series	Distal tibia/ fibula	23(19 solitary)	(Y)	(Y)	(Y)	Age 2–48	n/a	Retrospective study. Age 8–48, Follow-up over 2.5–19 years. 19 had surgery. State that 'deformity' improved after excision, but no documentation of type of deformity. Only 'pronation' mentioned and fibular or tibial indentation or bony shortening. Tables do not quantify deformity, or correction, or association with exostosis position
[16]	IV case Series	Radius & ulna	4	Y	n/a	n/a	n/a	n/a	4 lengthenings with excision exostoses. No separated procedures
[17]	V case report	Distal tibia	1	Y	n/a	n/a	n/a	n/a	Fibular indentation
[18]	IV – case series	Shoulder	145	n/a	n/a	n/a	n/a	n/a	N = 172, clinical examination only. No correlation of exostosis number or location with deformity
[19]	IV case series	Stature	172	n/a	n/a	n/a	n/a	n/a	N = 172 all ages with MO. Height is normal until the age of 10 when there is a diminution. Cross-sectional study is main weakness
[20]	IV – case series	Whole body	143	n/a	n/a	n/a	n/a	n/a	143 patients all ages, Genotype-phenotype with number of exostoses at different sites as main disease parameter.. No assessment of deformity. Clinical examination only

[21]	IV case series	Radius & ulna	106	n/a	n/a	n/a	n/a	n/a	Adults. Clinical examination only. Exostosis number linked to range of movement and to presence of radial head dislocation
[22]	IV case series	Radius & ulna	168	Y	(Y)	n/a	n/a	n/a	N = 172, clinical examination only. Number of distal femoral exostoses are independent factors for deformity (degree of valgus) on multivariate analysis
[23]	IV case series	Knee	3	(Y)	(Y)	(Y)	N	N	Fibular bowing remodelled
[24]	IV case series	Radius & ulna	6	Y	n/a	n/a	n/a	n/a	Study of ex fix in 6 children age 9–14. forearms all had ulnar exostoses. All had negative ulna variances of at least 10 mm. Only one had a distal radial exostosis
[25]	V case report	Distal femur	1	Y	Y	n/a	n/a	n/a	9/12 girl, genu valgum osteochondroma medial femoral condyle. Deformity only here (genu valgum)
[26]	IV case series	Distal tib & fib	19	Y	n/a	n/a	n/a	n/a	Temporary hemiepiphyseodesis only. Never removed exostoses
[27]	IV case series	Hip	18(36)	Y	n/a	n/a	n/a	n/a	Observational study of deformity. No correlation of exostosis site/size with deformity
[28]	V case report	Hip	1	n/a	n/a	n/a	n/a	n/a	Case report of CDH and MO
[29]	IV case series	Distal tibia	5 (3 solitary, 2 HME)	Y	n/a	n/a	n/a	n/a	Tibial exostoses. All indent fibular. Pre-op ‘ankle deformity’ and post-op ‘correction of angular deformity of fibula’ not quantified
[30]	V case report	Hip	1	N	n/a	n/a	n/a	n/a	Case report of hip joint exostosis

(continued)

Table 49.2 (continued)

Reference	LoE	Location	N	Exostosis associated with deformity	Exostosis and deformity linked by causality	Excision exostosis stopped deformity?	Timing of surgery and deformity?	Complications	Summary
[31]	IV case series	Radius & ulna	48 (76)	Y	Y	n/a	n/a	n/a	Retrospective x-ray review. Unselected so selection bias (e.g. age) possible. 102 MO patients, but only 48 (76 forearms) available for x-ray analysis. Forearm x-rays grouped into 5 groups BASED ON LOCATION. Group 1 (distal ulna only N = 33), Group 3 (distal radius only N = 9); these had different deformity-associated characteristics (ulnar variance, radial articular angle, radial bow all worse in ulna-only group, but not statistically so. Authors state they believe they behave differently. 13/14 radial head dislocations in ulna only group. Group 4 (diaphyseal exostoses) had least ulnar shortening but these were younger (median age 7 vs 13 for distal ulna alone), so may be due to age-related effects. Unclear what surgery done during period of radial head evaluation. FU 7 years, and based on this a qualitative appraisal of deformity in ulna-only group is described due to ulnar tethering of radius. Radial and ulnar shortening is measured and the DIFFERENCE between the two is least in radial exostoses (2 mm) cf 7 mm for both bones and 14 mm for ulna only, but no statistical analysis done
[32]	V case report	Distal tibia	1	(Y)	n/a	n/a	n/a	n/a	Excision osteochondroma. Deformity is thin fibula only
[33]	IV case series	Radius & ulna	6	Y	Y	n/a	n/a	n/a	Forearm lengthening in 4 MO and 2 distal unlar osteochondroma (implication is they are SOLITARY). Indication for surgery is radio-ulnar length discrepancy of > 2cm

[34]	V case report	Prox femur	1	N	n/a	n/a	n/a	n/a	Age 24. Large femoral neck exostosis – solitary
[35]	IV case series	Radius & ulna	6	Y	n/a	n/a	n/a	n/a	6 children ulnar lengthening in Masada I (short ulna, ulnar exostoses, radial head located)
[36]	IV case series	Radius and ulna	13(14)	Y	Y	Y	Effective ages 4–12 (mean age 7)	Tumour recurrence in 8/14. No others volunteered	Natural history of MO following excision exostoses at distal ulna. Group 1 only distal ulna exostosis (6 forearms in 6 children, Mean age 7, FU 4 years). Group 2 touching distal ulna and distal radius exostoses (8 forearms in 7 children, mean age 8 years, FU 5 years). After surgery, group 1 showed significant improvement in % ulnar shortening (8.3–6.5 %) and % radial bow (10.2–7.3 %), but not radial articular angle or degree of carpal slip. Group 2 showed no significant improvement in any parameter and a deterioration in radial bow and radial articular angle. Tumour recurrence in 2/6 in group 1 and 6/8 in group 2, but recurrence not related to likelihood of continuing deformity. Possible that surgical adequacy had some effect
[37]	IV case series	Hip	2	(Y)	n/a	n/a	n/a	n/a	2 cases MO & hip subluxation. Surgical result not assessed for deformity improvement other than hip joint reduction
[38]	IV case series	Distal radius & ulna	10(12)	Y	n/a	n/a	n/a	n/a	Children with MO. Longitudinal radiographic study Combination of procedures including 4 excisions alone. Masada classification recorded, but small numbers and incomplete follow-up prevent analysis of long-term consequences
[39]	V case report	Distal tibia	1	Y	n/a	n/a	n/a	n/a	Short fibula but varus ankle
[40]	V case report	Distal humerus	1	N	n/a	n/a	n/a	n/a	Large exostosis, no deformity

(continued)

Table 49.2 (continued)

Reference	LoE	Location	N	Exostosis associated with deformity	Exostosis and deformity linked by causality	Excision exostosis stopped deformity?	Timing of surgery and deformity?	Complications	Summary
[41]	V case report	Distal tibia	1	N	n/a	n/a	n/a	n/a	Histological osteochondroma after raising a flap. No deformity
[42]	V case report	Phalanx	1	Y	Y	n/a	n/a	n/a	4th toe Proximal phalanx deformity in a SOLITARY exostosis
[43]	IV case series	Distal tib/fib	12	Y	? (see comment)	n/a	n/a	n/a	Lengthening fibular for valgus and talar shift. Table describes site of exostoses. 4 Fibula alone, 6 Tibia and Fibula. 2 Tibia alone. Authors state uncertain why an enormous tibial exostosis alone may cause fibular undergrowth
[44]	V case report	Prox femur	1	Uncertain	Uncertain	n/a	n/a	n/a	Solitary exostosis at surgical neck
[45]	IV case series	Radius & ulna	3	Y	n/a	n/a	n/a	n/a	Only 3/27 forearms had isolated exostosis removal (all radial). Remainder osteotomies/ lengthenings
[46]	IV case series	Hip	6	n/a	n/a	n/a	n/a	n/a	6 cases – qualitative descriptions including natural history. No assessment of deformity in relation to specific site, size or number of exostoses
[47]	IV case series	Radius & ulna	30(36)	Y	Y	Y	Y (single case appraised)	None volunteered	Classic paper with Masada deformity Classification. Type I distal ulna exostosis wthout ulnar dislocation; type IIa proximal radius exostosis causing radial head dislocation. Type IIb as type I but with radial head dislocated; type III (N = 7) distal radius exostosis and ulna is long. Single case (case 6) of excision alone in type III (distal radius). After surgery radial shortening improved from 10 to 4 mm after 19 months (Fig. 6). Opposite hand had ulnar shortening and had lengthening
[48]	IV case series	Radius & ulna	7	Y	n/a	n/a	n/a	n/a	7 legthenings with excision exostoses. No separated procedures
[49]	IV case series	Hip & Knee	2	n/a	n/a	n/a	n/a	n/a	2 cases of lower limb arthroplasty. Deformity not assessed in relation to exostoses

[50]	V case report	Distal femur	1	N	n/a	n/a	n/a	n/a	Small osteochondroma
[51]	IV case series	Prox fibula	6 (4 solitary, 2 MO)	n/a	n/a	n/a	n/a	n/a	No comment on deformity
[52]	V case report	Distal humerus	1	N	n/a	n/a	n/a	n/a	Large soft tissue osteochondroma – no deformity described
[53]	IV case series	Knee	8(16)	Y	n/a	n/a	n/a	n/a	Longitudinal study over 8 years from mean age 8 years. Valgus knee deformity markedly worsened from age 10.Tibia mainly resonsible.No assessment of exosotsis site
[54]	IV case series	Distal tib/fib	38	Y	n/a	n/a	n/a	n/a	Natural history adult
[55]	IV case series	Radius & ulna	39	Y	n/a	n/a	n/a	n/a	Functional outcome cross-sectional study in adults. Only 4 had previous exostosis removal
[56]	IV case series	Prox and distal tibia/fibula	6	Y	n/a	n/a	n/a	n/a	Ilizarov for ankle and knee valgus
[57]	V Case report	Hip	1	n/a	n/a	n/a	n/a	n/a	As other hip subluxation cases, only deformity is subluxation perhaps due to physical pressure of exostoses
[58]	IV case series	Foot	20 HME, 24 solitary	n/a	n/a	n/a	n/a	n/a	Large series of foot tumours in general
[59]	IV case series	Radius & ulna	10	Y	Y	n/a	n/a	n/a	Radiographic and clinical study. Clinical study showed shorter forearms had more exostoses. Direct correlation of ‘relative osteochondroma load’ on either radius or ulna with the relative length of that bone compared with its neighbour
[1]	V review	Whole body	n/a	n/a	n/a	n/a	n/a	n/a	Review of pathogenesis theories and especially neoplastic evidence
[60]	IV case series	Hip	12(24)	Y	Y	n/a	n/a	n/a	Radiographic study, proximal femoral exostosis ‘load’ related to coxa valga. Presence of periacetabular exostoses related to acetabular index

(continued)

Table 49.2 (continued)

Reference	LoE	Location	N	Exostosis associated with deformity	Exostosis and deformity linked by causality	Excision exostosis stopped deformity?	Timing of surgery and deformity?	Complications	Summary
[61]	IV case series	Radius & Ulna	8(10)	Y	n/a	n/a	n/a	n/a	10 forearms with ulnar lengthenings. No record of exostosis management
[62]	IV case series	Prox femur	6	Y	Y	n/a	n/a	n/a	All solitary, age 20–66. Patient age 20 with short leg 2.3 cm and coxa valga. One further patient with coxa valga age 28. No uncovering. No reason for coxa valga in some but not others
[63]	IV case series	Distal tib/fib	9(15)	Y	n/a	n/a	n/a	n/a	Screw epiphysiodesis only, youngest patient (age 9) had recurrent valgus afterwards
[64]	IV case series	Distal Tibia & Fibula	12(21)	Y	n/a	n/a	n/a	n/a	Screw epipyhsiodesis – rebound in 50 %
[65]	IV case Series	Distal radius & ulna	?	n/a	n/a	n/a	n/a	n/a	Ulna shortening in 7(11). Unclear how many, if any, had osteochondromas
[66]	IV case series	Knee	n/a	n/a	n/a	n/a	n/a	n/a	2 % rate of incidental exostosis on knee x-ray
[67]	V review	Whole body	n/a	n/a	n/a	n/a	n/a	n/a	Review of theories of pathogenesis and new evidence of a neoplastic pathogensis. Implications of a neoplastic aetiolgy on mangement includes justification for excision to prevent deformity
[68]	V case report	Tibia & Fibula	1	Y	n/a	n/a	n/a	n/a	Case of Ilizarov correction ankle valgus in an adult. Exostoses in both ankles. Site unclear

[69]	IV case series	Radius & ulna	22	Y	n/a	(N)	Mean age 9	No major	22 patients (mean age 9 years) had excision of exostoses alone. Indications pain, cosmesis, limitation of movement. Follow-up 2–9 (mean 3.5 years). Supination improved significantly. 'The radiological parameters did not change significantly' after excision exostoses alone. No tabular data. Abstract states that simple excision 'will not halt the progression of disease'. Radiographic parameters – carpal slip, RAA, linear axis, ulnar variance, radial bow. Unclear why this is said when radiographic parameters did not change significantly. Perhaps due to 36 % exostosis recurrence rate
[70]	V case report	Proximal femur	1	Y	(Y)	n/a	n/a	n/a	Adult case. One side relatively normal. Other side large exostosis with coxa valga and acetabular dysplasia
[71]	IV case series	Proximal femur	4 (2 HME, 2 solitary)	Y	Y (strong because solitary)	n/a	n/a	n/a	All age 20–40. One patient with solitary exostosis had a valgus neck deformity (age 30). This patient had reduced CE angle and uncovering cf opposite hip. Large exostosis at medial femoral neck. One patient with HME also had neck valgus
[72]	V case report	Distal tibia	1	N	n/a	n/a	n/a	n/a	Age 38 solitary?
[73]	V case report	Radius	1	n/a	n/a	n/a	n/a	n/a	Case report of regression after ulnar lengthening
[74]	IV case series	Radius & ulna	16(23)	Y	n/a	n/a	n/a	n/a	23 lengthenings with excision exostoses. No separated procedures
[75]	V case report	Distal fibula	1	Y	Y?	n/a	n/a	n/a	Age 51 ?solitary. Fibular exostosis. Valgus not mentioned, but present
[76]	V case report	Distal fibula	1	Y	Y	n/a	n/a	n/a	18/12 girl. No other deformity or exostosis. Short fibula

(continued)

Table 49.2 (continued)

Reference	LoE	Location	N	Exostosis associated with deformity	Exostosis and deformity linked by causality	Excision exostosis stopped deformity?	Timing of surgery and deformity?	Complications	Summary
[77]	IV case series	Stature	50	n/a	n/a	n/a	n/a	n/a	Cross sectional study. 50 children. Under age 12 skeletal age lower than chronological age. After age 12 the opposite. Authors suggest early skeletal maturation leads to shorter stature
[78]	IV case series	Upper limb	28(56)	Y	n/a	n/a	n/a	n/a	Adults and children Functional and x-ray cross-sectional study. Exostosis site & number & type of surgery not examined
[79]	V review	Hip	n/a	n/a	n/a	n/a	n/a	n/a	Review of Hip pathology in skeletal dysplasias. No analyisis of deformity or exostoses
[80]	IV case series	Distal tib/fib	33(62)	Y	Y	n/a	n/a	n/a	Natural history in children – longitudinal data. Never had exostosis excision or correction. Those with Taniguchi grade III (shortening of radius or ulna) had more distal tibia & fibula exostsoses than other groups. Those with involvement of both distal tibia and fibula had worse valgus (no volume noted). Shortening of fibula recorded but analysis in relation to site of exostosis not comprehensive (ie girls with no shortening did not have the site of exostosis included in the results). However Fig. 4 can be used to extrapolate. 17 patients had longitudinal data. Only tibia exostoses(n = 7) had change of 0.3°/year varus; only fibular exostosis (n = 3) had change of 1.6°/year valgus. Both tib and fib exostoses had change 1.5°/year into valgus. Start point for all patients – tib exostoses alone, 5 in valgus, 13 in neutral or varus – mean 0°; fib exostoses alone, 5 in valgus, 2 in neutral or varus – mean 4° valgus

[81]	IV case series	Radius & ulna	14	Y	n/a	n/a	n/a	n/a	14 lengthenings with excision exostoses. No separated procedures
[82]	IV case series	Distal radius and ulna	42	Y	Y(?)	n/a	n/a	n/a	8 patients group 1 (no involvement), 11 patients group II (exostoses, no shortening), 22 patients group III (exostsoses and shortening. IIIA radius shortening (n = 2), IIIB ulna shortening (n = 20). No record of whether exostoses on ulna associated with ulna shortening or vice versa, but Fig. 1 shows x-ray with radius shortening with only a radius exostosis. Classification associated with disease severity
[83]	IV case series	Radius & ulna	<30	n/a	n/a	n/a	n/a	n/a	Ulnar shortening in 30. Unclear how many, if any, had osteochondromas
[84]	V case report	Distal fibula	1	Y	n/a	n/a	n/a	n/a	Exostoses on both tib and fib
[85]	IV case series	Radius & ulna	12	Y	n/a	n/a	n/a	n/a	12 lengthenings with 5 synchronous excision exostoses. All Masada I and II
[86]	V review	Radius & ulna	n/a	Y	n/a	n/a	n/a	n/a	Review of single-bone forearm surgery in MO. No numbers. No exostosis site or size details
[87]	IV case series	Radius & ulna	142	Y	n/a	n/a	n/a	n/a	107 Adults, 35 children age <15. Clinical examination only. Exostosis number significantly negatively associated with proportional ulnar length and range of movement. Specific exostosis site and deformity not compared
[88]	V case report	Radius & ulna	1	Y	n/a	n/a	n/a	n/a	Description of forearm deformity in MO
[89]	V case report	Radius & ulna	1	Y	n/a	n/a	n/a	n/a	Description of forearm deformity in MO
[90]	IV case series	Prox femur	25(42)	Y	n/a	n/a	n/a	n/a	No tables. All femurs said to show identical coxa valga. Measurement said to be impossible due to distorting effect of exostoses

(continued)

Table 49.2 (continued)

Reference	LoE	Location	N	Exostosis associated with deformity	Exostosis and deformity linked by causality	Excision exostosis stopped deformity?	Timing of surgery and deformity?	Complications	Summary
[91]	IV case series	Hip	2	n/a	n/a	n/a	n/a	n/a	As other hip subluxation cases, only deformity is subluxation perhaps due to physical pressure of exostoses
[92]	IV case series	Distal ulna	2	n/a	n/a	n/a	n/a	n/a	Both ulnae short
[93]	IV case-control	Prox femur	11(21)	Y	n/a	n/a	n/a	n/a	Ischio-femoral impingement (distance between ischium and lesser trochanter) on MRI greater in HME than controls
[2]	IV case series	Whole body	23	n/a	n/a	n/a	n/a	n/a	Mutations in MO and solitary exostoses may involve non EXT-pathways

N number of patients (number of limbs or exostoses), *Y* yes, *N* no, *n/a* not applicable

or ulnar exostosis [5, 21], fibular indentation effects [17, 23, 32, 72, 84] or hip joint subluxation [37, 57, 91]. A further 37 reported deformity in MO without pressure-effects, but in which a detailed description of local exostoses was absent and hence no assessment of exostosis effect could be made. These reports included 2 papers about stature in MO [19, 77] and 35 in which MO was associated with ulnar shortening [8, 9, 11, 16, 24, 35, 38, 45, 48, 55, 61, 78, 81, 82, 85–89, 92, 95], fibular shortening [10, 26, 29, 39, 54, 56, 63, 64], coxa valga [27, 46, 90, 93] or genu valgum [53, 56].

Solitary exostoses are very common, and have no heritable component. Six papers indentified 8 patients with solitary exostoses who also exhibited local deformity. These included coxa valga [62, 71], antecurvatum and genu valgum in the distal femur [4, 25], ulnar shortening [33] and phalangeal valgus [42]. Their association with deformity is moderately strong evidence in favour of a local exostosis effect.

One study measured forearm length in MO compared with control patients. The authors found both short radius and short ulna compared to controls, and that the presence of exostoses did not correlate with the degree of shortening, suggesting that in MO there is a general field-change effect whch results in shorter limbs. However forearms which had only isolated radial exostoses had normal forearm lengths compared to controls [13]. This paper provides moderately strong evidence for a field-change effect on forearm growth in MO, but does not exclude the possibility that local exostoses might also exert differential growth effects on the radius and ulna.

A study of fibular shortening in MO identified 2 cases with an enormous tibial exostosis alone [43]. This local effect might be explained by lateral tethering but provides weak evidence against a local effect of osteochodromas on growth at the ankle.

MO is a germline condition. An association of local deformity and local exostoses in paired bones of the forearm or leg, where no deformity occurs in the absence of exostoses is evidence of a CAUSAL association since the effect is difficult to explain by means of a field-change. In the upper limb, a Taniguchi type IIIA forearm has a short radius rather than ulna. Several authors correlate the presence of radial exostoses with this type of deformity [12, 31, 47, 59]. One series of 76 forearms for radiographic analysis showed shorter ulnae with in patients with ulnar exostoses compared to those with radial exostoses, although no statistical analysis was performed [31]. In a study of 10 forearms, relative shortening of forearm bones depended on the relative exostosis burden on each bone [59]. At the ankle, the presence of fibular exostoses was associated with a greater degree of valgus and more rapid deterioration in deformity [80]. The longitudinal nature of this study provides moderately strong evidence that fibular exostoses resulted in more rapid fibular shortening

A similar differential effect around the hip was seen in one study; the degree of acetabular deformity was directly associated with peri-acetabular exostosis burden, whereas the degree of coxa valga was associated with proximal femoral exostosis burden [60].

Reports which show different effects on left and right sides of the body related to the presence or absence of exostoses also provide weak evidence in favour of a local growth-disturbing effect [47, 70].

In a reports derived from a large cross-sectional study of 172 patients with MO, number of distal femoral exostoses was an independent factor for the degree of knee valgus in a multivariate analysis where germline mutation and gender were included [22]. This constitutes weak evidence for a CAUSAL association.

Does Excision of Exostoses Result in Amelioration of Growth Disturbance in MO?

Publications which can address this question require a longitudinal component in which the only treatment is exostosis excision. No studies could be found with a control group.

One study of 11 forearms in 10 patients found that the mean values for wrist anatomical parameters of deformity deteriorated (but not-significantly) following excision surgery. Mean age was 11 (range 5–16). Length of follow-up was unknown [7]. This study constitutes weak evidence against excision for deformity amelioration.

One study of 22 patients with forearm exostoses (mean age 9 and follow up 2–9 years) states in the abstract that excision of exostoses ‘will not halt progression of the disease’. In the results section it is stated that ‘radiological parameters did not change significantly’. There was no tabulated data [69]. This study constitutes weak evidence against excision for deformity amelioration.

In contrast longitudinal evidence of an ameliorating effect of excision on future deformity was found in two studies:

One study of forearm deformity found that where single ulnar exostoses were the main radiographic features, (6 cases) their removal resulted in a significant improvement in relative ulnar shortening and radial bow after follow-up 24–97 months. However in forearms with touching radial and ulnar exostoses (8 cases) excision did not result in improvement [36]. In a separate paper a further single case of radial exostosis excision resulted in improvement in radial shortening after 19 months [47].

Is Early Removal Better than Awaiting the Onset of Disturbance Prior to Excision?

The evidence from studies quoted above is of poor quality and does not allow a confident assertion on timing of surgery. The patients in whom parameter of deformity improved after excision already had marked deformity at the time of surgery. Age ranged from 4 to 12 years [36, 47].

Even If Excision Does Ameliorate the Pace of Deformity, Are There Balancing Risks to Consider?

Studies are lacking which report symptoms in patients who had excision to prevent future deformity. One study in which 36 MO patients had forearm exostoses removed for pain rather than 'prevention of abnormal growth patterns' resulted in generally satisfied patients by telephone questionnaire at minimum 2 year follow-up [8]. Risk of recurrence is significant, especially if the exostosis is close to the growth plate. Risk of recurrence in the forearm is 36–57% [36, 69].

Future Questions

With only three level IV publications observing the result of excision on growth disturbance, the primary question has not yet been answered with satisfaction. The inclusion of a control group, or a contra-lateral control would add to the strength of evidence.

Summary

There is no evidence that a policy to remove widespread exostoses as they form can arrest the development of short limbs or short stature which is a manifestation of MO. However the balance of evidence is in favour of a local effect of osteochondroma growth on deformity, which also allows for the possibility of an additional more general 'field change' effect on overall skeletal growth.

The effect of excision of exostoses on developing deformity has only been studied in the forearm. There is some evidence that local excision surgery in childhood can arrest or even improve established local deformity, but by no means in all cases. It is not clear whether failure is due to inadequate excision, a defective physis which is already established or other unknown factors. Parents should be counselled that excision for prevention of future deformity has only a low to moderate chance of success, but occasionally has the potential to reverse deformity which has already occurred. Apart from failure to achieve the desired improvement in deformity, risks are only those which pertain to excision of exostoses in general (Table 49.3).

Table 49.3 Summary of grades of recommendation

Recommendation	Level of evidence	Grade
Excision of exostosis may arrest or reverse the development of deformity	IV	C
Optimal time for surgery (patient age and stage of deformity) is not established	IV	C
Risks of excision surgery for deformity are similar to those for excision for other reasons	IV	C

References

1. Porter DE, Simpson AH. The neoplastic pathogenesis of solitary and multiple osteochondromas. J Pathol. 1999;188(2):119–25.
2. Zuntini M et al. Genetic models of osteochondroma onset and neoplastic progression: evidence for mechanisms alternative to EXT genes inactivation. Oncogene. 29(26):3827–34.
3. Abdel MP et al. Surgical management of 121 benign proximal fibula tumors. Clin Orthop Relat Res. 468(11):3056–62.
4. Abd-el-Naby W, Hammond T, Court-Brown CM. Atypical osteochondroma of the distal femur. Orthopedics. 2000;23(7):725–6.
5. Abe M, Koyama S. Developmental anterior dislocation of the radial head caused by solitary osteochondroma of the proximal ulna. J Shoulder Elbow Surg. 1998;7(1):66–70.
6. Aguiar T, Dantas P. Arthroscopic Resection of Intra-Articular Osteochondromas of the Hip. Arthroscopy Techn. 3(3):e347–50.
7. Akita S et al. Long-term results of surgery for forearm deformities in patients with multiple cartilaginous exostoses. J Bone Joint Surg Am. 2007;89(9):1993–9.
8. Arms DM et al. Management of forearm deformity in multiple hereditary osteochondromatosis. J Pediatr Orthop. 1997;17(4):450–4.
9. Auregan JC et al. Children ankle valgus deformity treatment using a transphyseal medial malleolar screw. Orthop Traumatol Surg Res. 97(4):406–9.
10. Beals RK. The treatment of ankle valgus by surface epiphysiodesis. Clin Orthop Relat Res. 1991;266:162–9.
11. Bilen FE et al. Correction of forearm deformities in children with multiple osteochondroma, by corrective radial osteotomy and ulnar lengthening by distraction osteogenesis. Acta Orthop Belg. 2009; 75(6):743–7.
12. Bock GW, Reed MH. Forearm deformities in multiple cartilaginous exostoses. Skeletal Radiol. 1991;20(7):483–6.
13. Burgess RC, Cates H. Deformities of the forearm in patients who have multiple cartilaginous exostosis. J Bone Joint Surg Am. 1993;75(1):13–8.
14. Cerrahoğlu L1, Ulusoy A, Akgül Ö. Ankle pain in hereditary multiple exostoses: a case report. Rheumatol Int. 2015;35(6):1107–9. doi: 10.1007/s00296-014-3176-y. Epub 2014 Nov 20.
15. Chin KR et al. Osteochondromas of the distal aspect of the tibia or fibula. Natural history and treatment. J Bone Joint Surg Am. 2000;82(9):1269–78.
16. Cho YJ, Jung ST. Gradual lengthening of the ulna in patients with multiple hereditary exostoses with a dislocated radial head. Yonsei Med J. 55(1):178–84.

17. Ciampolini J, Gargan MF, Newman JH. Arthrodesis of the distal tibiofibular joint for a large osteochondroma in an adult. Foot Ankle Int. 1999;20(10):657–8.
18. Clement ND, Ng CE, Porter DE. Shoulder exostoses in hereditary multiple exostoses: probability of surgery and malignant change. J Shoulder Elbow Surg. 20(2):290–4.
19. Clement ND et al. Skeletal growth patterns in hereditary multiple exostoses: a natural history. J Pediatr Orthop B. 21(2):150–4.
20. Clement ND, Porter DE. Hereditary multiple exostoses: anatomical distribution and burden of exostoses is dependent upon genotype and gender. Scott Med J. 59(1):35–44.
21. Clement ND, Porter DE. Forearm deformity in patients with hereditary multiple exostoses: factors associated with range of motion and radial head dislocation. J Bone Joint Surg Am. 95(17):1586–92.
22. Clement ND, Porter DE. Can deformity of the knee and longitudinal growth of the leg be predicted in patients with hereditary multiple exostoses? A cross-sectional study. Knee. 21(1):299–303.
23. Danielsson LG, el-Haddad I, Quadros O. Distal tibial osteochondroma deforming the fibula. Acta Orthop Scand. 1990;61(5): 469–70.
24. Demir B et al. Single-stage treatment of complete dislocation of radial head and forearm deformity using distraction osteogenesis in paediatric patients having multiple cartilaginous exostosis. Arch Orthop Trauma Surg. 131(9):1195–201.
25. Donaldson JS et al. Osteochondroma of the distal femoral epiphysis. J Pediatr. 1953;43(2):212–6.
26. Driscoll M et al. Correction and recurrence of ankle valgus in skeletally immature patients with multiple hereditary exostoses. Foot Ankle Int. 34(9):1267–73.
27. El-Fiky TA et al. Hereditary multiple exostoses of the hip. J Orthop Surg (Hong Kong). 2009;17(2):161–5.
28. Evans EL. Case of multiple cartilaginous exostoses associated with congenital dislocation of hip. Proc R Soc Med. 1921;14(Surg Sect): 166.
29. Gil-Albarova J, Gil-Albarova R, Bregante-Baquero J. Fibular rotational osteotomy for the treatment of distal tibial osteochondroma: A technical modification for deformity correction and improved outcomes. J Foot Ankle Surg. 2007;46(6):474–9.
30. Gore DR. Intra-articular osteochondromas of the hip joint in a child with multiple osteochondromas. Case report. Clin Orthop Relat Res. 1985;199:173–8.
31. Gottschalk HP et al. Effect of osteochondroma location on forearm deformity in patients with multiple hereditary osteochondromatosis. J Hand Surg Am. 37(11):2286–93.
32. Herrera-Perez M et al. Osteochondromas around the ankle: report of a case and literature review. Int J Surg Case Rep. 4(11): 1025–27.
33. Hill RA et al. Forearm lengthening by distraction osteogenesis in children: a report of 22 cases. J Bone Joint Surg Br. 93(11): 1550–5.
34. Hussain W et al. Solitary osteochondroma of the proximal femur and femoral acetabular impingement. Orthopedics. 33(1):51.
35. Ip D et al. Reconstruction of forearm deformities in multiple cartilaginous exostoses. J Pediatr Orthop B. 2003;12(1):17–21.
36. Ishikawa J et al. Tumor location affects the results of simple excision for multiple osteochondromas in the forearm. J Bone Joint Surg Am. 2007;89(6):1238–47.
37. Jellicoe P et al. Surgical hip dislocation for removal of intraarticular exostoses: report of two cases. J Pediatr Orthop. 2009;29(4): 327–30.
38. Jiya TU, Pruijs JE, van der Eijken JW. Surgical treatment of wrist deformity in hereditary multiple exostosis. Acta Orthop Belg. 1997;63(4):256–61.
39. Johnston AJ, Andrews CT. Fibular lengthening by Ilizarov method secondary to shortening by osteochondroma of distal tibia. Strateg Trauma Limb Reconstr. 2008;3(1):45–8.
40. Karakurt L et al. Solitary osteochondroma of the elbow causing ulnar nerve compression: a case report. Acta Orthop Traumatol Turc. 2004;38(4):291–4.
41. Keskin M et al. Formation of osteocartilaginous exostosis resulting from the raising of a lateral supramalleolar flap. J Plast Surg Hand Surg. 46(1):49–51.
42. Kurashige T, Suzuki S. Valgus toe deformity of fourth proximal phalanx due to osteochondroma treated with closing wedge osteotomy. Foot Ankle Surg. 20(4):e51–5.
43. Lee DY et al. Fibular lengthening for the management of translational talus instability in hereditary multiple exostoses patients. J Pediatr Orthop. 34(7):726–32.
44. Li M et al. Operative treatment of femoral neck osteochondroma through a digastric approach in a pediatric patient: a case report and review of the literature. J Pediatr Orthop B. 21(3):230–4.
45. Litzelmann E et al. Forearm deformities in hereditary multiple exostosis: clinical and functional results at maturity. J Pediatr Orthop. 32(8):835–41.
46. Malagon V. Development of hip dysplasia in hereditary multiple exostosis. J Pediatr Orthop. 2001;21(2):205–11.
47. Masada K et al. Operations for forearm deformity caused by multiple osteochondromas. J Bone Joint Surg Br. 1989;71(1):24–9.
48. Matsubara H et al. Correction and lengthening for deformities of the forearm in multiple cartilaginous exostoses. J Orthop Sci. 2006;11(5):459–66.
49. Mesfin A et al. Total hip and knee arthroplasty in patients with hereditary multiple exostoses. Orthopedics. 35(12):e1807–10.
50. Mintzer CM, Klein JD, Kasser JR. Osteochondroma formation after a Salter II fracture. J Orthop Trauma. 1994;8(5):437–9.
51. Mootha AK et al. Modified resection technique for proximal fibular osteochondromas. Orthop Traumatol Surg Res. 97(5):569–73.
52. Narula R et al. Soft tissue osteochondroma in elbow joint: A case report. Malays Orthop J. 6(1):53–6.
53. Nawata K et al. Knee deformities in multiple hereditary exostoses A longitudinal radiographic study. Clin Orthop Relat Res. 1995; 313:194–9.
54. Noonan KJ et al. Natural history of multiple hereditary osteochondromatosis of the lower extremity and ankle. J Pediatr Orthop. 2002;22(1):120–4.
55. Noonan KJ et al. Evaluation of the forearm in untreated adult subjects with multiple hereditary osteochondromatosis. J Bone Joint Surg Am. 2002;84-A(3):397–403.
56. Ofiram E, Eylon S, Porat S. Correction of knee and ankle valgus in hereditary multiple exostoses using the Ilizarov apparatus. J Orthop Traumatol. 2008;9(1):11–5.
57. Ofiram E, Porat S. Progressive subluxation of the hip joint in a child with hereditary multiple exostosis. J Pediatr Orthop B. 2004;13(6): 371–3.
58. Ozdemir HM et al. Tumors of the foot and ankle: Analysis 196 cases. J Foot Ankle Surg. 1997;36(6):403–8.
59. Porter DE et al. Clinical and radiographic analysis of osteochondromas and growth disturbance in hereditary multiple exostoses. J Pediatr Orthop. 2000;20(2):246–50.
60. Porter DE, Benson MK, Hosney GA. The hip in hereditary multiple exostoses. J Bone Joint Surg Br. 2001;83(7):988–95.
61. Pritchett JW. Lengthening the ulna in patients with hereditary multiple exostoses. J Bone Joint Surg Br. 1986;68(4):561–5.
62. Ramos-Pascua LR et al. Solitary proximal end of femur osteochondroma. An indication and result of the en bloc resection without hip luxation. Rev Esp Cir Ortop Traumatol. 56(1):24–31.
63. Rupprecht M et al. Temporary screw epiphyseodesis of the distal tibia: a therapeutic option for ankle valgus in patients with hereditary multiple exostosis. J Pediatr Orthop. 31(1):89–94.
64. Rupprecht M et al. Rebound of ankle valgus deformity in patients with hereditary multiple exostosis. J Pediatr Orthop. 2015;35(1): 94–9. doi: 10.1097/BPO.0000000000000224.

65. Salon A, Serra M, Pouliquen JC. Long-term follow-up of surgical correction of Madelung's deformity with conservation of the distal radioulnar joint in teenagers. J Hand Surg Br Eur Vol. 2000;25(1): 22–5.
66. Seo SG et al. Incidental findings on knee radiographs in children and adolescents. Clin Orthop Surg. 6(3):305–11.
67. Schmale GA, Conrad 3rd EU, Raskind WH. The natural history of hereditary multiple exostoses. J Bone Joint Surg Am. 1994;76(7): 986–92.
68. Shawen SB, McHale KA, Temple HT. Correction of ankle valgus deformity secondary to multiple hereditary osteochondral exostoses with Ilizarov. Foot Ankle Int. 2000;21(12):1019–22.
69. Shin EK, Jones NF, Lawrence JF. Treatment of multiple hereditary osteochondromas of the forearm in children: a study of surgical procedures. J Bone Joint Surg Br. 2006;88(2):255–60.
70. Shinozaki T et al. Rotational acetabular osteotomy in a dysplastic hip with femoral neck osteochondromas. Orthopedics. 1998;21(5): 588–90.
71. Siebenrock KA, Ganz R. Osteochondroma of the femoral neck. Clin Orthop Relat Res. 2002;394:211–8.
72. Skyrme AD et al. Osteochondroma of the distal tibia leading to deformity and stress fracture of the fibula. Foot Ankle Surg. 2003;9(2):129–30.
73. Song KS. Spontaneous regression of osteochondromatosis of the radius after lengthening of the ulna: a case report. J Pediatr Orthop. 2000;20(5):689–91.
74. Song SH et al. Modified Ilizarov technique for the treatment of forearm deformities in multiple cartilaginous exostoses: case series and literature review. J Hand Surg Eur Vol. 38(3):288–96.
75. Sorensen BW, Mikkelsen P. Arthrodesis of the distal tibiofibular joint for an osteochondroma in the fibula encroaching on the distal tibia and involving the talocrural joint: a case report. J Foot Ankle Surg. 51(5):664–5.
76. Spatz DK, Guille JT, Kumar SJ. Distal tibiofibular diastasis secondary to osteochondroma in a child. Clin Orthop Relat Res. 1997; 345:195–7.
77. Staal HM et al. Skeletal maturity of children with multiple osteochondromas: is diminished stature due to a systemic influence? J Child Orthop. 9(5):397–402.
78. Stanton RP, Hansen MO. Function of the upper extremities in hereditary multiple exostoses. J Bone Joint Surg Am. 1996; 78(4):568–73.
79. Stelling FH. The hip in heritable conditions of connective tissue. Clin Orthop Relat Res. 1973;90:33–49.
80. Takikawa K et al. Characteristic factors of ankle valgus with multiple cartilaginous exostoses. J Pediatr Orthop. 2008;28(7):761–5.
81. Tang ZW et al. Management of forearm deformities with ulnar shortening more than 15 mm caused by hereditary multiple osteochondromas. Eur J Orthop Surg Traumatol. 23(5):611–8.
82. Taniguchi K. A practical classification system for multiple cartilaginous exostosis in children. J Pediatr Orthop. 1995;15(5): 585–91.
83. Tatebe M et al. Clinical, radiographic, and arthroscopic outcomes after ulnar shortening osteotomy: a long-term follow-up study. J Hand Surg Am. 37(12):2468–74.
84. Thakur GB et al. Transfibular excision of distal tibial interosseous osteochondroma with reconstruction of fibula using Sofield's technique -A case report. Journal of Clinical Orthopaedics and Trauma. 3(2):115–8.
85. Vogt B et al. Reconstruction of forearm deformity by distraction osteogenesis in children with relative shortening of the ulna due to multiple cartilaginous exostosis. J Pediatr Orthop. 31(4):393–401.
86. Waters PM. Forearm rebalancing in osteochondromatosis by radioulnar fusion. Tech Hand Up Extrem Surg. 2007;11(4):236–40.
87. Watts AC et al. The association between ulnar length and forearm movement in patients with multiple osteochondromas. J Hand Surg. 2007;32(5):667–73.
88. Weber FP. Diaphysial aclasis or periosteo-osteo-dysplasia (multiple exostoses) with shortness of forearms. Proc R Soc Med. 1924; 17(Clin Sect):40–5.
89. Weber FP. A developmental deformity of the right forearm, similar to that sometimes met with in cases of multiple exostoses ("Diaphysial Aclasis" of Keith). Proc R Soc Med. 1925;18(Clin Sect):25–7.
90. Weiner DS, Hoyt Jr WA. The development of the upper end of the femur in multiple hereditary exostosis. Clin Orthop Relat Res. 1978;137:187–90.
91. Woodward MN et al. Subluxation of the hip joint in multiple hereditary osteochondromatosis: report of two cases. J Pediatr Orthop. 1999;19(1):119–21.
92. Yang YF et al. Reconstruction of the distal ulnar epiphysis with vascularized proximal fibula including epiphysis in children after osteochondroma resection: report of two cases. Plast Reconstr Surg. 132(5):784e–9e.
93. Yoong P, Mansour R, Teh JL. Multiple hereditary exostoses and ischiofemoral impingement: a case-control study. Skeletal Radiol. 43(9):1225–30.
94. Clement ND. Letter regarding "Effect of osteochondroma location on forearm deformity in patients with multiple hereditary osteochondromatosis". J Hand Surg Am. 38(7):1447–8.
95. Aur?gan JC et al. Children ankle valgus deformity treatment using a transphyseal medial malleolar screw. Orthop Traumatol Surg Res. 97(4):406–9.

Evidence-Based Treatment for Musculoskeletal Disorders in Children with Down's Syndrome

50

Christopher Talbot and Sattar Alshryda

Abstract

Down's syndrome is one of the most common chromosomal abnormalities in humans, occurring in approximately 1 in 1000 live births. Of the orthopaedic manifestations that can occur in these patients, hip instability, patellofemoral instability and atlanto-axial instability.

Ligamentous laxity, hypotonia, and joint hypermobility are thought to be primary causes. However, other patho-anatomical factors seem to play important roles. In the following chapter, we examined the evidence behind current treatments of the hip and patellofemoral instabilities.

Keywords

Trisomy 21 • Down's syndrome • Hip dysplasia • Hip dislocation • Hip instability • Dislocation • Pelvic osteotomy • Patellofemoral instability • Patellar instability • Patellar dislocation • Patellofemoral dislocation

Background

Down's syndrome or trisomy 21 is the commonest chromosomal disorder (1 in 1000) and sufferers have learning disabilities and a cluster of medical problems including cardiac anomalies, musculoskeletal problems, thyroid disorders, immunological and haematological disorders. Not all patients show every disorder and there is variation in the severity of these conditions among children with Down's syndrome.

Among the several musculoskeletal problems that a child with Down's syndrome can have, three are considered the most significant namely hip instability, recurrent patellar dislocation and atlanto-axial instability. The main focus of this chapter is on hip instability and patellofemoral instability. Nevertheless, surgeons and anesthetists should consider atlanto-axial instability in every child with Down's syndrome (Fig. 50.1).

Hip Instability in Down's syndrome

Hip dysplasia and instability is not common in children with Down's syndrome; about 1 to 7 % develops hip instability between walking age and adolescence. Although this is usually attributed to the generalized ligament laxity and hypotonia, other anatomical abnormalities may play roles [1]. Femoral anteversion is moderately increased and there is usually a near normal neck shaft angle [2–4]. The acetabulum is retroverted in part due to a deficient posterior wall (Fig. 50.2). Over time the centre-edge angle reduces and the tear drop widens as the hip starts to sublux [2, 4, 5]. If the hip remains untreated, a stiff, dislocated and painful hip will cause significant functional impairment (Fig. 50.3) [6, 7].

Hresko et al. [6] studied the natural history of hip dysplasia in Down syndrome in 65 adults and found walking ability decreased markedly with age. Even in those with normal hips, 12 of 13 were still able to walk at the age of 40 years,

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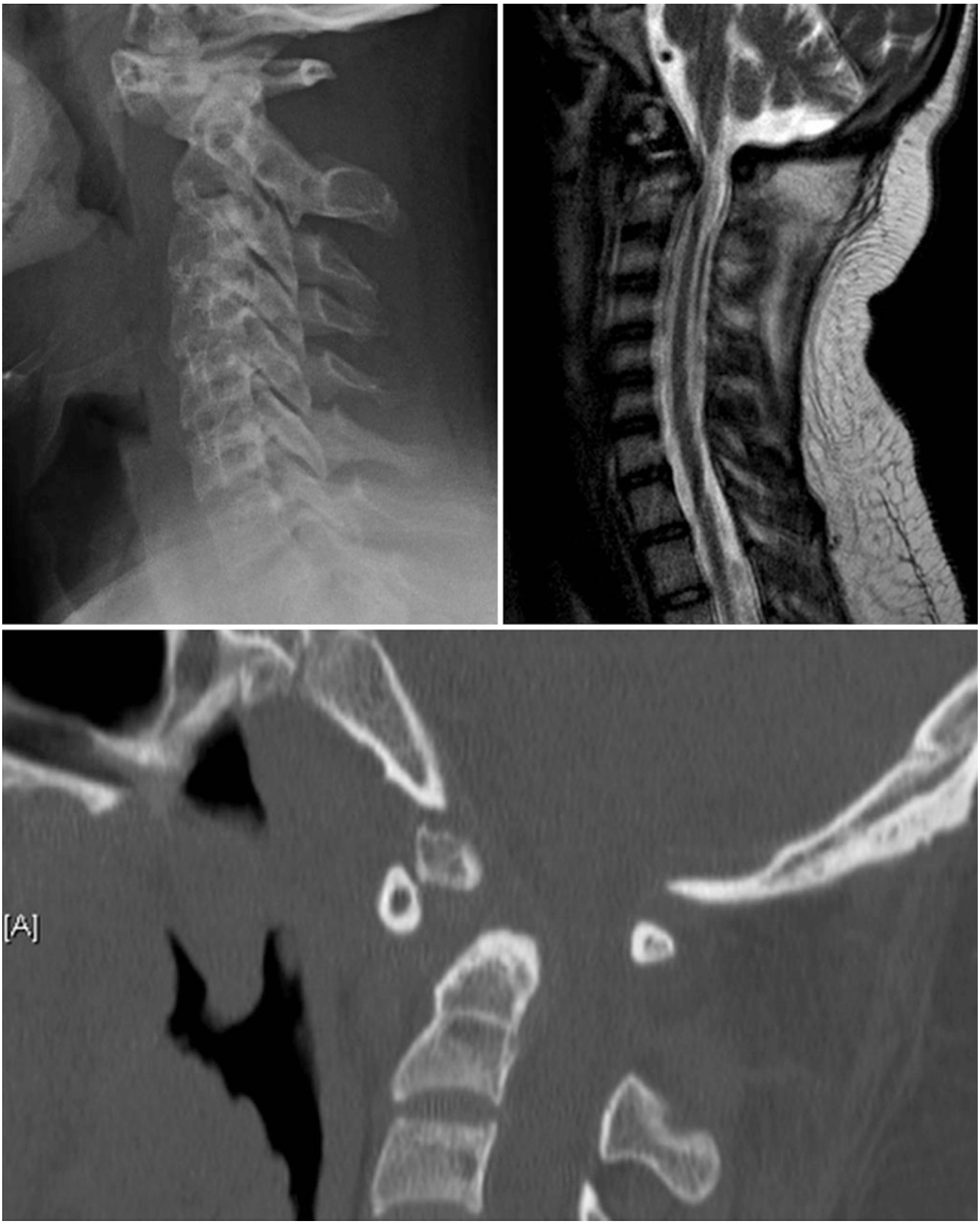

Fig. 50.1 Atlanto-axial instability in a child with Down's syndrome

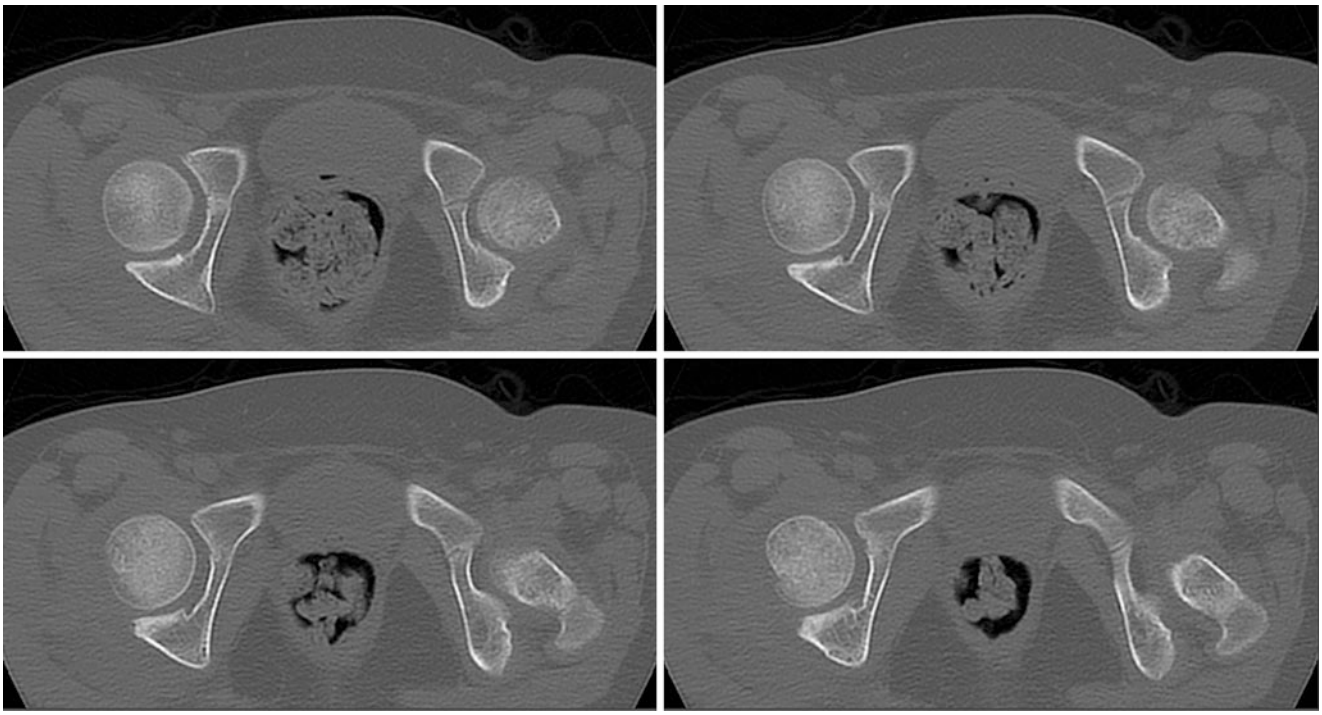

Fig. 50.2 Left acetabular dysplasia in a child with Down's syndrome

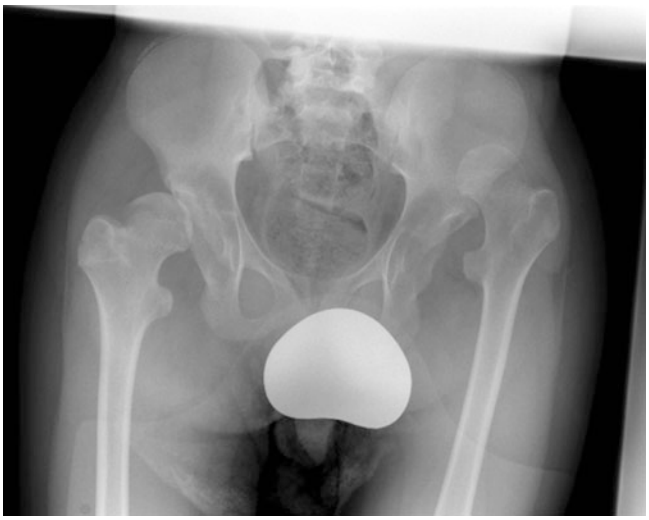

Fig. 50.3 Long-standing dislocated hips in a child with Down's syndrome

but by the age of 60 to 70 years only half were community ambulators. In contrast, patients with hip disease could not function as community ambulators after the age of 30 years.

Bennet et al. [7] recognised three phases in the natural history of hip subluxation in Down's syndrome. Initial phase (<2 years old): the hips are stable but there are features of hypermobility, and walking is usually delayed. Dislocation phase (2–10 years): the hip spontaneously begins to dislocate in a particular position without trauma and tends to spontaneously reduce (Fig. 50.4). Children may be brought for treatment at this stage because of clicking, an increasing limp or a complaint of giving way. Subluxation phase (>10 years): the hip starts to decentre and progressive acetabular dysplasia develops. Fixed phase (>15 years): if untreated, the hip invariably develops a painful fixed dislocation by the time the patient is in his late 'teens or early twenties.

Bennet reviewed a series of 28 patients (45 hips) with Down's syndrome who were treated for hip dislocations. A variety of methods were utilized including closed reduction and hip spica [5], capsular plication [2], femoral osteotomy and capsular plication [3]; Innominate osteotomy and capsular plication [4], Chiari osteotomy [4], Schantz osteotomies [1] femoral osteotomies [9], one in conjunction with a double pelvic osteotomy. Infection rate was high at 19 % and treatment was successful in only half of cases. They concluded that a bony procedure in isolation is insufficient. Capsular plication combined with femoral +/− pelvic osteotomy yielded the best results but even this was associated with a 50 % failure rate. They also found open operative intervention is required following poor results with closed reduction and spica and that Knight et al. [8] published a single surgeon series of 9 children (16 hips) with Down's syndrome. Patients were 5–7 years old at surgery with a mean follow-up of 5 years. All had a femoral varus derotation osteotomy to reduce the neck shaft angle (NSA) from a mean of 166° to 106°. In 2 hips, intra-operative instability

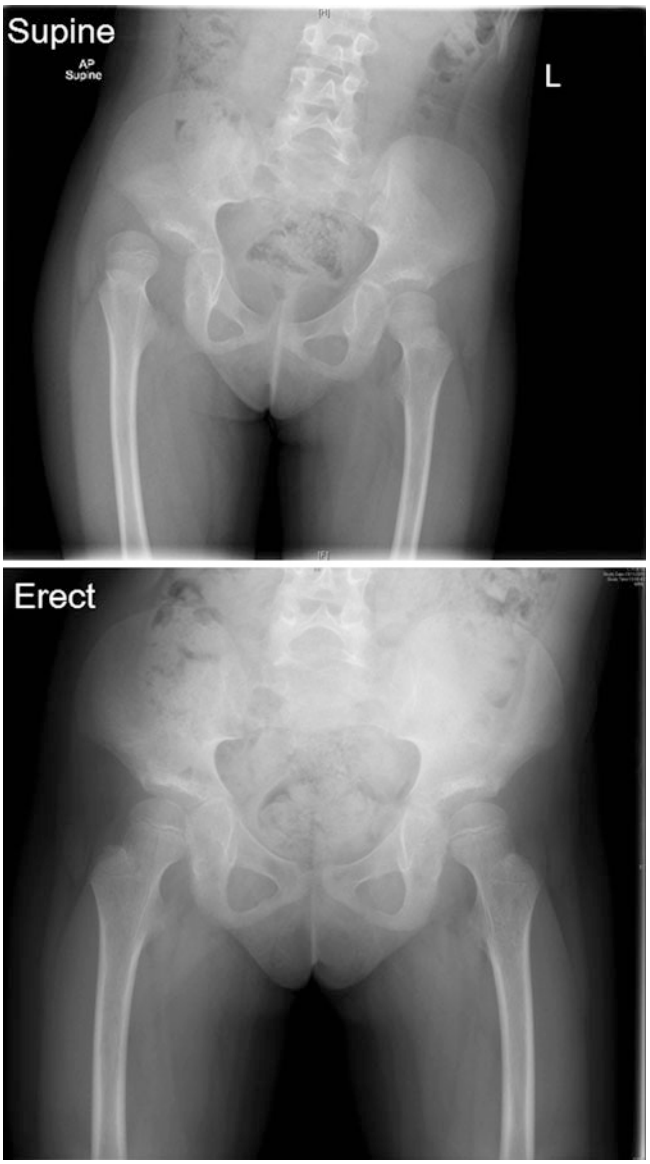

Fig. 50.4 Dislocatable right hip in a child with Down's syndrome

remained, requiring immediate periacetabular osteotomy and capsulorraphy. Postoperatively, all patients demonstrated an asymptomatic waddling gait, which persisted in 1 individual. Two patients had implant-related fractures, one hip developed arthritis and no hips redislocated. They concluded that performing a varus osteotomy to reduce the NSA to around 105° is effective treatment for hip dislocation in children with Down's syndrome who are under 7 years old. Implants should be removed at the appropriate time.

Sankar et al. [9] reviewed their experience of 35 hips in 29 patients (mean age 11.8 years). Twenty-five hips underwent a redirectional pelvic osteotomy (periacetabular or triple osteotomy) and 10 hips underwent a varus femoral osteotomy +/– Dega or shelf acetabuloplasty (Fig. 50.5). Although hips that underwent a redirectional pelvic osteotomy were radiologically worse, they had a better outcome with 92 % remaining stable. In contrast to 50 % only of dislocated hips remained stable following femoral osteotomy +/– acetabuloplasty.

In summary, hip instability and subsequent dislocation in children with Down's syndrome is an extremely challenging condition. Strong evidence to support various interventions or no intervention is lacking. Ambulation would be poor if hips are left unreduced (grade C). Closed reduction of the dislocated hip is insufficient, as is isolated capsular plication (grade B/C). In a young child (<7 years of age), a femoral varus osteotomy with judicious derotation, combined with an acetabular procedure (should instability persist) is recommended. In the older child, a redirectional pelvic osteotomy is required due to the deficient posterior wall and acetabular retroversion +/– femoral varus osteotomy (grade C). Post-operative infection is higher than normal and implants should be removed to minimize the risk of an implant-related fracture.

Patellofemoral Instability in Down's Syndrome

Patellofemoral instability prevalence children with Down's syndrome ranges between 4 and 8 % and it can be debilitating [10, 11]. It often causes recurrent falls, pain, poor quality of life. As with many other musculoskeletal manifestations of Down's syndrome, ligamentous laxity, hypotonia, and joint hypermobility are thought to be primary causes. An increase in the Q angle and dysplasia of the trochlear groove may also play a role [11, 12].

Dugdale and Renshaw [11] divided patellofemoral instability in patients with Down's syndrome into different grades by the degree of laxity (Table 50.1). Aside from this original paper, other have used this classification system [13, 14] when studying patellofemoral instability/dislocation in Down syndrome.

Is There Any Role for Non-operative Treatment?

There is limited published evidence regarding non-operative treatment for this condition. In a review article detailing orthopaedic manifestations of Down's syndrome, Caird et al. [15] generally recommend non operative treatment for patellofemoral instability in Down's syndrome patients as a first line treatment, especially in the initial phases, with low degrees of patellofemoral instability (grade II), no pain, and scarce functional disability. The treatment modalities include the knee sleeve and activity modification. Additionally, if pes planus is a component of the problem, arch supports are recommended. Quadriceps strengthening exercises and patellar stabilising braces may be of benefit to some individuals.

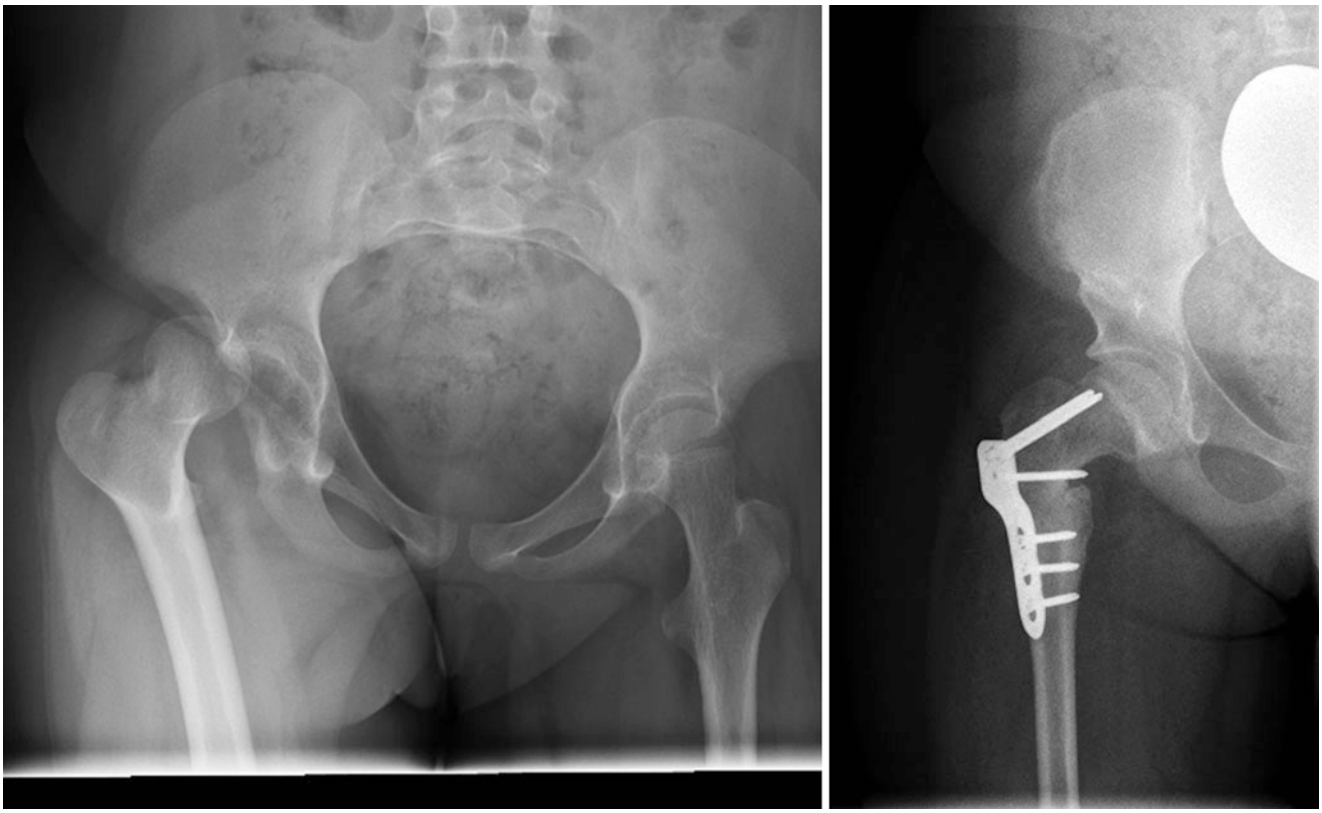

Fig. 50.5 Dislocatable right hip in a child with Down's syndrome treated with femoral VDRO and Dega pelvic osteotomy

Table 50.1 Patellofemoral stages according to Dugdale and Renshaw [11]

Grades	Description
I	Stable patellofemoral joint
II	Unstable or subluxed patella: patella subluxates laterally more than one half the patella width but does not dislocate
III	Dislocatable patella: the patella may be dislocated during examination
IV	Dislocated reducible patella: the patella is already dislocated, but the condition reversed manually
V	Dislocated irreducible patella: permanent loss of normal patellofemoral articular relationship, that cannot be reduced manually

Mendez et al. [16] found that nonsurgical treatments, including physical therapy and orthotics, were effective in maintaining or improving ambulation in most patients who were ambulatory before treatment. In this particular retrospective study, twenty-six dislocatable or dislocated patellae (type III, IV and V) were seen in 16 patients with Down's syndrome. Non operative treatment either maintained or improved the ambulatory status in 67 % of these knees with either fair or good ambulation. However, 80 % of the knees with poor ambulation did not improve. Operative treatment resulted in good ambulatory ability in 86 % of the knees with fair or poor preoperative ambulatory status. As such, in knees without significant deformity, the authors recommended surgical treatment, with attention paid to soft-tissue balancing and repositioning of the insertion of the patellar tendon. They did caution that degenerative arthritis eventually developed in patients with underlying deformities, despite the method of treatment of patellofemoral instability.

What Are the Operative Outcomes in Treating Patellofemoral Instability and Dislocation in Down Syndrome?

Despite the relative frequency of patellofemoral instability in Down's syndrome, there is little analysis in the literature, especially with regards to its surgical treatment. Several surgical techniques have been proposed, and the series reported in the literature are scarcely homogeneous in terms of procedures performed. A summary of operative outcomes is presented in Table 50.2. This represents the studies undertaken within the last 10 years.

As previously mentioned, in most knees presenting with fair or poor ambulatory status, conservative treatment has shown to be ineffective [16], and surgery is indicated. A

Table 50.2 Summary of results from literature within last 10 years for operative treatment of patella instability in Down syndrome

Study	Number of knees	Age (y)	Treatment	F/U (months)	Average Kujala	Modified Lysholm knee score	Patellar instability	Outcome	Complications
Joo et al. 2007	2	5.6	Four-in-one procedure	47	92	–	–	Good/excellent	50 %
Bettuzzi et al. 2009	10	10	Modified Roux Goldthwait Campbell	104	–	57.5 pre op 91 post op	–	–	0 %
Kocon et al. 2012	10	7.8	Green's quadricepsplasty (8 knees) Green's quadricepsplasty augmented with modified Galeazzi procedure (Semitendinosus tenodesis) (2 knees)	39	–	–	80 % (8/10) improvement in patella instability grade	Green's alone (6/8 satisfactory) Green's and modified Galeazzi (2/2 satisfactory) Pre-operative pain relieved in 33 %	No early post op complications related to surgery or wound healing 30 % limp post operatively

lateral retinacular release (LRR) can be performed [17]. Nevertheless, the long-term results reported in the literature for cases of unstable or hypermobile patellae treated by LRR are not satisfactory in about 50 % of the cases [18, 19]. This indicates that, particularly in patients with marked ligamentous laxity, LRR may not be sufficient, since it may actually worsen patellar instability.

In a growing child, surgical options for the transfer of the origin of the patellar tendon are limited by the open growth plate. Some authors have commented that depending on the degree of quadriceps dysfunction, lateral retinacular release, medial vector augmentation and patellar tendon alignment should be combined. Joo et al. [20] found the lateral patellar retinaculum, fascial bands and vastus lateralis to be very tight and vastus medialis was so deficient, that sufficient muscle advancement was not possible. The Insall technique [21] provided a secure repair which reduced the tension of the suture line to a minimum in their patients.

In addition, Joo et al. [20] reported their early results of the 'four-in-one' procedure in younger children in whom patellar dislocation with ligamentous laxity and trochlear dysplasia. The procedure included lateral release, proximal 'tube' realignment of the patella, semitendinosus tenodesis, and transfer of the patella tendon. Of the patients involved in the study, two patients had Down's syndrome. Mean follow up was 47 months. Post operatively, both had normal patella tracking. Outcomes were good and excellent, with both patients at final follow up having Type A trochlea dysplasia (Dejour classification) (see Fig. 12.1). One of the two patients had a dislocatable patella with forceful lateral stress. However, no dislocations occurred during voluntary full knee movement. Complications occurred in one patient, this was marginal skin necrosis. This healed after debridement and secondary closure.

Bettuzzi et al. [13] published on six children with Down's syndrome treated with a modified Roux Goldthwait Campbell procedure, in which parents of five children were satisfied with the outcome. Complete relief was achieved in one, and decrease of frequency of complaint was achieved in another. Falls during daily activities disappeared or decreased in all cases. However, limping persisted in two cases. No signs of recurrence of dislocation were noted over a mean follow up of 8 years and 8 months. An improved modified Lysholm Knee Scoring Scale was seen overall but care must be taken with this interpretation, as this scoring system is not validated in this cohort of patients. Furthermore, the authors found that the more severe grades of patella instability were associated with worse functional scores in comparison with less severe cases, consistent with other reports [20, 22]. With this finding the authors concluded that surgery should be recommended in cases presenting with good/fair function and little or no pain, particularly in younger children, to avoid progression towards higher grades of instability and functional worsening.

In addition to surgical intervention discussed, Kocon et al. [14] undertook a study to evaluate the mid-term results surgical intervention. Their inclusion criteria were as follows:

1. Irreducible or recurrent patellar dislocation
2. Unsuccessful conservative treatment
3. Knee pain
4. Significant limitation of locomotors abilities because of patellar instability
5. Associated diagnosis of Down's syndrome

Operative treatment involved Green's quadricepsplasty in six patients (eight knees). This involved a LRR, transfer of the medial head of quadriceps on the lateral part of the patella and a duplication of the medial patella retinaculum and joint capsule. In two cases, this was augmented with a dynamic correction of the knee extension mechanism – modified Galeazzi technique [23]. The two cases augmented were slightly older than the other patients in the study. Overall, 80 % of operated knees had a satisfactory result, defined when the correction of the position of the patella was achieved, resulting in an improvement in gait and mobility. Uneventful wound healing and no complaints during follow up were additional requirements for a satisfactory result. Eight knees had Green's procedure, in which two (25 %) were unsatisfactory. Both patients who underwent Green's and modified Galeazzi procedures had satisfactory results. The follow up for the latter was only 14 and 13 months, respectively. Whereas the average follow up for the patients having undergone Green's procedure was 45 months. Eight out of ten knees operated on had improved regarding patellar stability according to the Dugdale classification. Two patients, both group IV, remained in the same group following surgery. Authors concluded that Green's procedure in younger children led to satisfactory mid-term results. Whereas, in older children, Green's combined with a modified Galleazzi procedure may give more favourable results. However, this statement was made on very small numbers and the authors acknowledge the need for further experience to support this.

In conclusion, few studies have investigated this condition; the majority Level IV evidence. Further studies with longer follow up in children with Down syndrome and patellar instability are needed to determine the indications for non-operative and operative management of this condition, as well as more valid outcomes.

References

1. Woolf SK, Gross RH. Posterior acetabular wall deficiency in Down syndrome. J Pediatr Orthop. 2003;23(6):708–13.
2. Schoenecker JG. Pathologic hip morphology in cerebral palsy and Down syndrome. J Pediatr Orthop. 2013;33(Suppl 1):S29–32.

3. Shaw ED, Beals RK. The hip joint in Down's syndrome. A study of its structure and associated disease. Clin Orthop Relat Res. 1992; 278:101–7.
4. Kelley SP, Wedge JH. Management of hip instability in trisomy 21. J Pediatr Orthop. 2013;33(Suppl 1):S33–8.
5. Sankar WN, Schoenecker JG, Mayfield ME, Kim YJ, Millis MB. Acetabular retroversion in Down syndrome. J Pediatr Orthop. 2012;32(3):277–81.
6. Hresko MT, McCarthy JC, Goldberg MJ. Hip disease in adults with Down syndrome. J Bone Joint Surg Br. 1993;75(4):604–7.
7. Bennet GC, Rang M, Roye DP, Aprin H. Dislocation of the hip in trisomy 21. J Bone Joint Surg Br. 1982;64(3):289–94.
8. Knight DM, Alves C, Wedge JH. Femoral varus derotation osteotomy for the treatment of habitual subluxation and dislocation of the pediatric hip in trisomy 21: a 10-year experience. J Pediatr Orthop. 2011;31(6):638–43.
9. Sankar WN, Millis MB, Kim YJ. Instability of the hip in patients with Down syndrome: improved results with complete redirectional acetabular osteotomy. J Bone Joint Surg Am. 2011;93(20):1924–33.
10. Herring JA, editor. Tachdjian's pediatric orthopaedics. Philadelphia: Saunders Elsevier; 2013.
11. Dugdale TW, Renshaw TS. Instability of the patellofemoral joint in Down syndrome. J Bone Joint Surg Am. 1986;68(3):405–13.
12. Beals RK, Buehler K. Treatment of patellofemoral instability in childhood with creation of a femoral sulcus. J Pediatr Orthop. 1997;17(4):516–9.
13. Bettuzzi C, Lampasi M, Magnani M, Donzelli O. Surgical treatment of patellar dislocation in children with Down syndrome: a 3- to 11-year follow-up study. Knee Surg Sports Traumatol Arthrosc. 2008;17(4):334–40.
14. Kocon H, Kabacyj M, Zgoda M. The results of the operative treatment of patellar instability in children with Down's syndrome. J Pediatr Orthop B. 2012;21(5):407–10.
15. Caird MS, Wills BP, Dormans JP. Down syndrome in children: the role of the orthopaedic surgeon. J Am Acad Orthop Surg. 2006;14(11):610–9.
16. Mendez AA, Keret D, Macewen GD. Treatment of patellofemoral instability in Down's syndrome. Clin Orthop Relat Res. 1988;NA(234):148–58.
17. Mizobuchi RR, Galbiatti JA, Neto FQ, Milani C, Fujiki EN, Oliveira HC, et al. Ultrasonographic study of the femoro-patellar joint and its attachments in infants from birth to 24 months of age. Part II: children with Down syndrome. J Pediatr Orthop B. 2007;16(4):266–8.
18. Dandy DJ, Griffiths D. Lateral release for recurrent dislocation of the patella. J Bone Joint Surg Br. 1989;71(1):121–5.
19. Panni AS, Tartarone M, Patricola A, Paxton EW, Fithian DC. Long-term results of lateral retinacular release. Arthroscopy. 2005;21(5): 526–31.
20. Joo SY, Park KB, Kim BR, Park HW, Kim HW. The 'four-in-one' procedure for habitual dislocation of the patella in children: early results in patients with severe generalised ligamentous laxity and aplasis of the trochlear groove. J Bone Joint Surg. 2007;89-B(12):1645–9.
21. Insall J, Bullough PG, Burstein AH. Proximal "tube" realignment of the patella for chondromalacia patellae. Clin Orthop Relat Res. 1979;NA(144):63–9.
22. Gordon JE, Schoenecker PL. Surgical treatment of congenital dislocation of the patella. J Pediatr Orthop. 1999;19(2):260–4.
23. Moyad TF, Blakemore L. Modified Galeazzi technique for recurrent patellar dislocation in children. Orthopedics. 2006;29(4):302–4.

Part XII

Epilogue

Epilogue

51

James S. Huntley, Sattar Alshryda,
and Paul Banaszkiewicz

Abstract

We have been delighted to learn so much by virtue of the evidence elucidated (and recommendations made) by our co-authors. Conversely, in a few areas where we thought the subject might have advanced, we have been reassured that we are at least up to date. In setting out the state of the evidence for our subspecialty, we hope to have also provided a resource for discussion and a basis for future research.

Keywords

Evidence-based medicine • Paediatric orthopaedics • Levels of evidence • Systematic review • PICO • Grade of recommendation

We have been delighted to learn so much by virtue of the evidence elucidated (and recommendations made) by our co-authors. Conversely, in a few areas where we thought the subject might have advanced, we have been reassured that we are at least up to date. In setting out the state of the evidence for our subspecialty, we hope to have also provided a resource for discussion and a basis for future research.

In the introduction, we sketched out the evidence-based approach to topic areas: 'A clearly defined relevant question is required, followed serially by (i) identification of the studies/evidence by a thorough search of the literature, (ii) a critical appraisal of available evidence and its applicability to the clinical situation, and (iii) a balanced conclusion to the clinical problem and particular patient' [1]. We have encouraged a pragmatic approach, not as exhaustive as a formal systematic review [2], to help practicing surgeons derive evidence-based answers to important clinical questions [3]. As in an earlier text, the *Evidence for orthopaedics* [4], we advocated appraisal of the literature with assignation of levels of evidence and, for each Chapter a focussed summary 'Grades of recommendation' table. The importance and practicalities of assignation of levels of evidence [5] were broached in Chap. 2 on critical appraisal [6].

The subject matter is diverse and, as with most multi-author texts, there is heterogeneity of approach and depth. Authors vary in their perceptions and experience of evidence-based medicine, and modes of analysis of the literature. For some topic areas, a major problem is formulating the right question(s) [7]. As put eloquently in *Think like a freak* [8]: '*Before spending all your time and resources, it's incredibly important to properly define the problem-or, better yet, redefine the problem.*' Depending on the material to be addressed (but especially if the topic area is broad), a rapid scoping review, '*mapping the existing literature or evidence base*' [9] may be a useful antecedent to defining the parameters of a more formal subsequent review (eg question based on PICO format: **p**opulation, **i**ntervention, **c**omparison, **o**utcomes). In addition to the PICO formula, it is worth recognising that advice for resource-rich environments may differ substantially from that for resource-poor ones ie that the study *setting* can be important too [10].

Since the early 2000s, the reporting of levels of evidence linked to paediatric orthopaedic studies has yet to be associated

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with an increase in the proportionate quality of evidence *per se* [11]. An analysis of data from the *Journal of Paediatric Orthopaedics* 2009–2013 was interpreted as reflecting a trend in overall improvement in methodology, though this was largely as a result of the increase of self assignation of level of evidence, rather than a demonstrable change in proportions of higher level studies performed [12].

Grades of recommendation are fundamentally linked to levels of evidence – but the level of evidence is not the only important factor. For instance, in the treatment of simple bone cysts [13], percutaneous injection of steroid is superior to bone marrow on the basis of level 1 evidence (Grade recommendation assigned: B). Other interventions (eg curettage with or without grafting) may be superior but are accorded only Grade C. How then to advise the patient with a bone cyst? Of course, surgical decision-making (itself prone to bias) places the patient at the centre: it depends on the particular patient, the situation, the alternatives, and the benefits/risks.

The point is that there can be a difference between quality of evidence and strength of recommendation. This has been addressed by the GRADE (**G**rading of **R**ecommendations, **A**ssessment, **D**evelopment and **E**valuation) group [7, 10, 14–16]. Within the GRADE system there are four levels of evidence (similar to those elaborated by [5]): *high*, *moderate*, *low*, and *very low*. However, beyond the initial level allocation dictated by study design, there are five reasons to downgrade quality of evidence: *high likelihood of bias, indirectness of evidence, inconsistency of results, imprecision of results, high likelihood of publication bias*) [10, 17]. Conversely, there are also reasons to upgrade quality of evidence: *large effect, dose-response, all plausible confounding would reduce a demonstrated effect or would suggest a spurious effect when results show no effect*. In any case, the grading of recommendations remains a subjective process. Certainly we have argued the distinction between B and C on several occasions, enough perhaps to propose the great fudge – an extra grade: 'the B/C borderline'. It may be that in future, there is a better way to formulate Grades of recommendation.

With the growth in the surgical literature, there will be an ongoing requirement to critically appraise and abstract the evidence. In the future, evidence-based tomes such as this may become more formulaic and protocol-driven [18], minimizing risks of error and bias. There may be benefits to a stricter data extraction and review methodology, such as the more intensive approach of the PRISMA guidelines for systematic review [2]. Equally such an extended approach may confer untenable costs.

In the opening chapter of *Think like a freak*, the authors' remark:

'The fact is that solving problems is hard… …it takes a lot of time to track down, organize, and analyse the data to answer even one small question well.' [19]. A conservative estimate of 60 h work per chapter suggests upwards of 3000 h for this book. It is not just time; it is the time of practitioners with relevant expertise, itself a tightly limited resource. We record our immense gratitude to the authors for their labour in thinking, drafting, arguing, revising, and proofing – all freely given. This has truly been a collaborative and collective endeavour.

References

1. Alshryda S, Huntley JS, Banaskiewicz P. Introduction, Chapter 1. In: Evidence-based paediatric orthopaedics. London: Springer; 2016.
2. Liberati A, Altman DG, Tetzlaff J, Mulrow C, Gotzsche PC, Ioannidis JPA, Clarke M, Devereaux PJ, Kleijnen J, Moher D. The PRISMA statement for reporting systematic reviews and meta-analyses of studies that evaluate healthcare interventions: explanation and elaboration. Ann Intern Med. 2009;151:W-65-W-94.
3. Phillips B. Towards evidence based medicine for paediatricians. Arch Dis Child. 2001;85:252–5.
4. Limb D, Hay SM, editors. Evidence for orthopaedic surgery. Shrewsbury: TFM Publishing; 2006.
5. Wright JG, Tolo VT. JBJS and evidence-based orthopaedics. J Pediatr Orthop. 2012;32:S101–3.
6. Pollock R, Sayed-Noor AS. Critical appraisal of a published paper, Chapter 2. In: Evidence-based paediatric orthopaedics. London: Springer; 2016.
7. Guyatt GH, Oxman AD, Kunz R, Atkins D, Brozek J, Vist G, Alderson P, Glasziou P, Falck-Ytter Y, Schunemann HJ. GRADE guidelines: 2. Framing the question and deciding on important outcomes. J Clin Epidemiol. 2011;64:395–400.
8. Levitt SD, Dubner SJ. What's your problem?, Chapter 3. In: Think like a freak. London: Penguin Random House; 2014. p. 49–64.
9. Armstrong R, Hall BJ, Doyle J, Waters E. 'Scoping the scope' of a Cochrane review. J Public Health. 2011;33:147–50.
10. Guyatt G, Oxman AD, Akl EA, Kunz R, Vist G, Brozek J, Norris S, Falck-Ytter Y, Glasziou P, deBeer H, Jaeschke R, Rind D, Meerpohl J, Dahm P, Schunemann HJ. GRADE guidelines: 1. Introduction – GRADE evidence profiles and summary of findings tables. J Clin Epidemiol. 2011;64:383–94.
11. Cashin MS, Kelley SP, Douziech JR, Varghese RA, Hamilton QP, Mulpuri K. The levels of evidence in pediatric orthopaedic journals: where are we now? J Pediatr Orthop. 2011;31:721–5.
12. Murphy RF. Levels of evidence in the Journal of Pediatric Orthopaedics: update and comparison to the Journal of Bone and Joint Surgery. J Pediatr Orthop. 2015;35:779–81.
13. Alshryda S, Wright J. Evidence on simple bone cyst, Chapter 44. In: Evidence-based paediatric orthopaedics. London: Springer; 2016.
14. Guyatt GH, Oxman AD, Kunz R, Falck-Ytter Y, Vist GE, Liberati A, Schunemann HJ, GRADE Working Group. Going from evidence to recommendations. BMJ. 2008;336:1049–51.
15. Guyatt GH, Oxman AD, Kunz R, Vist GE, Falck-Ytter Y, Schunemann HJ, GRADE Working Group. What is "quality of evidence" and why is it important to clinicians? BMJ. 2008;336:995–8.
16. Guyatt GH, Oxman AD, Vist GE, Kunz R, Falck-Ytter Y, Alonso-Coello P, Schunemann HJ, GRADE Working Group. GRADE: an emerging consensus on rating quality of evidence and strength of recommendations. BMJ. 2008;336:924–6.
17. Bayat Z, Lang E. Guidelines helping clinicians make the GRADE. Intern Emerg Med. 2015;10:87–92.
18. Phan K, Tian DH, Cao C, Black D, Yan TD. Systematic review and meta-analysis: techniques and a guide for the academic surgeon. Ann Cardiothorac Surg. 2015;4:112–22.
19. Levitt SD, Dubner SJ. What does it mean to think like a freak?, Chapter 1. In: Think like a freak. London: Penguin Random House; 2014. p. 1–18.

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