Ian A. Trail
Andrew N. M. Fleming
Editors

Disorders of the Hand

Volume 4: Swelling, Tumours, Congenital Hand Defects and Surgical Techniques

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Preface

In recent years there have been significant advances in the understanding and treatment of disorders of the hand and wrist. This has resulted in a significant improvement in the quality of life for many patients. The authors who have produced this text were chosen as they are hand surgeons who have led many of these exciting developments in the management of both elective and trauma hand surgery. All are internationally respected.

The topics covered are well illustrated with images, radiographs and line drawings and provide practical guidance on surgical procedures. The references at the end of each chapter have been chosen as they are either classic papers or are the most relevant to modern surgical management.

Thus we hope that we have produced a book that will enable improved care for current patients with hand and wrist complaints and inspire surgeons to think in greater detail about treatment options that will provide even better care in the future.

Finally, we would like to thank all the contributors as well as Diane Allmark for her help, but also our families for their patience and support.

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We are also particularly grateful to Springer for allowing us to pursue this project and would like to especially thank Rachel Glassberg for all her helpful advice and prompting.

Finally we would like to thank our secretaries, particularly Diane Allmark, and respective families who, for longer than we dare think, have put up with us reading and re-reading manuscripts on what they think is only a small part of the body!

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Part III Surgical Techniques

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Part I

Swelling and Tumours

Ganglia of the Hand and Wrist

1

Joseph Dias

Keywords
Pathology • Diagnosis • Dorsal wrist ganglion • Aspiration • Excision • Recurrence • Palmar ganglion • Complications

Introduction

A ganglion is a peculiar disorder, which presents principally with a swelling of the wrist or finger. It is so common that many are treated with good advice and reassurance and in the United Kingdom around half the patients who present in orthopaedic or hand clinics with this condition no longer undergo surgery.

Incidence

A ganglion cyst accounts for between 50 and 70 % of soft tissue hand swellings and is the second most common elective disorder presenting within the United Kingdom with an incidence rate of between 44 and 50 per 100,000 per year. This accounts for over 25,000 cases each year. It occurs predominantly in women in the third and fourth decades of life.

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Distribution

In an audit of ganglia, conducted in the Trent region of the United Kingdom, 858 consecutive cases presenting to hand clinics were included. Forty six per cent of these affected the dorsum of the wrist, 28 % affected the palmar aspect of the wrist, 15 % were related to the sheath of the flexor tendon near the head of the metacarpal and 4 % were mucous cysts at the base of the nail in one of the fingers. The remaining ganglia had miscellaneous locations either in the palm or on the ulnar side of the wrist. Occasionally ganglia also appeared on the front of the finger or extended to the dorsum of the middle phalanx. This however was very uncommon. This distribution is similar to other reports [1].

Clinical Pearl

Typically present in females in the third and fourth decades of life.

Appearance is the predominant presenting complaint

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Pathology

Although the aetiology is not clearly defined the pathology is well known. A ganglion arises from collagenous tissue from either a ligament, capsule or tendon sheath. The ganglion itself has a wall and a stalk and contains clear mucinous fluid of jelly like consistency. It presents as a small well-defined hemispherical lump. This lump illuminates when light is shone through it.

The wall of the ganglion is made up of compressed collagen with sparse flat cells without any epithelial or synovial cells lining its surface [2]. The wall is created by the compression of stromal tissue and the cyst contains jelly. The jelly itself is usually clear unless there has been previous bleeding into the ganglion as a result of trauma; this would include a failed needle aspiration. The jelly itself is mucinous and contains glucosamine, mucin, hyaluronic acid with albumin and globulin. The consistency of the jelly is varied. The cyst usually has a stalk and this leads to a collagenous structure which is either a sheath, ligament or capsule. In the vicinity of the attachment of the stalk changes within the underlying structure may be present with clefts containing mucin [3]. There may be daughter ganglion cysts, which are much smaller than the main ganglion and which surround the attachment of the stalk.

Aetiology

The exact cause of a ganglion cyst has not been properly established. Over the years, various theories have been proposed. It has been suggested that a ganglion is a herniation from within the joint or is due to degeneration in the joint [4]. As far back as 1893, they were considered to be a cyst [5] caused by mucoid degeneration (Fig. 1.1).

A ganglion may be caused by mucoid degeneration [5] with cells somehow triggered to form the ganglion content. Fibrillation of collagen, mucin lakes within clefts, absence of synovial cells lining the ganglion cyst and that only 50 % communicate with the joint are all factors that support this hypothesis. The factors against

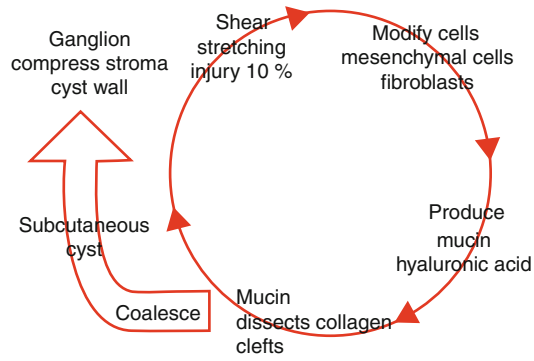

Fig. 1.1 Pathophysiology as described by Carp and Stout, 1928

mucoid degeneration include that it is usually a self-limiting disorder, ganglia are usually solitary and they can occur in young adults, including children, who may also get a recurrence after intervention.

In most cases the onset is spontaneous but in some 10 % of cases the onset of a ganglion may be triggered by an injury. There is probably excessive shear between planes of a collagenous structure in some patients, particularly those with joint laxity. This may initiate metaplasia of cells in the collagenous structure and the fibroblasts and mesenchymal cells start producing more hyaluronic acid and mucin. As this substance is produced and extruded from the cell it dissects neighbouring collagen and causes clefts within which this jelly-like material accumulates. These clefts then coalesce. The jelly like material compresses the surrounding stroma forming a cyst wall, thereby forming a small ganglion [2]. This ganglion grows in size and may herniate out of the capsule and come to lie in the subcutaneous tissue.

Clinical Presentation

The typical patient with a ganglion is a young female in her 30s or 40s. Westbrook et al. [6] explored the reasons why patients presented to hospital. They found that around 40 % presented because they did not like the appearance of the swelling, whereas 28 % thought it was sinister. A quarter had some discomfort related to the gan-

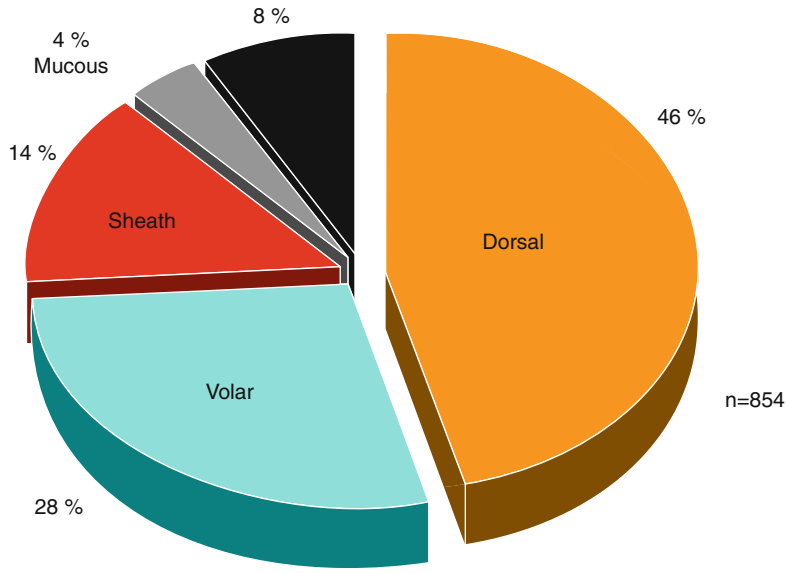

Fig. 1.2 Types of ganglia in the wrist and hand and their distribution. The distribution of wrist and hand ganglia is shown. Mucous cysts of the terminal phalanx account for the least and dorsal wrist ganglia are the most common. The *black pie* represent 8 % of miscellaneous ganglia. These formed the basis of two reports [1, 2]

glion while only 8 % presented with either altered sensation or a significant impact upon their ability to perform tasks. It is important to be aware of the reasons why patients present with a ganglion as very often all they need is reassurance.

The patients usually present with a spontaneous onset swelling either over the wrist or at the base of a finger (Figs. 1.2 and 1.3). The swelling then slowly grows in size. The size of the swelling fluctuates over time. Patients often say that the swelling can increase after a spell of increased activity. The swelling, when it is large, can cause a clear cosmetic blemish. Women, especially when they are young, do not like the appearance as it makes their wrist and hand stand out and look abnormal.

Although patients do not spontaneously complain of weakness, they do admit to it when asked. There is usually a feeling of weakness, particularly when the ganglion is large, and just before the ganglion appears. This weakness changes depending upon the patients' level of activity. Some patients, when asked, also complain of a feeling of stiffness in the involved joint. One in four patients described experiencing discomfort and pain, usually preceding the appearance of the ganglion. The pain is mild and aching in character without any obvious exacerbating or relieving factors. This pain is eased when the ganglion shrinks in size.

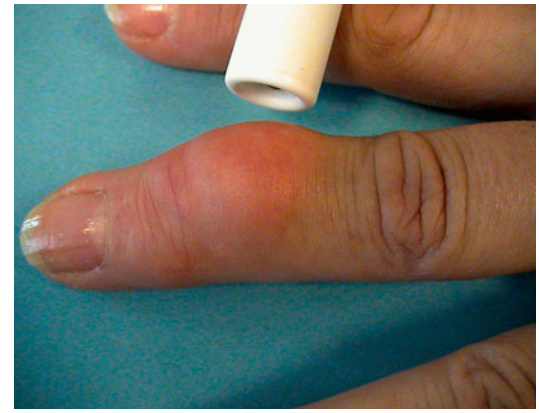

Fig. 1.3 Finger ganglion presenting on the dorsal surface. This is an uncommon site for a finger ganglion. It can often arise from the A4 pulley at its side but herniate through the fascia to present on the dorsum of the digit. It transilluminates thereby confirming the diagnosis in most cases

Dorsal Wrist Ganglion

Presentation

A typical patient who presents with dorsal wrist ganglion is usually female in the second, third or fourth decades of life. In the audit of dorsal wrist ganglia, conducted in the Trent region, women outnumbered men four to one.

Patients usually say that the swelling becomes more prominent when the wrist is palmar flexed. The swelling is just distal to Lister's tubercle and is hemispherical in shape without any inflammatory signs around it. It can often feel quite tense. The lump is, however, rarely tender or associated with any restriction of wrist movement. Often the patient has generalised joint laxity assessed by the Beighton's score [7]. Trans-illumination of the ganglion with a light source will result in the cyst "lighting up". Otherwise, whilst patients may have joint laxity, it is unusual for them to have definitive scapholunate instability.

Further investigations are generally not needed. If, however, the wrist is stiff when compared to the contralateral side, then obtaining a radiograph of the wrist is important to ensure that there is not a more generalised disorder, such as an arthritic wrist.

A diffuse non-hemispherical swelling, located only on the dorsum of the wrist usually suggests teno-synovitis of the common extensor synovial sheath. When examined this swelling can be made to fluctuate across the extensor retinaculum. It may also trans-illuminate and therefore it could be confused with a dorsal wrist ganglion. Another swelling that is often hemispherical, soft and may trans-illuminate is a lipoma. This however tends to be much more mobile in two planes and its consistency is softer. A diffuse swelling on the radial side of the wrist can also be related to degenerative arthritis of the radio scaphoid joint but this can be usually identified due to the restriction of wrist movement compared to the opposite side. A ganglion of the wrist should not be confused with DeQuervain's tenosynovitis or peritendinitis crepitans. Another disorder that can be confused with a ganglion is a bony carpal boss [8, 9] which lies at the base of the index or middle finger metacarpals. This too, presents as a swelling on the back of the wrist, although it is usually distal from the usual location of a ganglion. There may, however, be an overlying ganglion cyst arising from the bony protuberance. Generally, however, the consistency of the swelling is hard. Circumduction may demonstrate the tendons snapping over this lump.

A ganglion may occasionally not trans-illuminate, particularly when there has been bleeding into the sack. If the swelling does not trans-illuminate therefore, further investigation is important.

Any ganglion can be clearly seen on an ultrasound scan [10] or on a magnetic resonance image [11]. On the ultrasound the appearance is of a discrete well defined hypoechoic area. On the MRI scan the appearances of a discrete well defined dark area on T1 scan which lights up and is white on a T2 scan.

Treatment

Once the diagnosis has been established, then the treatment options can be discussed with the patient. These treatment options include reassurance, aspiration or surgery to excise the ganglion. Often once patients are told that they have a ganglion and that it is not sinister, they are happy to live with the symptoms and just be reviewed after an interval to ensure that they do not have any other significant disability. If, however, pain or appearance is of significant concern, then the surgeon may consider intervention, either aspiration with or without the injection of chemical substances or alternatively surgical excision.

Clinical Pearl

Differential diagnosis;
- DeQuervain's tenosynovitis
- Peri-tendinitis crepitans
- Lipoma
- Osteoarthritis of the radio-carpal joint
- Carpal bossing

Recurrence rate after aspiration, approximately 60 %.

Recurrence rate after surgery between 35 and 40 %

Aspiration (Fig. 1.4)

The aspiration of the dorsal wrist ganglion is undertaken using a wide bore needle. Very often, no local anaesthetic is required. A quick stab into

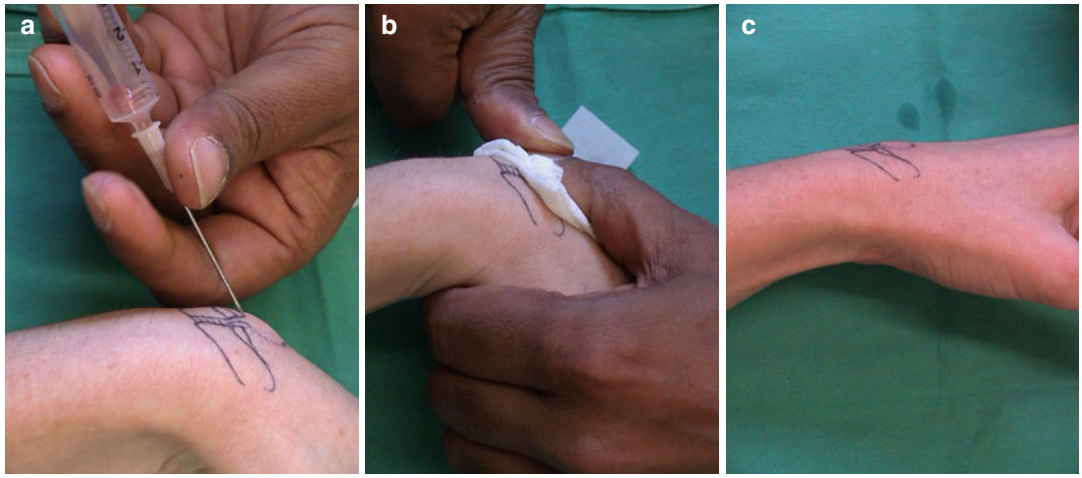

Fig. 1.4 Aspiration of the dorsal wrist ganglion. A dorsal wrist ganglion being aspirated (**a**) dispersed after multiple punctures (**b**) and showing the ganglion disappeared. (**c**) We usually maintain pressure with a pressured pad and a crepe bandage for the first day

the ganglion and aspiration will draw out the jelly from the ganglion collapsing the swelling. The ganglion disappears immediately. It is possible to do multiple perforations of the ganglion wall by perforating it with the needle and then compressing it firmly using gauze over the perforation site. The pressure should be maintained for a period of a few minutes. The principle here is to cause the ganglion walls to collapse and hopefully with sustained pressure obliterate the space.

Rarely substances have been injected into the ganglion cyst. These include the instillation of steroid. However, studies have not demonstrated any better outcome when steroid has been injected into the ganglion [12]. Hyaluronidase has also been used [13, 14]. There is certainly no established benefit in aspirating recurrent ganglia [15].

After aspiration a third of ganglia reappear within 2 weeks. Another third reappear within 6 months and 15 % each would reappear between 6 months and a year and after a year.

Excision of a Dorsal Wrist Ganglion

The alternative intervention is excision of the ganglion cyst. This can be done either openly through a cut on the dorsum of the wrist or alternatively arthroscopically. The excision can either be a simple excision of the swelling alone or include identification of the stalk and tracing the stalk back to its origin.

Surgical excision is straightforward and is conducted either through a zigzag longitudinal incision or a transverse incision centred over the swelling itself. Any large veins are diathermised. The walls of the ganglion are then carefully defined. The tendons are moved away from the ganglion. Which tendons need to be moved depends upon the position of the ganglion. It is usual, however, for the ganglion to lie between the third and fourth extensor compartments.

The ganglion is carefully dissected off the capsule and then an arthrotomy is undertaken to identify the stalk of the ganglion. This usually arises from the dorsal scapholunate ligament and often there are daughter ganglion cysts surrounding its attachment, which need to be scraped off the ligament. Care must be taken that the ligament itself is not excised or divided [16]. Usually a rongeur or acurette will help dissect the stalk from the dorsal scapholunate ligament. Haemostasis is then obtained. The capsule does not need to be formally closed. The skin is closed using absorbable sutures and steri-strips. At the end of the surgery local anaesthetic is infiltrated into the wound around the scapholunate ligament and in the subcutaneous tissue. The wrist and hand is wrapped in a bulky wool and crêpe

bandage. For a couple of hours after surgery the wrist and hand is elevated in a Bradford sling. The patient is encouraged to use the hand from the very outset.

Immediate after-care includes instructing the patient to use the hand without restrictions. We prefer to retain the bandages for about a week.

If the patient is very lax jointed with a high Beighton score, we often advise them to use a Futuro splint for the first 3–4 weeks to restrict wrist movement in the initial post-surgical period, in the hope that this may promote some stiffness and reduce movement at the scapholunate interval. We hope that this may decrease recurrence. However, this hypothesis has not been proven.

Arthroscopic Excision of a Dorsal Wrist Ganglion

This ganglion may be excised arthroscopically [17]. Generally authors recommend excision of the ganglion stalk as well as the intra-articular part of the cyst. Other authors [18] have recommended the excision of 1 cm of the dorsal capsule at the same time.

In 2001, Nishikawa et al. [19] described the technique of arthroscopic diagnosis and treatment of dorsal wrist ganglion in 37 patients with a mean follow-up of 20 months and without any complications. They approached this through two arthroscopic portals into the radiocarpal joint using a 2.7 mm arthroscope introduced between the first and second extensor compartment and a probe introduced into the wrist joint from the ulnar side between the fourth and fifth extensor compartments. The radial portal does risk damaging the superficial branches of the radial nerve.

Using an arthroscopic shaver the area over the scapholunate ligament was debrided and a 1 cm diameter section of the dorsal capsule resected, even if the ganglion itself could not be clearly identified. No attempt was made to remove the ganglion sac in the superficial layers. In some ganglia the stalk was clearly seen, in others the stalk ballooned into the wrist joint with external compression but in others the stalk could not be identified. It is, however, some times difficult to identify the stalk [20]. As a consequence, other authors [21] have suggested injecting colour dye into the ganglion to improve visualisation. Others have used intraoperative ultrasonography [22] to aid with identification of ganglion.

In the same study [20] the authors have recommended that the mid carpal joint should be explored routinely, as this was required to completely excise any dorsal wrist ganglion.

Others have noted a high prevalence of TFCC degenerative changes when a ganglia is present [23].

Generally, the rate of recurrence following arthroscopic excision of ganglia is extremely low [20, 24, 25]. The exact reason for this, when compared to open excision, remains unclear. A prospective randomised comparison between arthroscopic and open dorsal ligament excision, found no difference between the recurrence rate at 12 months [25].

Persistence and Recurrence

Regardless of treatment a ganglion may either persist or recur. The symptoms of pain, weakness and stiffness can persist after all interventions including reassurance, aspiration or surgery. Patients can also continue to regard their ganglion as unsightly.

Results

Reassurance

If the ganglion is left untreated then almost 40 % will disappear spontaneously.

In 1954, McEvedy [26] reported the outcome of 21 dorsal wrist ganglia which did not have any surgery. These were reviewed at 10 years and 41 % had resolved spontaneously with no intervention. In one report [27] 40/101 ganglia treated with reassurance disappeared when assessed at a mean of 6 years after presentation. The rate of spontaneous resolution in children is much greater with 22/28 (76 %) ganglia disappearing [28]. In another study [29] 55 dorsal wrist ganglia were treated without surgery at 6 years. It was noted that 23 had resolved spontaneously. It therefore appears that, when patients

are reassured, that two out of every five dorsal wrist ganglia disappear without treatment in adults and this rate increases to three out of every four in children.

Of the ones that disappear it is very uncommon for this to happen early. We found that 56 % disappear within 6 months, 19 % between 6 months and a year and 13 % take over 1 year to disappear. As a consequence, most that will disappear spontaneously do so within 1 year.

The remaining 60 % which persist behave in two ways. Either they increase and decrease in size, or they remain unchanged merely changing somewhat in size. Thirty eight per cent of the ganglia that persist when nothing is done change size in an episodic fashion usually becoming smaller while 50 % are always present.

Clinical Pearl

Approximately 40 % of ganglions disappear spontaneously.

In children, 75 % of ganglions disappear spontaneously.

After reassurance, 7 % continue to feel that their lump is unsightly and around a quarter of patients, regardless of intervention, will continue to complain of some discomfort in the dorsum of the wrist and hand.

Aspiration

The recurrence rate is 61 % with aspiration [29]. After aspiration alone McEvedy [26] reported on 43 dorsal wrist ganglia at 10 years and noted an 18 % recurrence rate. Nelson et al. [30] reported a 40 % recurrence rate after aspiration with or without injection of steroids and or hyaluronidase. Dias et al. [29] reported 61 % recurrence after aspiration alone, regardless of whether substances were injected in addition.

Excision

The recurrence rate is 36 % after surgery [29]. The recurrence rate after surgery has been reported extensively in the literature over a long period of time [26, 27, 29–31]. McEvedy [26] did a 10-year follow-up of patients who had a dorsal wrist ganglion excised and found that 40 % of these recurred. Zachariae and Vibe-Hansen [27] reported 29 % recurrence rate, following open surgery, undertaken by experienced surgeons compared to 37 % when done by inexperienced surgeons. A more recent study documented a recurrence rate of 36 % at 6 years [29].

Studies have demonstrated that ganglia that do reappear after surgery do so within 6 months, with half appearing within 6 months and another 20 % appearing between 6 months and a year. Only one third of ganglia reappear after a year.

The patterns of recurrence after intervention is that 57 % recur slowly, only 5 % recur suddenly after a single identifiable episode of trauma and 38 % recur suddenly but without injury.

Symptoms After Intervention

Regardless of treatment the number of patients complaining of pain, weakness, stiffness and concern about the appearance all decreased over 6 years. More patients complained of weakness after surgery, than after aspiration or reassurance. In the audit conducted in the Trent region 60 % of patients with a dorsal wrist ganglion complained of pain, 34 % of a feeling of weakness and 15 % felt it was significant. Eighteen per cent complained of some weakness and 12 % said that the wrist felt stiff.

Disability assessed by the Patient Evaluation Measure after intervention was similar at 19 % where zero represents no disability, after surgery, 15 % after aspiration and 15 % after reassurance alone [29].

Satisfaction With Treatment

The surgical group had higher satisfaction [29] but also had a higher incidence of residual symptoms. As the study was not a prospective randomised study it is difficult to know whether there was selection bias, which may explain this.

It is of note that, even if the ganglion recurs, patients are more satisfied after a surgical excision with 88 % being satisfied with the outcome of the intervention, 80 % are satisfied after aspiration and only 46 % are satisfied when they are merely reassured. This reflects the expectations patients have when they attend a surgical clinic.

There was no difference between men and women on the degree of satisfaction expressed. As expected, recurrence after intervention was associated with more dissatisfaction with 32 % of patients who had a recurrence after treatment expressing dissatisfaction while 17 % of those who did not have a recurrence expressed dissatisfaction with treatment [29].

Complications

We identified an 8 % complication rate. These were usually related to the wound with very few nerve injuries. There were 3 % complications after aspiration but these were minor and short lived. If steroids are used then patients run the risks of lipoatrophy, telengiectasis and depigmentation if the steroid leaks into superficial tissues.

It is clear that one in five patients regardless of intervention will continue to have some pain and weakness at 5 years, although the rate improves over this period of time. More patients who are merely reassured continue to remain concerned about the appearance of the hand. If surgery is done then 8 % have some complication, usually minor. Particular care needs to be taken not to damage the dorsal divisions of the superficial radial nerve.

Prior to treatment, we advise patients of the pros and cons of the different interventions and, in particular that the symptoms of pain, stiffness and weakness may be related to whatever is causing the ganglion rather than to the ganglion itself. We also warn our patients of the recurrence after intervention, but reassure them that the lump is not sinister.

Occult Wrist Ganglion

Occasionally patients, usually young women, present with pain on the dorsum of the wrist without an obvious cause. The possibility of an occult dorsal wrist ganglia needs to be considered. These are easily investigated either using an ultrasound or MRI scan [32–36]. Depending upon the level of symptoms patients may need open or arthroscopic excision of the ganglion or an ultrasound guided aspiration.

In a retrospective review, Steinberg et al. [37] reported that 18 out of 21 patients having surgical exploration for dorsal wrist pain with normal radiographs had an occult scapholunate wrist ganglion and all were improved after surgery.

It must be noted, however, that occult ganglia may be asymptomatic and more are related to the palmar scapholunate ligament than the dorsal scapholunate ligament. Lowden et al. [38] reported 53/103 asymptomatic wrist ganglia picked up on MRI scans of the wrists of asymptomatic volunteers, only 14 % of which were dorsal.

Palmar Wrist Ganglia

A palmar wrist ganglion is the second most common ganglion around the wrist and hand. It accounts for between 23 % [30] and 38 % of wrist ganglia.

Presentation

These present with a swelling sometimes associated with pain in the front of the wrist usually on the radial side and usually proximal to the scaphoid tuberosity. Two thirds of these ganglia arise from the palmar aspect of the scapholunate joint and one third from the scaphotrapezium joint. Patients can complain that the wrist feels a bit weak. Like a dorsal wrist ganglion, these are more common in women.

The ganglion cyst usually lies between the radial artery, which can be palpated on the radial aspect of the ganglion and the flexor carpi radialis tendon, which lies on its ulnar side. The history and examination findings are similar to those described for the dorsal wrist ganglion.

Reassurance

After reassurance alone of 38 patients with a palmar wrist ganglia, who were followed up for 6 years, 47 % had a persistent ganglion. That is

exactly similar to the rate of persistent ganglia after aspiration [39].

The feeling of weakness was similar between those that had their ganglion excised, aspirated or those patients who were merely reassured; this did not alter over 6 years.

Aspiration

Just as for dorsal wrist ganglia injection of steroid after aspiration does not alter the outcome.

The recurrence rate after aspiration with or without injection of steroids or other substances has been reported to be between 25 % in 67 % [15] per annum. The complications are very low but include discomfort from a while which occurs in about 5 % of those aspirated.

Excision

The surgical excision of a palmar wrist ganglion is conducted through a zigzag or longitudinal incision over the flexor carpi radialis extended distally towards the radial aspect of the base of the thumb. This is the usual approach to the palmar aspect of the wrist. The flexor carpi radialis is reflected ulnarwards. The radial artery and its distal superficial division are identified and protected. The ganglion is then carefully dissected. Usually it runs through an interval between the radioscaphoid and the long radiolunate ligament to its origin on the palmar scapholunate ligament. This stalk needs to be identified and traced down to this ligament. The ganglion along with its stalk are then excised, care being taken not to damage the scapholunate ligament. Any daughter cysts in the region of the attachment of the stalk are scraped off using a curette or rongeur.

A third of the ganglia arise from the scaphotrapezium joint. Once again, the origin is scraped off the palmar ligaments of the scaphotrapezium joint. Any daughter cysts are also removed. Haemostasis is then undertaken. The skin is closed and the wound infiltrated with local anaesthetic. The main advantage of excising the palmar wrist ganglion is that in a proportion of patients the lump disappears immediately after surgery. The main disadvantages are that just over 40 % of these recur and 27 % may continue to have discomfort. Added to that, a small minority end up with damage to the palmar cutaneous branch of the median nerve or very rarely to the radial artery itself. Patients should be made aware of this potential prior to surgery.

After excision of the palmar wrist ganglion in 79 patients, 42 % had a recurrence [39].

Complications

The complications after excision of a palmar wrist ganglia have been reported in 7–28 % of patients and include damage to the palmar cutaneous branch of the median nerve, keloid formation, wound infection and stiffness. Dias and Buch [39] found 20 % of patients reported some form of complication. However, these were usually minor related to the scar, although there were patients who had symptoms related to the palmar cutaneous branch of the median nerve, and one patient who had damage to the radial artery [40].

Jacobs and Goevers [41] reported 28 % of 71 palmar ganglia who had some damage to the palmar cutaneous branch of the median nerve. These risks and benefits need to be clearly explained to patients prior to surgical intervention.

Six patients (of 78 who had surgery) in one study [39] felt that the hand was worse after treatment. Conversely, in none of the patients who had the ganglia aspirated or where it was left untreated felt that their hand had been made worse.

Clinical Pearl

Palmar wrist ganglia have a similar outcome regardless of intervention. The recurrence rate is high and one in five will have a surgical complication.

Sheath Ganglia

Sheath ganglia account for 14 % of ganglia of the wrist and hand [42].

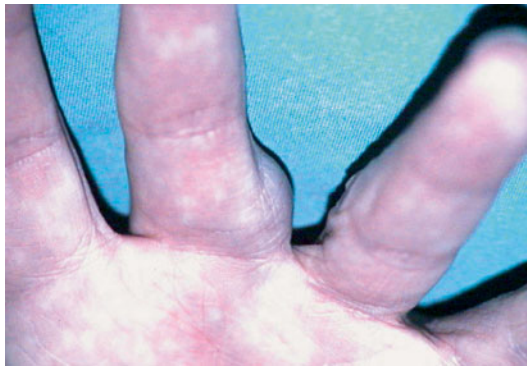

Fig. 1.5 A sheath ganglion. Usually the sheath ganglion is related to the proximal part of the A1 pulley and is not always obvious. The neurovascular bundle will be lateral to the ganglion. When aspirating the ganglion, the needle should be introduced from the midline, with the needle pointing dorsally. Thus avoiding the neurovascular structures

This ganglion usually presents as a hemispherical and slightly tender swelling which is usually on one or the other side at the base of the finger, usually proximal to the proximal finger crease. (Fig. 1.5) Forty per cent affect the middle finger and 26 % the index finger. Both hands are equally affected. As with other ganglia, women are affected three times more often than men [43].

Patients describe discomfort when holding onto objects such as the steering wheel of a car or the handles of a shopping bag. These symptoms come on spontaneously and gradually. Occasionally, this sheath ganglion can be associated with triggering of the finger. Usually, ganglions that present with symptoms tend to be around one centimetre in diameter and tender to touch. Added to that, they arise from the collagenous surface of the first annular pulley and, on occasion, the second annular pulley. An ultrasound scan of the finger will clearly identify the ganglion and its origin. Uncommonly, sheath ganglia can occur in the middle of the finger arising out from the distal end of the A2 or from the A4 pulley.

When they are big enough they may transilluminate; others are identified using an ultrasound or a MRI scan. Sheath ganglia may be treated with either reassurance, aspiration or surgery.

Reassurance

The natural history of the sheath ganglion is that a large number of these will resolve spontaneously without any intervention and regardless of intervention.

Aspiration

They can be treated by aspiration, under ultrasound guidance if very small. After aspiration at 7 years only 12 % had recurred. Complications occurred in a small number of these and these can be avoided by making sure that the entry of the aspirating needle is from the midline and thereafter directed into the ganglion and thus avoiding the neurovascular structures.

Any gelatinous fluid is aspirated and a firm compression applied to disperse the ganglion. Generally, aspiration of a sheath ganglion results in a better outcome than aspiration of wrist ganglia. This may reflect the different rate of development the cell changes that lead to the formation of the ganglia.

The most cost-effective treatment for recurrent flexor sheath ganglion after two aspirations was excision [44].

Excision

Surgical excision is conducted either through a transverse incision centred over the ganglion itself or a longitudinal zigzag incision. A zigzag incision, however, can heal with thickening at the apex resulting in pain and potential contracture. As a consequence, this incision is best avoided. Dissection needs to be meticulous, with care taken to avoid damaging the neurovascular structures which lie on or outside the sheath ganglion. The cyst is dissected to the pulley and removed taking with it a small window from the sheath. If there is any triggering then the A1 sheath is released. Local anaesthetic is infiltrated and the wound closed using dissolving sutures. A bulky bandage is applied but the hand is used from the very outset after elevation for the first hour or so.

Mucus Cyst

Mucus cysts account for 4 % of all ganglia of the wrist and hand. These are associated with degenerative change in the distal interphalangeal joint and often there are Heberden's nodes. These present as a translucent swelling at the back of the terminal phalanx between the nail and the joint. It usually does not appear in the midline as the ganglion bulges out from one or other side of the extensor tendon. When the ganglion extends distally it encroaches upon the germinal matrix of the nail bed leading to the formation of the longitudinal groove in the nail plate in 23 % [45] (Fig. 1.6). Occasionally, the ganglion can rupture and become secondarily infection. This can leave a sinus or punctum on the dorsum of the cyst. Added to that, patients may have restrictions on distal interphalangeal joint movements and occasionally complain of an aching pain secondary to the degenerative arthritis.

Treatment includes reassurance, aspiration or excision. Forty per cent recur after aspiration alone [45]. My personal preference with these is that if the cyst is troublesome or the skin is at risk then these are excised through a Y shaped incision. The ganglion is traced to the joint and any secondary cysts excised. If there is a large osteophyte then it is trimmed back using a small narrow rongeur. However, this is not essential as osteophyte removal does not have an impact on the recurrence rate after excision of the mucus cyst which is generally low. Matthews [45] reported no recurrence after excision of 54 mucous cysts while up to 3 % recurrence rate has been observed in another study [46]. The main risk is stiffness in the distal interphalangeal joint and problems with wound healing when the walls are thin. The condition of the nail ridge, however, improved in 80 % of 31 cases [45]. If there is a risk of skin loss, then either Wolfe graft [47], a bilobed flap [48] or a rotation flap [49] can be used. Surgeons need to be aware of all these techniques. Complications of surgery include infection in 3 % and nail deformity in 7 %. Patients also complain of more pain after surgery [46].

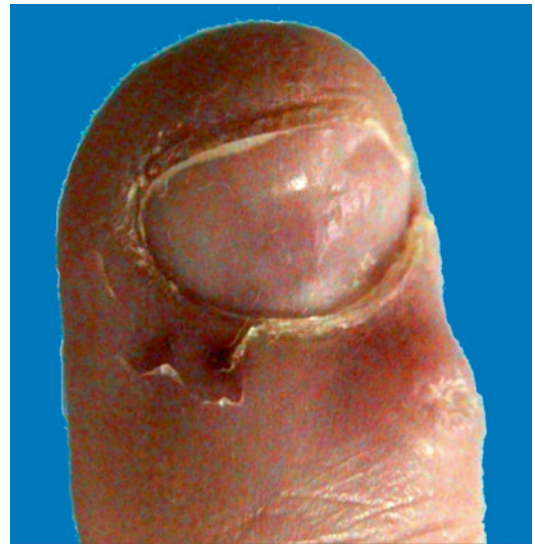

Fig. 1.6 A mucous cyst on one side of the dorsum of the distal interphalangeal joint with a punctum where a previous clinician attempted aspiration. The ganglion has pressed on part of the germinal matrix causing a nail ridge (Dias et al. [29, 39])

Intraosseous Ganglia

Intraosseous ganglion cysts occur in the proximal scaphoid or radial part of the lunate, adjacent to the scapholunate joint and usually, but not always towards the dorsum. These are commonly treated by excision of the ganglion and curettage of the cyst wall with or without cancellous bone grafting [50] with reasonably good outcomes [51]. The natural history of this disorder has not been established.

Nerve Compression by Ganglia

In certain locations ganglia encroach upon nerves and cause either local pressure or infiltrate the nerve itself. This is a particular issue with the ganglia that encroach Guyon's canal. In patients presenting with weakness of the first dorsal interosseous muscle with good preservation of the abductor digiti minimi and with no sensory symptoms in the little and ring fingers, the surgeon must consider the possibility of a ganglion pressing on the deep branch of the ulnar nerve within

this canal. These ganglia can arise from the pisohamate ligament or from within the joint just distal to the hook of the hamate. These can either be aspirated under ultrasound or excised to relieve the pressure on the ulnar nerve. This is one of the few instances of a ganglion where intervention is mandatory to protect nerve function.

Summary

Ganglion cysts are common, accounting for the second most common reason for attendance at hand clinics. Removal of these ganglia is not mandatory, as once the diagnosis is made, many patients are happy simply to monitor the situation. If the ganglia are removed, however, it is important to note that there is a significant recurrence rate, particularly with the palmar wrist ganglia. As such, following reassurance, careful discussion needs to take place regarding the natural history of these swellings and the advantages and disadvantages of either aspiration or surgical excision.

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Pigmented Villonodular Synovitis in the Hand

2

S.R. Murali

Keywords
Giant cell tumour • Pigmented villonodular synovitis • Fibrous xanthoma • Sclerosing haemangioma, benign synovioma • Multinucleated giant cells • Xanthoma cells

Introduction

The first description of pigmented villonodular synovitis (PVNS) was by Chassaignac in 1852 [1]. However the actual term PVNS was not introduced into the literature until 1941 by Jaffe, Lichtenstein and Sutro [2]. PVNS is an idiopathic, benign, hyperplastic proliferation of the synovium, which occurs within joints with characteristic pigmentation from haemosiderin. A focal extra-articular form commonly affects the hands, possibly arising from the tendon sheath, and is called a giant cell tumour of the tendon sheath (GCTTS). There are many similarities in the histology between these two conditions, although the clinical presentation can be different. Other terms used are pigmented villonodular bursitis or tenosynovitis, nodular tenosynovitis, fibrous xanthoma, sclerosing haemangioma and benign synovioma. The use of several terms can sometimes be misleading.

Clinical Pearl
Pigmented Villonodular Synovitis (PVNS) arises from intra-articular synovium
Giant Cell Tumour of tendon sheath (GCTTS) arises from tendon sheath

Incidence

The average annual incidence has been estimated to be 1.8 per 1 million population for the intra-articular form and 9.2 per million for the extra-articular form [3]. The GCTTS is the second commonest soft tissue tumour affecting the hand after ganglia [4]. Presentation is typically between age 30 and 50 years. Female predominance ranges between 1.5 and 3:1. The radial digits tend to be more affected. There are reports of this condition affecting all the broad racial groups.

Pathophysiology

Pigmented villonodular synovitis is a condition affecting synovial membranes intra and extra

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articular. The exact aetiology of PVNS is still unknown but hypotheses include inflammatory, neoplastic and metabolic processes. Experimental and epidemiological studies have suggested that PVNS is a reactive process although recent studies have shown that these lesions are capable of autonomous growth thereby suggesting the involvement of a neoplastic process. This theory has been further supported by studies suggesting that heterogeneous proliferating cells, such as fibroblasts, histiocytes, multinuclear cells, and chronic inflammatory cells, might be neoplastic, with other cell types being reactive in nature [5].

This is a benign non-metastasizing condition but can recur locally if not completely excised. Malignant transformation in the hand is rare and reports describe a good clinical outcome providing there are clear resection margins [6].

Clinical Presentation in the Hand

The clinical presentation can be grouped into three categories:

(a) The commonest form is a **discreet subcutaneous nodule**, usually multi-lobulated, in the digit or palm. Typically these are painless and have been present for several months or even years. They can sometimes interfere with hand function because of their size or can produce neurological symptoms, like paraesthesia, in the affected region. The most frequent location of the tumour is in the index finger (30 %), although other digits are also commonly affected: thumb (13 %), the long (25 %), ring (17 %) and little (16 %) fingers [7]. Lesions are found twice as often on the volar aspect of the hand compared with the dorsal side. Lesions are small and usually vary between 1 and 2 cm in size, but can get bigger if the presentation is late. It is also possible for these lesions to surround the digit and breach through fascial intervals and can be palpated on the dorsal and volar sides of the palm or digit (Figs. 2.1 and 2.2).

(b) The **diffuse form** can involve a large area of the hand. The margins are ill defined and they present as a soft tissue mass. This is thought to represent the intra articular form occurring in other areas of the body such as the knee joint as mentioned below. The soft tissue structures of the hand, like the extensor tendons can be engulfed in the tumour mass and can be difficult to dissect free (Figs. 2.3, 2.4, and 2.5).

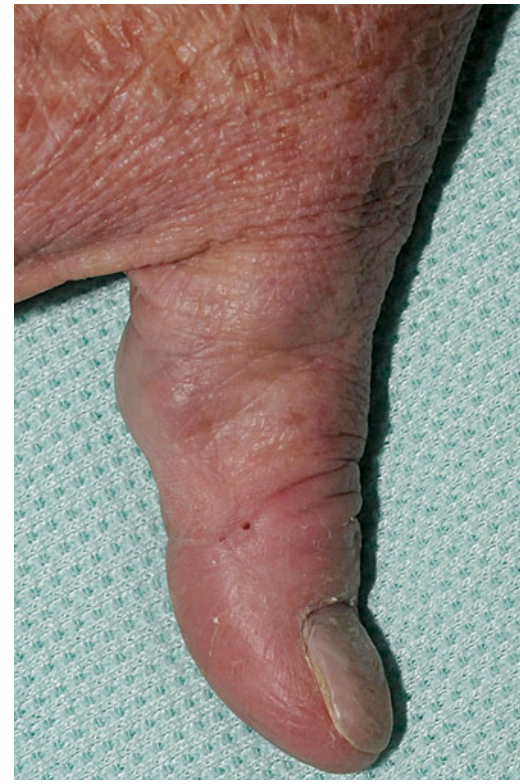

Fig. 2.1 Swelling in the Thumb

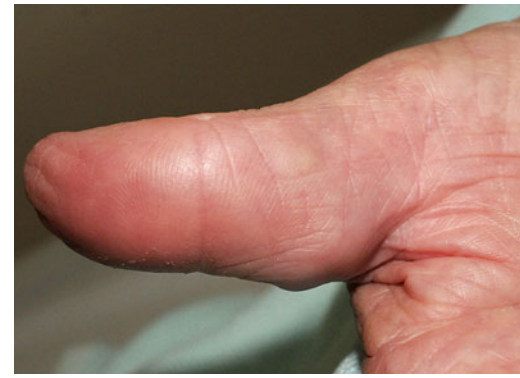

Fig. 2.2 Swelling in the Thumb

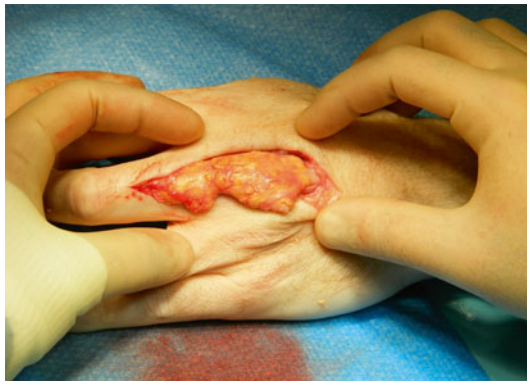

Fig. 2.3 Diffuse form of PVNS infiltrating into the extensor tendon

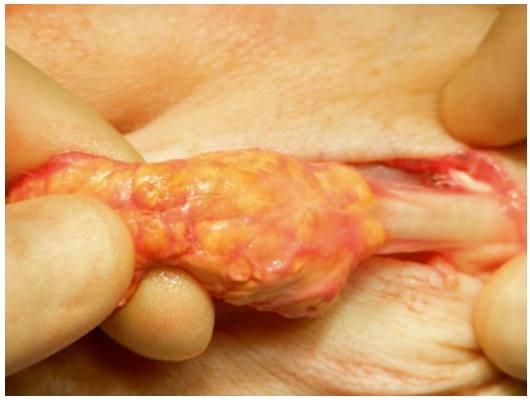

Fig. 2.4 Diffuse form of PVNS infiltrating into the extensor tendon

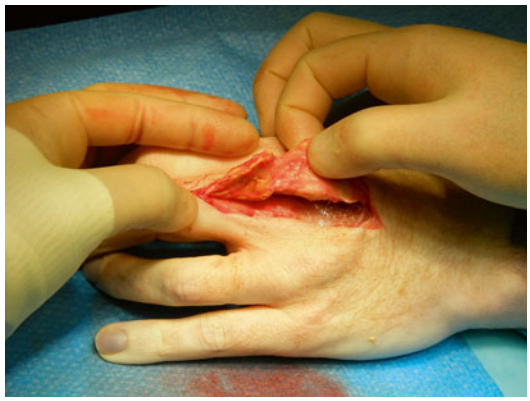

Fig. 2.5 Diffuse form of PVNS infiltrating into the extensor tendon

(c) The **intra articular form** is not common in the hand (usually presents in this form in the knee and foot). These can present with joint pain and stiffness in the digit. The proximity of the joint synovium to the tendon sheath around the small joints in the hand can make it difficult to determine the precise origin of the lesion.

Imaging of PVNS in Hand

X-Ray

Typically plain x-rays will show a non-specific soft tissue swelling. Bony involvement is uncommon, occurring in perhaps 10–20 %. These cases show external pressure erosion due to the soft tissue tumour.

Ultrasound

Ultrasound most commonly shows a solid homogeneous well-defined mass. The presence of internal echoes and internal vascularity on colour or power Doppler will indicate that this is a soft tissue, and not a cystic, mass [8].

CT Scan

This is not the imaging modality of choice due to poor contrast resolution between different soft tissues. The presence of a soft tissue mass will be detected, which may have increased density if there is increased iron content due to haemosiderin

MRI

This is the imaging modality of choice for the diagnosis of a soft tissue mass in the hands or fingers. Giant cell tumours of the tendon sheath show a well-defined nodular mass of low/intermediate signal on T1 weighted images. There is typically low signal on T2 –weighted scans due to haemosiderin, but they will often show increased signal STIR scans [9, 10]. The mass typically surrounds the flexor tendon sheath and shows prominent homogeneous contrast enhancement (Figs. 2.6, 2.7, 2.8, 2.9, and 2.10).

Clinical Pearl
MRI is imaging of choice for a soft-tissue mass in the hand

Pathology of PVNS

Macroscopic Pathology

An intra-articular lesion appears as a diffuse mass which extensively involves and thickens the synovium of the entire joint, whereas a GCTTS is well circumscribed, lobulated and a nodular soft tissue mass which arises from the tendon sheath. PVNS forms a collagenous, relatively well circumscribed mass, pale in colour. The cut surface is characteristically brown in colour due to haemosiderin deposition. The lesion is not strictly encapsulated but usually forms a well demarcated mass. If such a mass is enucleated this may leave residual areas of PVNS behind tending to increase the risks of local recurrence (Figs. 2.11 and 2.12).

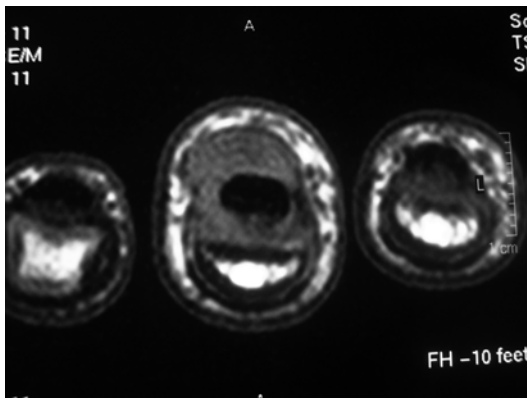

Fig. 2.6 MRI Axial T1 weighted image

Microscopic Pathology

The features of both the diffuse and localized forms are similar. There is a background population of uniform appearing, round or polygonal cells with moderate amounts of eosinophilic cytoplasm. These are thought to be derived from synovial cells which in turn are derived from macrophages/histiocytes. These cells fuse together to form variable numbers of multinucleated giant cells interspersed with the background population of polygonal cells. Also present are xanthoma cells which are cells having clear or foamy cytoplasm due to the ingestion of lipids, although this condition is not associated with hyperlipidaemia. There are variable degrees of

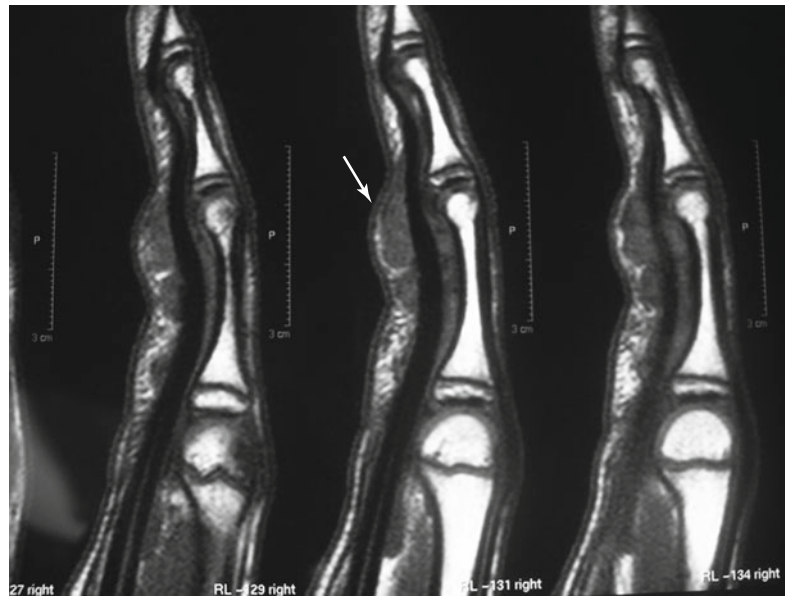

Fig. 2.7 MRI Sagittal T1 weighted image

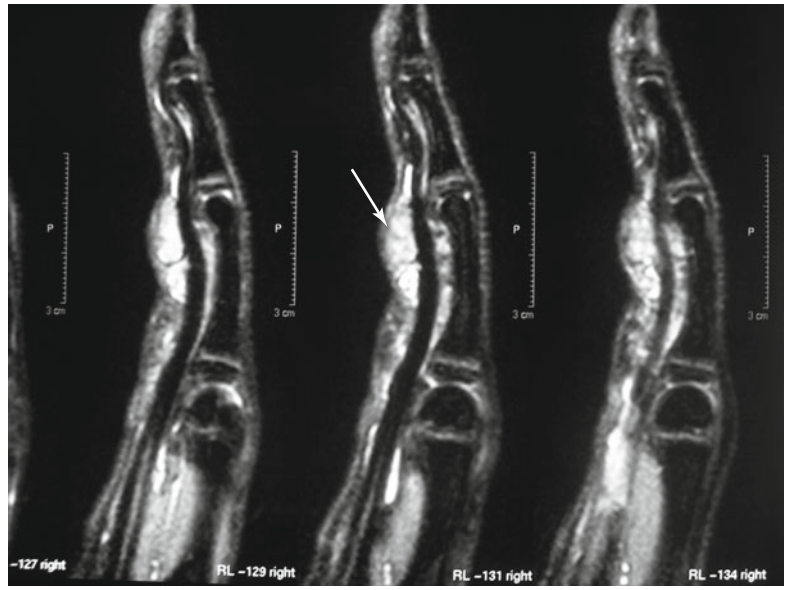

Fig. 2.8 MRI Sagittal STIR image

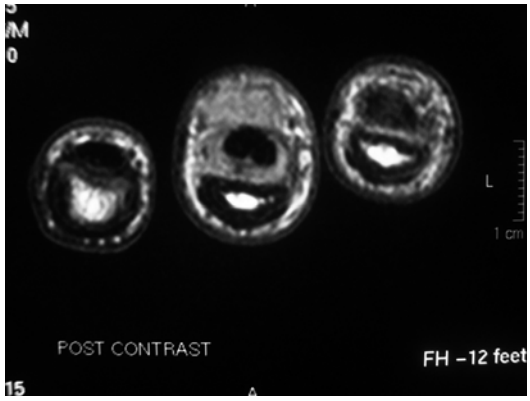

Fig. 2.9 MRI Axial T1 weighted image with contrast

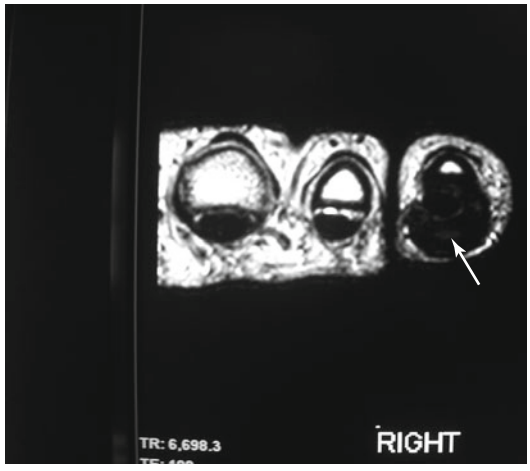

Fig. 2.10 MRI Axial T2 weighted image

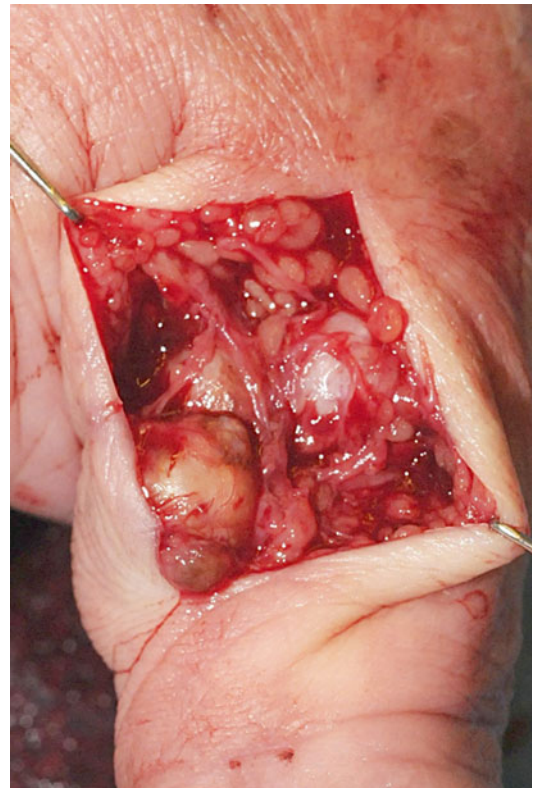

Fig. 2.11 Intraoperative photograph showing multiloculated swelling

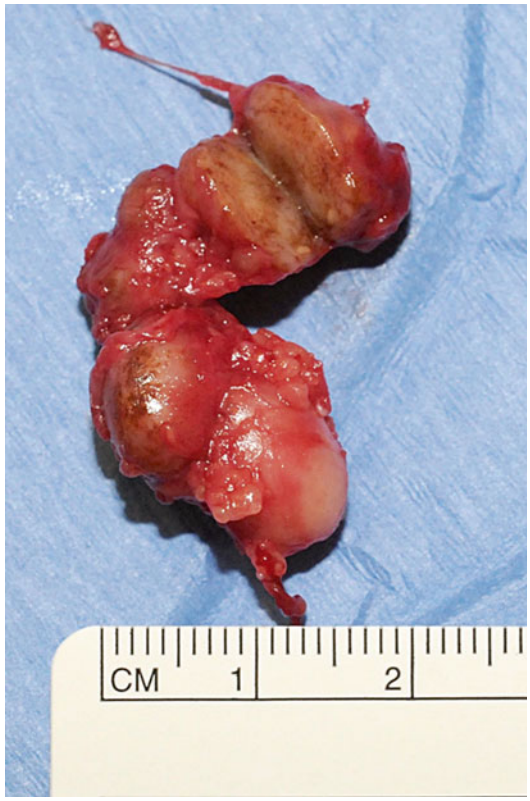

Fig. 2.12 Cut section of the Tumour

haemosiderin pigmentation from haemorrhage. The proportions of background polygonal cells, giant cells, xanthoma cells and areas of pigmentation vary from tumour to tumour so that some may have virtually no giant cells or abundant giant cells. There may be dense or minimal pigmentation and the background polygonal cells may be abundant or few in number.

Treatment and Prognosis

Surgical resection is the gold standard of treatment, although recurrence rates can be high (12–44 %) [11–13]. The nodular variety can usually be easily exposed and dissected free of the surrounding structures. The recurrence rate is very low if the tumour has a good pseudo capsule around it as it makes the complete excision more likely. However because the tumour can surround a significant portion of the digit, separate incisions may be required. The recurrence rate is higher when the capsule is not well formed and there are separate discrete nodules. In the diffuse variety (Fig. 2.5) it may be possible only to remove parts of the tumour or sacrifice a block of soft tissue and plan reconstruction. Adjuvant post-operative radiotherapy has been recommended to reduce recurrence, reducing the recurrence rate in a series of 48 cases to 4 % [14]. Recurrence can be successfully treated with repeat surgical resection.

Garg et al. has defined two clinical groups with respect to their risk of recurrence. They demonstrated that a well encapsulated lesion, confined to either the dorsal or volar aspect of the digit had a negligible risk of recurrence. They predicted a high recurrence rate in all other lesions and these were therefore targeted with a postoperative radiotherapy regime (local irradiation at 20 Gy in divided doses: 2 Gy surface dose daily at 200 kV). Their recurrence rate overall in a series of 106 lesions (mean follow up 12 months) was 3.8 % in all patients and 8 % of the patients in the high risk group [7].

Pharmacological therapy with TNFα blockade in larger joints affected with PVNS have been reported but not in the hand, although this may be a future avenue for diffuse intra-articular disease or in the treatment of recurrence after surgery. Tyrosine kinase inhibitors such as imatinib, which target the over expression of the *CSF-1* gene seen in the intra-tumoural cells, may have a role in the primary treatment of diffuse intra-articular disease. Early studies have been able to demonstrate a good response to treatment in a small numbers of patients [15].

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Vascular Anomalies of the Upper Limb

3

Steven E.R. Hovius and Christianne A. van Nieuwenhoven

Keywords

Congenital vascular malformations • Vascular tumors • Hemangioma • Congenital hemangioma • Infantile hemangioma • Rapidly involuting congenital hemangioma • Non-involuting congenital hemangioma • Capillary malformations • Venous malformations • Arteriovenous malformations • Lymphatic malformations • Low flow vascular malformations • High flow vascular malformations

Introduction

Vascular anomalies are localized defects characterized by an increase in the number of vessels, which are tortuous and enlarged, and can affect the capillaries, arteries, veins and/or lymphatics. The estimated prevalence is 4.5 % [1] and diagnosis is made during infancy or childhood. With a vast variation in presentation they might lead to disfigurement, bleeding, destruction of surrounding tissues, dysfunction, infection, thrombosis and pulmonary embolus [2], ulceration and pain.

Historically, the terminology of the anomalies were descriptive, their names derived from foods (e.g., strawberry, port-wine, cherry, salmon patch) to characterize the different anomalies. In 1982, Mulliken introduced a biological classification that correlated with clinical features, natural history and cellular characteristics [3]. Two main groups were identified; the vascular tumors and the vascular malformations. The International Society for the Study of Vascular Anomalies (ISSVA) adopted Mulliken's classification in 1996 and modified it in 2007 [4]. The vascular tumors are endothelial neoplasms with an increased endothelial turnover compared to an abnormal development of the vascular system during embryogenesis and maturation in vascular malformations.

Hemangioma is the most common in the tumor group, completed with the kaposiform hemangioendothelioma, the pyogenic granuloma, and rare tumors like angioblastoma (tufted angioma), hemangiopericytomas, hemangioendothelioma and giant-cell angioma (Table 3.1). The malformations derive their name from the involvement of the predominant vessel type: capillary,

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Table 3.1 Classification of vascular anomalies

Tumors	Vascular malformations Simple	Vascular malformations Combined
Infantile haemangioma	Capillary (C)	Arteriovenous fistula
Congenital haemangioma	Lymphatic (L)	Arteriovenous malformation
Tufted angioma	Venous (V)	(AVM)
Kaposiform haemangioendothelioma		CVM
Haemangiopericytoma		CLVM
Pyogenic granuloma		LVM
Spindle-cell haemangioendothelioma		CAVM
		CLAVM

The International Society for the Study of Vascular Anomalies (ISSVA) adopted Mulliken's classification in 1996 and modified it 2007 [3 and 4]

Table 3.2 Classification of vascular anomalies by vascular dynamics for ease of investigation and treatment

Classification of vascular anomalies by vascular dynamics
I. Haemangioma
II. Vascular malformations
(a) Low-flow (VM)
(b) High-flow (AVM)
III. Lymphatic malformation (LM)

Modified by Jackson et al. [6]

lymphatic, venous, arteriovenous and complex forms. When an arterial component is part of the malformation, the malformation will be a high-flow lesion; in the remainder of cases the malformation will be a slow-flow lesion. Jackson et al. [6] modified the classification of vascular anomalies by vascular dynamics for ease of investigation and treatment (Table 3.2).

Tumors and malformations must be distinguished to be able to implement the proper management. Therefore, a proper medical history and physical examination are the prerequisites for an accurate diagnosis, and consequently the proper treatment.

Vascular Tumors

Hemangiomas

Hemangiomas are the most common vascular tumors, presenting in 1–3 % of neonates of all races and account for 65 % of all childhood tumors [8]. They are benign tumors and can be divided into infantile and congenital hemangiomas. Reviewing the literature has been complicated by the use of hemangioma as a synonym for both vascular tumors and malformations. Furthermore, the term hemangioma is regularly used for infantile hemangioma, not comprising the rapidly and non-involuting congenital hemangiomas. The differences in these two tumors will be explained.

Infantile Hemangioma

The infantile hemangioma (IH) is a benign, endothelial tumor with a unique biological behavior. The natural history of hemangiomas involves a rapid growth phase, followed by a static phase and a slow involution phase consecutively. After this spontaneous regression, fibrofatty tissue can be deposited at the site of the involuted hemangioma. The incidence is 4–10 % in Caucasian infants [9, 10]. They comprise 86 % of all vascular tumors [1]. The IH are unusual in dark-skinned infants. The prevalence increases in premature children (23 % in children with a birth weight less than 1,200 g) [11]. Females are affected more commonly with a ratio between 3:1 and 5:1. Thirty to fifty percent of the hemangiomas are present at birth, with a median age of appearance of 2 weeks after birth [12]. When an IH is superficial it will appear red, the skin is raised and firm. If situated beneath the skin, it may be noted as a bluish deformity, slightly elevated and warm. Only after a couple of months after birth, when

it becomes large enough, will it be truly visible. All of the superficial IHs might overlay a deeper component.

The IH is found as a single tumor in 80 % of the cases; head and neck are involved in 60 %, the trunk in 25 %, and the extremity in only 15 % [13]. If multiple IHs are present, an association with IHs in other organs might be present, especially the liver.

In the **proliferating** phase, the endothelial cells of the IH grow out of proportion to the growth of the child and 80 % of its volume is achieved after approximately 3–4 months [14], stabilizing the growth after 9 months. In this phase, the IH consists of rapidly proliferating endothelial cells, forming a tumor with tightly packed sinusoidal, capillary-sized, vascular channels. The endothelial cells highly express angiogenic factors. Glucose transporter protein GLUT-1 is a useful histopathological marker for IH, and is expressed at all stages of the evolution [15].

After 9–12 months the IH grows in the same proportion as the child, and after 12 months, the **involuting** phase begins. This phase is characterized by a decrease of the endothelial proliferation and increase of apoptosis, with diminution of the number of channels and luminal enlargement. As a result, the tumor decreases in volume, its color fades, and flattens, with softness of the overlying skin. Clinically, the signs of regression are: a pale skin, typically at the center of the lesion, and a patchy grayish discoloration. The IH is softer to palpation (Fig. 3.1).

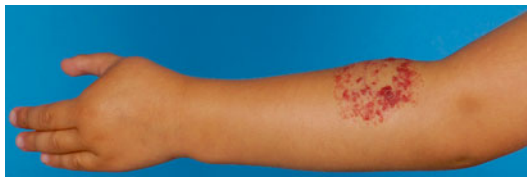

Fig. 3.1 Involuted hemangioma on the proximal forearm. Note the pollicisation, the thumb was absent

The involuting phase extends up untill the age of 5–7 years. In this period, regression is expected, however this process is not related to the appearance, depth or size of the IH. Therefore, it is important to realize that a large and bulky IH can regress totally, while a superficial IH can leave an irregular, atrophic scar.

In the differential diagnosis, mostly the hemangiomas are misdiagnosed. A deep hemangioma might resemble a lymphatic malformation, and a macular IH might be mistaken for a capillary malformation. Furthermore, a fibrosarcoma might be misdiagnosed as an IH.

Congenital Hemangioma

Congenital hemangioma (CH) can be distinguished from the IH because they are fully developed at birth. It has a different appearance (Table 3.3). There are two subsets, the rapidly involuting congenital hemangioma (RICH) and the noninvoluting congenital hemangioma (NICH). The GLUT-1 marker, present in IH, is not found in the CH. The CH occurs in approximately 8 % of all vascular tumors [1].

In contrast to the IH, the CH can be associated with coagulopathy like thrombocytopenia, low fibrinogen levels, and increased levels of fibrin degradation products and D-dimers. Coagulopathy is mostly associated with the RICHs, and is self-limiting and usually not associated with bleeding complications [16].

RICH

The RICH is a red-violaceous coloured tumor with telangiectasiae, and an average diameter of

Table 3.3 Clinical symptoms of hemangiomas

	Infantile haemangioma	Congenital haemangioma	
		RICH	NICH
Growth	Most appear after birth, then proliferate, then regress	Fully formed at birth, rapidly involutes	Fully formed at birth, no involution
Colour	Red/blue depending on depth	Red, violaceous	Pale grey
Coagulopathy	Rare unless multiple/visceral	Yes	No

5 cm. It mostly has a central pallor region and a pale halo. It is more common in extremities and the male to female ratio is equal. Signs of superficial ulceration and occasionally, signs of arteriovenous shunting, simulating an arteriovenous malformation, might be present.

The RICH involutes directly after birth, with a completed regression after 7 months in 50 % of all lesions. The remainder is fully involuted by 14 months after birth. The head and neck are involved in 42 % of cases, the limbs in 52 %, and the trunk in 6 %.

NICH

The NICH typically is an ovoid, macular, or slightly raised tumor with a pale gray coloration and prominent telangiectasiae. It is warm when palpated.

They persist up till late in childhood, and rarely involute. The head and neck is involved in 43 %, the limbs in 38 %, and the trunk in 19 % [17].

Clinical Pearl

Red fully formed hemangiomas, which are present at birth, involute rapidly in the first year of life (RICH).

Infantile Hemangioma's grow after birth and can be accompanied by bleeding and ulceration, followed by regression. They regress mostly well on propranolol medication.

Other Vascular Tumors

The kaposiform hemangioendothelioma and the pyogenic granuloma are rare tumors. The kaposiform hemangioendothelioma is a tumor, which can be associated with Kasabach Merrit syndrome or lymphangiomatosis [18]. The pyogenic granuloma is a small vascular tumor that bleeds easily and rises often at a site of recent injury. Its name is misleading since it is a type of hemangioma of traumatic origin, and therefore not pyogenic nor a true granuloma. The tumor may heal spontaneously, but in chronic cases, treatment may be needed, ranging from surgical resection, topical phenol [19], bleomycine injection [20], laser therapy [21], topical cream [22], and curettage to silvernitrate treatment [23]. For the scope of this chapter even more rare vascular tumors like the angioblastoma (tufted angioma), hemangiopericytomas, hemangioendothelioma, and giant cell angioma will not be further discussed.

Diagnostic Imaging in Vascular Tumors

Most infantile hemangiomas do not require additional diagnostic imaging since the clinical course through the different stages is predictable, and points to the diagnosis itself.

However, when the clinical features are atypical or the anatomical extend of the lesion is uncertain, ultrasonography or magnetic resonance imaging (MRI) can be helpful.

The ultrasonography appearance is dependent on the stage of the tumor. A proliferating hemangioma will be visualized as a dense, well-circumscribed homogeneous tumor with hypervascularity. There is a low arterial resistance and increased venous flow. Most hemangiomas are hypoechoic (82 %) [24]. When involution progresses, the more diffuse infiltrating soft tissue and decreased vessel flow will show. Differentiation from, for instance, an arteriovenous malformation is made by the presence of solid parenchymal tissue. The RICH and NICH are distinguished by their clinical course from IHs, but cannot be differentiated from IHs by imaging alone.

Differentiating factors in ultrasonography are the presence of more visible vessels, intravascular thrombi, calcification, vascular aneurysms and AV shunting in the CHs.

On MRI studies, the CHs will be less defined than IHs [25].

Treatment of Infantile Hemangiomas

Most IH hemangiomas in the upper extremity are small and solitary. They do not cause functional problems in the majority of the cases. However,

one should take care of rapidly growing lesions causing ulceration, maceration, crusting or bleeding in 10 % of cases [26]. Because of decreased oxygen delivery and relatively ischemic skin a superficial hemangioma may be predisposed to skin ulceration and loss of skin. Therefore, minimal trauma, like finger sucking or scratching, may lead to wound formation. Regular cleaning and **wound care** with non-adherent gauze should be used in (ulcerative) lesions to keep the lesion moist, and prevent bleeding from the surface when removing the dressing. If a cellulitis is present, parenteral antibiotics might be considered. Superficial lesions and infections should heal within 2 weeks; full thickness lesions will take longer. When lesions are present, patients are followed closely in the proliferative phase.

Since the natural course of IH's generally is complete regression, the rule is to wait patiently. It mostly is a challenge to translate this policy to parents.

In upper limb hemangioma, medical or surgical treatment is only necessary in highly exceptional cases. Therefore, these treatments will be concisely discussed.

Corticosteroids are not often used systemically in the treatment of hemangioma in the upper extremity [27]. Local, intra-lesional corticosteroid injection of a hemangioma can be given at 6–8 week intervals. Complications in the upper extremity are lightening of the skin, subcutaneous skin atrophy, and increased residual scarring [28, 29]. A minimal effect on IH's has been obtained when using topical corticosteroids.

Interferon Alfa-2a or Alfa-2b is proposed when corticosteroid treatment is complicated by side effects or a failure to respond to corticosteroids. The interferon is given at an early age and continued for 9–14 months for the most optimal treatment result. It is described as being successful in 80 % of patients, including those patients not responding to systemic corticosteroids [30]. Several side effects are reported, of which the most serious long-term side effect is spastic diplegia, reported in 10 % of patients treated with interferon [31].

Several **lasers** have been used to treat hemangioma, with the main drawback being the restricted penetration of approximately 1 mm, leaving the deeper component untreated. Although controversial, the advantage in treatment will be in an early stage before the hemangioma starts to grow, or when treating residual defects for esthetic reasons, or in treating defects with a high risk for complications [28].

Recently, **propranolol** has been used for the medical treatment of hemangiomas with an oral dose of up to 1 mg per kg every 12 h [32]. The mechanism is poorly understood, but may influence the vasoconstriction with a color change in the early stage, with a down regulation of pro-angionenic factors, and triggering of apoptosis in a later phase. To what extent it should be used for upper limb IH's is not clear.

Surgical treatment, if ever needed, consists of primary closure, transposition flaps or skin grafts.

Treatment of Congenital Hemangioma

As the RICH regresses surgical intervention in infancy is hardly ever necessary. In a later phase, the atrophic, abundant skin and subcutaneous tissue can be reconstructed. Care must be taken to improve scarring. The NICH will not be a problem in the majority of cases, and do not cause a significant deformity. Resection may lead to a more conspicuous scar. To improve appearance of the deformity, treatment with pulse-dye laser will remove the telangiectasiae [33].

Vascular Malformations

The treatment of vascular malformations is a greater challenge for the hand surgeon than hemangiomas are.

Many congenital malformations are present at birth, but will go unnoticed until the first 5 years of life and grow in proportion with the child. The growth may increase, especially in girls during adolescence, or during pregnancy [29]. As it is believed to result from errors in vasculogenesis and angiogenesis, they will have no tendency to involute. A small number of these

malformations can be recognised at birth. Most malformations will be low-flow lesions, with a ratio of 7:1 compared to high-flow lesions [34]. Of all vascular anomalies, approximately 65 % are malformations [1].

Diagnosis of vascular malformation is mostly easy to make, extent of disease however is not. Venous malformations are the most common type within the group of vascular malformations. Extensive lesions in the upper arm can also involve thorax, mediastinum and abdomen on the ipsilateral side. Combined vascular malformations can comprise all different combinations, between capillary, lymphatic and venous or arterio-venous. Combined vascular malformations are mostly complex lesions, which can be associated with skeletal overgrowth. Well known are the Klippel-Trénaunay Syndrome, Parks-Weber Syndrome, Proteus Syndrome, and the Sturge-Weber Syndrome. The latter is a syndrome typically present in the facial area with leptomeningeal vascular malformations, but might show lesions on the upper extremity as well.

Capillary Malformations

Capillary malformations (CM's) used to be referred to as port-wine stain. Birthmarks or fading capillary stains (salmon patch, angel's kiss, stork bite) may be mistaken for CM's, these, however, will fade in the first weeks after birth. In contrast, the CM's will darken and are not always visible at birth. This so-called true CM is seen in 3 of 1,000 infants, with an equal distribution among male and female sex [9] and comprise 11 % of all vascular malformations [1]. Histopathologically, the CM's change from dilated capillaries in the papillary dermis of young children to a random arrangement of distended vessels in the papillary and sometimes the reticular dermis [35]. Syndromes associated with CM's are Sturge-Weber syndrome, capillary malformation-arteriovenous malformation syndrome, cutis marmorata telangiectatica congenital, Klippel-Trénaunay syndrome (Fig. 3.6), and occasionally in Proteus syndrome.

Clinical Presentation

Clinically, CM's present as cutaneous stains or patches, which can be bright red or pink to purple. Initially they are flat and with ageing they thicken and become nodular. In the first months after birth they tend to lighten, an expression of the decreased circulating blood haemoglobin level, instead of an indication of spontaneous involution. The CM will grow in proportion with the child and presents at any part of the body. Facial CM's usually have a dermatomal distribution, in contrast to involved limbs or the trunk. Limb CM's can be associated with hypertrophy of underlying soft tissue and bone [36]. In upper extremity CM's, an associated venous malformation or lymphatic malformation should be ruled out.

Treatment

Many treatment modalities have been tried. The pulsed-dye laser will lighten the colour and decrease the blush. This effect is more outspoken in treated facial CM's than in involved extremities. Therefore, most CM's involving an extremity will stay untreated. The timing of the pulsed-dye laser treatment is disputed with better results reported during infancy [37], while others found no difference in age of therapy [38]. To minimize the psychosocial impact for a child with a major deformity, early treatment may be preferred. At this moment, the pulsed-dye laser is the gold standard treatment for aesthetically sensitive (mostly facial) CM's, with an effective treatment of complete lightening in 20 % of the cases. However, 20–30 % do not respond to treatment with the pulsed-dye laser, and this is related to the location of the CM and concurrent soft tissue overgrowth. In CM's that do not respond, other laser modalities and combinations of lasers, and combinations of laser and topical treatment have been used [39]. Occasionally excisional surgery is planned in patients with a soft tissue or bony hypertrophy to restore the 'normal' anatomy. When a pyogenic granuloma arises in the CM, excision is needed to prevent repeated bleeding. Quite often camouflage ointments are used.

Venous Malformations

Ninety-five percent of venous malformations are sporadic [5], without a preference regarding sex, with an incidence of 1–2 per 10,000 births and a prevalence of 1 % [40]. Approximately 37 % of all vascular malformations will be venous malformations [1].

Clinical Presentation

Extensive venous malformations and other lesions are easily detected in the upper limb at birth. However, small venous lesions can stay unrecognised for years. Clinically a lesion may only be partly apparent. For instance a finger can be clearly involved, however extension to the palm of the hand and forearm are not easy to detect. Patients complain of pain, a tired feeling at the end of the day and swelling, especially when the arm is in a dependent position. This is relieved as soon as the arm is elevated. Thrombosis can occur, resulting in sudden pain and firmness of the swelling. These malformations extend mostly along anatomical planes. They can easily extend along the axilla to the thorax. They are able to infiltrate along all tissues, however this is not common. Also ulceration is not common. Skin maceration and infection can be encountered.

In puberty, after exercise and after hormonal changes (especially pregnancy) expansion can occur, and even enlargement. After pregnancy the lesion does not decrease to its original size.

Clinical diagnosis is nearly always apparent. Bluish discoloration and filling of the tumour in dependent position are the clearest signs. However the extent is mostly underestimated.

X-rays of the hand, arm and chest wall can demonstrate calcifications, inter-osseous involvement and growth changes.

An ultrasound can delineate the well-defined sponge like collection of vessels, as well as the presence of phleboliths and presence of thickened subcutaneous tissue [41]. Furthermore, it can be used in estimating the success of sclerotherapy. This treatment modality is more successful in lesions with mainly vascular or lymphatic spaces [42].

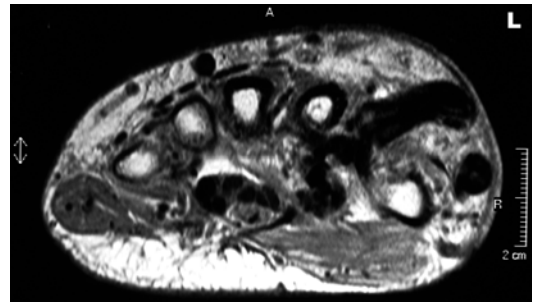

Fig. 3.2 Venous malformation, MRI with venous lakes and dilated veins at the radial side of the hand

MRI is the best investigation at present to determine the extent and infiltration of the lesion and to distinguish between slow flow and fast flow malformations. It will show multilocular, lobulated and septated masses. The lesions infiltrate into the adjacent tissue (Fig. 3.2). Intravenous gadolinium will delineate vascular channels. Dynamic contrast-enhanced MRI increases specificity up to 83 % when combined with conventional MRI. Conventional MRI has a sensitivity of 100 % but a specificity of 24–33 %. Angiography is only used as a preoperative measure in very large resections.

Treatment

Venous malformations are mostly treated conservatively. Quite often compression garments are used to diminish swelling and pain due to filling in a dependent position. Indications for surgery are mostly related to size and depth of infiltration, location and/or local nerve compression. Diffuse malformations cannot be completely excised but can be debulked. Multiple stages are often necessary to preserve neurovascular bundles, tendons and joints. Decompression of compartments in the hand and forearm can be useful to relieve pain. At present most venous malformations are reduced with repeated injections with sclerosing agents. In our institution radiologists inject the lesions. In a series of 66 adult patients treated at our hospital partial or total relief of symptoms was obtained in 58 %. Symptoms were pain in 89 % and swelling in 91 %. The extremities were involved in 44 patients. Of the injected vascular malformations

83 % were of the low flow venous type. In these cases ethanol or polidocanol was injected under ultrasound guidance. The mean follow-up was 30 months (range 6–147 months). In 40 % complications were reported of which nearly 50 % needed additional treatment. Polidocanol treated patients had less problems. Complications in this study were mainly tissue necrosis, compression syndromes and deep infection [7].

Complications of surgery are mainly due to damage during operation, like scar formation, tendon adhesions, neuromas, stiffness and skin necrosis. Management of venous malformations combined with limb overgrowth is the most difficult. Mostly these children require multiple debulking operations. They are treated symptomatically.

Lymphatic Malformations

The lymphatic malformation (LM) is usually present at birth, but may only become visible during late childhood or adolescence. The LM is formed by malformed lymphatic channels and is microcystic, macrocystic or a combination of both. Of all vascular malformations, 28 % will be lymphatic [1].

Clinical Presentation

A lymphatic malformation can be very large and infiltrate through all tissues. Most are in the skin and subcutaneous tissue and feel spongy. The skin overlying these lesions may be covered with epidermal vesicles, which can leak lymph fluid, which may become infected, or the skin may be normal. The lesions can vary in size due to variable filling and size of the cysts. When opening lymphatic malformations surgically smaller and larger cysts can be seen as well as conduits filled with lymph fluid.

Symptoms are related to location and size. Lesions can become indurated and can restrict motion of joints as well as producing imbalance of the fingers due to muscle infiltration (Fig. 3.3a–e). Compression syndromes are rare.

As in venous malformations, plain X-rays, ultrasound and MRI are the investigations, which provide most information. An ultrasound can differentiate between a LM and VM since flow is detectable in 85 % of the VMs, compared to no flow in LMs [42]. In macrocystic lesions, cystic lesions with thin septae can be seen. In the microcystic variant, an ill-defined hyperechoic lesion is seen [41]. Again the MRI is very important to evaluate the extent of the lesion. Clearly smaller and larger cysts can be seen. On MRI, a microcystic lesion will show as a solid lesion with a minimal enhancement on administration of contrast. Furthermore, the lesion is difficult to differentiate from a soft tissue mass [41].

Treatment

Treatment is usually conservative. Compression garments can be used; however they are not as effective as they are in venous malformations. Infections are common and should be treated with antibiotics. In infections lymphatic masses can become indurated and firm.

Indications for surgery are similar to those for venous malformations; size, weight and recurrent maceration. Localised masses can be completely removed together with the indurated skin. Often residual malformations give rise to complaints years after initial surgery. In large malformations surgery can cause extensive collateral damage and post-operative lymphedema is very common. Care must be taken to prevent infections post-operatively. Re-operations in scarred area are very difficult, therefore an operation should be planned carefully, and care should be taken that extension of surgery should not go beyond a semi-circular approach in the upper limb to prevent too much damage of normal structures. Large cysts located near the brachial plexus are very difficult to excise and can result in extensive scarring and nerve damage, giving rise to pain syndromes. Microsurgical lymphatic venous anastomoses have not been successful in these malformations. In massive lesions amputation is occasionally necessary.

Complications are mostly related to fluid leakage and infections. Postoperatively complications are often related to surgery.

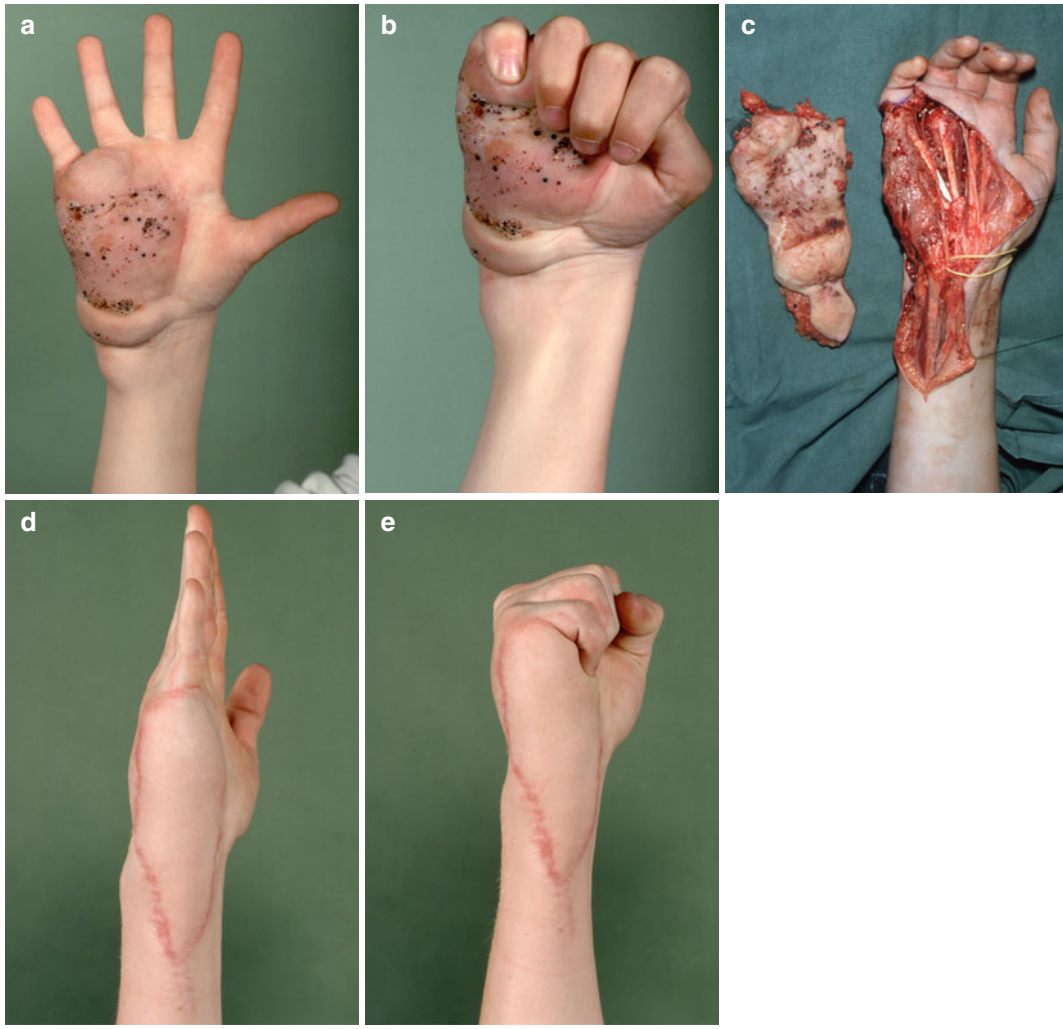

Fig. 3.3 (**a**) Lymphatic malformation of hand and wrist: extension preoperatively. (**b**) Lymphatic malformation of hand and wrist: flexion preoperatively. (**c**) Lymphatic malformation of hand and wrist: peroperative. (**d**) Lymphatic malformation of hand and wrist: extension postoperatively. (**e**) Lymphatic malformation of hand and wrist: flexion postoperatively

Arteriovenocus Malformations

In arteriovenous malformations (AVMs), because of an error in vascular development, blood is shunted directly from the arteries to the venous bed through a fistula or nidus. Of all vascular malformations, only 14 % are AVMs. The AVM is most common in the central nervous system, followed by head and neck, limbs, trunk and viscera [43].

Clinical Presentation

About 40 % of all AVMs are present at birth. The other 60 % present themselves in later childhood or adolescence. Swellings can be warm with thrills and bruits, due to fast flow. Pain following exercise is common. Elevation of the arm does not always prevent pain, like in venous malformations. Upton et al. have differentiated fast flow lesions into type A, B and C lesions [44]. Type A lesions (quiescence), being lesions with single

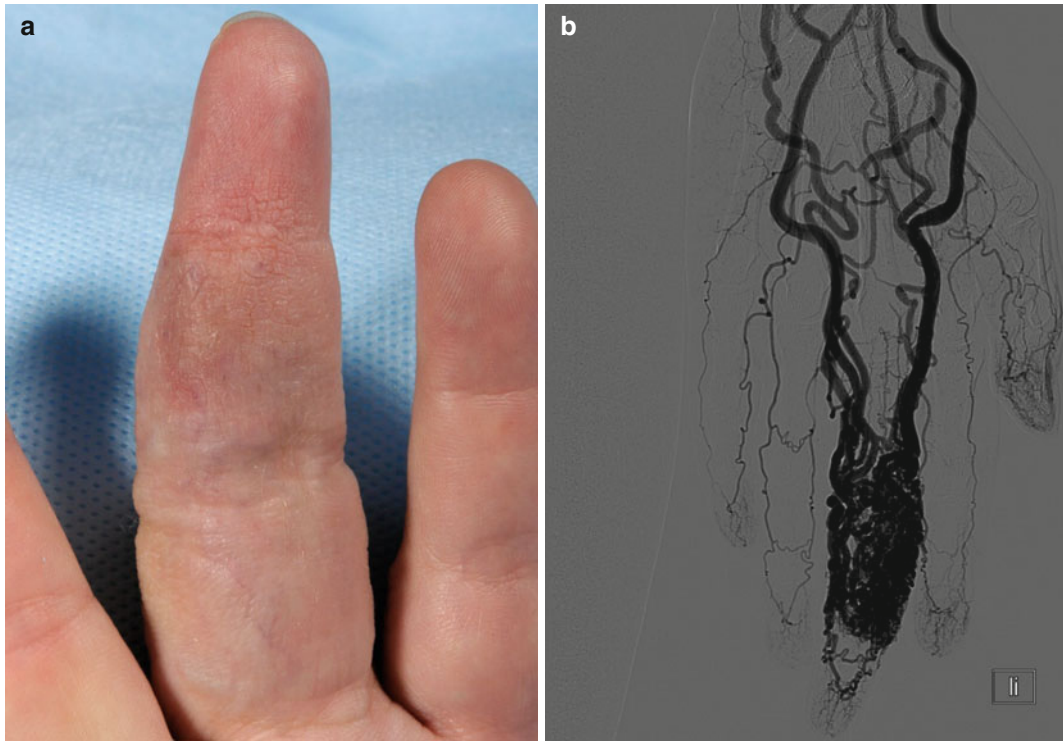

Fig. 3.4 (**a**) Arteriovenous malformation (type A) of the middle finger, with pain and dysfunction. (**b**) Angiogram of arteriovenous malformation of the middle finger of the same patient

or multiple arterio-venous fistulas in which more than one major artery can be involved. They will grow proportionately with the child and are not associated with a distal steal phenomenon (Fig. 3.4a, b). Type B lesions (expansion) have micro- and macro fistulas in a single artery of a limb or digit. These lesions can expand into previously uninvolved tissues with progressively increased flow characteristics. Type C lesions (destruction) are diffuse arterial malformations with macro and micro fistulas, involving all tissues of the limb. Type C lesions are progressive with fast flow characteristics and can develop distal steal symptoms. They can lead to ischemic pain and congestive heart disease, due to shunting of cardiac output. Compression syndrome, compartment syndromes and ulceration with bleeding are common (Fig. 3.5a, b). Hormonal influence, like pregnancy and anti-ovulate medication can aggravate symptoms and enlargement.

Diagnostic evaluation can consist of X-rays, ultrasound, MRI and angiography. X-rays can reveal bone involvement. Ultrasound will show a poorly defined hyper-vascular lesion with multiple tortuous feeding arteries. MRI studies can provide information concerning the extent of the lesion and distinguish between combined or other malformations. Soft tissue thickening and flow voids can be identified. Fast flow lesions can be distinguished from slow flow lesions. Gadolinium can be used to delineate the feeding and draining vessels, connected with enlarged central channels. Angiography is a good study to visualise extent, size and haemodynamic characteristics of shunting within the extremity. Fast flow lesions are covering a large area and cross tissue plains. A central network can mostly be detected with abnormal vascular channels with feeding arteries and draining veins.

Treatment

In the first instance treatment is conservative. Mostly compression garments are provided, however children often do not use them. On the

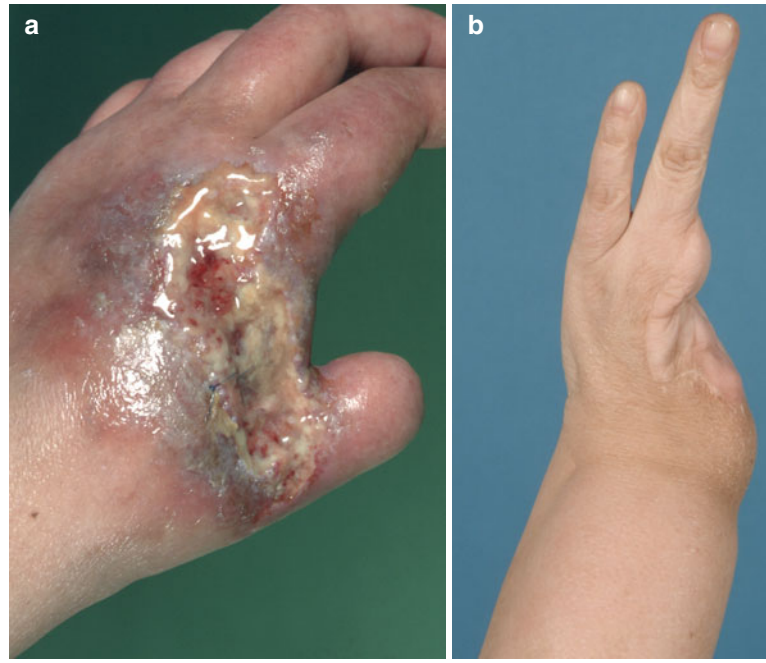

Fig. 3.5 (**a**) Arteriovenous malformation (type C) with ulcer preoperatively, excruciating pain and dysfunction of hand. Note prior distal amputation of the thumb was not successful as the nidus of the malformation was not removed. (**b**) The hand in extension postoperatively, the ulnar part of the middle finger was used as coverage of the radial side of the hand

contrary adults know exactly when to wear them. In outpatient clinics garments are torn and worn as an indication that they are used. If symptoms, especially pain, progress then arterial embolisation can be a treatment of choice in type A and type B lesions. Complications of embolisation can be distal ischemia with subsequent necrosis. Other indications for surgery are bleeding and compartment syndrome. Bleeding especially can be very troublesome with multiple admissions to ligate bleeding vessels. Type A and type B lesions can be resected as they are mostly localised. It must be stressed that there is no indication for localised arterial ligation, as dormant vessels will open and recurrence will occur in a few months. In contrast to type A and type B, type C lesions are very extensive and are very difficult to treat. Lesions tend to extend and progress more proximally from digit or hands to forearm and arm. Increase in size and severe shunting with excruciating pain makes it necessary in these cases to perform an amputation. Partial resections are not successful in these lesions. Aggressive resections are limited because of extensive anatomical damage. If undertaken the nidus of the malformation should be resected.

Diagnostic Imaging in Vascular Malformations

Most vascular malformations can be diagnosed by clinical examination. Since the visible malformation is ‘the tip of the iceberg’, additional investigation may be necessary to delineate the extent and flow characteristics of the malformation (Table 3.4).

An accurate delineation of the malformation with ultrasound is not possible, therefore an MRI is recommended (Fig. 3.2). The MRI is superior in visualizing the extent of the malformation. Furthermore, it enables one to differentiate between the different flow characteristics. Dynamic contrast-enhanced MRI increases specificity when combined with conventional MRI [7]. However, in high-flow and combined flow lesions, the MRI is inferior to angiography in depicting the nidus, fistula and supporting vessels (Fig. 3.4b). If therapy is indicated, an angiogram can be performed, possibly in conjunction with sclerotherapy. The disadvantage of an angiogram is its invasive character, with the need for ionizing radiation and administration of contrast material. Therefore, we rec-

Table 3.4 Diagnostic investigations

	Capillary malformation	Venous malformation	Arterio-venous malformation	Lymphatic malformation
X ray	No	If bone involvement is suspected	If bone involvement is suspected	No
Ultrasound	Can be useful for depth of lesion	Can be useful for extent of lesion and sclerotherapy	Not first choice	Not first choice
MRI ± contrast	Only if syndrome is suspected	Preferably, for extent, venous lakes, low-flow	Yes, for extent and distinction between mixed and 2 high flow lesions	Preferably, for extent, cysts variable in size, low-flow
CTA/angiography	No	No	Yes, for nidus, fistulas and supporting vessels and embolization	No

ommend to perform an angiogram only when treatment of the lesion is necessary [45].

Clinical Pearl

In congenital vascular malformations, excluding capillary malformations, MRI is the first choice diagnostic test.

CTA/angiography should only be used in arteriovenous malformations for extent or for different methods of embolization or prior to surgery.

Associated Syndromes

Hemangiomas are listed as a part of a syndrome in many cases. However, since the lesions occur frequently in children, the occurrence of hemangioma concurrent with other anomalies may be coincidental [46]. Generally, the vascular anomalies in the upper limb that go with syndromes will be malformations instead of hemangioma.

Klippel-Trenaunay Syndrome (*KTS*) is a rare slow-flow complex condition, present at birth and should be suspected in all children with a capillary venous malformation of the limb. It is distinguished by capillary malformations in association with varicose veins and soft tissue and bony overgrowth of the affected limb (Fig. 3.6) [47]. The involvement of the upper and lower limb is usually ipsilateral. Furthermore, deep venous malformations may be present, as well as lymphatic malformations, not only in the affected limb, but also in the viscera. The KTS should be differentiated from the Parkes-Weber syndrome, which is characterized as a high-flow complex combined vascular malformation. In the KTS,

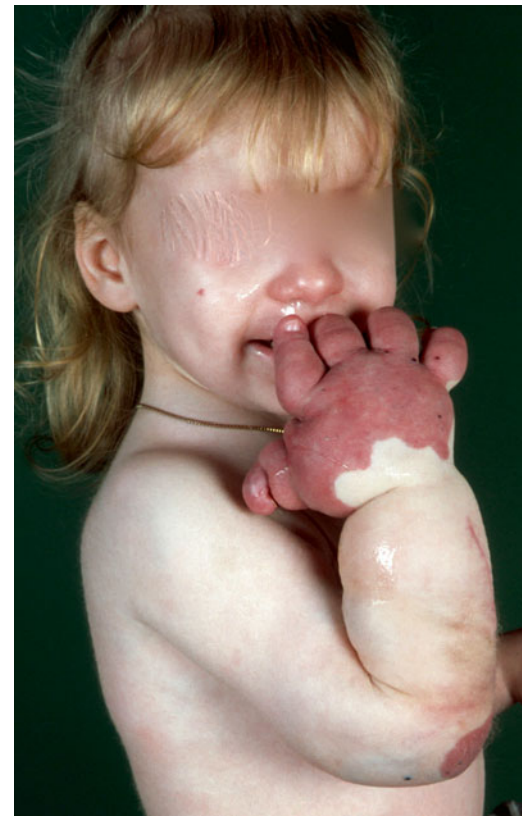

Fig. 3.6 Klippel-Trénaunay-Wilson Syndrome. Next to the visible capillary malformation, also venous malformations of the same arm, right breast and right leg are present. The other hand has macrodactyly of the thumb, index and middle finger. Furthermore venous malformations of various internal organs were identified with a tendency of venous thrombosis

life-threatening complications may occur due to thrombosis, embolism, sepsis and coagulopathy. Fortunately, most patients can be treated with pressure garments. Pulsed-dye laser gives less satisfying results as in truncal or facial lesions. Surgical treatment is preserved for symptomatic malformations as described above.

Proteus Syndrome (*PS*) is characterized by cerebriform thickening of the palms and soles, alongside the presence of epidermal nevi, hemihypertrophy, and vascular malformations, which may be identical to the presentation seen in KTS [4]. In 2004, diagnostic criteria, developed in 1998 by the workshop on Proteus syndrome, were revised [48]. Management is restricted mainly to conservative treatment of the symptoms.

Summary

Vascular anomalies of the upper extremity are rare, with diagnosis made during early childhood. Mulliken [3] introduced a biological classification, dividing the anomalies into tumours and malformations. Ultrasound and MRI are the diagnostic tools of choice; an angiogram is not often needed. With their own distinct characteristics and presentation, the anomalies need a careful clinical assessment and diagnostic imaging to suit the most appropriate, personalized treatment. Treatment can vary from nothing, pressure garments, medication, lasers, and radiotherapy to more extensive surgery.

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Glomus Tumour

4

Andrew N.M. Fleming

Keywords
Glomus • Tumour • Sub-ungual • Glomangioma • Sucquet-Hoyer canal • Glomangiosarcoma • Love's sign • Cold intolerance • Hildreth's test

Introduction

Glomus tumours of the hand were first described by Wood in 1812, calling it "painful subcutaneous tubercle." However, it was Masson in 1924 who coined the descriptive term "glomus" – Latin for "ball" – and recognised their neuro-myo-arterial origin [1]. Touraine in 1936 first described multiple hereditary glomangiomas [2].

There can be few more satisfying conditions to treat, for patient and surgeon, than a subungual glomus tumour. The patient has usually lived with a painful, debilitating finger for years, has sought multiple medical opinions and often interventions, with no apparent relief in sight. They come before their surgeon, who makes a quick and accurate diagnosis on purely clinical grounds and offers them a complete cure, with a simple local anaesthetic day-case procedure!

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Background and Pathology

Many surgeons will be familiar with the terms "glomus tumour of the carotid body" and "glomus jugulare", now more correctly termed paragangliomas – these tumours arise from chromaffin cells in paraganglia (chromaffin-negative glomus cells) of neural crest origin. They are specialised chemoreceptors of the sympathetic nervous system and are **not** histologically related to the classic subungual glomus tumour, which is correctly classified as a peri-vascular tumour of the neuromyo-arterial apparatus or "glomus body". Familial extra-digital multiple glomus tumours (glomangiomas), however, may also be related to failures of neural crest cell migration [3]. To further complicate discussions on the cellular origin and potential genetic patterns of inheritance of glomus tumours, is the rare but recognised link between digital glomus tumours and neurofibromatosis Type 1 [4].

The glomus body is a microscopic arteriovenous shunt pathway at the capillary level, which is innervated by autonomic nerve fibres and surrounded by modified smooth muscle cells or glomus cells. The afferent or arterial end is

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called the Sucquet-Hoyer canal. The glomus body is thus intimately involved with the body's thermoregulatory system and regulates blood flow to the skin. They occur throughout the body at the dermal-subdermal level, but are concentrated in areas exposed to the elements as the body's front-line temperature-receptors. They are especially numerous in the supra-periosteal layer of the nail-bed of fingers and toes and the pulps of these digits; they are thus only separated from the weather elements by an often very thin nail-bed matrix and nail-plate.

Glomus tumours are small vascular tumours arising from smooth muscle components of a glomus body and are highly organised, encapsulated and usually benign. Tumour size is usually of the order of 3–5 mm, although tumours up to 1 cm in size are not unknown. The tumour has a light brown, well encapsulated appearance, which is quite distinctive from either surrounding fat or similar tender lumps like a neuroma – this is usually white or pale.

Ninety per cent of glomus tumours are single, digital and in the sub-ungual plane of the distal phalanx of fingers or toes. They are rare and represent only 1–5 % of all soft tissue tumours of the hand [5] and most full-time hand surgeons will only see a few glomus tumours annually. They occur in all ages [6] although there is a slight female preponderance in presentation in the 30–50 year age group.

Solitary lesions may, however, also occur intra-osseously [7, 8] and at other anatomical sites such as tongue, stomach, lung, rectum, mesentry and mediastinum [5, 9] where they tend to be more common in men.

Ten per cent of glomus tumours are multiple and occur in all anatomical areas [1, 9, 10] or in multiple digits [11].

Glomus tumours have been classified clinically and pathologically – see Tables 4.1 and 4.2.

Multiple/Familial tumours These tend to be multiple, subcutaneous, pinkish-blue nodules and extra-digital in the younger, often male, patient and may have a familial inheritance – so called "familial glomangiomas"[2, 6, 13]. These are inherited in an autosomal dominant pattern with incomplete penetrance – the number of individuals displaying the trait – and variable expressivity – the degree of clinical expression of the trait in the involved person [14]. There are some families with high penetrance and expressivity and Rudolph describes one where the family had a name for it – "The Fowlie Curse" [15].

They tend only to involve the skin and subcuticular layer of the extremities (Fig. 4.1). They are often mistaken for vascular lesions and may have a raised hyperkeratotic appearance. They do not involve muscle and joints; they are not

Table 4.1 Clinical classification Iqbal [2]

Solitary	Non-hereditary, usually painful – Masson type
Grouped	Painful, non-hereditary
Multiple	Usually painless, hereditary type

Table 4.2 Pathological classification

Benign glomus tumour	Dense glomus cells, normal mitotic figures, no pleomorphism
Glomangiomyoma	Typical glomus tumour but smooth muscle cells present
Malignant glomus tumour	Deep location and size >2 cm, or atypical mitotic features, or moderate to high nuclear grade with >5 mitoses/HPF
Symplastic glomus tumours	High nuclear grade, but no other features of malignancy
Glomus tumours of uncertain malignant potential	Lack malignant criteria and superficial, but have high mitotic rate, or large size or deep location
Glomangiomatosis	Histologic features of diffuse angiomatosis and excess glomus cells

Modified from Folpe [12]

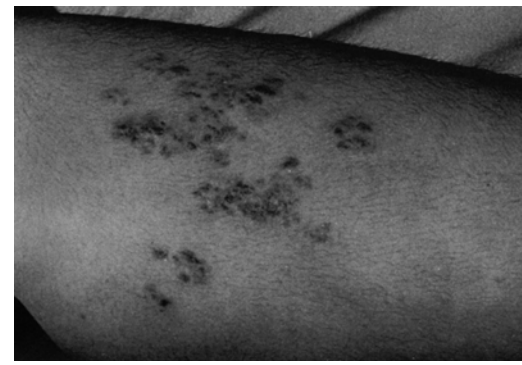

Fig 4.1 Glomangiomas of forearm (with thanks to Peter Davenport FRCS)

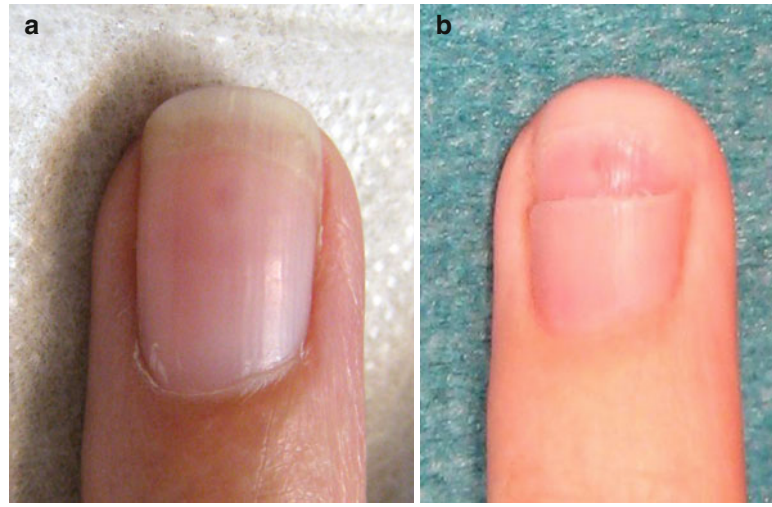

Fig. 4.2 Small distal glomus tumour showing red-ish tinge under nail-plate (**a**) and partial nail-plate removal to gain access (**b**)

compressible and may or may not be painful – they do become painful if they are subject to compression therapy, in the mistaken belief that they are venous malformations [16]. Glomangiomas may appear clinically identical to another inherited cutaneous vascular lesion; the Blue Rubber Bleb Naevus syndrome [17] – these typically appear in early infancy and may be associated with gastrointestinal bleeding. Histologically they lack glomus cells and only biopsy differentiates them.

Malignant transformation (see classification above) of digital tumours is extremely rare and usually restricted to case reports [18, 19] or small series of extra-digital tumours [12]. Hand based "glomangiosarcomas" would appear to rarely metastasize, usually occurring in the older age group and are usually larger at initial presentation, >1 cm in size.

Extra-digital malignant tumours are also larger, average >2 cm, deeper and may metastasize – 38 % in Folpe's series. These tumours may arise within an existing benign glomus tumour and are typically locally invasive, or arise de-novo as a primary malignant neoplasm.

Presentation and Investigation

The average duration of symptoms in the classic benign sub-ungual lesion is 10 years [20] with an average 2.5 doctor visits before diagnosis.

The classic presentation is of a triad of clinical symptoms and signs:

1. Paroxysmal lancinating **pain**, often with proximal radiation up the arm and exacerbated by local trauma.
2. Love's sign [21] – exquisite **pin-point tenderness** – usually elicited with the tip of a paper-clip.
3. **cold intolerance** and exacerbation of symptoms in cold weather or under a cold water stream. The use of ethyl chloride spray is a less kind way of confirming cold intolerance!

   This cold intolerance is especially so with digital tumours but much less common with single tumours in other anatomical areas [9].

Clinical signs can be otherwise frustrating although a blue-ish/red spot or blush in the common sub-ungual plane provides confirmatory evidence (Figs. 4.2 and 4.3). Nail plate abnormalities are also diagnostic (Fig. 4.4) and seen in 47 % of Van Geertruyden's series [20].

Hildreth's test [22] is a simple, clinic-based diagnostic test which has high specificity and sensitivity [23]. This is performed by exsanguinating the affected digit or arm – I personally use a cut-off glove finger, with a small distal aperture and roll this from distal-proximal, thereby exsanguinating the finger. A positive Hildreth test occurs when Love's paper-clip test then elicits no pain, followed by often intense resumption of pain on releasing the tourniquet.

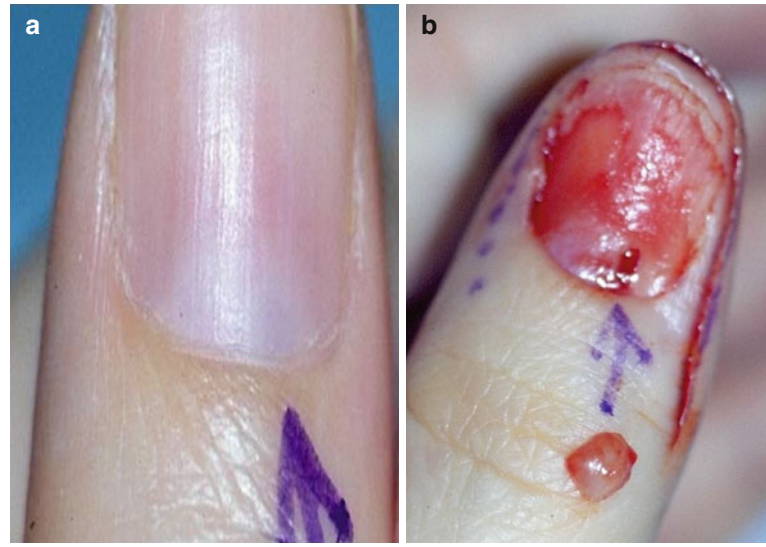

Fig. 4.3 Small proximal tumour illustrating bluish tinge under nail-plate (**a**) and fish-mouth incision to gain access to the tumour (**b**)

Extra-digital tumours are prone to even further diagnostic delay, due to their deeper location, milder symptoms and relatively small size [1, 9, 10].

Clinical Pearls: Diagnosis of Glomus Tumour
Intermittent pain
Pin-point tenderness
Cold intolerance

Special Investigations

X-Ray may reveal scalloping of the distal phalanx in up to 36 % of patients [20] (Fig. 4.5), or enlargement of the soft tissue of the sub-ungual space on lateral views when compared to the opposite normal digit [24].

High Resolution Ultrasonography (5–9 MHz) and Colour Doppler Ultrasonography (Fig. 4.5) have shown excellent value in assessing tumour size (range 3–6 mm) and location in pre-operative studies [25, 26]. The tumour presents as a hypoechoic focal mass with marked vascularity on colour Doppler .

High resolution MRI, with or without Gadolinium enhancement, will clearly demonstrate the tumour in many cases [27] (Fig. 4.6). These are better seen on a T2 weighted sequence after gadolinium injection; due to the paucity of vascular lumen they may be difficult to see on T1 images [28]. The absence of a tumour on MRI scan does not preclude the presence of a glomus tumour, as this very much remains a clinical diagnosis, however, and the need to resort to expensive investigations remains rare [27].

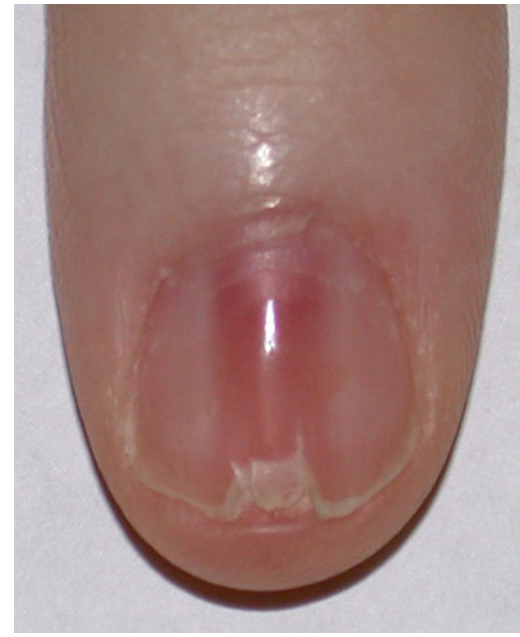

Fig 4.4 Proximal glomus tumour with nail-plate deformity

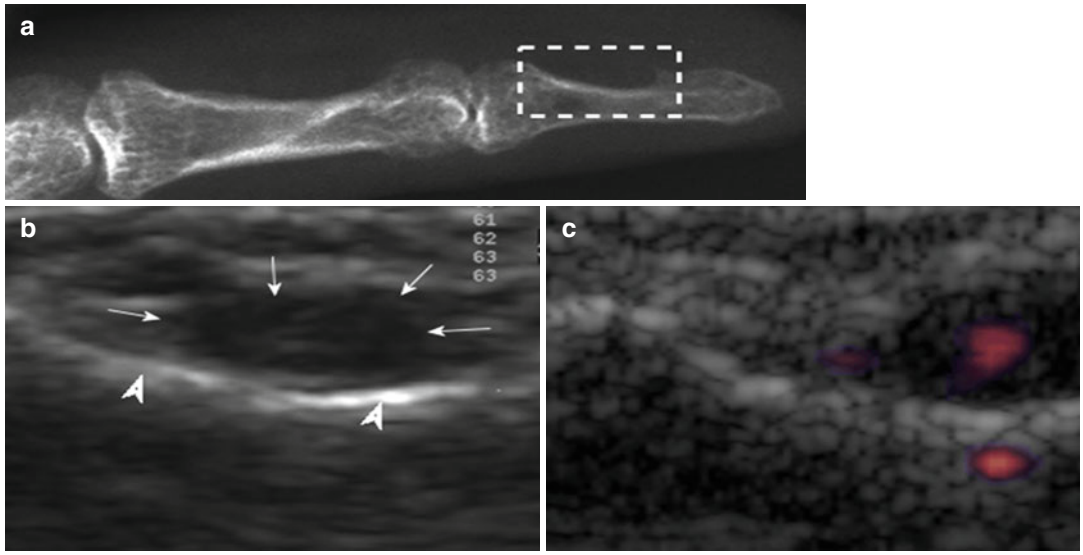

Fig. 4.5 (**a**) Lateral radiograph of the left fifth digit in a 16 year old female with a tender lump within the dorsal aspect of her finger tip; showing early bony scalloping along the flexor surface of the distal phalanx. (**b**) High resolution ultrasound focussing on the area outlined by *white rectangle* in image (**a**), showing a well defined low reflective nodule (*arrows*), adjacent to the reflective periosteal-cortical interface (*arrowheads*). (**c**) Power Doppler study demonstrates vascularity within the lesion (with thanks to Dr James Pilcher FCS (Radiol)

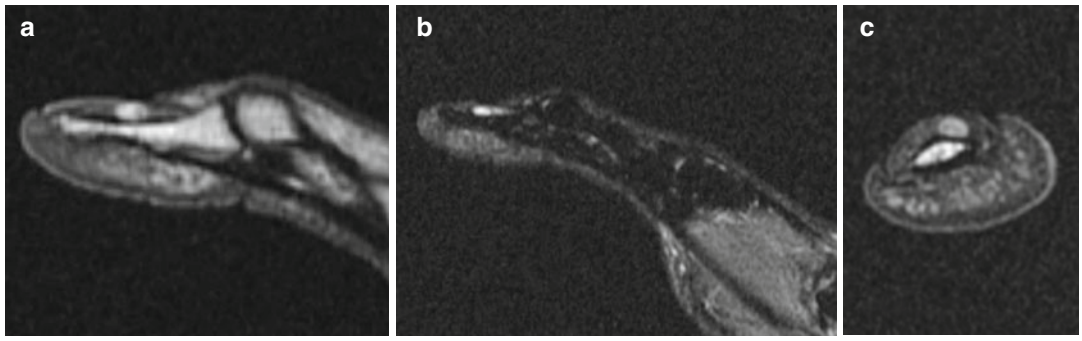

Fig. 4.6 MRI images of a sub-ungual Glomus tumour. (**a**) T1 weighted post-contrast. (**b**). sagittal and (**c**) axial STIR images of the same tumour

Treatment Options

Solitary Sub-Ungual Lesion

There are no proven conservative treatments available for the treatment of these tumours once diagnosis is established or presumed – surgical exploration and tumour extirpation remains the mainstay of therapy. There are reports of a chemotherapeutic drug, Paclitaxel, exacerbating the symptoms of a glomus tumour [29], but no reports of any sustained symptomatic relief from any drug group.

Exposure: for the common solitary sub-ungual or finger pulp tumour, surgery is performed on a day-case basis under digital block anaesthesia, digital tourniquet and loupe or microscope magnification. Many authors recommend the use of the operating microscope to carefully assess tumour margins and to see small satellite lesions (thought to be implicated in early recurrences). Once anaesthesia is established, all

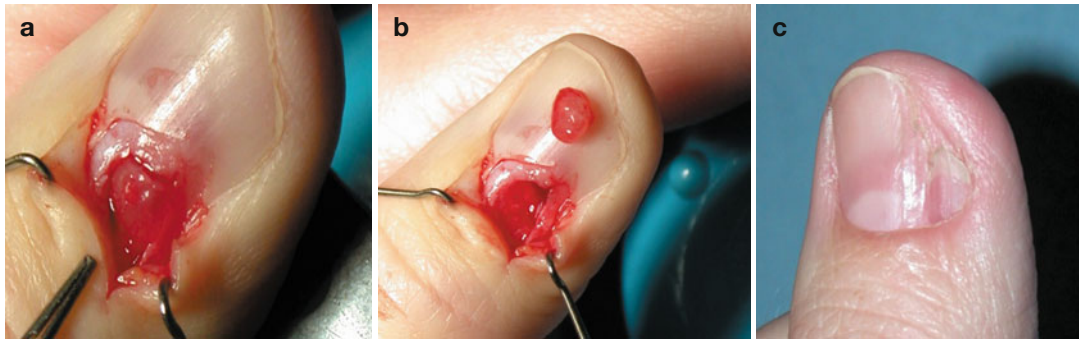

Fig 4.7 Large proximal glomus tumour (**a**) with poor preservation of nail-bed on extirpation (**b**) and resultant nail-plate deformity (**c**)

procedures commence with removal and retention of the nail plate.

The approach thereafter depends on the size and precise anatomical location of the tumour. Larger tumours will often leave the overlying nail-bed extremely thin and friable. Overly robust dissection from the classic **dorsal trans-ungual approach**, with direct incision of the nail-bed, with either linear incision or "H" or "L" flaps (Figs. 4.3, 4.7, 4.8, and 4.9) [6], may then lead to nail-bed damage and subsequent chronic nail plate abnormalities (Fig. 4.7). For more proximal germinal matrix area and larger tumours, this dorsal approach is, however, usually unavoidable. Some authors still report no nail-plate sequelae with this method [30].

For smaller and more distal and pulp tumours the **volar (or lateral) sub-periosteal approach** (Figs. 4.10 and 4.11) [24, 31, 32] is very satisfactory and the approach I will generally try and employ. Exposure may be slightly less optimal than the dorsal approach, but the risk of nail-bed injury less. An ulnar or radial hemi fish-mouth incision, depending on the tumour's laterality, with a rim of normal skin is incised and then proceeds straight to the periosteum of the distal phalanx. This is then incised and elevated dorsally, carrying a flap of periosteum and nail-bed as a continuous layer. The tumour will then usually be exposed.

Dissection: once the tumour is exposed gentle, sharp dissection with a 15 blade and minimal traction to avoid disruption of the tumour capsule, proceeds with the help of gentle teasing, using a Mitchell's periosteal elevator (dental digger). The tumour will usually pop out with just a few tendrils of connective tissue and slender vessels restraining it and needing sharp incision and cautery.

Once the tumour is removed, the nail-bed is repaired if incised or damaged – I will generally use a 7/0 vicyrl rapide with micro instruments – and the nail-bed replaced and fixed with either suture or glue. After skin closure, a simple finger bandage is applied (Adaptec) and the tourniquet then released.

Pain relief is dramatic and durable.

Multiple Lesions/Glomangiomas

The treatment of multiple glomangiomas depends very much on the clinical presentation, anatomical location and complexity of the lesions. Surgical excision remains the mainstay of treatment [33] for troublesome lesions but laser, sclerotherpay and radiotherapy have all been used for more complex lesions [2].

Malignant Glomus Tumours

For sub-ungual tumours and, depending on their location and size, ray amputation is usually indicated and curative. Surgery also remains the mainstay of treatment of extra-digital malignant tumours.

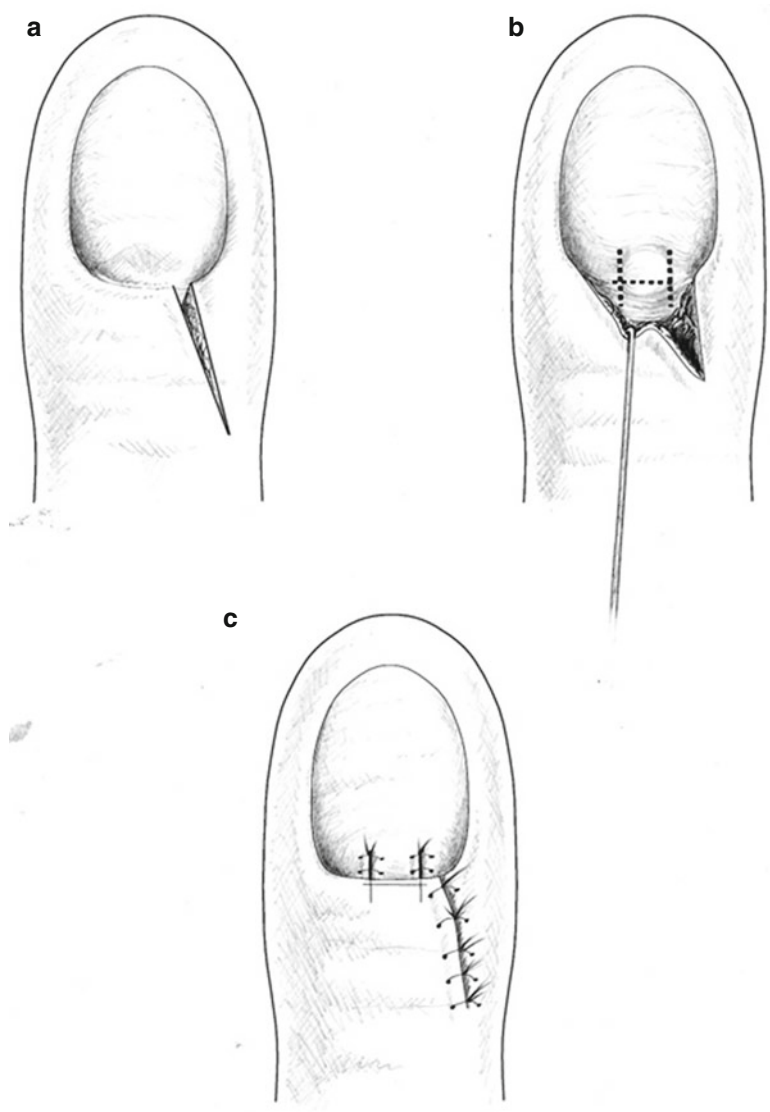

Fig. 4.8 Dorsal transungual approach. Nail-fold incision (**a**) and elevation with "l" or "H" shaped nail-bed incision (**b**) and repair with absorbable sutures (**c**)

Outcome Including Literature Review

There are no prospective trials exploring alternative treatment options for these tumours; at present all series represent retrospective Level II studies and case reports. The literature would suggest that history, examination and sound clinical awareness remains the mainstay of diagnosis. MRI and Ultrasound are useful adjuncts and will develop further as technology enables smaller lesions to be better visualised.

Surgical removal of these tumours provides immediate relief of all symptoms, with little down-time for recovery.

Complications of Treatment

Recurrence

These tumours are prone to local "recurrence" for a number of reasons. Rates of local recurrence have been quoted as high as 17 % [34].

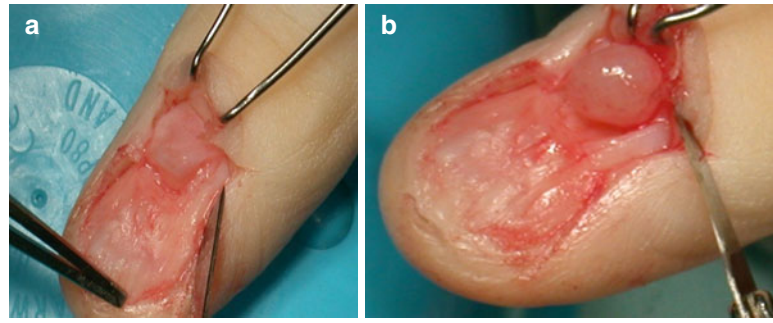

Fig. 4.9 (**a**, **b**) Proximal sub-ungual glomus tumour with "L"-shaped nail-bed incision to gain access

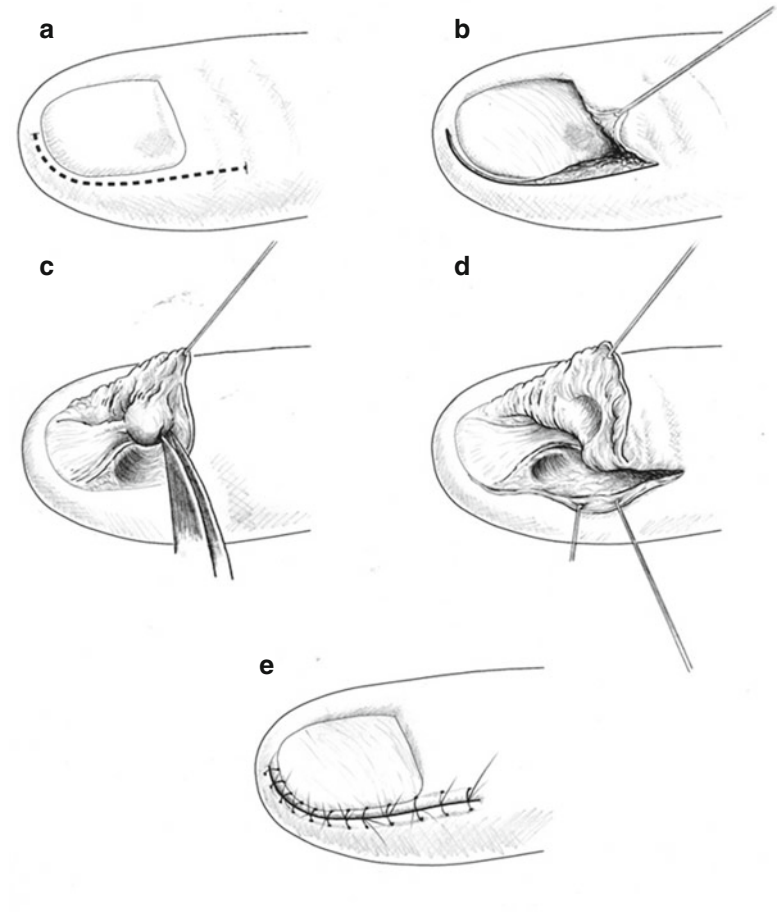

Fig. 4.10 Lateral or sub-periosoteal approach. Skin incision (**a**), nail-fold elevation (**b**), elevation of periosteum leaving glomus tumour attached to base of nail-bed (**c**). After tumour removal scalloping of distal phalanx obvious (**d**). Closure with absorbable sutures (**e**)

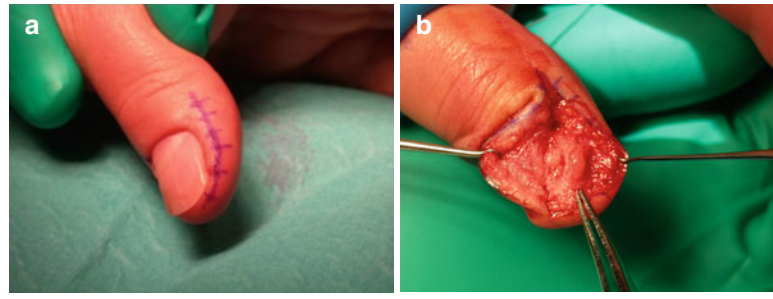

Fig. 4.11 Recurrent Glomus tumour extirpated by lateral sub-periosteal approach (**a**) preserving nail-bed, elevated on skin hook (**b**) showing scalloping of dorsal bony cortex and recurrent glomus tumour

The reasons quoted for this are, because these tumours are small, benign and in an anatomically sensitive area, limited exposure is employed for surgical removal. No patient in their series who had pre-operative MRI or Ultrasound imaging suffered a recurrence. These imaging studies are presumed to assist surgical planning to assess tumour size, exclude the presence of small synchronous lesions and help with precise anatomical tumour location intra-operatively. Their retrospective study provides no evidence for this assertion.

The presence of small, unrecognised, asymptomatic synchronous lesions at the time of the initial surgery probably accounts for many recurrences. The careful anatomical study of Gandhi [35] in their 5 "recurrences" out of 12 patients would tend to support this hypothesis. They suggest the use of the operating microscope and wider exposure of the sub-ungual plane to deal with this possibility at the time of primary surgery. Maxwell's series [36] confirms the presence of more than one localised tumour at the time of primary extirpation in 3 of 28 patients and also suggest that most recurrences are in fact due to synchronous lesions.

Most authors believe then that recurrence in the same anatomical area, which is the usual pattern, is probably from a new tumour at this site – either unrecognised at the time of primary surgery or true re-growth -especially if the lag time is measured in years, rather than weeks or months [5, 36].

Nail Plate Irregularities

This occurs especially with large, proximal tumours, where the nail-plate may be abnormal at presentation and the germinal matrix very thin. In these cases, and in those where overly robust surgical technique leads to matrix damage, subsequent nail plate deformity may persist. Where possible the volar sub-periosteal approach should thus be used. Autologous fat transfer at the time of defect creation has been suggested as a method of supporting the nail-bed repair [37] although this report lacks a clinical series.

Clinical Pearl: Surgical Approach

Use lateral sub-periosteal (dorsal) approach to sub-ungual plane wherever possible; together with use of operating microscope this may limit recurrences and nail plate deformities.

Conclusions/Personal View

These small, rare, benign tumours may often be consigned to trainee-led, local anaesthetic day-case operating lists. There is no doubt, however, that sound knowledge of tumour behaviour, nail-bed anatomy and the possible presence of satellite lesions demands consultant/attending-led surgery, with careful dissection perhaps under the operating microscope. Improved surgical technique and awareness should lead to fewer recurrences and nail-plate deformities.

With these caveats, a very happy patient invariably meets their surgeon for their post-operative visit.

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Tumours of the Hand

5

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Keywords

Hand Tumours • Enchondroma • Bizarre Parosteal Osteochondromatous Proliferation • Giant Cell Tumour • Acrometastases • Epidermoid Cysts • Nodular Fasciitis • Mohs Micrographic Surgery • Soft Tissue Sarcoma • Squamous Cell Carcinoma • Melanoma • Synovial Sarcoma • Epithelioid Sarcoma

Introduction

The hand is the site of many different types of benign and malignant neoplasms. These lesions may originate in either soft tissues or bone. The most common benign tumours of the hand include enchondromas, ganglions, giant cell tumours of the tendon sheath, and epidermoid cysts while squamous cell carcinoma represents the most common hand malignancy. This chapter describes the clinical, radiological and histological findings, in addition to the treatment strategies of the most common tumours of the hand and wrist.

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Benign Bone Tumours of the Hand

Table 5.1.

Enchondroma

Enchondroma is a benign hyaline cartilage neoplasm of medullary bone. It is the most common primary bone tumour of the hand. The phalanges are affected in 80 % of hand cases,

Table 5.1 Common benign bone tumours of the hand

1. Enchondromas.
2. Bizarre parosteal osteochondromatous proliferation.
3. Osteoid osteoma.
4. Osteoblastoma.
5. Aneurysmal bone cysts.
6. Giant cell tumours.

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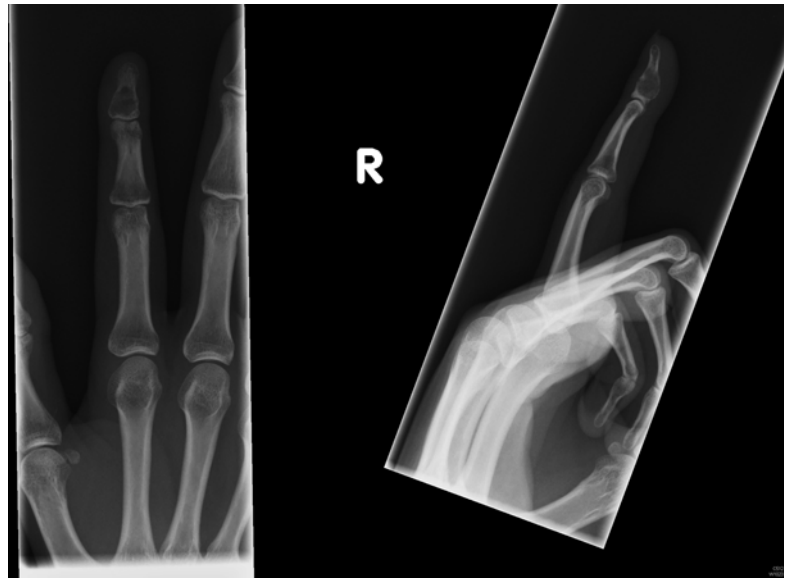

Fig. 5.1 Enchondroma of the distal phalanx. Posteroanterior and lateral radiographs show a lytic intramedullary lesion containing thin internal trabeculations and no visible chondroid calcifications

the metacarpals in 20 %, while enchondromas of the carpal bones are rare and account for less than 1 % of cases. The proximal phalanx is the most commonly involved tubular bone of the hand (40–50 %), while the thumb is the least affected digit. Patients present with painful swelling (40–51 %), pathological fracture (15–38 %), while 11–17 % present as an incidental finding on radiographs obtained for unrelated reasons [1–4].

Radiographs show a well-defined cystic, radiolucent intramedullary lesion containing thin internal trabeculations. Cortical thinning and medullary expansion are commonly seen, while chondroid calcifications *i.e.* rings-and-arcs, popcorn, flocculent, or stippled calcifications are seen less often than in enchondromas at other skeletal sites (Fig. 5.1). Magnetic resonance imaging shows multiple lobules of high signal intensity on T2-weighted and low signal intensity on T1-weighted images. Calcified foci appear as low signal intensity on both T1 and T2-weighted images. Macroscopically, enchondromas appear as lobules of blue-white hyaline cartilage with gritty yellow-white areas of calcification or ossification. Histologically, the tumour is hypocellular with abundant hyaline cartilage with chondrocytes that reside within lacunae. Enchondromas of the tubular bones of the hands and feet are generally more cellular and cytologically atypical than long bone tumours [5, 6].

Simple curettage without augmentation is the treatment of choice of symptomatic enchondromas while small asymptomatic lesions could be observed [7, 8]. Schaller et al. [8], found no significant difference in the bone density or the functional results in those patients who had no bone grafting of their enchondromas as compared to those who had. Local recurrence rates of 1.4–27.2 % have been reported in the literature including late recurrences of up to 17 years postoperatively [2–4, 9]. Malignant transformation has been observed particularly in recurrent cases [2, 10].

Clinical Pearl

Simple curettage without augmentation is the preferred treatment modality of enchondromas. Asymptomatic lesions need no treatment.

Enchondromatosis

Ollier's disease is a rare, non-familial, developmental disorder caused by failure of normal enchondral ossification and is characterized by multiple enchondromatosis with an asymmetric distribution and varying degrees of bone deformity. **Maffucci's syndrome** is distinguished from Ollier's disease by the presence of haemangiomas in the soft tissues

and/or viscera. **Metachondromatosis** is another autosomal dominant incompletely penetrant disorder leading to multiple enchondromas and osteochondromas [6, 11].

The hand is the most common site of involvement followed by the foot, femur, humerus, and forearm bones. The disease is frequently confined to one side of the body. Clinical manifestations include localised pain, palpable bony masses, leg length discrepancy, pathological fractures, and bowing deformities. Changes in the symptoms, cortical destruction, and soft tissue extension are suspicious for malignant transformation of these syndromic enchondromas as opposed to solitary enchondromas. Malignant transformation occurs in 25–30 % of syndromic cases [6, 11, 12].

Bizarre Parosteal Osteochondromatous Proliferation of Bone

These are best described as surface-based osteocartilaginous lesions typically affecting the hands and feet. Histologically, the lesion consists of a cartilage cap and underlying bone. The cartilaginous component is highly cellular with irregular bony-cartilaginous interfaces, and enlarged, bizarre, and binucleate chondrocytes, mimicking chondrosarcoma [13–16]. Radiologically, Nora's lesions are well-defined calcium containing masses adjacent to the cortical surface of the affected bones [17]. The lack of continuity with the underlying medullar cavity differentiates the lesion from an osteochondroma. It usually affects the proximal and middle phalanges and is most commonly seen in the third and fourth decades [18, 19]. Wide resection is the treatment of choice for bizarre parosteal osteochondromatous proliferation. These lesions have a high tendency for local recurrence after excision and recurrence rates between 51 and 67 % have been reported in the literature [13, 19, 20].

Osteoid Osteomas

Osteoid osteomas are benign bone-forming tumours that constitute 12 % of all benign bone neoplasms. These lesions usually become symptomatic in the second and third decades of life. Osteoid osteomas have limited growth potential and their size rarely exceed 1.5 cm in maximum diameter. Approximately 10 % of osteoid osteomas occur in the hands and feet, and these can be extremely difficult to diagnose as the typical pain pattern may be absent, and the radiographic and histologic features may vary from the classic osteoid osteomas which occur in the long bones. Most of the hand lesions occur in the proximal phalanges, followed by the distal and middle phalanges, carpal bones, and the metacarpals [21–24].

Pain and local tenderness are the most common complaints. Pain tends to be more severe at night and usually is relieved by salicylates. The pain is probably due to the high levels of Prostaglandin E2 and prostacyclin found in the tumour. Rarely, osteoid osteoma may be painless and these patients usually present with localized swelling of the affected digit. Slight erythematous changes that mimic tenosynovitis or infection can be observed in some patients [21–23].

The typical radiographic appearance is that of a small central radiolucent lesion, often referred to as the 'nidus', usually surrounded by an area of dense, fusiform, reactive osteosclerosis [21, 22] (Fig. 5.2.).

Radiofrequency ablation has now replaced surgical excision of the nidus as the treatment of choice of osteoid osteoma. This technique should be used cautiously in the treatment of superficial lesions and those lesions in close proximity to important structures as thermal burns and soft-tissue reactions with pain have been reported [25, 26].

Clinical Pearl

Osteoid osteomas typically present with night pain that is relieved by simple analgesics or anti-inflammatories. CT scan is often needed to identify the nidus. Radiofrequency ablation should be used with caution as thermal burns of important structures can occur.

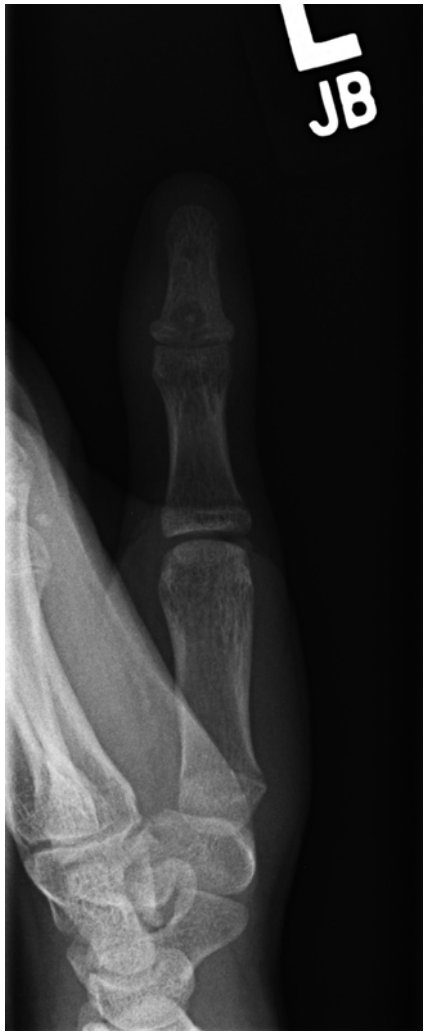

Fig. 5.2 Osteoid Osteoma of the distal phalanx. Posteroanterior radiograph shows a small central radiolucent nidus in the base of the distal phalanx

Osteoblastoma

Osteoblastoma is a rare, benign bone-forming neoplasm which produces woven bone spicules, which are bordered by prominent osteoblasts. It accounts for less than 1 % of primary bone tumours. Osteoblastomas are histologically identical to osteoid osteoma, however they are larger in size (≥2 cm in diameter), and usually do not respond to NSAIDs. Osteoblastomas of the hand bones are very rare. Osteoblastomas of the carpal scaphoid, hamate, metacarpals, and phalanges were reported in the literature [27–31].

Radiographs often demonstrate an expansile, lytic well circumscribed oval or spherical lesion surrounded by a periosteal shell of reactive bone. Focal areas of calcification may be observed in some cases indicating tumour bone mineralisation. Curettage and bone grafting is the preferred treatment modality of the primary cases. Recurrent lesions, and cases associated with severe bone destruction are best treated by en bloc resection and reconstruction [27, 29, 30].

Aneurysmal Bone Cysts

Aneurysmal bone cyst (ABC) is a benign osteolytic expansile cystic and haemorrhagic lesion of bone composed of blood filled spaces separated by connective tissue septa containing fibroblasts, osteoclast-type giant cells and reactive woven bone. Aneurysmal bone cysts of the hand account for only 3–5 % of all ABCs, mainly affecting the metacarpals and the phalanges [32–35].

Patients present with pain and swelling, which are usually present for an average period of 2–5 months. Roentgenograms show a lytic, eccentric, expansile lesion surrounded by a thin shell of subperiosteal reactive bone. MRI shows septated multilobulated lesion with characteristic fluid-fluid levels. Aneurysmal bone cyst is the only benign bone lesion which can extend across a growth plate into the epiphysis [32, 33, 36].

Although some authors reported satisfactory results with intralesional injections of calcitonin, methylprednisolone or Ethibloc (a sclerosing agent that has been used in the treatment of haemangiomas, lymphangiomas, aneurysmal bone cysts, and other hypervascular lesions), surgery remains the treatment of choice of aneurysmal bone cysts of the hand. Curettage, with or without bone grafting, is usually recommended. En-bloc excision of the primary lesion with structural bone grafting should be reserved for cases when full bone invasion of the phalanx or the metacarpal had occurred. Local recurrence was reported in 33–67 % of those cases treated with intralesional curettage while no recurrences were reported in

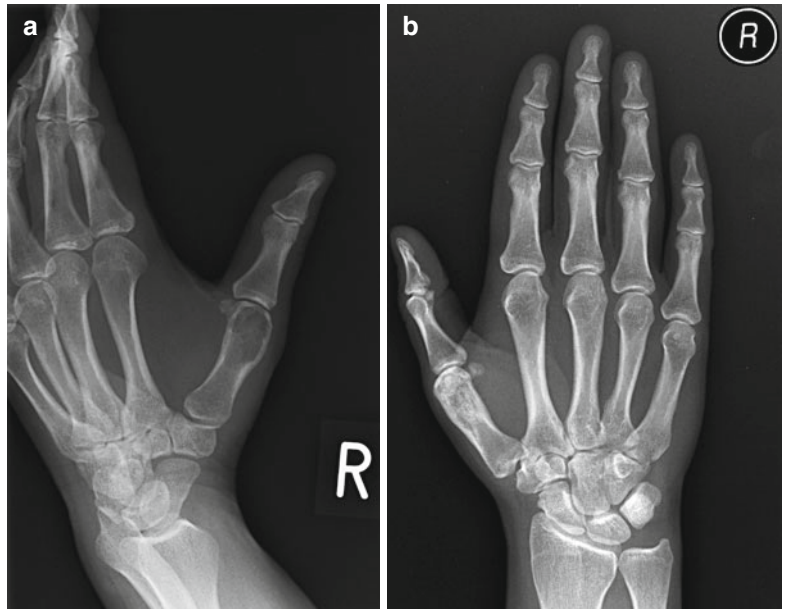

Fig. 5.3 (**a**, **b**) Giant cell tumour of the first metacarpal in a 36-year-old patient. (**a**) Preoperative radiograph shows a centrally located lytic lesion with internal trabeculations and a narrow zone of transition. (**b**) Follow-up radiograph 12 months after treatment by extended curettage shows evidence of consolidation of the lesion

those cases treated by surgical excision. Repeat curettage or resection and reconstruction may be performed for recurrent cases [33, 37–40].

Giant Cell Tumour of Bone

Giant cell tumour (GCT) of bone is defined as a benign, locally aggressive neoplasm which is composed of sheets of neoplastic ovoid mononuclear cells interspersed with uniformly distributed large, osteoclast-like giant cells [41]. Primary involvement of the bones of the hand and wrist is rare [42]. The metacarpals and phalanges are more commonly affected while less than 10 % of the reported cases involved the carpal bones [43–45]. The tumour occurs in a more central location, which differs from the eccentric location of giant cell tumour at other sites possibly due to the limited volume of bone [46]. Radiographs demonstrate a centrally located lytic lesion with internal trabeculation and a narrow zone of transition at the metaphyseal margin of the lesion [46] (Fig. 5.3). Giant cell tumours of the small bones do occur in younger patients and appear to have greater propensity for local recurrence than lesions in the long bones [42, 47, 48] (Fig. 5.4). Curettage was found to be an ineffective treatment method with recurrence rates of 79–87 % reported in the literature [43, 44]. Wittig et al. [49], reported no local recurrences after curettage, cryosurgery, and cementation of 3 cases of GCT of the tubular bones of the hand. However, most authors recommend en bloc resection and reconstruction or ray resection as primary treatment modalities of GCT of the hand with significantly lower rates of local recurrence when compared to intralesional procedures [42–44].

Clinical Pearl

En bloc resection and reconstruction or ray resection is the treatment of choice of GCT of the tubular bones of the hand as curettage is associated with high rates of local recurrence.

Primary Malignant Bone Tumours of the Hand

Bone sarcomas are rare and represent only 0.2 % of all neoplasms. While most patients with benign bone tumours present with an intermittent

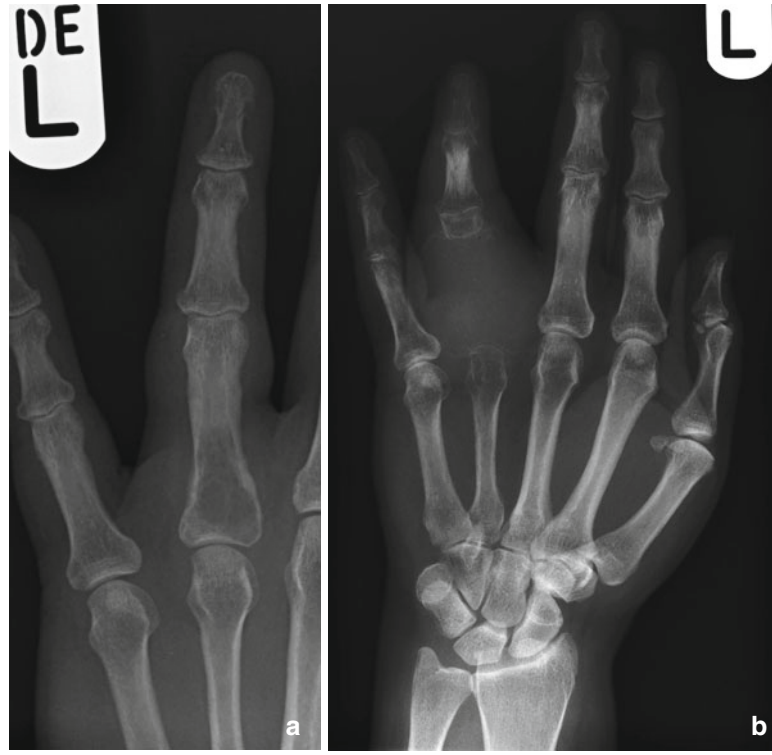

Fig. 5.4 (**a**, **b**) Giant cell tumour of the proximal phalanx in a 41-year-old patient. (**a**) Preoperative posteroanterior radiograph demonstrates a typical giant cell tumour of the phalanx. (**b**) Posteroanterior radiograph six months following treatment by extended curettage of the lesion shows extensive local recurrence. This was treated by ray amputation

aching pain or occasionally a pathological fracture, malignant bone tumours typically present with non-mechanical or night pain. Radiological features can help in distinguishing benign from malignant neoplasms. These include tumour size, location, pattern of bone destruction, periosteal new bone formation, cortical disruption and soft tissue involvement. Once the diagnosis of a malignant neoplasm is suspected, the patient should be referred to the local sarcoma unit for treatment. Needle biopsy will confirm the nature of the lesion. MRI is the main imaging modality for local staging while chest CT and radionuclide bone scan will complete the systemic staging of the tumour [50] (Table 5.2).

Table 5.2 Common primary malignant bone tumours of the hand

1. Chondrosarcoma.
2. Osteosarcoma.
3. Ewing's sarcoma.

Chondrosarcoma

Chondrosarcoma is the most common primary malignant bone tumour of the hand [51, 52]. One to eight percent of chondrosarcomas are located in the bones of the hand [53, 54]. Palmieri [55] found that 78 % of chondrosarcomas of the hand originated without a pre-existing lesion. Secondary chondrosarcomas usually develop in patients with multiple enchondromas. A chondrosarcoma arising on top of a solitary enchondroma is exceedingly rare [52, 55]. Patients are usually 60–80 years of age and present with painful swelling, often of long duration (up to 30 years) [56–58].

Chondrosarcoma of the hand almost always affect the epiphyseal region of the proximal phalanx or the metacarpal. Typical radiographic features include cortical destruction, scattered punctate densities of dystrophic matrix calcification, wide zone of transition, pathological fracture, and soft tissue extension [5, 55] (Fig. 5.5). Histologically, chondrosarcomas are composed of hyaline cartilage with hypercellularity and double nucleated cells. Bertoni et al. [56], stated

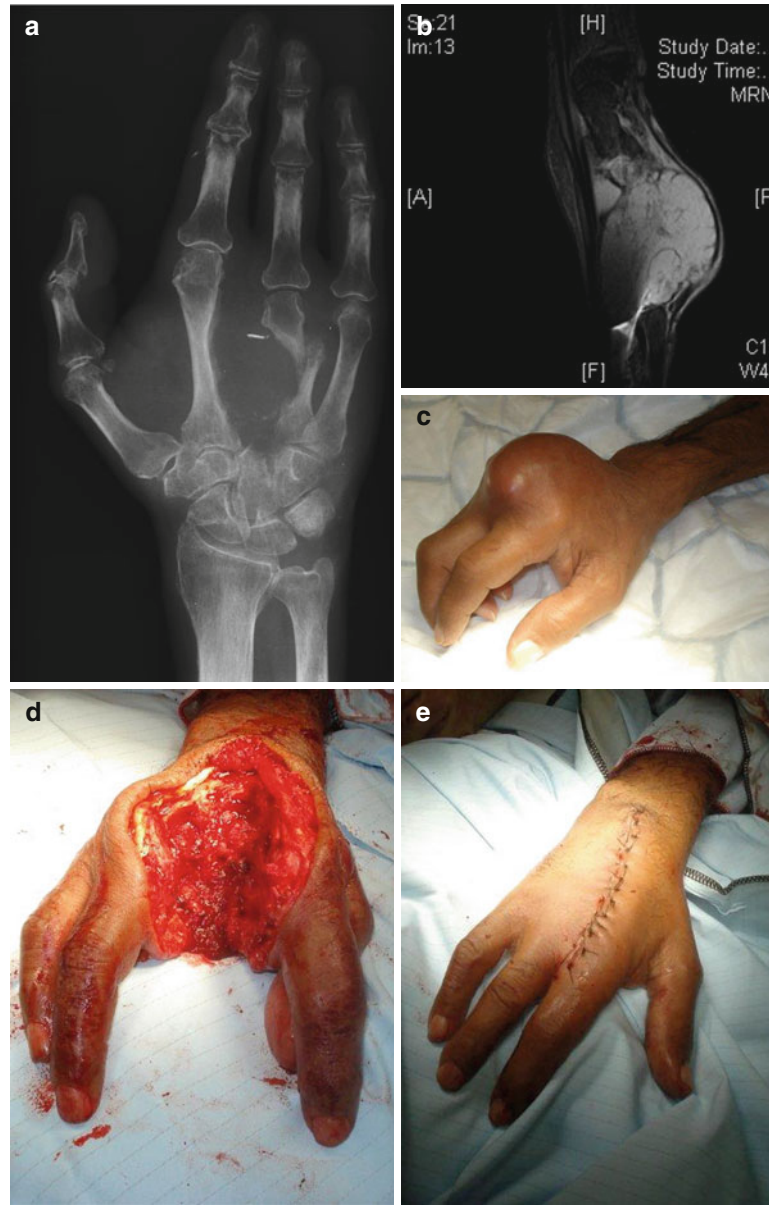

Fig. 5.5 (**a**–**e**) Chondrosarcoma of the third metacarpal in an 80-year-old patient. (**a**) Posteroanterior preoperative radiograph demonstrates a pathological fracture through the lesion. (**b**) Sagittal T2-weighted MR image demonstrates the intraosseous and extraosseous extent of the lesion. (**c**–**e**) Pre-, intra-, and post-operative pictures of the hand showing that local control of the lesion was achieved by resection of the third ray

that the histological findings of increased cellularity, binucleated cells, hyperchromasia and myxoid change may all be present in enchondromas of the small bones of the hands and feet. They found that the most significant histologic feature of chondrosarcoma in this location is permeation through the cortex into soft tissues and a permeative pattern in the cancellous bone [56].

Chondrosarcomas of the hand have significantly lower metastatic potential when compared to chondrosarcomas of other locations. However, ray resection is still recommended to minimize the incidence of local recurrence [55, 57, 59].

Osteosarcoma

Osteosarcoma of the hand is exceedingly rare with less than 50 cases reported in the literature (0.18–0.39 % of all osteosarcomas). Twenty per

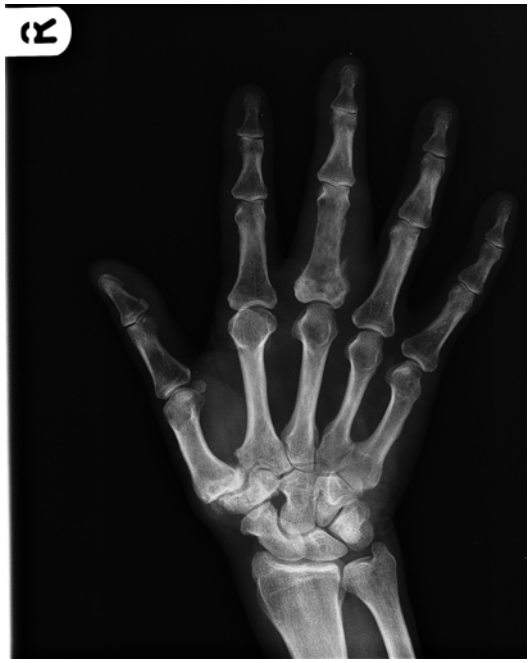

Fig. 5.6 Osteosarcoma of the proximal phalanx of the middle finger. Posteroanterior radiograph shows areas of sclerosis, periosteal reaction, and other areas of bone destruction

cent of these reported cases represented secondary osteosarcomas after prior irradiation, Paget's disease or as metastatic deposits from osteosarcomas elsewhere in the skeleton [58, 60, 61].

The site-specific distribution of these reported cases show that more than 70 % of the cases occur in either the metacarpal heads or the bases of the proximal phalanges. Osteosarcoma of the hand tends to occur at a relatively older age than in other skeletal sites, with more than 50 % of the reported cases occurring in patients 40 years of age or older [52, 60].

Pain and/or swelling are the most common presenting complaints with most cases showing an identifiable mass at the time of diagnosis. Radiographs show areas of sclerosis and periosteal reaction as well as areas of bone destruction (Fig. 5.6). The extent of extraosseous soft tissue extension generally corresponds to the size of the intramedullary tumour and the extent of cortical destruction [52, 58, 60, 61]. A complete tumour workup should include radiographs of the involved extremity, MRI of the involved region with gadolinium enhancement to evaluate the osseous and soft tissue extension of the lesion, a CT scan of the chest, and a bone scan to identify any skip lesions or possible metastatic disease.

The diagnosis of osteosarcoma is based on the identification of tumour osteoid produced by atypical cells. Conventional osteosarcoma accounts for the majority of the cases while high grade surface osteosarcoma, periosteal osteosarcoma, and low grade parosteal osteosarcoma were rarely reported in the hand [52, 58]. Treatment of high-grade osteosarcoma should consist of neoadjuvant chemotherapy, surgical excision, followed by adjuvant chemotherapy. Ray amputation is the surgical procedure of choice for osteosarcoma of the phalanges while osteosarcoma of the metacarpals usually requires more extensive resections and reconstructions [52].

Ewing's Sarcoma

Ewing's sarcoma is the second most common primary malignant bone tumour of children and adolescents. The short tubular bones of the hands are rarely involved and these represent less than 1 % of all cases of Ewing's sarcoma reported in the literature [58, 62].

Patients usually present with pain and swelling in the involved digit which may be associated with constitutional symptoms i.e. fever, leucocytosis, and malaise. Histologically, the tumour is composed of uniform round cells with small eosinophilic to clear cytoplasm, containing glycogen, and large round nuclei with fine chromatin and small nucleoli [58]. Radiographs demonstrate permeative lytic lesion with periosteal reaction and cortical destruction. Atypical radiographic findings have been reported in some cases including bony expansion, a cystic or honeycomb pattern, and lack of laminated or speculated periosteal reaction [62]. Treatment involves systemic chemotherapy in addition to local treatment in the form of radiotherapy, surgery, or a combination of both. Wide surgical excision with local reconstruction or ray amputation is the preferred local treatment modality with or without

adjunctive postoperative radiotherapy. Although chemotherapy is the mainstay of treatment, postoperative radiation therapy should be considered as an essential part of the treatment programme, especially when wide margins cannot be achieved surgically or the response to chemotherapy, as noted in the resected specimen, is incomplete [63]. The overall survival rate and the event-free survival rate in patients with Ewing's sarcoma and primitive neuroectodermal tumour (PNET) of the distal upper extremity was found by the cooperative Ewing's sarcoma study group to be remarkably high as compared to other anatomical locations. This has been attributed to the average low tumour volume and the low incidence of metastatic disease at presentation [64].

Hand Metastasis

Metastatic lesions to the hand are rare and account for 0.1–0.2 % of all metastasis. The lung is the most common primary site (50 %), followed by the breast (15 %), and the kidney. The parenchyma of lung or liver tends to act as a filter in other malignancies. The absence of a filter may account for the relative frequency of lung cancer as a source in metastasis to the hand [65–68].

Acrometastases (metastases distal to the elbow and the knee) usually occur as rare, preterminal events and often are part of a widespread dissemination of metastases but it can be the first presenting sign of an occult carcinoma in about 15 % of the patients. The age of presentation is generally after the fifth decade, although a few patients as young as 18 months of age have been reported in the literature. The clinical findings commonly include pain, swelling, and erythema. Patients may occasionally present with pathological fracture. The terminal phalanges are the most frequent sites of metastasis, followed by the metacarpals and the proximal phalanges while the carpal bones are only rarely affected. The dominant hand is more commonly involved. This could be attributed to the fact that the dominant hand receives greater blood flow than the nondominant one and may also be more prone to trauma [67–69].

Radiographs usually show a destructive permeative lesion without a marginal rim of sclerosis. Thyroid and renal carcinomas frequently produce lytic lesions, and prostate carcinoma is classically associated with blastic secondaries. Mixed lytic and blastic deposits are most often seen with malignancies of the lung and breast [67, 69].

Staging studies should be performed, since the extent of spread of primary tumour plays an important part in the prognosis of the patient. The aim of treatment of acrometastases is usually palliative. An exception is patients with solitary metastasis of renal cell carcinoma. These patients are appropriate candidates for wide surgical excision to improve survival [68–70].

Treatment modalities of acrometastases include radiotherapy, systemic chemotherapy, ray amputations, and curettage or limited marginal excision with adjuvant radiotherapy. Surgical excision of the lesion usually results in prompt relief of symptoms and early restoration of function. Ray resections to achieve wide surgical margins are often required. Patients with large unresectable lesions at presentation may be candidates for radiotherapy or intralesional surgery and adjuvant radiotherapy. Curettage of the lesion should be considered in patients with metastases in the carpal bones, or the thumb for whom amputation would result in an unacceptable degree of functional disability [67–69].

Tumours of the Soft Tissue

The large majority of soft tissue tumours are benign, with a very high cure rate after surgical excision. It is often very difficult to distinguish benign from malignant tumours of the soft tissue. Grimer et al. [71], recommended that any patient with a soft tissue mass that is increasing in size, has a size >5 cm or is deep to the deep fascia, whether or not it is painful, should be referred to a diagnostic centre with a suspected soft tissue sarcoma (STS). A core needle biopsy will confirm the nature of the lesion, however an excisional biopsy maybe the best option for superficial lesions

<5 cm in diameter. MRI scan is the preferred imaging modality on investigating a soft tissue mass. Staging studies should be completed once the diagnosis of a STS is confirmed and these should include a high resolution CT scan of the chest to exclude pulmonary metastases prior to definitive treatment.

Clinical Pearl

Any patient with a soft tissue mass that is increasing in size, has a size >5 cm or is deep to the deep fascia, whether or not it is painful, should be referred to a diagnostic centre and managed by a specialist sarcoma multidisciplinary team for a suspected STS.

Benign Soft Tissue Tumours

Table 5.3.

Epidermoid Cysts

Epidermoid cysts or epidermal inclusion cysts are cysts lined by squamous epithelium and containing keratin [72–74]. These cysts can occur in subcutaneous, intratendinous, subungual, or intraosseous locations [73]. They are believed to be caused by traumatic implantation of epidermal fragments into the dermal tissue. They are therefore more commonly seen in adult males, especially manual workers. About 70–80 % of these cysts are located on the palm or the palmar aspect of the fingers [75].

Table 5.3 Common benign soft tissue tumours of the hand

1. Epidermoid cysts.
2. Infantile digital fibromatosis.
3. Nodular fasciitis.
4. Fibroma of tendon sheath.
5. Benign adipocytic tumours.
6. Giant cell tumour of tendon sheath[a].
7. Glomus tumours[a].
8. Haemangiomas[a].

[a]See other relevant chapters

Inclusion cysts are very common and may represent up to 16 % of all tumours of the hand [72, 73]. They are usually asymptomatic although symptoms of pain, tenderness, redness, and swelling are not infrequently seen [76]. Intraosseous epidermal inclusion cysts usually present as a well-defined unilocular lytic lesion with sclerotic margins. They are more commonly seen in the terminal phalanges [72–76].

Surgical excision is only indicated in symptomatic cases where the entire cyst, its intact lining, and any overlying scar tissue from any previous penetrating injury are marginally excised. Controversy exists as to whether the intraoperative spillage of the cyst contents increases the risk of local recurrence. Intraosseous epidermoid cysts should be treated with excision and curettage of the wall of the cavity [72, 73, 75]. Local recurrence rates of 11–17 % after surgical excision have been reported [72, 75].

Infantile Digital Fibromatosis

Infantile digital fibromatosis (IDF) is an uncommon benign proliferation of myofibroblasts with characteristic inclusion bodies in the cytoplasm of the neoplastic fibroblasts [77, 78]. Clinically, patients present in the first year of life with asymptomatic smooth nodular swellings, which may resemble a keloid, on the dorsal aspect of the fingers or toes. The tumour may undergo a spontaneous decrease in the number of inclusion bodies and becomes fibrotic with time [79, 80].

IDF does not have an aggressive nature and spontaneous involution is the rule. Wide local excision has been associated with a recurrence rate of up to 60 %. These lesions should therefore be observed without any aggressive treatment unless it causes marked dysfunction of the affected digit [77–80].

Clinical Pearl

IDF should not be excised as aggressive local recurrence could be seen in up to 60 % of the patients. Biopsy should be followed by watchful waiting as spontaneous involution is the rule.

Nodular Fasciitis

Nodular fasciitis is a rapidly growing benign, self-limiting, reactive lesion that can be mistaken for a malignant neoplasm [81, 82]. This lesion is commonly seen on the volar surface of the forearm but is rare in the hand. It is most commonly seen in young adults between 20 and 40 years of age. Nodular fasciitis of the hand seems to have a close association with trauma as compared to other locations. Macroscopically, it appears as a solitary round to oval well circumscribed nodule usually measuring less than 2 cm in diameter. Histologically, lesions show marked hypercellularity and high mitotic activity but lack nuclear hyperchromatism and pleomorphism. They are composed of spindle-shaped fibroblasts or myofibroblasts separated by intercellular myxoid material mixed with areas of prominent eosinophilic hyalinised stroma and incompletely formed storiform pattern. These lesions might grow much faster than Dupuytren nodules and therefore are often initially misdiagnosed as sarcomas. Diagnosis of nodular fasciitis requires histologic confirmation, and both diagnosis and treatment are accomplished by excisional biopsy. Nodular fasciitis has a very low recurrence rate of 1–2 % [81–83].

Fibroma of Tendon Sheath

They are more commonly seen in male patients in their fourth decade. They usually present as well-defined multilobated lesions, measuring less than 3 cm in diameter. Symptoms of nerve entrapment, finger triggering, or pain are not infrequently seen. Histologically, the lesion is composed of a collagenous stroma containing spindle fibroblasts in a moderate degree of cellularity with numerous slit-like vascular channels. Magnetic resonance imaging reveals a focal nodular mass adjacent to a tendon sheath with decreased signal on all pulse sequences and little or no enhancement. The aim of surgical excision is to relieve symptoms while preserving the function of the involved tendon. Recurrence has been reported in 11–24 % of cases [84–87].

Benign Adipocytic Tumours

Benign fatty tumours of the upper extremity can be classified based on the cell origin and the location of the fatty tumour. There are three histologically distinct types of fat cells: the immature fat cells (lipoblasts) which give rise to lipoblastomas, the mature brown fat cells which give rise to hibernomas, and the mature white fat cells (lipocytes) which give rise to lipomas. Lipomas only comprise 1–3 % of hand tumours. They usually present as a gradually enlarging, soft and resilient non-tender mass. Localised pain and symptoms of nerve compression are not infrequent complaints at presentation. Magnetic resonance imaging shows homogeneous and high signal intensity on T1-weighted images and an intermediate intensity on T2-weighted images. The intramuscular variety can be divided into two types: the infiltrative type and the well-circumscribed type. Local recurrence after excision is not uncommon in the infiltrative type. Malignant transformation has not been reported in lipomas of the hand [74, 88–90].

Malignant Soft Tissue Tumours

Soft Tissue Sarcomas

Table 5.4.

The management of soft-tissue sarcomas of the hand is controversial to both the hand surgeon and the oncologist. Wide surgical excision is often difficult to achieve in the hand because of the complex anatomy and the lack of readily expendable soft tissues [91]. Wide excision may also result in significant functional loss. Marginal excision, aiming to preserve the function can compromise the oncological outcome. Moreover, radiotherapy

Table 5.4 Common soft tissue sarcomas of the hand

1. Epithelioid sarcoma.
2. Synovial sarcoma.
3. Clear cell sarcoma.
4. Rhabdomyosarcoma.
5. Dermatofibrosarcoma protuberans.
6. Malignant fibrous histiocytoma.

to the hand is poorly tolerated and could be associated with long term toxicity and poor functional outcome [92]. The importance of early diagnosis cannot therefore be overemphasized. Pradhan et al. [91], have shown that inadequate excision margins result in a 12 times greater risk of local recurrence, a three times greater risk of developing metastases and a five times greater risk of death than for those with clear margins. Bray et al. [92], and Jyothirmayi et al. [93], demonstrated that limb-salvage surgery, with adjuvant radiotherapy when necessary, is an effective alternative to amputation in the majority of patients with sarcoma of the forearm and hand. Post-operative radiotherapy to a dose of 55–60 Gy is well tolerated, with a low incidence of late toxicity [93, 94].

Local recurrence of soft tissue sarcomas is related to surgical margins, grade, and the use of postoperative radiotherapy. The overall 5-year survival rate for STS in the limbs is now being in the order of 65–75 % and is based on patient's age, grade, depth, size, and histological diagnosis. Patients with intermediate/high grade tumours should be followed every 3–4 months in the first 2–3 years, then twice a year up to the fifth year, and once a year thereafter. It is recommended that patients with low grade tumours should be followed up every 4–6 months for 35 years, then annually thereafter [52, 71, 91, 95].

Approximately 5 % of soft tissue sarcomas occur in the hand region [94]. The most common histologic types are epithelioid sarcoma, clear cell sarcoma, synovial sarcoma, and malignant fibrous histiocytoma [91, 96, 97].

Clinical Pearl

En bloc excision with clear margins is the standard treatment for all patients with adult-type, localised STS. In some situations amputation may be the most appropriate surgical option to obtain local control and offer the best chance of cure. Postoperative radiotherapy should be considered in all cases of intermediate to high-grade STS despite the anticipated significant morbidity and poor functional outcome.

Epithelioid Sarcoma

Epithelioid sarcoma is one of the most common soft tissue sarcomas of the hand [52, 91, 96, 98–100]. It was first described by Enzinger in 1970 [101]. These tumours may represent up to 30 % of all malignant soft tissue tumours of the hand in the 16–25-year-old group [97]. Males are more commonly affected with a male-to-female ratio of 2:1 [52, 98]. The volar aspect of the fingers, hand, and wrist are most commonly involved. Epithelioid sarcoma usually presents as a painless, slowly growing mass or multiple palpable nodules [52, 96]. Ulceration of the epithelioid sarcoma is frequently seen, and thus can be initially misdiagnosed as an infection [52]. The macroscopic appearance of epithelioid sarcoma is that of multiple, ill defined, indurated, greyish-white nodules [52, 96, 102]. Histologically, epithelioid sarcoma has a characteristic nodular growth pattern and is composed of a mixed proliferation of eosinophilic epithelioid and spindle cells exhibiting slight nuclear atypia, vesicular nuclei, and small nucleoli [102]. Epithelioid sarcomas are highly malignant tumours that spread in an unpredictable fashion along the lymphatic system, tendons, nerve sheaths and fascial planes [52, 102]. The tumour also carries a high risk of local recurrence (35–70 %), regional and distant metastases (40 %) [97, 101, 102]. The most frequent sites of metastasis are the lungs, regional lymph nodes, scalp, bone, and brain [95, 102]. Aggressive surgical management with wide margins of excision is therefore indicated and amputation may be necessary in selected patients. Sentinel lymph node biopsy is recommended because of the frequent regional treatment failures. The roles of chemotherapy and radiation remain unclear [52, 96, 98, 100–102].

Synovial Sarcoma

Synovial sarcoma is a rare and aggressive soft tissue tumour that accounts for 5–10 % of all soft tissue sarcomas. They may represent 8–35 % of all soft tissue sarcomas of the hand and wrist, and are predominantly seen in males under the age of thirty [91, 97, 100, 103, 104].

Clinically, synovial sarcomas are similar to epithelioid sarcomas in that they can be initially misdiagnosed as benign lesions, owing to their usual presentation as painless, slowly-growing masses that have been present for months or even years. Focal or irregular calcifications may be visible on plain radiographs [52, 105].

Histologically, synovial sarcoma is either monophasic or biphasic. Biphasic synovial sarcoma has epithelial and spindle cell components while the spindle cell component occurs alone in the monophasic type [52, 105].

Synovial sarcomas are locally aggressive with unexplained natural history of late recurrence and distant metastasis (up to 30 years after diagnosis) [105, 106]. Complete surgical excision with negative margins is the treatment of choice. The difference between marginal and wide tumour resection appears to influence the outcome. Positive microscopic margins increases the risk of local recurrence associated with increased risk of metastatic spread and decreased disease free-survival. Radiotherapy is often required for local control of the disease but the role of adjuvant chemotherapy remains controversial [107, 108].

Clear Cell Sarcoma

Clear cell sarcoma is defined as a soft tissue sarcoma of young adults with melanocytic differentiation, typically involving tendons and aponeuroses [109]. These tumours usually present as a slowly growing mass that has been present for long duration of up to several years. Macroscopically, clear cell sarcomas have uniform fascicular growth pattern with lobular arrangement of cells delimited by delicate fibrous septa intimately bound to tendons or aponeuroses. Histologically, tumoral cells are round or spindle-shaped with abundant clear to slightly basophilic cytoplasm, with round vesicular nuclei and prominent nucleoli [109, 110]. Surgical excision is the preferred treatment modality. Tumour size, local recurrence and the presence of necrosis are statistically significant predictors of prognosis. Three-year, 5-year, and 20-year survival rates averaging 72, 62, and 10 % were reported in the literature [109–111].

Rhabdomyosarcoma

Rhabdomyosarcoma represents the largest category of soft tissue sarcomas in children and adolescents, accounting for more than 50 % of soft tissue sarcomas in this age group, with an annual incidence of 4–4.6 cases per million people under 15–20 years of age [112, 113]. The alveolar rhabdomyosarcoma is the type more commonly seen in the hand and more often occurring in adolescents and young adults (Fig. 5.7), while the embryonal subtype is typically seen in children less than 10 years of age [52, 112, 113].

Rhabdomyosarcoma is an aggressive tumour that tends to invade contiguous structures and becomes disseminated via the lymphatics and blood stream [112]. Histologically, embryonal rhabdomyosarcomas are composed of primitive mesenchymal cells in various stages of myogenesis, while the alveolar subtype exhibits round cell cytological features reminiscent of lymphomas but with primitive myoblastic differentiation [113, 114].

Patients are usually treated using a multimodality approach including surgery, chemotherapy, and radiotherapy. Radiotherapy should only be considered in patients at risk of local failure (e.g. marginal/incomplete resection, alveolar subtype) [112]. Due to the technical difficulties in achieving complete surgical resection without amputation in the hand region, there has been a recent report which recommends either definitive radiotherapy or surgical resection that maintains form and function as primary local therapy with no difference in the overall survival between the two groups [115].

Dermatofibrosarcoma Protuberans

Dermatofibrosarcoma protuberans (DFSP) is a rare fibroblastic slowly growing low-grade soft tissue sarcoma of the dermis layer of the skin. The

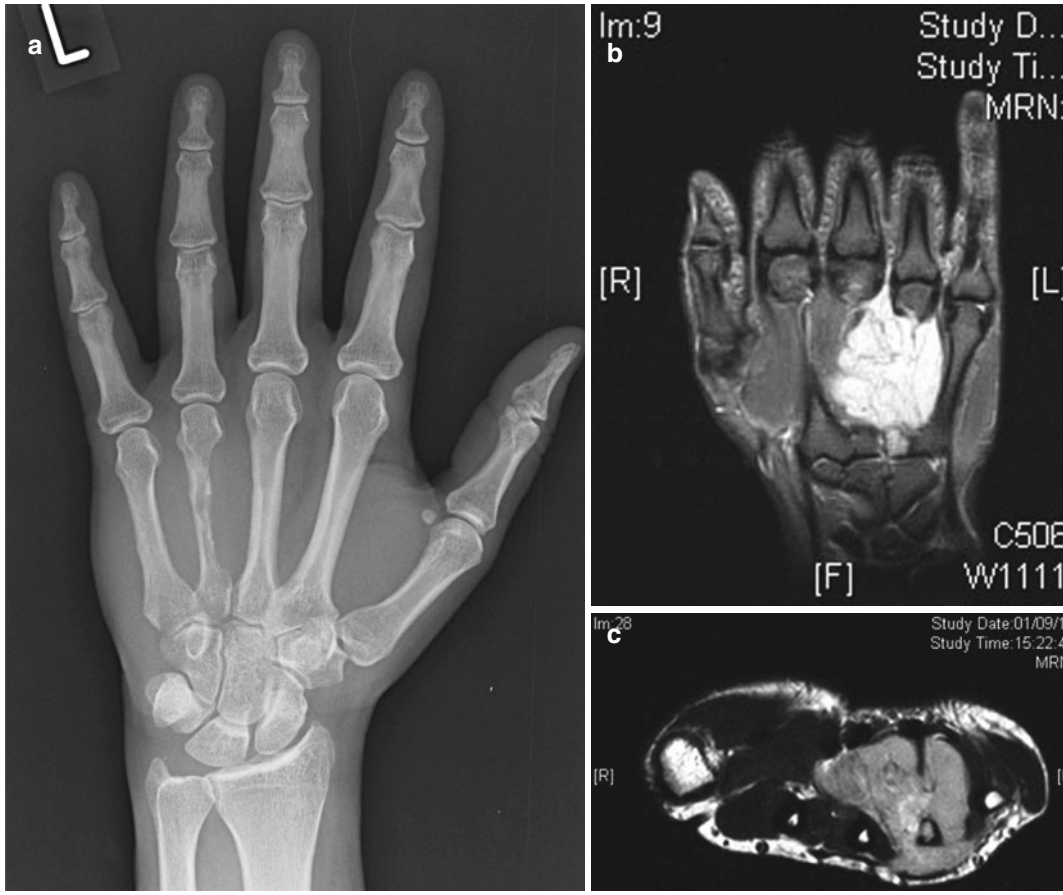

Fig. 5.7 (**a**–**c**) Alveolar Rhabdomyosarcoma of the hand in a 41-year-old male patient. **a** Posteroanterior radiograph, **b** Coronal fat-saturated T2-weighted MR image and **c** axial T1-weighted MR image demonstrate the extent of the lesion

lesion appears as a painless nodule that develops into a multinodular red-blue plaque. [52, 96] It is difficult to differentiate between DFSP, keloids, and hypertrophic scars on the basis of clinical presentation and therefore a biopsy is recommended. The locally infiltrative growth pattern features clinically inapparent extensions which often extend for long distances in a horizontal direction [116]. Consequently, the recurrence rate following excision of these tumours historically has been high, with reported recurrence rates of up to 60 % [117, 118]. Although adjuvant radiotherapy has been used in patients with positive margins, the preferred treatment modality is re-excision of the tumour aiming at a 3 cm margin. The use of Mohs micrographic surgery (MMS) has significantly decreased the recurrence rate of DFSP with a mean recurrence rate of 2.4 % reported in a literature review of 169 cases [116, 118–121].

Malignant Fibrous Histiocytoma

Malignant fibrous histiocytoma (MFH) is one of the three most prevalent histologic forms of soft tissue sarcomas in the hand [96]. MFH was found to represent 13 % of the distal upper extremity lesions in a large demographic study of malignant soft tissue tumours [97]. These are high grade malignant neoplasms that are best treated with wide local excision, combined with adjuvant radiotherapy. The role of chemotherapy remains controversial.

Malignant Skin Tumours

Table 5.5.

Squamous Cell Carcinoma

Squamous cell carcinoma (SCC) is the most common malignant skin tumour of the hand, representing 35–90 % of malignant hand tumours [122–125]. Multiple risk factors have been identified including ultraviolet radiation, immuno-compromised individuals, irradiated skin, human papillomavirus infection, chemical exposures, chronic wounds, previous burn injury, and certain genetic diseases such as xeroderma pigmentosum [126].

SCC of the hand appears to have higher rates of local recurrence, and metastasis, usually to regional lymph nodes. Schiavon et al. [124], reported a 22 % local recurrence rate, and a 28 % incidence of distant metastasis. Rayner [127] described the "danger zone" of the hand to include the dorsal skin of the proximal phalanges, interdigital clefts, and the first webspace. Tumours of this region of the hand are associated with an increased incidence of local recurrence and distant metastasis.

Many authors have recommended wide excisions with 1- to 2-cm margins. This radical treatment can lead to severe functional morbidity, undesirable cosmetic appearance with the potential need for amputation [126].

In order to minimize the functional morbidity caused by the wide surgical excision and to prevent the significant surgical injury to the surrounding tissues, other authors have favoured Mohs micrographic surgery (MMS) for treatment of cutaneous squamous and basal cell carcinomas [128]. MMS is a surgical technique that allows precise microscopic marginal control by using horizontal uninterrupted frozen sections. Joyner et al. [126], recommended marginal excision of squamous cell carcinoma of the hand with a 2- to 4-mm normal margin of tissue based on gross appearance by intraoperative evaluation with loupe magnification. Both techniques have been associated with 0 % incidence of local recurrence.

Table 5.5 Common malignant skin tumours of the hand

1. Squamous cell carcinoma.
2. Keratoacanthoma.
3. Basal cell carcinoma.
4. Melanoma.

Keratoacanthoma

Keratoacanthoma is a relatively common low-grade malignant skin lesion of the head and neck. It seldom presents in the hand and may be difficult to differentiate from squamous cell carcinoma. Keratoacanthoma is characterized by rapid growth over a short period of time, followed by spontaneous resolution in most cases. Despite this spontaneous resolution, surgical excision is the treatment of choice and large or recurrent cases can be treated with Mohs micrographic surgery. Intralesional injections of methotrexate (MTX), bleomycin and steroids have been used with success in patients who are poor surgical candidates because of patient, tumour, or treatment-related factors [96, 129–133].

Subungual Keratoacanthoma is a rare benign neoplasm which most commonly occurs in middle-aged Caucasians [134]. It is a squamoproliferative neoplasm arising at the nail bed. Radiography consistently demonstrates a well-defined cup-shaped erosion of the underlying bone [134, 135]. Local surgical excision with curettage of the underlying bone is the preferred treatment modality of these rather aggressive lesions [134, 135].

Basal Cell Carcinoma

Basal cell carcinoma (BCC) is the most common overall cutaneous malignancy. Although extremely frequent elsewhere on the body, basal cell carcinomas are infrequent on the hands (<1 % of all basal cell carcinomas). Chronic sun exposure is the main etiologic factor but the rare occurrence in the hand cannot be explained [136, 137]. BCC is a slowly-growing tumour that patients tend to neglect for years. Males are more commonly

affected and most patients would report a history of multiple skin malignancies. Metastatic spread is rare and local recurrence rate is influenced by the treatment method and the adequacy of the resection margins. Small lesions can be treated with curettage and electrodessication, laser therapy, or cryosurgery. Surgical excision with negative margins is the preferred treatment modality for BCC of the hand [96, 136, 137].

Melanoma

Melanoma is a highly malignant skin tumour that arises from melanocytes. Melanoma accounts for approximately 3 % of primary malignant hand tumours. Risk factors include ultraviolet radiation, fair skin, congenital nevi, multiple freckles, and positive family history of melanoma. These lesions have a variable clinical appearance and the diagnosis is often delayed resulting in more advanced stages of disease at presentation [138, 139]. Hove et al. [140], reported on 19 cases of melanoma of the hand with 21 % of these patients presenting with local spread of the disease (clinical stage II) and another 21 % presenting with regional lymph node metastases (clinical stage III).

Histologically, some features of the benign pigmented lesions of the palms may raise the suspicion of melanoma if considered alone. The presence of severe melanocytic atypia is possibly the most valuable feature in distinguishing between naevi and melanomas [141].

Most patients can be treated without radical amputations, even for melanomas of the digits. Roh et al. [139], found no significant difference in the survival rate between the patients treated by amputation versus wide local excision. Biopsy of all unexplained pigmented lesions on the hands can lead to early diagnosis thus improving the overall survival [140, 142, 143].

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Part II

Congenital Hand Defects

Failure of Formation of Parts Transverse and Longitudinal

6

Wee Lam and Gillian D. Smith

Keywords

Transverse arrest • Symbrachydactyly • Phocomelia • Radial club hand • Radial ray dysplasia • Thumb hypoplasia • Absent thumb • Pollicisation • Cleft hand • Ectrodactyly • Ulnar club hand • Ulnar dysplasia • Absent fingers • Poland's syndrome

Introduction

Congenital anomalies of the upper limb affect approximately 0.1 % of all newborns [1, 2]. The birth of a child with a visible difference invariably has a significant impact on the family. For the child to adjust well to their difference, the family needs to come to terms with the differences in their child early on so they can support them as they grow.

To aid in this process, the hand surgeon plays a unique role as counselor, physician and link. As a counselor, he may be instrumental in relieving the parental guilt that accompanies the birth of a child with anomalies, removing the sense of isolation felt by most parents, and providing reassurance about the functional outcome of the child's hand. As a physician, he should formulate a management plan that aims to time surgery in order to optimize long term outcome and avoid progressively increasing deformity with growth. Finally, as a link, he provides access to a wider group of professionals including physiotherapists, occupational therapists, nurse specialists, geneticists, psychologists, and established charity organisations where parents can meet other parents who have children with similar anomalies.

On the whole, unilateral anomalies are well tolerated functionally with minor adaptations but bilaterality and syndromic associations can significantly alter the prognosis for the long-term functional ability of the child. Following initial assessment by a general paediatrician, limb examination of the child must be repeated on several occasions before surgery is commenced and must include examination of the whole of both upper limbs and the feet. The physician must

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be wary of overlooking less obvious anomalies indicating a unifying diagnosis that may alter the prognosis for the child.

Clinical Pearl

Treating the patient involves establishing a relationship with the family

Address the mother's feelings of guilt directly

Do not operate on child after a single meeting with family

Do not assume other abnormalities have been detected

Aetiology and Classification

Embryogenesis of the upper extremity commences with formation of the upper limb bud on the lateral wall of the embryo 4 weeks after fertilization and is complete at 8 weeks. The majority of upper extremity congenital anomalies occur during this period of rapid limb development, when the mother is frequently unaware of the pregnancy. A thickened layer of ectoderm, the apical ectodermal ridge [3], condenses over the limb bud. This acts as a signaling center producing fibroblast growth factors that initiate and guide the underlying mesoderm to differentiate into appropriate structures in a proximal-to-distal direction. Within the posterior margin of the limb bud resides an additional signaling center, the zone of polarizing activity (ZPA), which is responsible for anterior-to-posterior (radio-ulnar) development, via the action of the sonic hedgehog protein (SHH). The Wnt (Wingless type) signaling pathway resides in the dorsal ectoderm and secretes factors, such as LMX1B, that induce the underlying mesoderm to develop dorsalization of the limb. The apical ectodermal ridge, the zone of polarizing activity, and the Wnt pathway all function in a complex orchestrated fashion to ensure proper limb patterning and growth during embryogenesis.

This fragile period of embryogenesis has been altered in animal models by experimental embryologists whose research has provided insight into mechanisms and factors responsible for limb malformation.

Table 6.1 Classification of congenital upper limb anomalies accepted by IFSSH [4]

Group		Examples
I	Failure of formation	Transverse arrest
	Transverse	Radial dysplasia
	Longitudinal	Cleft hand
		Ulnar dysplasia
II	Failure of differentiation	Syndactyly
		Camptodactyly
III	Duplication	Mirror hand
		Thumb duplication
		Ulnar polydactyly
IV	Overgrowth	Macrodactyly
		Limb gigantism
V	Undergrowth	Brachydactyly
VI	Constriction ring syndromes	
VII	Generalised skeletal defects	Arthrogryposis
		Madelungs deformity
		Aperts craniosynostosis

The current classification (Table 6.1) was first published by Swanson [4], later accepted by the IFSSH, and with our improved knowledge of limb development, is likely to be superseded soon by the OMT classification (Table 6.2) [5]. Group 1 anomalies deal with conditions arising due to transverse or longitudinal failure of formation of the upper limb. Interruption of these various signaling pathways leads to a truncation of distal or segmental development of the upper limb, with varying degrees of necrosis of mesenchymal tissue. This embryological maldevelopment can occur with or without regeneration. The attempted regeneration of tissues explains the presence of distal structures, such as a thumb in radial deficiency or nubbins in congenital amputations. Exposure to teratogens, such as thalidomide or the retinoids, early in pregnancy may produce defects in the AER with resultant major limb defects, exposure to Misoprostol or chorionic villus sampling may produce more minor but significant defects. Presentations of similar anomalies can also occur after the 8 weeks of intrauterine formation as a result of amniotic band syndrome, trauma and more recently,

Table 6.2 OMT classification of congenital hand anomalies [5]

Group	Description	Examples
I	Failure of axis formation/ differentiation	Radial longitudinal deficiency Radio-ulnar synostosis Transverse deficiency
II	Failure of hand plate formation/differentiation	Syndactyly Cleft hand Camptodactyly Clasped thumb
III	Duplication	Radial polydactyly Ulnar dimelia Ulnar Polydactyly
IV	Overgrowth	
V	Amniotic band sequence	
VI	Generalised skeletal abnormalities	

iatrogenic injury from the use of laser therapy for twin-to-twin transfusion.

This broad group includes diverse abnormalities but includes those congenital upper limb anomalies most likely to be detected during routine antenatal screening. This chapter deals the aetiology, incidence, history, presentation, treatment and outcomes of these varied conditions.

Transverse Failure of Formation

Incidence and Diagnosis

A transverse arrest refers to a shortened upper limb where, beyond a defined level, there is skeletal absence (Fig. 6.1). Attempts at regeneration often mean that vestigial distal appendages are frequently present. The overall incidence of true congenital amputations is around 7 % of all congenital anomalies [1, 6], the majority being at the hand/digit level (2.9 %) and arm/forearm (2.8 %) with the remainder at the wrist (1.1 %) [2].

Most proximal congenital deficiencies are unilateral without any genetic basis and most likely represent a disruption in development, rather than a malformation, with an abnormality in function of the AER/FGF at a time consistent with the level of the defect [5, 7]. The AER, stimulated by SHH, produces FGF-2, FGF-4 and FGF-8 to regulate and maintain proximal to distal development of the limb bud. Thalidomide probably produces phocomelia in humans by causing localized cell death in chondrogenic precursors in AER during a time window when proximal differentiation is compromised but distal differentiation has not yet begun. The diagnosis is usually straightforward in the proximal cases, but confusion exists over terminology when the failure of formation is at the metacarpal level. Traditionally, if there have been distal vestigial ectodermal elements with nails or nail ghosts present, this has been termed peromelic symbrachydactyly whereas, in the absence of these elements, the term transverse arrest at the metacarpal level has been used. The two conditions are likely to be identical with similar aetiology and pathogenesis. There is usually no difference in the management of these conditions.

Treatment of Proximal Transverse Deficiencies

The mainstay of treatment in the proximal transverse arrest is with the use of prosthetics and orthotics. The surgeon may be asked to remove distal appendages to aid prosthetic fitting, or more frequently, at the parents' request. Children with unilateral upper extremity deficiencies generally cope very well functionally without the use of prostheses [8]. Myoelectric prostheses are often discarded, as they grow older due to the weight of the devices and limitations in dexterity and sensibility. Those that continue to use prostheses,

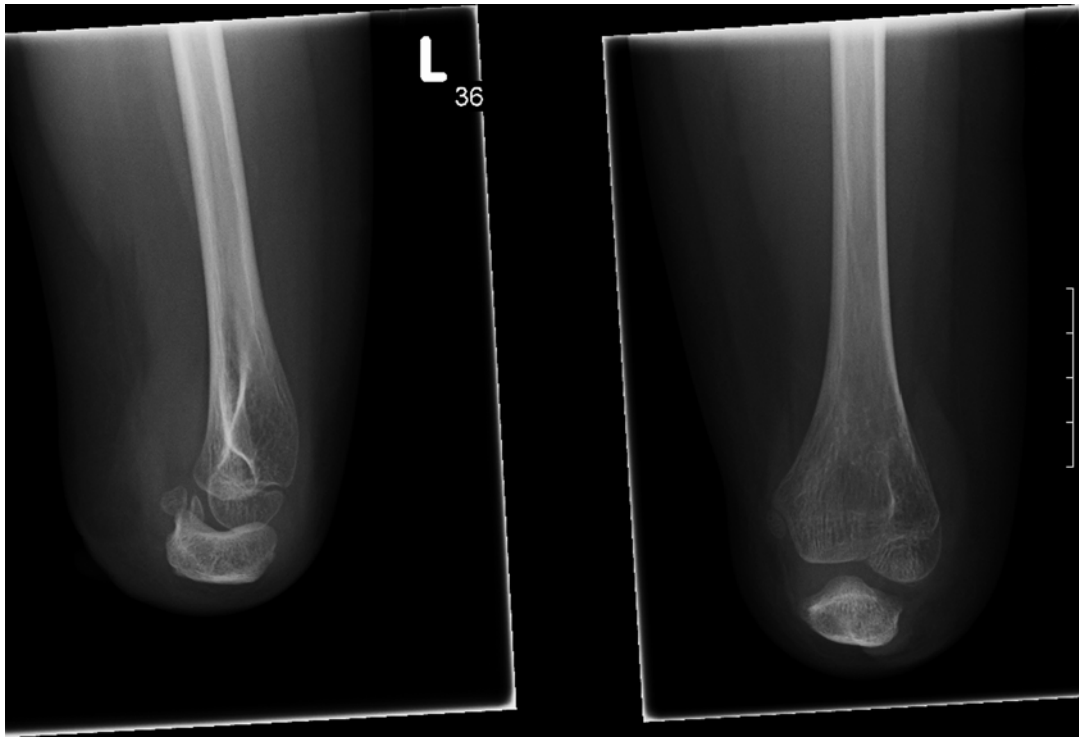

Fig. 6.1 Transverse arrest

functional or cosmetic, are inclined to do so for specific tasks or events, rather than continually. Patient Reported Outcome Measures (PROMS) may be beneficial in assessing the usefulness of upcoming future devices, which have arisen from technological advances stimulated by war. When the arrest is at the metacarpal level, the objectives move from creating an assisting hand to producing a basic functioning independent hand.

Treatment of Distal Deficiencies (Transverse Arrest at the Metacarpal Level; Symbrachydactyly)

The goals of treatment in a distal deficiency is to add or improve prehension; most importantly, tip to tip pinch, lateral pinch and large cylinder grasp, raising the child's hand function up the Great Ormond Street Ladder of functional ability [8].

Symbrachydactyly fits uncomfortably into the IFSSH classification. It is generally divided into four main types: (1) short finger type (Fig. 6.2); (2) oligodactylous (cleft hand) type (Fig. 6.3) [8]; monodactylous (Fig. 6.4); and (4) peromelic type (Fig. 6.5). Any of these or other unilateral upper limb anomalies may be associated with Poland's syndrome, where there is an abnormality of development of the pectoral girdle. Care must be taken to examine the shoulder girdle for size and presence of latissimus dorsi and pectoral muscles, and the chest wall for deformity and nipple asymmetry.

Short finger type symbrachydactyly, where brachymesophalangy (small middle phalanx) is present with or without symphalangism and syndactyly, is effectively a segmental deficiency and so is described here. No treatment may be required for short finger type symbrachydactyly, except to correct any deviation in a delta phalanx

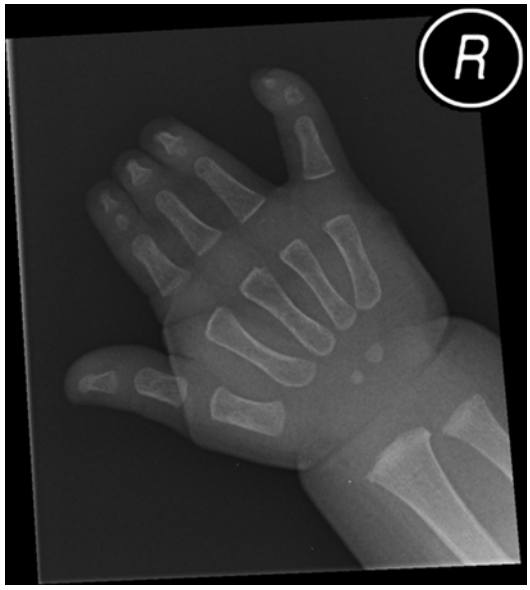

Fig. 6.2 Symbrachydactyly – short finger type

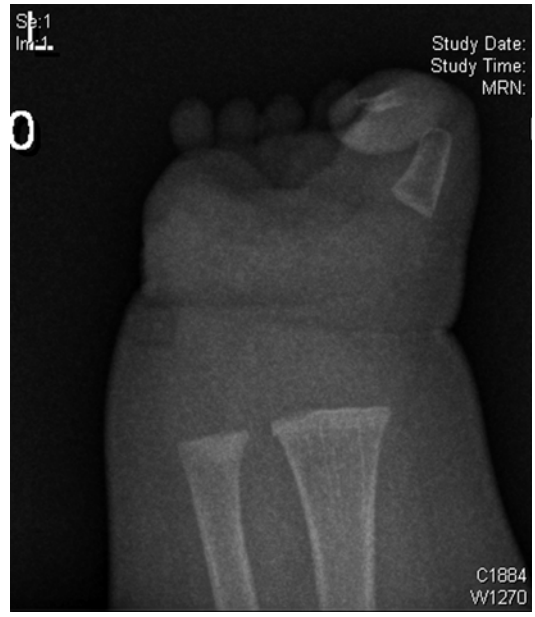

Fig. 6.4 Symbrachydactyly – monodactylous type

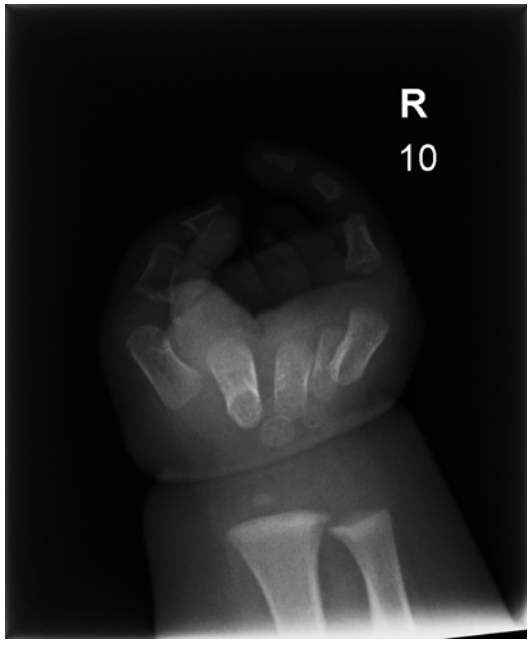

Fig. 6.3 Symbrachydactyly – cleft hand type

or to separate syndactyly. In the remaining three types, the usual approach is to ensure there is a sufficient 1st web space to create a lateral pinch, assuming sufficient radial elements are present, i.e., an adequate thumb and thenar muscles. To improve function further, any existing fingers may need to undergo lengthening in order to obtain greater precision pinch and a larger cylinder grasp. In the absence of fingers, two options are available: the use of free phalangeal transfers with or without distraction augmentation manoplasty (finger lengthening by distraction and bone grafting) or the use of free vascularized toe transfers.

Free Phalangeal Transfer with or without Distraction Augmentation Manoplasty

The use of free phalangeal transfer works on the concept of transplanting a non-vascularised whole phalanx from a toe (usually the third or fourth toes) to fill an empty soft tissue envelope extending beyond the metacarpal heads. Crucial to the whole process is the stability but associated mobility of any neo-joint created between the phalanx and the metacarpal. This requires a suitable configuration of metacarpal heads, ideally

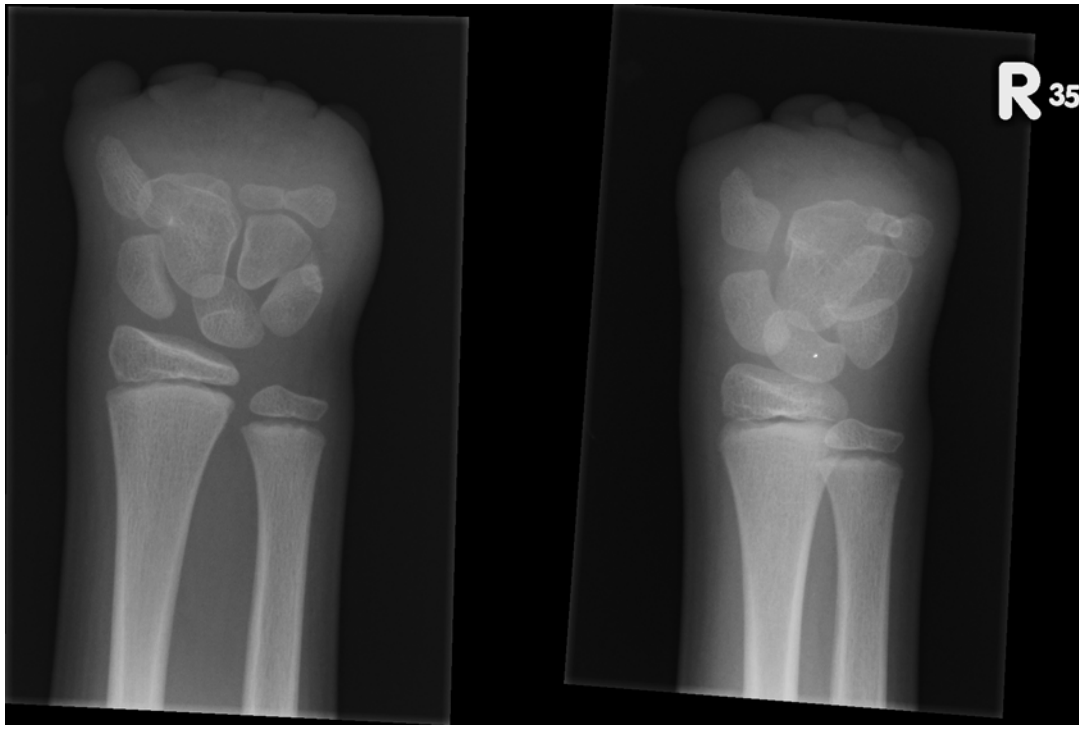

Fig. 6.5 Symbrachydactyly – peromelic type

similar to a normal hand, and sufficient soft tissue to insert a whole phalanx without undue tension.

Clinical Pearl

It is not necessary to harvest the volar plate and collateral ligaments with the toe phalanx

Sufficient soft tissue envelope and an appropriate bony configuration of metacarpal heads is necessary

The surgical approach to the hand requires release of the longitudinal fibrous tissue that extends from the flexor-extensor complex over the metacarpal head to the tip of the soft tissue nubbin, so that the size of the phalanx which will fit can be assessed.

The whole toe phalanx, usually a proximal phalanx, is harvested with an intact periosteal envelope to maximize the growth potential. It is necessary to harvest a whole phalanx since a partial phalanx inserted into the pocket, unless used as an interposition graft, will resorb. The phalanx is inserted into the soft tissue envelope, sutured onto the intact flexor-extensor hood and secured with a single K-wire passed through the phalanx and metacarpal head. Four weeks later, this is removed and active and passive movement is encouraged in the new metacarpophalangeal (MCP) joint.

Distraction augmentation manoplasty is considered as a secondary procedure, if requested, when the child is old enough to participate in the decision and comply with treatment, rarely before the age of 8 years. The length of the bone to be distracted must be a minimum of 10 mm long, be stable on the metacarpal and the MCP joints must possess a range of >60° of motion. In the thumb, the carpometacarpal joint must be stable. A distraction frame is applied to the bone with two Kirschner (K) wires or threaded pins fixing each bone proximally and distally. A subperiosteal corticotomy is performed between the proximal and distal fixation points and the bones are

immediately separated by 4 mm. Postoperatively, distraction is begun after 1 week at a rate of 1 mm/day, with weekly radiographs to monitor progress. Once the required soft tissue length is obtained, a further procedure is performed to bone graft the resultant defect with bone graft from the metatarsals, which gives a cylindrical shape to fit within the envelope of callus, which is easy to fix with a single longitudinal k wire. Although distraction osteogenesis may be successful at the metacarpal level, our experience is that callotaxis (rapid distraction with subsequent bone grafting) is required in the digits, particularly if distracting free phalangeal transfers since the period required for bone formation exceeds the length of the time that the fixation will hold or the child and family will tolerate it.

Free phalangeal transfer offers a straightforward solution to create valuable digital length, a crude grasp and lateral pinch, largely due to movement at the MCP joint, which may reach 90° of motion. From the parental point of view, it allows reconstruction of a four digit hand, albeit with extremely short straight fingers. The donor site morbidity, however, may be problematic as, with growth, particularly into teenage years, significant deformities become visible with marked shortening and deformity of the donor toes and secondary deformities in the adjacent toes [9]. Functional issues arise which may result in eventual requests for toe amputation. The use of interposition iliac bone grafts to the donor site [10] is promising, but long-term studies are necessary to assess its efficacy.

This long-term donor morbidity should be explained to parents who have to weigh the initial benefits of a non-vascularised versus vascularised toe transfer.

Free Vascularised Toe Transfer

Free toe transfer offers an alternative paradigm for improving functional abilities in congenital hand anomalies and provides a mechanism for a more sophisticated opposition pinch. The transfer of a whole composite toe with its accompanying skin, bones, joints, tendons, and nerves allows for the potential of creating new digits that are mobile and sensate, and which potentially can harness unused muscles to increase the strength of the affected hand. With vascularized transfers and careful preservation of the epiphyseal plates, an average of 90–105 % growth approximation to the unoperated toe may be expected [11, 12]. The second toe is the preferred donor and no more than one toe from each foot should be harvested if a normal gait is to be preserved.

The goal of free toe transfer in reconstruction of the congenital hand is to add at least one new prehension pattern to the hand. The reconstruction of a thumb, or partial thumb is the most obvious indication [13]. Pollicisation of the index finger still remains our first option when three or more digits are present. In defects with fewer fingers, or where there is inadequate index finger volume (or mobility) to be pollicized, toe-to-thumb transplantation remains an option to reconstruct a permanent radial post, strength and mobility being dependent on the recipient motors which may be deficient. The best indication for toe-to-thumb transplantation in congenital hand surgery is in constriction ring syndrome where the recipient base, vasculature and musculotendinous units are in pristine condition, with the expected outcome superior to pollicisation [14].

In toe to finger transfer, the goal should be to reconstruct an opposable digit to the thumb to create a tip-to-tip pinch, or transfer of two toes to create a tripod pinch, either as a synchronous double toe transfer or asynchronous double toe transfer [11].

Kay [15] classified the indications for toe-to-hand transplantations into either transverse or longitudinal defects; in the formal, there is a higher percentage of suitable residual structures for toe-to-hand transplantations. In contrast, longitudinal deficiencies often present several challenges with missing structures, often necessitating the additional transfer of adjacent tissues such as vessels or a stable skeletal base, or other treatment modalities like distraction lengthening. Angiography of the affected hand and donor foot should be considered when there are grossly abnormal defects, as it may highlight the absence of available vessels that may preclude microsurgical procedures, or require more detailed

planning such as the harvest of a longer pedicle for end-to-side anastomosis to more proximal vessels.

During the procedure, the surgeon dissects the hand first and confirms the availability of adequate recipient structures before proceeding with the foot. Harvesting of the toe begins on the dorsum of the foot to find a suitable vein, and then proceeds to the first web space to identify the dominance of the supplying artery. In general, the dorsal system is dominant in up to 70 % of cases [15, 16], which allows for a much easier dissection in a retrograde fashion. Removal of the whole of the second metatarsal gives better access when dissecting the toe and also allows easy closure of the resultant cleft in the foot to leave an acceptable donor site. The toe is K-wired in place on the hand, tendon and nerve repairs are performed and microsurgical anastomoses of arteries and veins are carried out. Both nerve and arterial anastomoses may have to be carried out in an end-to-side fashion due to congenitally deficient or absent recipients. Tourniquet time should be kept to a minimum as well as the ischemic time between harvest and revascularization, due to the relative sensitivity of the growth plates to ischemia [12, 17].

With modern technology and experience in microsurgery, success rates of survival of transferred toes should be more than 95% in experienced centres. Care of the post-operative microsurgical pediatric patient is identical to the adult patient in that every effort must be made to prevent vasospasm or compression of the transplanted vessels. The patient must therefore be kept in an appropriate setting with 24-h monitoring of the patient and the flap. There should also be a low threshold for re-exploration at the first sign of arterial or venous compromise, again due to the extremely sensitive nature of the epiphyseal plates to ischemia [17]. The use of indwelling regional local anesthetic catheters or ongoing sedation may also be beneficial to prevent vasospasm [15].

One of the advantages of toe to hand transfer is the mobility of the transferred joints in the toes that allow for improved prehension, including precision pinch and a stronger grip. Despite secondary procedures like tenolysis, the active range of motion remains significantly less than the passive range [11]. The recommendation by Kay [15] to repair a second extensor tendon in order to allow some degree of independent movement or the use of prolonged night splintage may help reduce this problem if it is related, as believed, to the pre-existing extensor lag in the toes.

Clinical Pearl

Where the defect is unilateral, overall function will be good – surgery is to upgrade the assisting hand

Where the problem is largely cosmetic – surgery will replace a naturally abnormal limb with an unnaturally abnormal one

Free phalangeal transfer may create a 5 digit hand but at the expense of a major donor site defect

Only free toe transfer will add strength to the hand, with minimal donor site morbidity but will not create a 5 digit hand

Phocomelia

In phocomelia, the limb has an intercalary segmental failure of formation resulting in the presence of distal segments such as a hand, but with absence of a proximal long bone [18]. The term means “seal-like” in Greek since the limb may appear as a flipper-like appendage (Fig. 6.6). Phocomelia includes Roberts syndrome, caused by mutation in the ESCO2 gene on chromosome 8, inherited in an autosomal recessive fashion, where the chromatids attach poorly and cell division is delayed, producing four short limbs. Research has suggested that thalidomide and X-irradiation, both known to cause similar defects in chick limb buds, may cause a time-dependent loss of skeletal progenitors, perhaps by preventing angiogenesis. There are three main types of phocomelia [19]

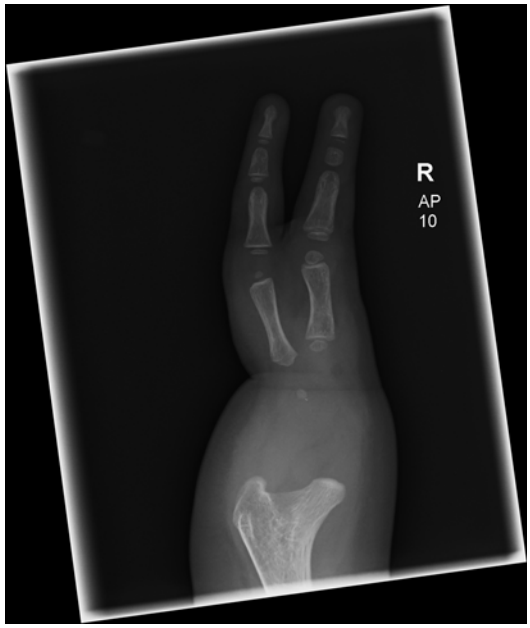

Fig. 6.6 Phocomelia

- Type I: hand attached directly to the trunk (complete phocomelia)
- Type II: short forearm attached to trunk (proximal phocomelia)
- Type III: short humerus attached to hand (distal phocomelia).

The incidence is rare, constituting less than 1 % of upper limb anomalies [2]. There is now some evidence that these defects largely fit within the most extreme of the longitudinal radial and ulnar defects [20]. In true phocomelia, there is little place for surgery and the majority of care centres on the use of prostheses. Exceptions are where confusion exists between a type III phocomelia and a radial or ulnar aplasia, as the latter could potentially be managed more expectantly (see below). In Roberts syndrome, humeral lengthening may assist hygiene functions.

In examining the child, it is important to start proximally as the clavicle and scapula may be abnormal. Although distal elements are always present, the hand is almost always abnormal and may require reconstruction, especially of the thumb [21]. Any other surgery carried out should be with the aim of allowing orthotic and prosthetic fitting and use as soon as possible afterwards.

In reality, many children with normal lower limbs will use these to perform tasks for which the normal child would use their upper limbs. Many will use prosthetics or orthotics for certain specific tasks and discard them in favour of the remains of their sensate upper limbs, supplemented by their lower limbs, for the rest of the time.

Clinical Pearl
Perineal hygiene is a significant problem for these patients

Longitudinal Failure of Formation

Radial Longitudinal Deficiency

Radial longitudinal deficiency, often referred to as "radial club hand" describes a range of conditions where there is total or partial absence of the bones and soft tissues of the preaxial border of the extremity. It is slightly more common in males than females and the right side is affected twice as much as the left [2]. The incidence is understood to be in the region of 1:30,000 to 1:100,000 [22]. While the etiology of radial club hand remains uncertain, it is likely that both genetic and environmental factors are likely to have an influence. The embryological abnormality is probably related to a combination of defects in the AER and ZPA signaling centres, as demonstrated in chick embryos experiments [23]. SHH secreted from the ZPA establishes an "ulnarizing" gradient and maintains ulnar proliferation. FGFs secreted from the AER promote proliferation in the underlying progress zone which adds to limb volume and width but progressive loss of FGFs leads to lack of development of radial structures. The hand plate develops in the fifth week of gestation. Digit number and identity are determined by a combination of the Homeobox transcription factors (HOX) and SHH. SHH results in a ulnar-to radial gradient of bone morphogenic

Table 6.3 Syndromes associated with radial club hand

VACTERLS association
Fanconi anemia
Holt-Oram syndrome
Ventriculoradial dysplasia
Craniosynostosis-radial aplasia syndrome (Baller-Gerold syndrome)
Nager sydrome hemifacial microsomia
Goldenhar syndrome (oculo-auriculo-vertebral syndrome)
Huberg-Haywood syndrome (oro-cranio-digital syndrome)
Rothmund-Thomson syndrome
Duane radial dysplasia syndrome
Levey-Hollister (LARD) syndrome
Seckel syndrome
Trisomy 18 (Edward syndrome)
Trisomy 21 (down syndrome)
Trisomy 13 (Patau syndrome)
Thrombocytopenia-absent-radius syndrome
Aase-Smith syndrome
Instituto Venzolano de Investigaciones Clentificas (IVIC) syndrome
Okihiro syndrome (Duane radial ray syndrome)

proteins (BMPs) that induce apoptosis in the interdigital spaces by suppressing FGF expression in the AER.

The majority of cases of radial dysplasia are sporadic, although exposure to teratogens such as thalidomide [24], valporic acid [25] and radiation can yield radial deficiencies. Bilateral cases are more likely to have a syndromic association (Table 6.3); the well-known ones include the haematological disorders of Fanconi's anemia and thrombocytopenia absent radius syndrome (TAR), as well as Holt–Oram syndrome (cardiac septal defects), and VACTERL syndrome (Vertebral abnormalities, Anal atresia, Cardiac abnormalities, Tracheoesophageal fistula, oEsophageal atresia, Renal defects, and Limb abnormalities). Patients with TAR syndrome may be distinguished by the presence of functional but abnormal thumbs, as well as bilateral involvement with disproportionately short forearms. It is crucial to exclude Fanconi's anaemia in all cases of radial dysplasia since this typically presents later in life (3–12 years) as aplastic anaemia which is fatal if left undiagnosed and untreated [26].

Clinical Pearl

Screening tests for radial ray dysplasia:
chromosome fragility test for Fanconi's anaemia,
echocardiogram,
renal ultrasound,
spinal x rays,
FBC if functional thumbs present

Functional Anatomy

The radial deficiency ranges from a mild deviation with some degree of tightness on the radial wrist aspect to a complete dramatic collapse where the hand in lies more than 90° of radial deviation, with carpal subluxation onto the volar aspect of the forearm. The forearm is shortened, with associated elbow and shoulder abnormalities, especially in VACTERL syndromes. The most popular classification is by Bayne and Klug [27]:

- Type I deficiency, the mildest type, is characterized by mild radial shortening of the radius
- Type II deficiency, is characterized by a shortened radius with distal and proximal physeal abnormalities and moderate deviation of the wrist.
- Type III deficiency is characterized by a partial absence of the radius (most commonly the distal portion) and severe wrist radial deviation.
- Type IV deficiency is characterized by a complete absence of the radius (Figs. 6.7 and 6.8)

James et al. suggested modifications [27] to include a group 0 and group 1 to include abnormalities affecting the carpus and distal radius.

Associated with the bony abnormalities, there is severe neuromuscular aplasia affecting the radial side of the hand and wrist, especially affecting muscles that originate or attach to the radius. This includes the extensor carpi radialis longus, extensor carpi radialis brevis, pronator teres, flexor carpi radialis, palmaris longus, flexor pollicis longus, pronator quadratus, and supinator muscles. The radial nerve may terminate at the elbow. The ulnar nerve is normal. An enlarged median nerve may substitute for the superficial radial nerve or both may exist but the median nerve is sited more

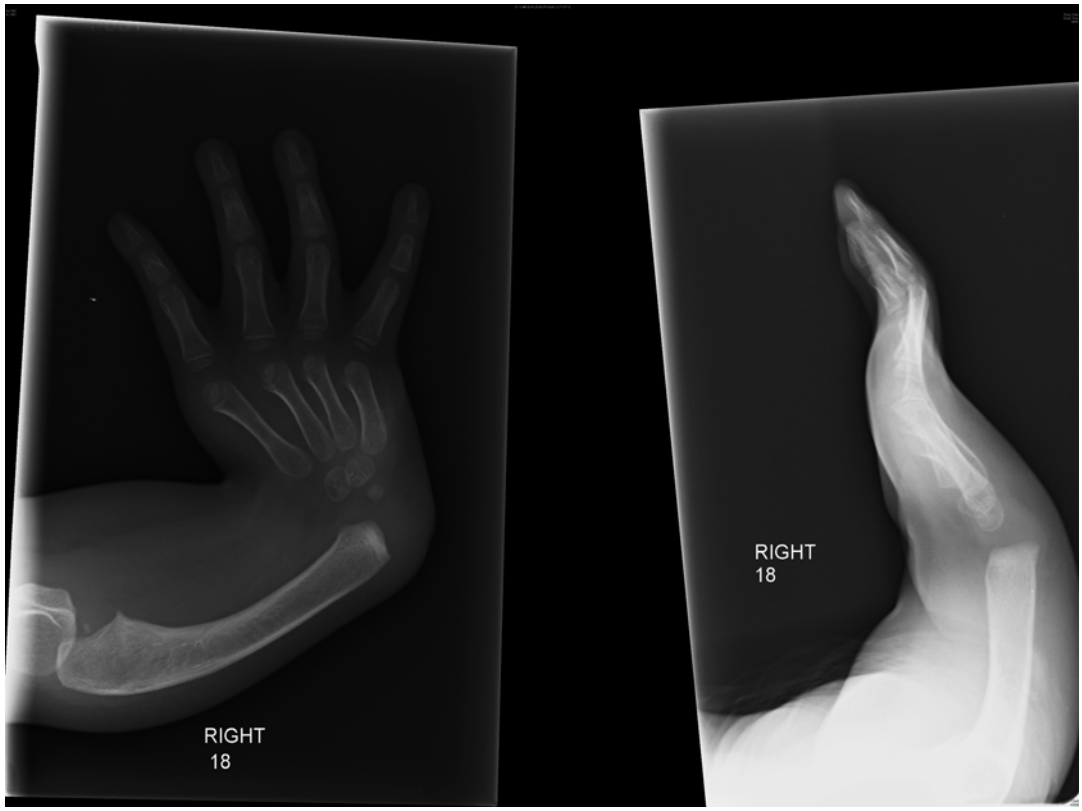

Fig. 6.7 Radial club hand – type IV

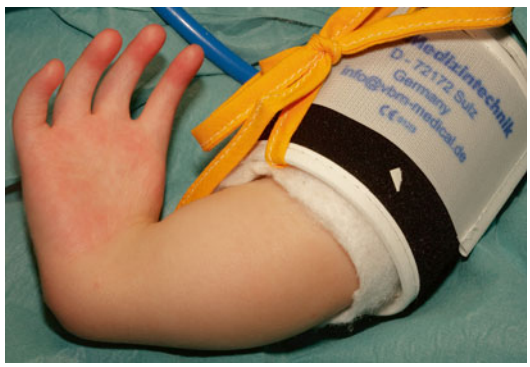

Fig. 6.8 Radial club hand

radially than normal and due to its aberrant anatomy is at risk of damage during surgery. The radial artery may be absent or deficient.

Treatment

Treatment of radial longitudinal deficiency may address both essential and desirable aims. The essential aims of surgery are: (1) to create a stable ulnocarpal relationship, (2) to correct the radial deviation by a combination of bony realignment, soft tissue releases and tendon transfers (3) to reconstruct the thumb, if possible and (4) to improve cosmesis. The desirable aims are: (1) to preserve as much range of motion as possible at the wrist and (2) maintain or improve forearm length. Surgical procedures should not be at the expense of compromising the already impaired growth potential of the ulna. They must also minimize the chances of recurrence. For most patients, the treatment remains difficult and controversial [28].

Contraindications to surgery include the presence of other life-threatening and urgent systemic conditions that require medical or surgical attention, and mild deformity that poses no functional limitations. The stiff elbow is often quoted as a strict contraindication to centralization of the wrist because this may compromise

the hand reaching the mouth [2, 28, 29]. The senior author's view is that this is unnecessarily conservative and that any active motion in the elbow increases once preoperative splintage is commenced on the wrist, a view similar to that of other authors [24, 30].

Early Presentation

Ideally seen soon after birth, the physician can initiate appropriate investigation for associated pathologies and treatment, consisting of stretching exercises by the parents and splintage of the wrist. Subsequent treatment depends on the grade of the deformity.

Clinical Pearl

Check shoulder stability

Elbow range of motion will improve with wrist splintage

Type I deformities: These mild deformities may be missed *per se* but are usually referred to the hand surgeon due to associated absence or hypoplasia of the thumb. Although these patients often improve without surgery to the wrist, it should be noted that a tendency to collapse into flexion and radial deviation with load bearing is persistent to some degree. The commonest form of treatment is splintage, although occasionally some form of tendon transfer may be necessary.

Type II deformities: These, the rarest group, have a residual radius present which actually complicates treatment decisions. While the temptation is to perform distraction osteogenesis with bone grafting, this is exceedingly difficult and may have to be repeated several times during growth. These are usually treated like Type III and IV deformities but an alternative approach is that of Vikki [31] where a free vascularized 2nd metatarsophalangeal joint is used to stabilize the hand after distraction and theroretically, these would be ideal cases for this technique.

Types III and IV: Continuing controversies surround the treatment of these more severe deformities, but the most frequent approach is as follows [29, 32–34]:

- Pre-operative distraction with an external fixator device for stretching of soft tissue
- Release of soft tissue deforming forces
- Centralisation, radialisation or other stabilizing procedures to create a new ulno-carpal joint with temporary ulnocarpal pinning and tendon transfers to prevent recurrence

Clinical Pearl

Centralisation = carpal slot + alignment with index metacarpal + tendon transfer

Radialisation = no carpal resection + alignment with middle metacarpal + tendon transfer

Pre-operative Distraction

Centralization procedure, without pre-operative distraction, requires the use of local flaps, extensive soft tissue release, extensive resection of the carpus, frequent ulna osteotomy and pressure on the ulna epiphysis from tension in the soft tissues, to position the wrist on the ulna, thus resulting in immediate shortening of the forearm length and compromise of the future growth potential. Soft tissue distraction may allow radialisation rather than centralization, an easier approach, less or no carpal resection, reduced pressure on the epiphysis and less need for ulna osteotomies – overall, it makes the procedure easier for the surgeon and may help preserve ulnar longitudinal growth.

A soft tissue distraction device (uniaxial fixator with a multiaxial joint or multiaxial frame) can be attached to the skeleton once the child has reached an adequate size to fit one in place (Fig. 6.9). A multi-axial frame, allows for a more precise control of 3 dimensional distraction, but is more cumbersome and rarely required in primary cases [35]. The authors use a mini Pennig™ fixator positioned on the radial side of the wrist, with two threaded pins inserted into the ulna and two into the index and middle finger metacarpals. Depending on the tightness of the wrist, much of the final distraction in the radio-ulnar plane can be achieved at the time of fixator application, but there is often residual overlap of the carpus and the ulna. The parents are taught pin site care and distraction before discharge

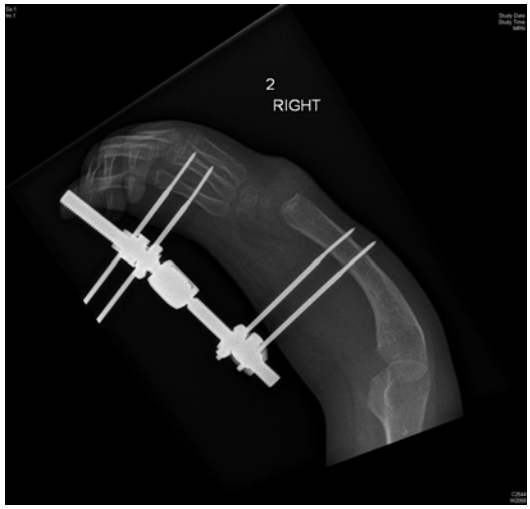

Fig. 6.9 Radial club hand – application of fixator to radial club hand

with distraction started after 48 h at the rate of 1 mm per day. Radiographs are obtained initially on a weekly basis, followed by fortnightly until a satisfactory position has been obtained, with a gap between the carpus and the ulna on radiographs (Fig. 6.10). Then the fixator is left in place for a minimum of 4 weeks to allow soft tissue recovery before definitive treatment is carried out.

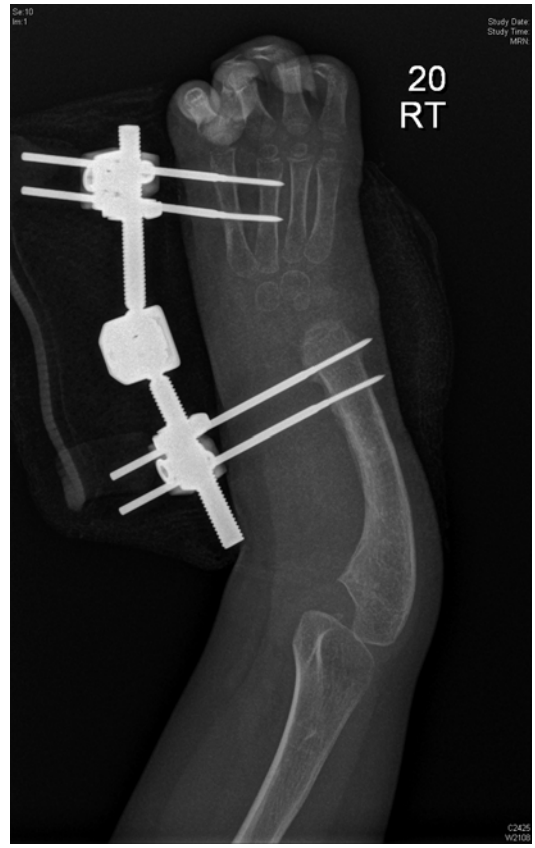

Fig. 6.10 Radial club hand – undergoing distraction with fixator in situ

Clinical Pearl

Remember that the median nerve is radially situated when applying fixator

Fixators applied to radial side will have less problems with pullout of pins

Leave threaded pins of fixator palpable from ulnar side in case of breakage to ease removal

Centralisation or Radialisation

Following satisfactory pre-operative distraction, the wrist is approached via a longitudinal incision, the use of local flaps being unnecessary after distraction. The decision is made at surgery whether to perform centralization or radialisation, depending on the quality of the dorsoradial muscle mass available. Whichever is performed, it is important for success, that an adequate release of the tight radial structures and the deep fascia is performed from mid volar to mid dorsal point and that the muscles that comprise the dorsoradial muscle mass (ECRL, ECRB, PT, BR, FCR) are transferred to pull ulnarly to the base of the 3rd metacarpal or to the extensor carpi ulnaris. If, at exploration, the dorso-radial muscle mass is found to be inadequate, then the surgeon should proceed to centralization.

The classic centralization procedure has become synonymous with the method described by Lamb [36] where a carpal slot is resected to accommodate the distal ulna, opposite the third metacarpal. For maximal stability, the length of the carpal slot should be identical to the width of the ulna [24]. The wrist is then stabilized with a longitudinal K wire that is left in-situ for at least 6 months (Fig. 6.11a, b). Even in the pre-distraction

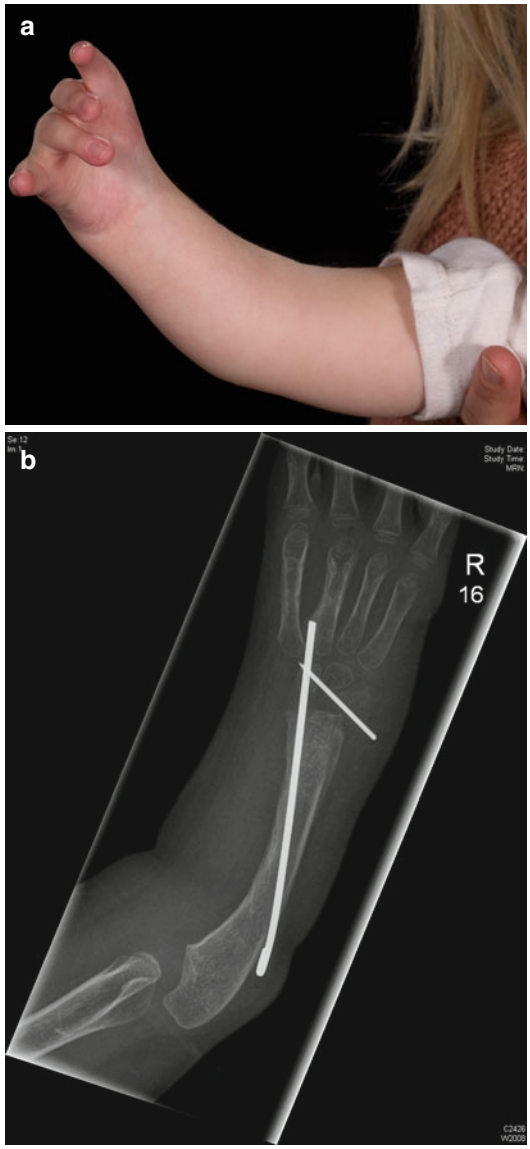

Fig. 6.11 (**a**, **b**) Radial club hand – post centralisation of radial club hand

era, this method has shown good consistency and reliability in achieving a long-term stabilization without premature closure of the ulnar epiphysis [36]. With the positioning of the ulna within the carpal slot, a partial fusion may ensue with long-term stability but little movement at the wrist joint, although the carpopmetacarpal joints may produce some useful motion.

In order to try to preserve motion, in 1985, Buck-Gramcko [37] suggested wrist stabilization without carpal resection, termed "radialisation". Unlike centralization, where the third metacarpal is positioned in line with the distal ulna, radialisation "overcorrects" the deformity by aligning the second metacarpal over the ulna, avoids any carpal resection and allows the temporary pin to be applied with the wrist extended. This also ensures that the dorsoradial muscle mass, which is transferred to the extensor carpi ulnaris (ECU) tendon, will exert a more ulnar pull. Radialisation will seem a more favourable option with its ability to more reliably preserve wrist motion and to "overcorrect" the deformity, but in our experience, approximately half those judged to be suitable will recur early. The reasons for this are multi-factorial, but fundamentally, are because it is essentially a balancing procedure and prone to destabilization due to uneven joint surfaces and poor muscle quality. In revision cases, where there is already some degree of spontaneous fusion, it may not be possible to perform radialisation. Our preference is to perform radialisation where the dorsoradial muscle mass is of good quality but we consider centralization as a good alternative and a salvage procedure when it is inadequate.

Clinical Pearl

K wire breakage is common so ensure both ends are accessible for removal

Recurrence of radial deviation and flexion remains a problem which increases in degree with time of follow up. In view of this others have sought alternative approaches.

Vilkki and colleagues [31, 38] pioneered a free metatarsophalangeal (MTP) joint to the radial side of the ulna, after soft tissue distraction. This abuts the carpus to create a more stable "Y" platform for the carpus, which will grow together with the distal ulna. Wrist motion is theoretically preserved and both joints can flex and extend as one. His long term outcomes show better wrist motion but entirely in the flexion range and a tendency for radial recurrence with time, necessitating distal transfer of the vascularized MTP joint with growth in several cases. These promising

results seem to suggest that this might be a step in the right direction but not the complete answer.

Others have decided that a stiff wrist is best and are either using serial pinning through the epiphysis with recurrent exchange of pins as the child grows or an early release of radial tight structures followed by wrist fusion if desired at skeletal maturity. This dichotomy of philosophies demonstrates the difficulty in deciding the best management for these children, with a successful outcome only being recognized 16 years later at the end of a surgeon's career and few surgeons having a large enough series to adequately assess even mid term results.

Thumb Hypoplasia

Radial longitudinal deficiency encompasses a spectrum that spans from cases so deficient in forearm length that they appear phocomelic to those with almost no difference in forearm length and only minor thumb deficiencies. All radial club hand patients have some degree of thumb hypoplasia and all patients with thumb hypoplasia have some measureable deficiency in the radial carpal bones, if not in the radius itself. Thumb hypoplasia can also be seen with other conditions including Poland's syndrome, ulnar club hand, constriction ring syndrome and thumb duplications. However, the common classification for thumb hypoplasia, the Blauth classification [39] with modifications by Manske [40], and Buck-Gramcko [41] (Table 6.4), takes no account of carpal or radial deficiencies and relates to those seen in radial deficiencies. This classification does directly relate to the treatment required, with Grade I, II and IIIA being appropriate for reconstruction whilst Grades IIIB and above are, within the Western world, considered too deficient for satisfactory reconstruction and planned for index finger pollicisation instead.

Grade I thumbs rarely need any treatment and the abnormalities in the thumb may not be noticed for many years. Grade II thumbs need first web release, opponensplasty and stabilisation of the first metacarpophalangeal (MCP) joint (Fig. 6.12). The techniques employed for all of these procedures are largely a matter of personal choice and experience. Stabilisation of the MCP joint may be for uni-axial or multi-axial instability (Fig. 6.13). The former is usually addressed by some form of tendon transfer, frequently a flexor

Table 6.4 Buck-Gramcko modification of Blauth classification of thumb hypoplasia [41]

Blauth grade	Soft tissue anomalies	Skeletal anomalies
I	Minor hypoplasia of entire thumb, APB/OP hypoplasia	Normal
II	Adduction contracture of 1st web, median innervated intrinsic muscles absent	Lax ulnar collateral ligament of MCP joint, slim intact skeleton
III	All of above + extrinsic muscle anomalies, absent intrinsic muscles	Partial aplasia of metacarpal base, absent CMC joint, unstable MCP joint
IV	Floating thumb, no muscles	Absent metacarpal
V	Absent thumb	Nil

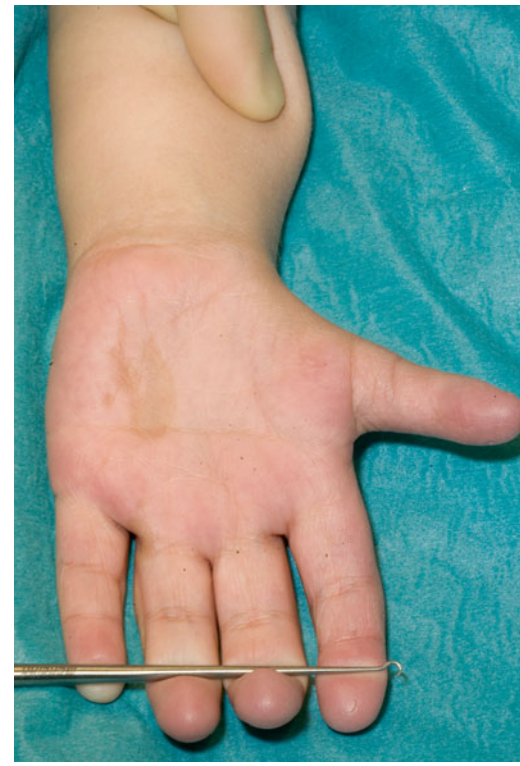

Fig. 6.12 Grade II thumb hypoplasia

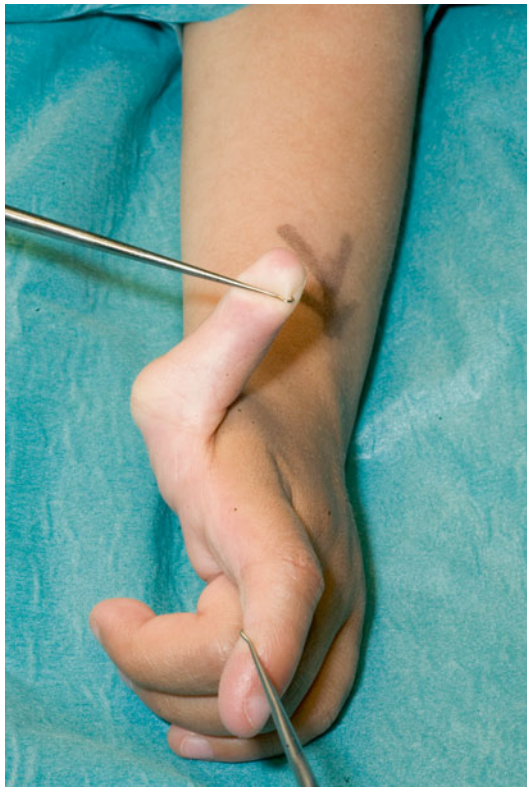

Fig. 6.13 Grade II thumb hypoplasia with multiaxial instability of the MCP joint

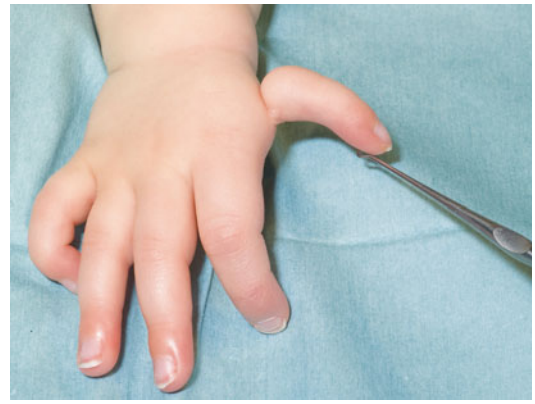

Fig. 6.14 Grade IV thumb hypoplasia

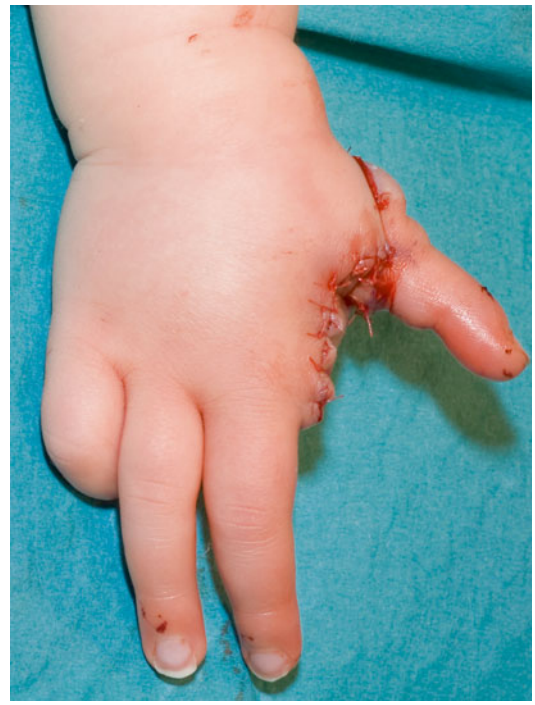

Fig. 6.15 Index finger pollicisation

digitorum superficialis tendon used for opponensplasty, passed through the bone to reconstruct the ulnar collateral ligament. The latter is probably best served by an epiphyseal sparing chondrodesis since other techniques are unlikely to provide sufficient stability. However, these thumbs, once chondrodesed, may later reveal themselves to be unstable at the 1st CMC joint and therefore would have been better, grouped together with Grade IIIB and IV thumbs (Fig. 6.14), for an index finger pollicisation.

Index finger pollicisation is an elegant procedure designed to convert an index finger into an opposable digit of an appropriate length and in a suitable position to substitute for a thumb. The procedure involves design of appropriate skin incisions, dissection of the ulnar digital artery to the index finger which will remain its principal supply, splitting of the common digital nerve, skeletal shortening with destruction of the metacarpal epiphysis, repositioning with hyperextension of the metacarpophalangeal head and rotation of the digit, rebalancing of the adductor and abductor, shortening of the extensor tendons and redraping of the skin without compression of the venous return (Fig. 6.15).

The results of index finger pollicisation are dependent on associated pathologies, so that those with radial club hand do less well, and the flexibility and musculature of the index finger.

Unfortunately, even with the best of results, its strength rarely exceeds 50 % of that of a normal thumb. However, when the parents agree to proceed, this is still superior to reconstruction of a Grade IIIB or greater thumb and to reconstruction of some Grade IIIA thumbs.

Clinical Pearl
The result of pollicisation is dependent on the quality of the index finger and of the surgeon

Central Ray Deficiency

Commonly referred to as the "cleft hand", Flatt's [42] comment that the deformity is a "functional triumph and a social disaster" astutely sums up the surgeon's dilemma with this condition. Although traditionally grouped under the IFSSH classification of 'failure of formation', the cleft hand differs from the other longitudinal deficiencies in that, although carpal bones may be affected [43], the deformities seldom involve more proximal structures and are usually confined to the hand plate (Fig. 6.16). The cleft hand includes a spectrum of abnormalities which aren't true failures of formation such as central syndactyly and radial polydactyly. Hence Ogino suggested that this group should be considered under the title "abnormal induction of rays" [44].

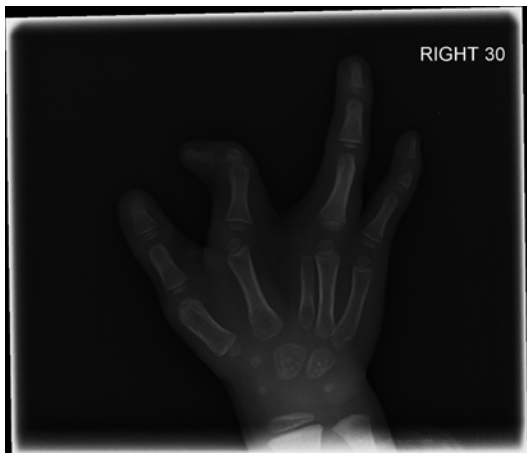

Fig. 6.16 Cleft hand with hypoplastic 3rd metacarpal

Although an isolated cleft hand is usually a sporadic deformity, the more common entity is as split-hand and split-foot (SHSF) that displays an autosomal dominant pattern with variable penetrance, with up to 70 % of subjects exhibiting some deformity [45, 46]. Genetic studies into the aetiology of SFSF deformities have identified defects in genes responsible for formation of the apical ectodermal ridge [3, 47]. Maisel's centripetal theory attempts to explain more precisely the location of the AER defects, suggesting that the central and radial portions are affected, which could explain for the tetratologic sequence and morphology of the cleft hand [48]. Typically, the deformity starts in the central portion and then progresses in a radial direction. Even in the most severe monodactylous cleft hand, the little finger is still preserved. However, this theory remains insufficient to account for some of the clinical associations, such as syndactyly and polydactyly. Ogino's experiments with teratogens in rats [44, 49], together with Blauth and Falliner's work [50] offers a more comprehensive explanation; interruptions of molecular signaling pathways and controllers of apoptosis lead to focal defects in the AER with either lack of differentiation (syndactyly) or exaggerated differentiation (polydactyly) or defect formation (clefting). In fact, Blauth and Falliner [50] suggested that apart from aplasia of the bones and soft tissue, synostosis accounts for the origin of clefting in at least 40 % of their cases. Suffice to say, our understanding of the aetiology of the cleft hand remains rudimentary, but emphasis must be placed on the osseous syndactylies and the central polydactylies and their close associations with the cleft hand. There is a potential argument therefore, that clefting should probably lie within failure of differentiation rather than failure of formation.

Clinical Presentation

A wide spectrum of presentations confront the surgeon dealing with the cleft hand, ranging from a minor soft tissue cleft, to a deep cleft with complete absence of the metacarpal, to the most severe form, where the little finger is the only ray preserved. In addition to the missing digits, various metacarpal and proximal phalangeal

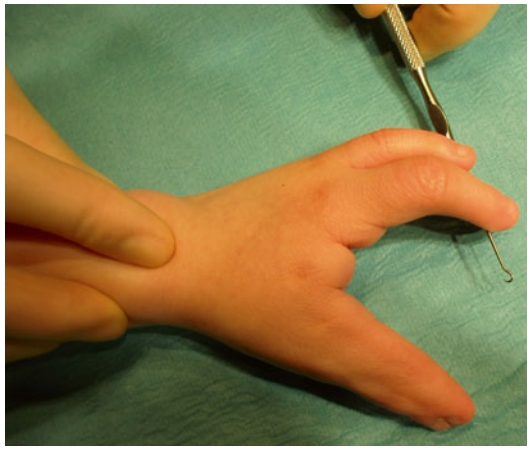

Fig. 6.17 Cleft hand

Table 6.5 Classification of cleft hand according to 1st web space [51]

Type	Description	Features
I	Normal web	
IIA	Mildly narrowed	Mild narrowing of first web
IIB	Severely narrowed	Severe narrowing of 1st web
III	Syndactylysed web	1st web complete syndactyly
IV	Merged web	Index ray deficient, 1st web merged with cleft
V	Absent web	Only ulnar rays present

variations are seen in the cleft, which may be missing, bifid and supporting two digits, or lying transversely. The distal part of the latter may be in communication with the adjacent metacarpophalangeal joint or proximal interphalangeal joint. Syndactyly and radial polyactyly may also be present (Fig. 6.17). The digits adjacent to the cleft display anomalies: they may be broader, have syndactyly, camptodactyly, and angulatory deformities. The first web space is always affected – leading to the classification of cleft hand as described by Manske (Table 6.5) [51].

Recognition of clefting associated with EEC or ectrodactyly-ectodermal dysplasia-clefting syndrome allows the detection and repair of a submucous cleft to prevent future speech and language deformities. Other associated syndromes are shown in Table 6.6.

Table 6.6 Syndromes associated with cleft hands

Ectrodactyly-ectodermal dysplasia – cleft syndrome
Split Hand foot malformation syndrome
Silver-Russell syndrome
Focal dermal hypoplasia
Cornelia de Lange syndrome
Ectrodactyly and cleft palate syndrome
Ectrodactyly/mandibular dysostosis

Management

Most commonly, this condition is familial, the parents are aware that the deformity is extremely functional and there may be reluctance in pursuing surgical solutions. If surgery is considered, the risk of losing some degree of function to improve aesthetics must be discussed. Where a parent is affected and they wish to pursue early surgery aggressively, there is a concern that this may relate to their guilt in having produced an abnormal child and their expectations may be unrealistic. Similarly, for older children who have requested surgery to improve aesthetics but who have functionally adapted successfully, it is important to approach the consultation with caution to curb unrealistic expectations. The functional indications for surgery are as follows:

Strong Indications

1. Excision of transversely lying bones – these cause the cleft to widen with growth and may affect adversely function and worsen aesthetics. In cleft feet, the progressive deformity causes major problems with obtaining footwear. In cases where the bone forms a common joint with the adjacent finger, reconstruction of the joint structures, including the collateral ligaments, must be undertaken if the whole bone is removed or partial removal may avoid the risk of joint instability.
2. Release of border syndactylies, or syndactylies in digits of unequal length. The thumb – index finger syndactyly release is especially important if the central cleft is to be closed but beware the presence of shared tendons and blood supply which may preclude this separation.
3. Widening of first web space – a natural progression from syndactyly release of the first

web space. The procedure to achieve a sufficient first web varies enormously with the degree of the deformity and the tightness of the web.

These three indications can often be combined within a single procedure, together with closure of the cleft, transposition of the index finger and recruitment of the cleft commissure to reconstruct a first web. The most well-known operation to achieve this is the Snow-Littler procedure, which involves closing the cleft, removal of any transverse lying bone, osteotomy to the index metacarpal and transposing a palmar-based flap (from the cleft) to reconstruct the first web. This is a technically demanding procedure and there is a significant risk of distal flap necrosis, as the length of this random pattern flap exceeds its breadth. Different authors have modified the technique to make it more reliable, including using a dorsal flap as described by Miura and Komada [52], or a combined dorsal and palmar flap as described by Upton [53]. The underlying principles are the same: excess skin from the cleft is used to augment the first web space, and the interval between the central digits is closed and reconstructed. The first dorsal interosseous and adductor pollicis may need extensive subperiosteal release to gain an adequate first web space.

Following removal of any transverse bones, a new transverse metacarpal ligament is reconstructed, either using redundant cleft-based tendons, adjacent A1 pulleys or both, taking care to avoid creating a rotational deformity with scissoring in flexion. If there is an absent or very short third metacarpal, an osteotomy is not required and a formal index finger transposition unnecessary.

Other Relative Indications or Additional Procedures

1. Correction of camptodactyly – this should be approached with caution as the position of the angulation may be beneficial for hand function, especially in more severe cleft hands with only border digits. It is, however, still worthwhile to examine tendon integrity during closure of the cleft to release any aberrant insertions to prevent progression of deformity.
2. Correction of syndactyly between digits of similar length – this is still a worthwhile procedure for both functional and aesthetic improvements. Skin grafts for the syndactyly release can be obtained from excess skin following cleft closure or from more traditional skin donor sites. In the presence of proximal metacarpal synostosis, release of complex syndactyly needs to be approached with caution, as there is frequently a shared flexor tendon and pulleys. Parents need to be advised that a full range of motion may not be possible in the released digits.
3. Toe-to-hand procedures – This may be helpful to improve prehension. In the monodactylous hand, where the little finger is often well preserved, it may be worth considering an ulnar pollicisation by stabilization, rotation and an opponenplasty procedure. An immobile radial post can then be reconstructed to permit opposition and tip-to-tip prehension [54]. This is especially in the case where the presence of proximal motors to power any subsequent toe transfers may be uncertain.
4. Rotational deformities – rotational osteotomy procedures to improve the orientation of the thumb can be considered to gain a better prehension pattern but is rarely needed if adequate first web release has been performed.

Clinical Pearl

Beware creating a rotational deformity with closure of the cleft hand

Cleft feet are easier to correct when younger and frequently cause problems with footwear

Ulnar Longitudinal Deficiency

Ulnar longitudinal deficiencies, termed “ulnar club hand”, are rare, up to ten times less common than radial longitudinal deficiencies. On the internet, the two conditions are often confused and parents come to the clinic severely misinformed. The clinical presentations of ulnar deficiencies are much more varied than that of the

Fig. 6.18 Bilateral ulna club hands

radial deficiencies. Abnormalities can affect any part of the upper limb with the severity of hand deformities not necessarily correlating with the proximal deformities (Fig. 6.18). The incidence is around 1 in 100,000 live births, although some have reported a much higher incidence [55].

In utero ulnar deficiencies are thought to occur earlier in gestation than radial deficiencies. The zone of polarizing activity (ZPA) is responsible for development and differentiation along the antero-posterior or radial-ulnar axis, via its action on the sonic hedgehog gene (SHH) [55]. This 'ulnarising' process is controlled by a feedback loop that maintains SHH expression at the ulnar side of the AER during limb growth [5]. Loss of SHH function therefore leads to both longitudinal and ulnar-associated deficiencies as a combined entity, with the wide spectrum of presentations a reflection of variation in the timing, degree, and duration of SHH disruption [56]. Furthermore, the loss of SHH can reduce FGF expression through interruption of the SHH–FGF loop and interfere with formation of radial structures, resulting in a surprisingly high number of concomitant thumb deficiencies [5, 57].

Most ulnar deficiencies are sporadic, but there are a few syndromic associations, mostly in an autosomal-dominant pattern (Table 6.7).

Table 6.7 Syndromes associated with ulnar dysplasia

Syndrome	Associated anomalies
Cornelia de lange	Microcephaly, cleft palate, cardiac defects, serve developmental delay
Schinzel's syndrome	VSD, pyloric stenosis, anal stenosis, reduced sweating
Weyer's unlar ray oligodactyly syndrome	Midline craniofacial anomalies, fibular defects, renal and splenic abnormalities
Ulnar mammary syndrome	Hypoplasia of breasts, nipples and apocrine glans, abnormal teeth and genitalia
Femoral-fibural-ulnar deficiency syndrome	Short stature, fibula hypoplasia, talipes equinovarus
Ulnar fibula dysplasia	Short stature, fibula hypoplasia, mandibular hypoplasia
Klippel-Feil syndrome	Short webbed neck, cervical vertebral abnormalities

Table 6.8 Classifications of ulnar dysplasia

Bayne and Klug		Paley and Herzenberg	
Type	Ulnar deficiency	Type	Ulnar deficiency
I	Ulnar hypoplasia	I	Ulnar hypoplasia with epiphysis intact
II	Partial ulnar aplasia	II	Partial ulnar aplasia – absent distal 1/3
III	Total ulnar aplasia	III	Partial ulnar aplasia – absent distal 2/3
IV	Radiohumeral synostosis	IV	Total ulnar aplasia
		V	Radiohumeral synostosis

Clinical Presentation

Despite the wide spectrum of presentation, classifications for ulnar deficiencies [58, 59, 62] are based on the degree of skeletal absence of the ulna, which is rarely the indication for treatment (Table 6.8). Havenhill et al described a type O ulnar longitudinal deficiency where the forearm bones were normal but there were deficiencies in the carpus and hand [60]. In the presence of radio-humeral synostosis, the ulna is totally absent [61]. Treatment is largely directed at the hand so Cole and Manske devised a classification based on the radial side of the hand, but this fails to address proximal deformity [57]. Perhaps a combination of different classification methods should be used, or one that is individualized to the patient and based more on function, as suggested by Al-Qattan [61].

Clinical Pearl

Radial and ulnar deficiencies may be present in the hand

Ulnar deviation of the wrist is rarely severe

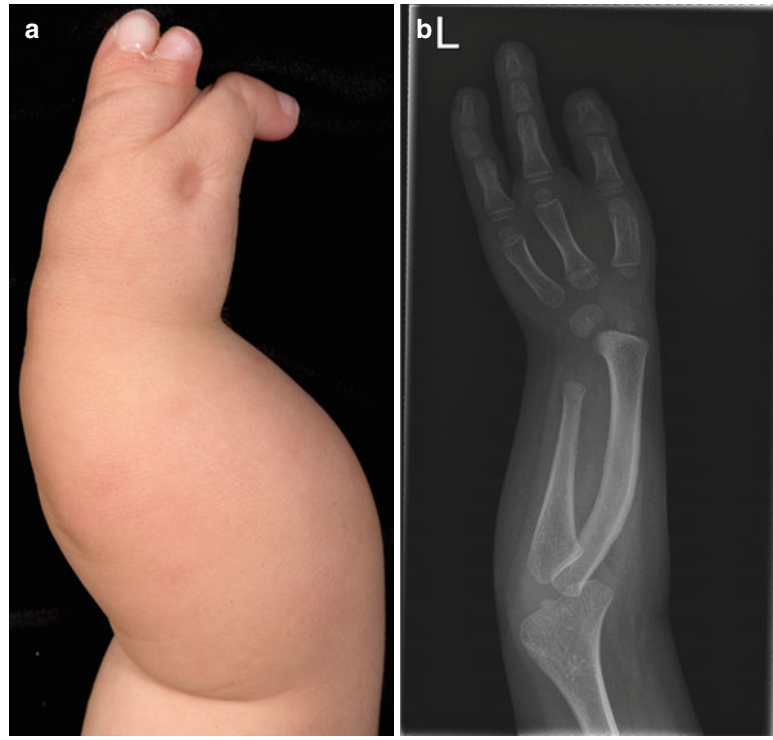

Fig. 6.19 (**a**, **b**) Ulnar club hand – type II

Treatment

The primary aims of treatment are to firstly position the hand in space, and subsequently to improve prehension as visualized on the Great Ormond Street ladder. Many procedures have been proposed for correction of the ulnar club hand, mainly anecdotally or in small series. The only surgery shown to be reliable is to the humerus and hand.

Shoulder

The upper limb may be positioned in such a way that, at rest, the hand faces backwards, the dorsum resting on the buttocks. Shoulder mobility is usually unrestricted and compensates significantly for the lack of elbow flexion. Humeral rotation osteotomies, either above, or more frequently, directly below the deltoid insertion, give great benefit both functionally and cosmetically in allowing the hand to lie in front of the body so objects held in it are visible.

Elbow

The elbow is the most severely affected joint in ulnar dysplasia and its treatment remains a challenging enigma. In most cases of ulna hypoplasia, the articular surfaces are congruent but the coronoid process is deficient. In more severe hypoplasia, there is deficiency of all articular parts (Fig. 6.19a, b). There may be a flexion deformity of the elbow with a pterygium. The radial head may be dislocated with the capitellum absent or flattened. In mild cases, the elbow functions reasonably well, but as the child reaches teenage years, a lump may appear on the radial aspect of the elbow. This is the radial head which has dislocated with growth. Excision before skeletal maturity is ill-advised but, if painful, may be considered in adulthood, although the effect on the wrist has not been fully established. Radial head replacement in adulthood is constrained by the abnormal capitellum and soft tissues surrounding it.

Finally, in the most severe form, the radius and humerus are fused with the forearm fixed in a pronated position, either in full extension or in marked flexion (>60° fixed flexion).

Soft tissue and bony releases at a young age to gain more range of motion have been disappointing with good results on table but gradual

recurrence. Relocating the radial head dislocation is possible but maintenance of it in joint is prone to failure, since the articular surfaces and soft tissues are deficient. Distraction lengthening of the ulna, with or without radial shortening provides the potential to provide a more stable platform for the radial head, but can disrupt the relatively good wrist function and needs to be repeated several times during childhood.

The creation of a one-bone forearm, although technically difficult, remains a valid option and may be combined with lengthening. The fibrous tissues between the two forearms are divided. Lengthening of either the ulna or shortening of the radius may be considered at this stage until the radial head is in a better position. The two bones are fused by osteosynthesis, forming the one bone forearm. This can then be lengthened again at a later stage, if necessary.

Forearm and Wrist

The forearm is short and may be bowed. A fibrocartilaginous remnant or anlage may connect the distal end of the ulna to the carpus especially in the Bayne type III ulnar deficiencies and was believed to produce increasing bowing with growth. The proponents of early excision of the anlage claimed that this prevented progressive deformities, but no evidence has supported this. If performing the excision, it is only necessary to excise the distal third of the anlage. Correction of the radial bow by osteotomy with or without ulna lengthening is worthwhile to improve the aesthetic appearance and may be done through the ulnar incision used for excision of the anlage (Fig. 6.20).

The ulnar deviation present at the wrist is typically not severe and rarely alters with growth. The indication for its surgical correction with tendon transfer is uncertain.

Hand

Most surgical procedures for ulnar deficiencies are focused on the hand. Unlike in radial longitudinal deficiencies, both preaxial and postaxial abnormalities are likely to occur. Ninety percent of hands have missing digits (ectrodactyly), 30 % have syndactyly and 70 % have

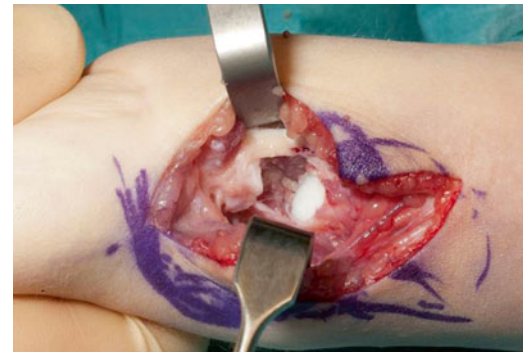

Fig. 6.20 Ulna club hand – Excision of anlage and radial osteotomy

with some form of thumb and first web deficiency [50, 53]. In his personal series of 164 patients, Buck-Gramcko found that only 8.5 % of patients presented with all five digits and the majority (43.5 %) presented with a three-fingered hand. The digits can be abnormal, with hypoplastic, missing or synostosed metacarpals, longitudinal bracketed epiphyses or delta phalanges, complex syndactylies and poorly formed joints with symphalangism or camptodactyly. If present, the little finger is always abnormal. In the most severe forms, the only movement possible is at the carpometacarpophlanageal (CMC) joints.

Surgery to the hand presents with the greatest potential for improving function and requires a return to first principles to gain a stable radial digit which can oppose to other digit(s) with an intervening adequate gap. First web space deepening may be achieved using local rotational flaps and skin grafts but, in the more severe cases, a free flap may be required. A tight complex syndactyly, present in up to one third of cases, may benefit from pre-operative soft tissue distraction to provide enough skin cover and to obtain more aesthetically pleasing fingers. In a two-digit hand, metacarpal osteotomy may need to be performed in both digits to obtain an adequate pulp-to-pulp pinch (Fig. 6.21). When separating metacarpal synostoses, it is important to fully divide the bones down to the CMC joint and provide some soft tissue interposition as this may be the only mobile joint.

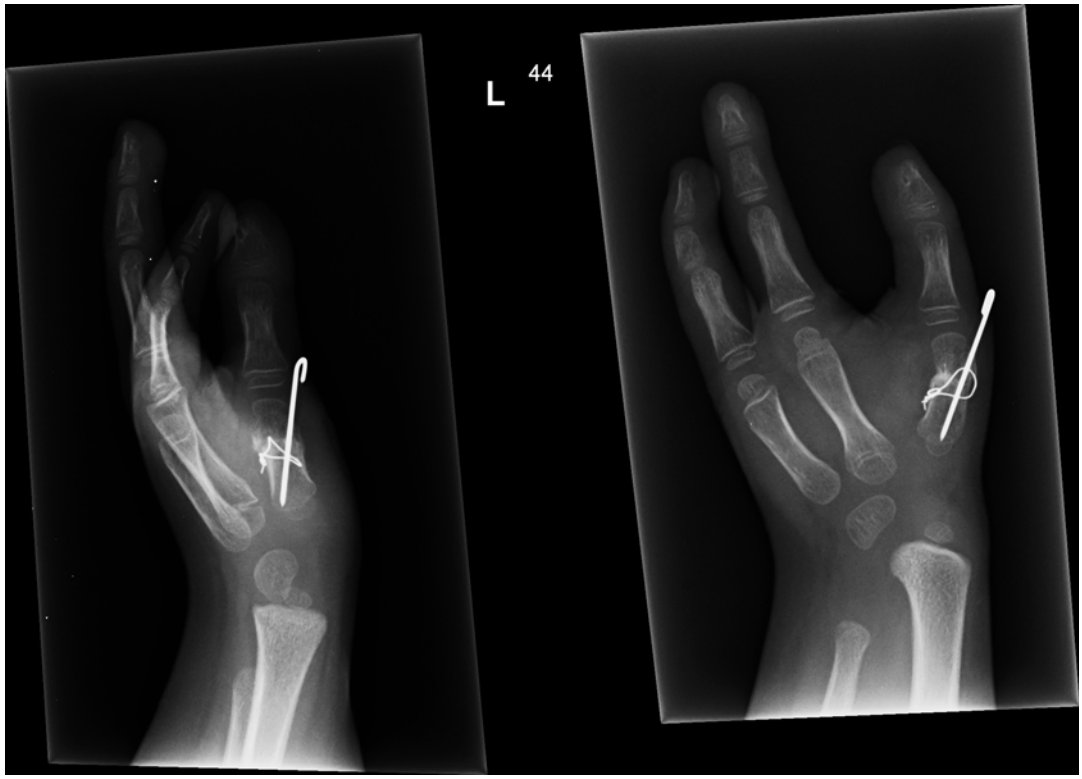

Fig. 6.21 Ulna club hand – post metacarpal rotation osteotomy

Free toe transfer is an attractive potential option for increasing the number of digits, but there are rarely sufficient motors to power the transfer and the arterial anatomical variations need to be considered. With dysplasia of the ulna, the radial artery may be absent in up to 16 % of cases, affecting the arrangement of the arches and digital arteries. In the absence of a thumb, pollicisation of a radial digit, assuming the digit is of good quality, will significantly improve function, but the parents are often reluctant to agree to this since the child already has a reduced number of digits. Here, the otherwise discarded metacarpal shaft may be used to lengthen another deficient ulnar digit or separate a metacarpal synostosis.

Clinical Pearl

Procedures dealing with the hand anomalies are those most likely to improve function

Conclusion and Future Directions

The embryology, classification, and management of hand anomalies listed under the IFSSH classification of "failure of formation" continues to intrigue and challenge the surgeon. With increasing knowledge regarding the molecular basis of limb patterning, the whole classification will soon be changed to differentiate between malformations and disruptions and this is likely to result in different grouping of these conditions. A greater understanding in the future may create the foundation for gene therapy and tissue engineering to correct these deformities.

War has always advanced medicine, and conflict in the Middle East and Afghanistan has produced many young multiple limb amputees. Upper limb prosthetic and orthotic devices that have previously heavy, cumbersome and difficult to wear and control are rapidly improving with this sudden increased demand.

Progress will occur with refinement and improvement in microsurgical transfers and distraction devices. However, long-term optimism should be tempered by the experience that many techniques have not stood the test of time, giving satisfactory short-term outcomes but withering under the essential goal of maintenance of the result until skeletal maturity. The paediatric hand surgeon should strive to seek out new solutions and to critically evaluate them over the long term. For those whose paediatric practice is limited, they should rely on the limited armamentarium of procedures that have been well-tested to provide limited but reasonable improvement of function and aesthetics.

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Failure of Differentiation of Parts

7

Scott H. Kozin

Keywords
Failure of differentiation • Syndactyly • Synostosis • Camptodactyly • Clinodactyly

Introduction

The most widely accepted classification of congenital limb anomalies was pioneered by Frantz and O'Rahilly [1] and published by Swanson [2]. This work eliminated much of the confusing Greek and Latin terminology and was accepted by the American Society for Surgery of the Hand, International Federation of Societies for Surgery of the Hand, and International Society for Prosthetics and Orthotics. This system is based on embryonic failure during development and relies upon clinical diagnosis for categorization. Each limb malformation is classified according to the most predominant anomaly and placed into one of seven categories (Table 7.1). Different clinical presentations of similar categories of embryonic failures are explained by varying degrees of damage within the organization of the limb mesenchyme [2, 3].

This chapter will focus on the Group II category defined as failure of differentiation or separation of parts. This terminology implies development of all the essential components, but failure of arrangement into a proper finalized form. This failure of differentiation can affect skeletal, dermal, fascial, muscular, ligaments, and/or neurovascular components of the limb. This category includes a heterogenous group of disorders that are further subdivided according to anatomical abnormalities.

Syndactyly

Introduction

Syndactyly is the most common type of a group II anomaly. Syndactyly is characterized by an abnormal connection between adjacent digits. Syndactyly is diagnosed when the distal margin of the inter-digital web extends beyond the midpoint of the proximal phalanx.

Web spaces are critical for unimpeded inter-digital abduction/adduction and digital

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Table 7.1 Embryologic classification of congenital anomalies

Classification	Subheading	Subgroup	Category
I. Failure of formation			
	A. Transverse arrest		
		1. Shoulder	
		2. Arm	
		3. Elbow	
		4. Forearm	
		5. Wrist	
		6. Carpal	
		7. Metacarpal	
		8. Phalanx	
	B. Longitudinal arrest		
		1. Radial deficiency	
		2. Ulnar deficiency	
		3. Central deficiency	
		4. Intersegmental	Phocomelia
II. Failure of differentiation			
	A. Soft tissue		
		1. Dissemenated	(a) Arthrogryposis
		2. Shoulder	
		3. Elbow & forearm	
		4. Wrist & hand	(a) Cutaneous syndactyly
			(b) Camptodactyly
			(c) Thumb-in-palm
			(d) Deviated/ deformed digits
	B. Skeletal		
		1. Shoulder	
		2. Elbow	Synostosis
		3. Forearm	(a) Proximal
			(b) Distal
		4. Wrist & hand	(a) Osseous syndactyly
			(b) Carpal bone synsostosis
			(c) Symphalangia
			(d) Clinodactyly
	C. Tumerous conditions		
		1. Hemangiotic	
		2. Lymphatic	
		3. Neurogenic	
		4. Connective tissue	
		5. Skeletal	
III. Duplication			
	A. Whole limb		
	B. Humeral		
	C. Radial		
	D. Ulnar		
		1. Mirror hand	
	E. Digit		
		1. Polydactyly	(a) Radial (preaxial)
			(b) Central
			(c) Ulnar (postaxial)

Table 7.1 (continued)

Classification	Subheading	Subgroup	Category
IV. Overgrowth			
	A. Whole limb		
	B. Partial limb		
	C. Digit		
		1. Macrodactylty	
V. Undergowth			
	A. Whole limb		
	B. Whole hand		
	C. Metacarpal		
	D. Digit		
		1. Brachysyndactyly	
		2. Brachydactyly	
VI. Constriction band syndrome			
VII. Generalized skeletal abnormalities			

flexion/extension. Normal inter-digital abduction between the thumb and index is approximately 70°. Between the fingers, inter-digital abduction is around 35° [4, 5].

The natural commissure is comprised of supple skin that gently slopes 45°–50° from dorsal and proximal to distal and palmar. The distal extent of the commissure is normally about one half the length of the proximal phalanx [3, 6]. The shape of the commissure varies slightly across the hand to accommodate varying function. The commissure is more rectangular in shape between the index/ long fingers and ring/ small fingers for greater inter-digital abduction. In contrast, the commissure is more V-shaped in configuration between the long and ring fingers to help stabilize the central rays.

Incidence/Etiology

Simple syndactyly occurs with an approximate incidence of 2–3 per 10,000 live births [3, 5–7]. Bilateral involvement occurs in approximately 50 % of patients. The long/ ring web space is most frequently involved, followed by the ring/ small web space. The thumb/ index web space is the least common and often associated with other congenital differences, such as central deficiency and Apert syndrome (Fig. 7.1).

Syndactyly often occurs sporadically without a familial history. In some cases (10–40 % of patients), syndactyly is inherited as an autosomal dominant trait with incomplete penetrance and variable expression [4, 7]. This indicates that the syndactyly may skip a generation and occur with varying extent (variable phenotype). The sequencing of the human genome has provided details about the chromosomal markers for syndactyly [7, 8]. The classic autosomal dominant syndactyly has been localized to a specific candidate region of a responsible gene (2q34-q36) [7]. Syndactyly combined with duplication of the ring finger, or synpolydactyly (Fig. 7.2), has been linked to a mutation of the HOX13 gene, which is also located on chromosome 2 (2q31) [8].

The etiology of syndactyly has been related to the apical ectodermal ridge (AER), which regulates proximal-to-distal limb axis configuration and secretes proteins that influence the development of the underlying tissues. The spaces between the fingers form in a distal-to-proximal direction to the level of the normal web [4, 9]. This process appears to be dependent on the presence of the AER and the molecular signaling of several cytokines [10]. Failure of the AER is the most common explanation for syndactyly.

Evaluation (Presentation, Investigation and Treatment Options)

The diagnosis of syndactyly is usually straightforward. A standard prenatal, birth, and familial

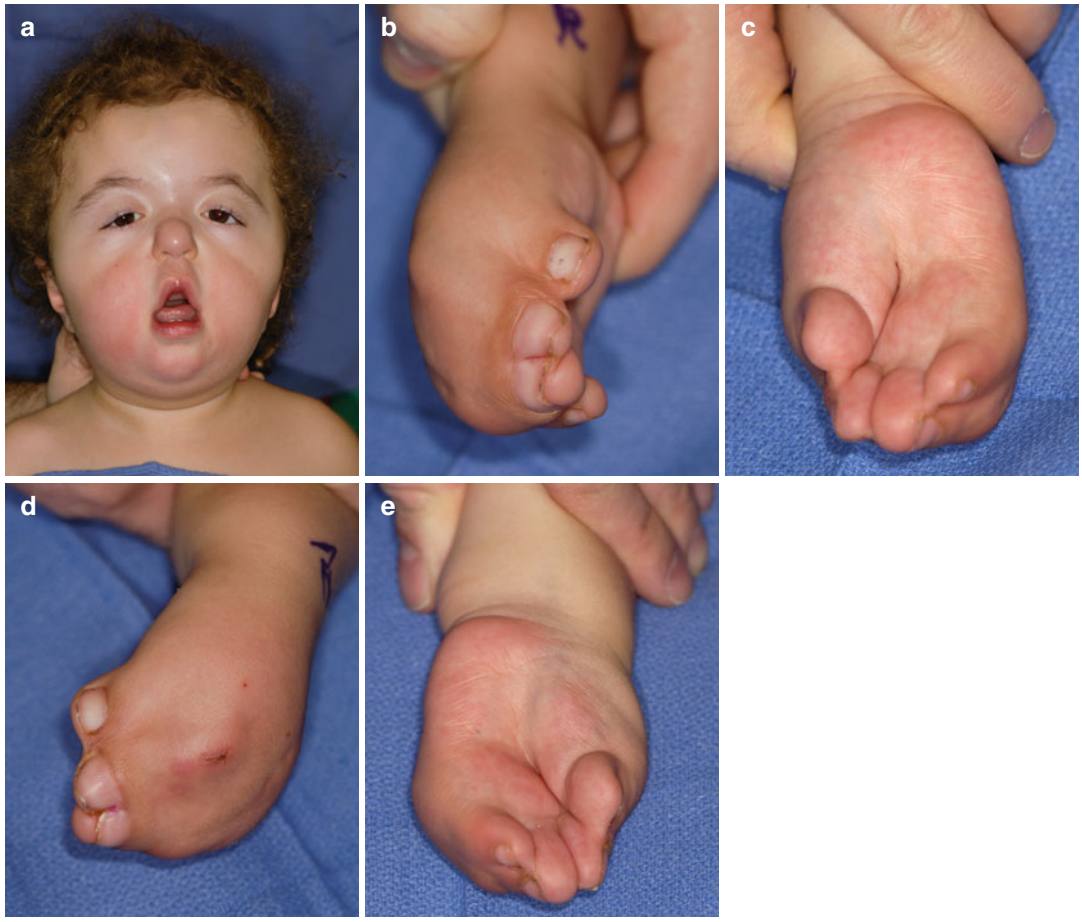

Fig. 7.1 2 year-old with Apert syndrome (Courtesy of Shriners Hospital for Children, Philadelphia). (**a**) Typical facies. (**b**) Dorsal view right hand with severe syndactyly including thumb/index web space. (**c**) Palmar view right hand with deep crevasses. (**d**) Dorsal view of left hand. (**e**) Palmar view of left hand

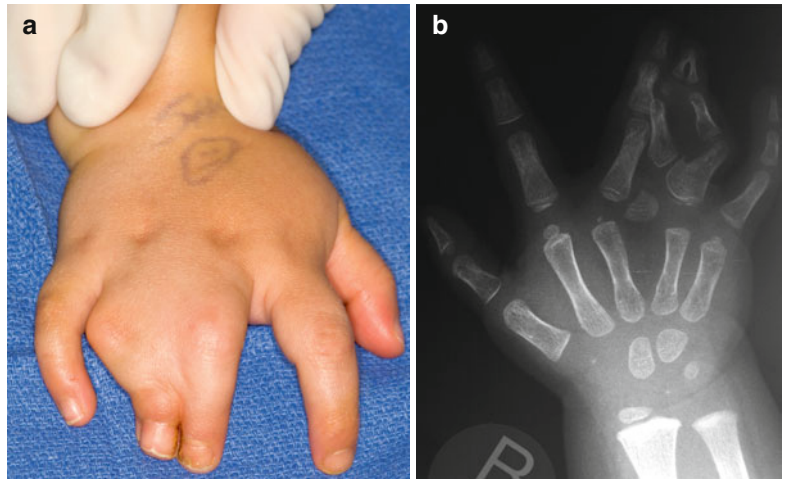

Fig. 7.2 2 year-old with syndactyly combined with duplication of the ring finger (synpolydactyly) (Courtesy of Shriners Hospital for Children, Philadelphia). (**a**) Dorsal view of hand with syndactyly. (**b**) X-ray reveals ring finger duplication with awkward interconnections

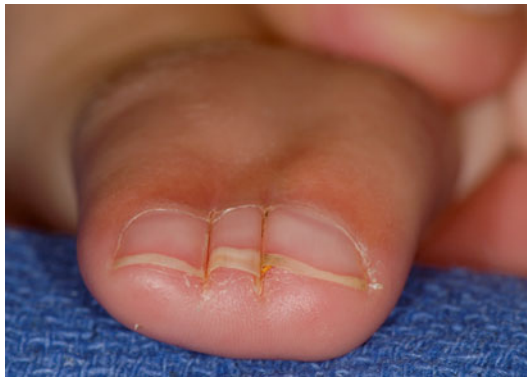

Fig. 7.3 3 year-old with ulnar deficiency and syndactyly. Distal phalangeal tufts are fused and a common fingernail or synonychia crosses the bony mass (Courtesy of Shriners Hospital for Children, Philadelphia)

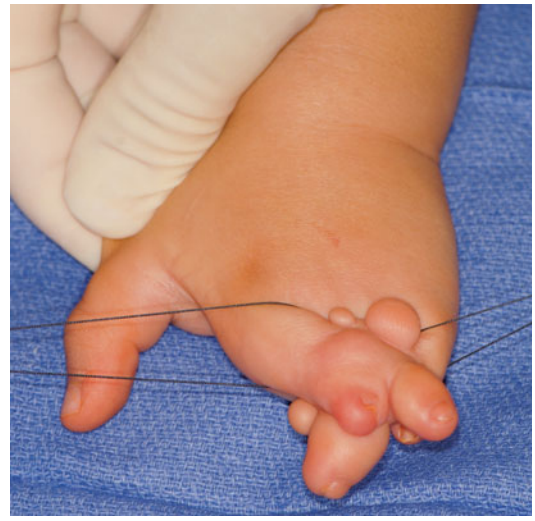

Fig. 7.4 3 year-old with amniotic band disruption sequence and complicated syndactyly (Courtesy of Shriners Hospital for Children, Philadelphia)

history are obtained. The physical examination should include observation and palpation of the affected hand and digits. In older children, the overall hand function is observed and the use of the interconnected digits is noted.

Simple syndactyly is a soft-tissue connection only and can be incomplete or complete. Complete syndactyly can result in a shared or common fingernail (synonychia). Complex syndactyly involves fusion of adjacent phalanges or interposition of accessory bones. When the distal phalangeal tufts are fused, the synonychia crosses the bony mass (Fig. 7.3). Atypical forms of syndactyly also exist. There are many atypical or complicated configurations of syndactyly that occur in conjunction with congenital syndromes, such as amniotic band disruption sequence (Fig. 7.4) [2, 4]. These complicated forms can cause bizarre soft-tissue interconnections, may contain a hodgepodge of abnormal bones, and often defy standard classification.

Passive and active motion between the connected digits is recorded. Lack of differential motion may indicate bony fusion. Palpation may also reveal an extra digit concealed within the conjoined digits (synpolydactyly).

Plain x-rays are routine to assess for skeletal fusion (complex syndactyly), to delineate a concealed extra digit (synpolydactyly), or to expose other skeletal deformities (Fig. 7.2b). Advanced imaging studies, such as MRI, are unnecessary.

Management

Nonsurgical Treatment

Nonsurgical treatment is recommended for mild incomplete syndactyly that does not interfere with function. Surgical intervention is also not recommended for adult patients that have accepted their deformity. Lastly, surgery is avoided if separation would lead to a loss of hand function. In certain circumstances, the connect digits rely on each other for stability and separation would result in unstable digits with less function.

Surgical Techniques

Simple syndactyly separation is a straightforward procedure as long as the surgeon abides by general principles listed below. In contrast, complex or polysyndactyly is far more challenging to treat, especially as the magnitude of bony union increases. Extensive complex or polysyndactyly

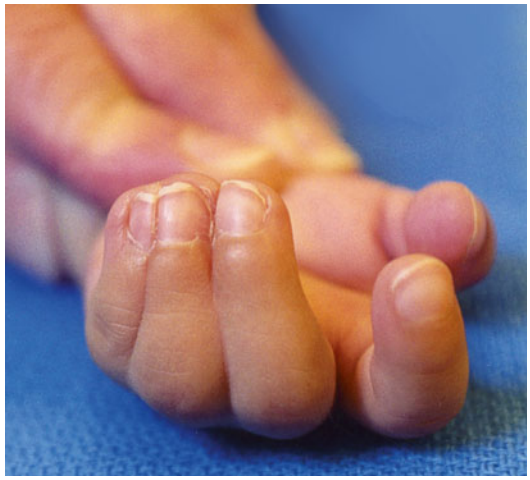

Fig. 7.5 Syndactyly between digits of unequal length leads to tethering of the longer digit, which results in a flexion contracture and rotational deformity (Courtesy of Shriners Hospitals for Children, Philadelphia)

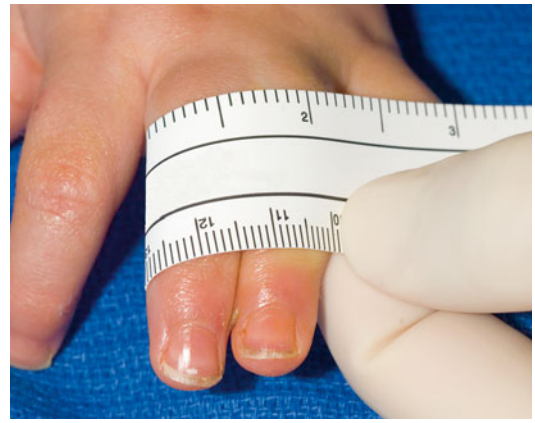

Fig. 7.6 Measuring the distance around the digits with a tape measure and comparing to the distance around individual digits exemplifies the skin deficiency in syndactyly (Courtesy of Shriners Hospitals for Children, Philadelphia)

is best treated by an experienced pediatric hand surgeon. The surgery is challenging and the complication rate is much higher compared to simple syndactyly separation.

Timing of Surgery

The timing of surgical release follows certain guidelines [4–6]. Digits of unequal length require early separation (3–6 months of age) to prevent deformity developing as the shorter digit tethers the longer digit from growing (Fig. 7.5). The principle applies especially to border digit syndactyly - thumb/ index and ring/ small syndactyly. Tethering can also occur in syndactyly associated with amniotic disruption sequence that causes digital truncation and connections between digits of unequal lengths.

Surgery for syndactyly of relatively equal length digits can be delayed (12–18 months of age). This delay allows the hand to be larger, which facilitates surgical reconstruction. Syndactyly release performed in patients older than 18 months has a lower incidence of complications and unsatisfactory results, such as web creep [11].

Extent of Surgery

Surgery should be performed only on one side of an affected digit at a time to avoid vascular compromise of the skin flaps or conjoined digits [3–6]. Therefore, complete separation of three or more connected adjacent digits requires staged surgical procedures usually separated by 3–6 months. Isolated release of the fingertips and distal phalangeal fusions of all digits can be performed during a single procedure to reduce the tethering effect.

Skin Grafting

Release of a complete syndactyly produces a skin deficiency that requires skin grafting. A skin graft is required because the circumference of two separated digits measured separately is 22 % greater than the circumference of two conjoined digits (Fig. 7.6) [4, 5]. Full-thickness skin grafts are preferred over split-thickness skin grafts to lessen graft contracture. There are multiple possible donor sites; I prefer the distal wrist crease, antecubital fossa, or medial arm. These sites have better skin match and are within the surgical field.

The amount of skin graft required is variable and lessened by defatting of the fingers [12].

Intense defatting should be avoided for fear of iatrogenic vascular injury and producing a thin finger following the involution of the infant fat [12]. Another factor that effects the amount of skin graft is the amount of coronal skin between the digits. Plentiful coronal skin lengthens the flaps and lessens the amount of skin graft. Another option to avoid skin grafting is to import skin from the dorsum of the hand and/or adjacent digits [13].

Incomplete syndactyly can be released and closed using local flaps without the need for supplemental skin grafts. A variety of flap options have been described including a simple Z-plasty, a four-flap Z-plasty, the double-opposing Z-plasty (butterfly flap), or other combinations [4, 14].

Commissure

Supple commissure skin is required to facilitate appropriate interdigital movement and digital flexion/ extension. Use of skin graft should be avoided. A local flap is designed to fill the commissure, preferably from the dorsum of the web.

Flap Design

There are a myriad of methods to design flaps for syndactyly reconstruction and most surgeons have their own individual preference [5, 6, 15]. I prefer a dorsal commissure flap that begins at the level of the metacarpal heads and extends two thirds the length of the proximal phalanx (Fig. 7.7). On the palmar surface, a rectangular flap is fashioned to resurface the proximal area of one of the digits adjacent to the commissure (Fig. 7.8). The proximal transverse incision for this rectangular flap denotes the level of commissure reconstruction, and the distal transverse edge equals the length of the dorsal commissure flap. Following design of the commissure flap, interdigitating zigzag dorsal and palmar flaps are constructed along the sides of the digits. The

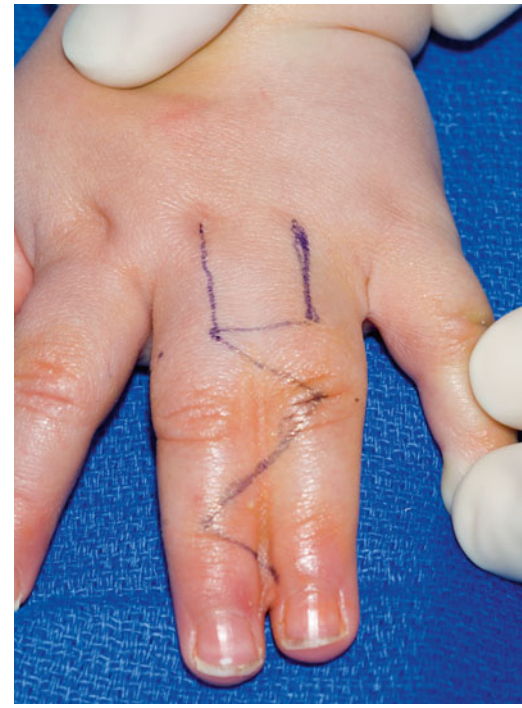

Fig. 7.7 Dorsal commissure flap begins at the level of the metacarpal heads and includes two-thirds the length of the proximal phalanx (Courtesy of Shriners Hospitals for Children, Philadelphia)

dorsal zigzag incision begins at one distal corner of the commissure flap and the palmar zigzag incision starts at the opposite distal corner. The dorsal zigzag incision extends to the midline of the proximal interphalangeal (PIP) joint of the adjacent finger and back across to the midline of the distal interphalangeal (DIP) joint. From this point, the incision extends distally between the tips of the interconnected digits. The palmar flaps are based opposite the dorsal flaps (i.e., mirror images) with the base of each flap centered over the opposite PIP or DIP joint to allow for interdigitation. The palmar rectangular flap and zigzag incision can be skewed to cover one digit entirely. Any remaining bare areas require full thickness skin graft.

The dorsal flaps are usually elevated first, preserving the paratenon over the extensor tendon. I prefer to visualize the volar neurovascular structures from the dorsal side before elevation

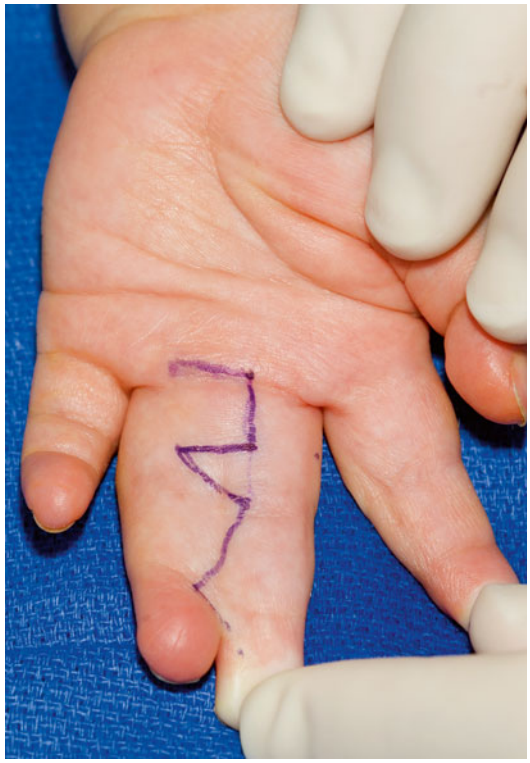

Fig. 7.8 Palmar rectangular flap is fashioned to resurface the proximal area of a digit adjacent to the commissure (Courtesy of Shriners Hospitals for Children, Philadelphia)

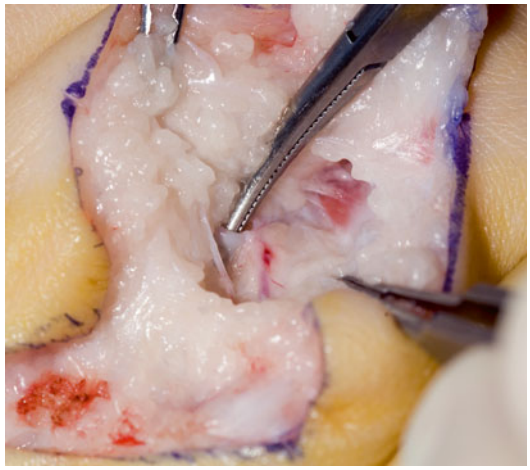

Fig. 7.9 From the dorsal side, the volar neurovascular structures are visualized (Courtesy of Shriners Hospitals for Children, Philadelphia)

of the palmar flaps (Fig. 7.9). This method allows for assessment of the neurovascular bundles and a double check of the palmar flap design

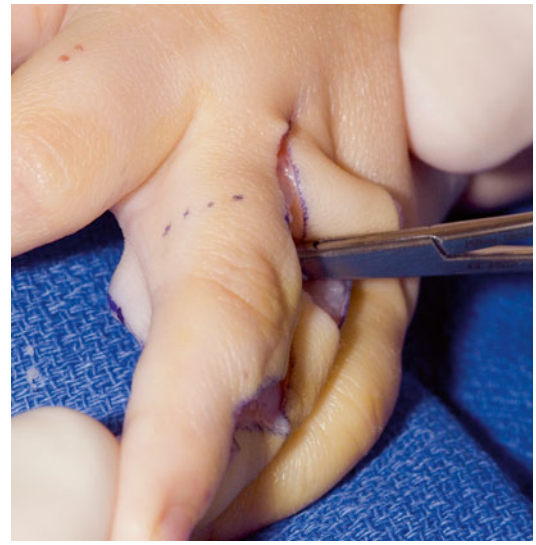

Fig. 7.10 The length and location of the volar flaps can be checked after elevation of the dorsal flaps (Courtesy of Shriners Hospitals for Children, Philadelphia)

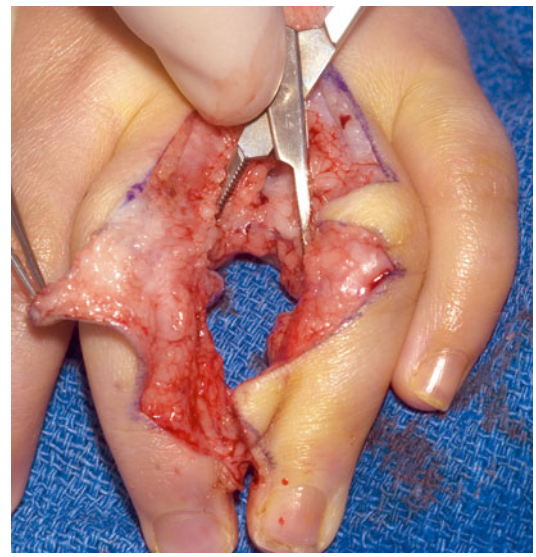

Fig. 7.11 Lateral spreading of the digits places the intervening tissue under tension and facilitates digital separation (Courtesy of Shriners Hospitals for Children, Philadelphia)

(Fig. 7.10). Following elevation of the dorsal flaps, the palmar flaps are elevated and the underlying neurovascular bundles are protected. The digits are separated from distal to proximal. Manual lateral spreading of the digits places the interconnecting tissue under tension, which eases digital separation (Fig. 7.11). Stout transverse

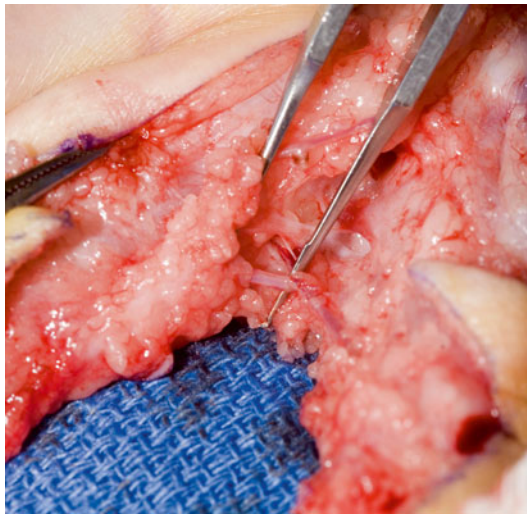

Fig. 7.12 The bifurcation between the common and proper neurovascular structures requires identification (Courtesy of Shriners Hospitals for Children, Philadelphia)

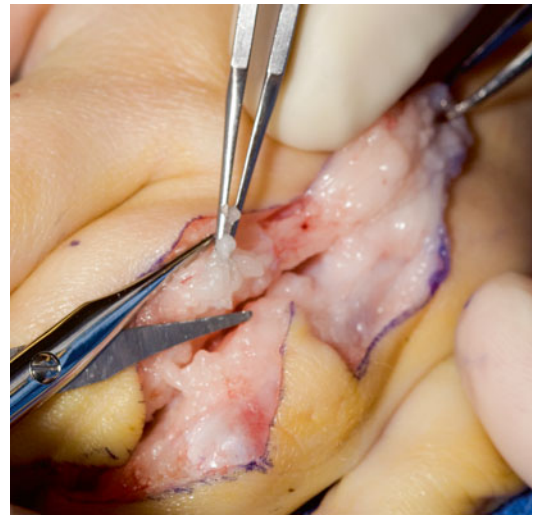

Fig. 7.13 The adjacent sides of the separated digits are defatted prior to insetting of the flaps (Courtesy of Shriners Hospitals for Children, Philadelphia)

fascial bands are incrementally incised toward the arterial bifurcation. The transverse intermetacarpal ligament is not divided because this may cause instability.

Neurovascular Considerations

The neurovascular anatomy requires surgical decision making (Fig. 7.12). If a common digital nerve divides distal to the proposed commissure, micro-dissection is performed to separate the nerve in a proximal direction. If a common digital artery divides distal to the proposed commissure, ligation of a proper digital artery may be necessary for commissure placement. Selection of the proper digital artery to ligate depends on the status of the proper digital arteries on the adjacent sides of the conjoined digits. If each digit has intact proper digital arteries on both sides, the smaller artery is usually ligated. However, if one of the digits still requires additional syndactyly surgery on its other border, then ligation of the larger artery may be appropriate. If the status of the opposite digital artery is unclear, vascular clamps are applied to the digital arteries and the tourniquet deflated to confirm adequate perfusion of each digit. Failure of either digit to perfuse is rare and requires preservation of both digital arteries. Options for this situation include moving the commissure more distal without vessel ligation or microsurgical reconstruction.

Closure

Prior to insetting of the flaps, the adjacent sides of the separated digits are defatted to minimize the amount of skin graft required (Fig. 7.13). A fast-absorbable 5–0 suture is used for insetting of the commissure flap, interdigitating zigzag dorsal and palmar flaps and full-thickness skin graft (Figs. 7.14 and 7.15). Following closure, a meticulous hand dressing is applied and the tourniquet is deflated to check for capillary refill. Subsequently, a long arm soft cast ® (3 M, St. Paul, MN, USA) is applied with the elbow flexed to more than 90° and the digits are covered. At 3 weeks, the dressings are removed and wound care initiated. Therapy is usually not required. Therapeutic modalities, including scar massage, silicone gel sheets, and elastomer putty can be used to treat areas of hypertrophic scarring.

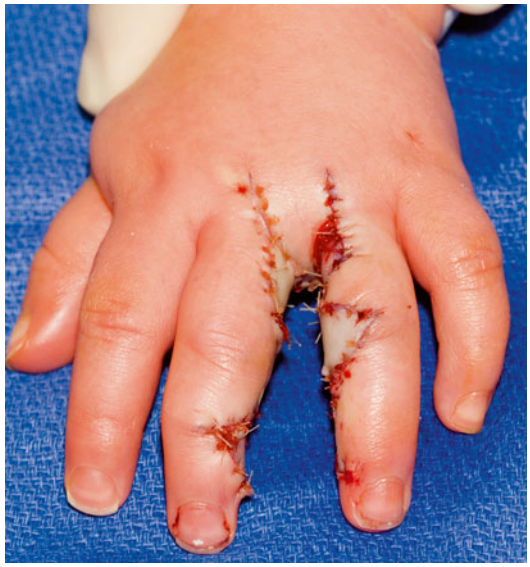

Fig. 7.14 Dorsal appearance after flap closure and full-thickness skin grafting (Courtesy of Shriners Hospitals for Children, Philadelphia)

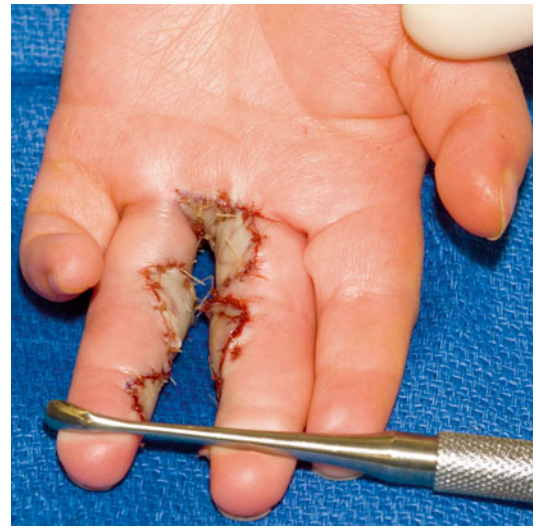

Fig. 7.15 Volar appearance after flap closure and full-thickness skin grafting (Courtesy of Shriners Hospitals for Children, Philadelphia)

Clinical Pearls

Dorsal flap for commissure reconstruction

Dorsal dissection to identify volar neurovascular structures

Vascular clamps can be applied to the proper digital arteries when the status of the other proper artery is unknown

Skin graft from distal wrist crease, antecubital fossa, or medial arm is within surgical field and provides a better match than groin graft

Soft cast immobilization negates the use of a cast saw

Outcome

Simple syndactyly separation that follows the principles and techniques outlined above reliably results in improved appearance and independent digital function [11]. The procedure must be done with careful technique and handling of the soft tissue. In contrast, the outcome for complex and polysyndactyly is less predictable and the complication rate is higher [4–6]. The adherent bony and soft tissue anatomy makes the surgery more difficult and the outcome less satisfactory. Despite one's best effort, residual mal-alignment, stiffness, and/ or instability are common.

Complications

Acute complications include infection, flap failure, and loss of skin graft. [4, 5] Preventative measures include preoperative antibiotics, careful tissue handling, and hemostasis combined with compressive post-operative dressings. A small amount of graft loss is inconsequential and will heal by secondary intention. Substantial graft loss is usually attributed to infection or inadvertent cast removal and requires repeat grafting to prevent hypertrophic scar formation. Digital circulation must be verified before cast immobilization. Circumferential tension around the digit can be caused by tight dressings within the web space or flap tightness that compresses the digital vessels. If loosening of the dressings does not result in capillary refill, release of the compressing skin flap is required. Additional skin grafting may be necessary to fill the deficit caused by flap release.

Chronic complications are more common and include web creep, residual nail deformity, and scar

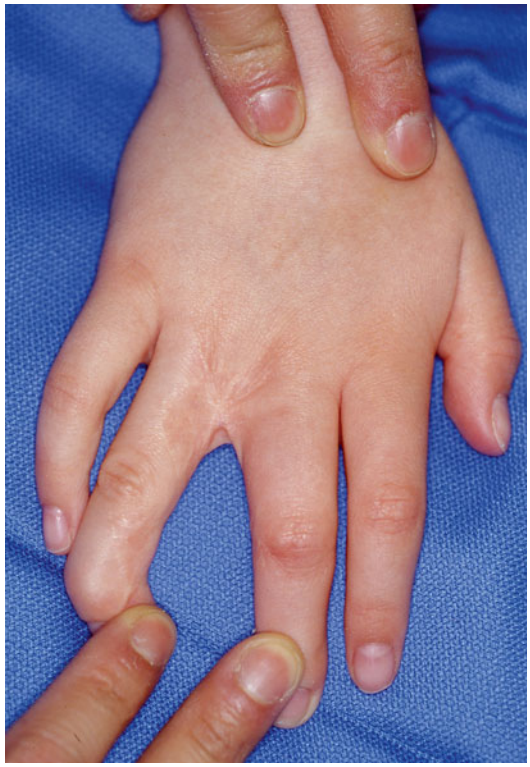

Fig. 7.16 Web creep after syndactyly separation (Courtesy of Shriners Hospitals for Children, Philadelphia)

contracture. Approximately one third of patients with simple syndactyly and approximately two thirds of patients with complex or complicated syndactyly will require additional surgery [4, 5]. Web creep is the distal migration of the reconstructed commissure that occurs with subsequent growth of the patient (Fig. 7.16) [16]. The development of web creep is multifactorial and is related to age at surgery, surgical technique, and quality of skin graft. Use of a split-thickness graft is associated with an eightfold increase in web migration compared with use of a full-thickness graft [5]. Patients younger than 18 months at the time of surgery tend to have more web creep [11]. Another common complication of complete syndactyly release is residual nail deformity. Despite valiant attempts at nail fold reconstruction using coronal Buck-Gramcko pulp flaps (Fig. 7.17) [17]. residual deformity of the nail is common especially with synonychia.

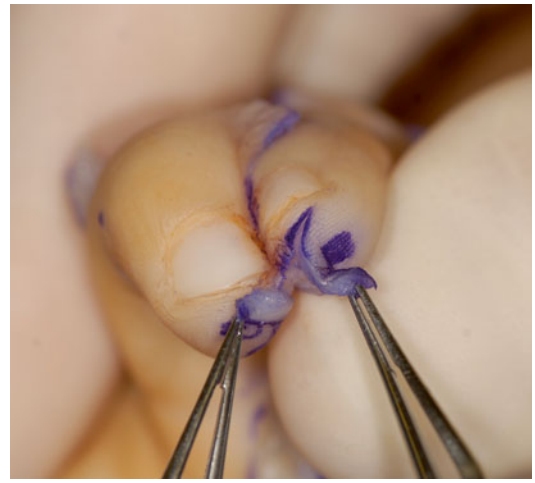

Fig. 7.17 Nail fold reconstruction using coronal Buck-Gramcko pulp flaps (Courtesy of Shriners Hospitals for Children, Philadelphia)

Synostosis

Introduction

Synostosis is a generic term that indicates an osseous union between bones that are normally separate [3, 18, 19]. Synostosis is primarily a failure of skeletal differentiation, although an associated soft tissue ptyergium may be present. The precise cause is unknown.

Synostosis can occur anywhere throughout the limb. Synostosis can involve the fingers (symphalagism or metacarpal synostosis), carpus (carpal coalitions), forearm (radioulnar synostosis), or elbow (radiohumeral or ulnohumeral synostosis) (Fig. 7.18). Restoration of motion for congenital synostosis has been uniformly unsuccessful. Treatment is primarily realignment or reorientation to improve function. This section will focus on radiohumeral, radioulnar, and metacarpal synostosis.

Elbow/Forearm Synostosis

Incidence/Etiology

Elbow synostosis can be longitudinal or transverse. Longitudinal synostosis can occur across the ulnar-humeral and/ or radial-humeral articulation. Transverse forearm synostosis can occur at the distal, middle, or proximal forearm.

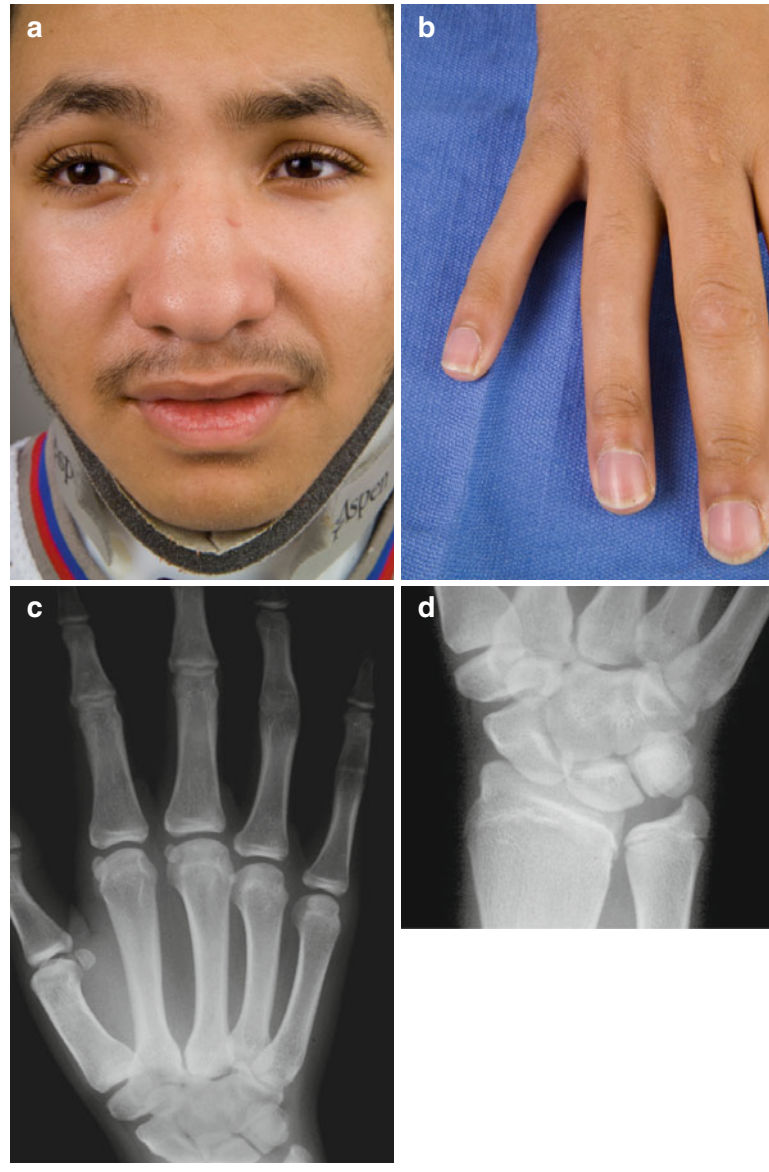

Fig. 7.18 14 year-old with Herrmann Multiple Synostosis Syndrome (Courtesy of Shriners Hospitals for Children, Philadelphia). (**a**) Characteristic facies with broad nose with hypoplastic nasal alae. (**b**) Right hand with absent proximal interphalangeal joint skin creases and symphalagism. (**c**) X-ray reveals longitudinal synostosis across the proximal interphalangeal joint. (**d**) Wrist x-ray with capitohamate carpal coalotion

Radiohumeral synostosis most commonly occurs in ulnar deficiency (Fig. 7.19) [3]. Radiohumeral synostosis can also be part of a synostosis syndrome, such as Herrmann multiple synostosis syndrome [3, 20]. Radioulnar joint synostosis may occur sporadically, may be inherited as an autosomal dominant trait, or can be found in a variety of syndromes including trisomy [13, 21] and fetal alcohol syndrome [3]. Radioulnar joint synostosis is bilateral in approximately 50 % of children [18, 19].

Evaluation (Presentation, Investigation and Treatment Options)

The child with radiohumeral synostosis presents at a young age with lack of active and passive elbow flexion and extension. There are absent elbow skin joint creases (a.k.a. joint creases). The synostosis is typically cartilaginous early in life, which accounts for the findings of a small amount of perceived motion. However, the connection becomes ossified over time leading to a complete lack of passive motion. An initial x-ray often reveals a small joint space that proposes

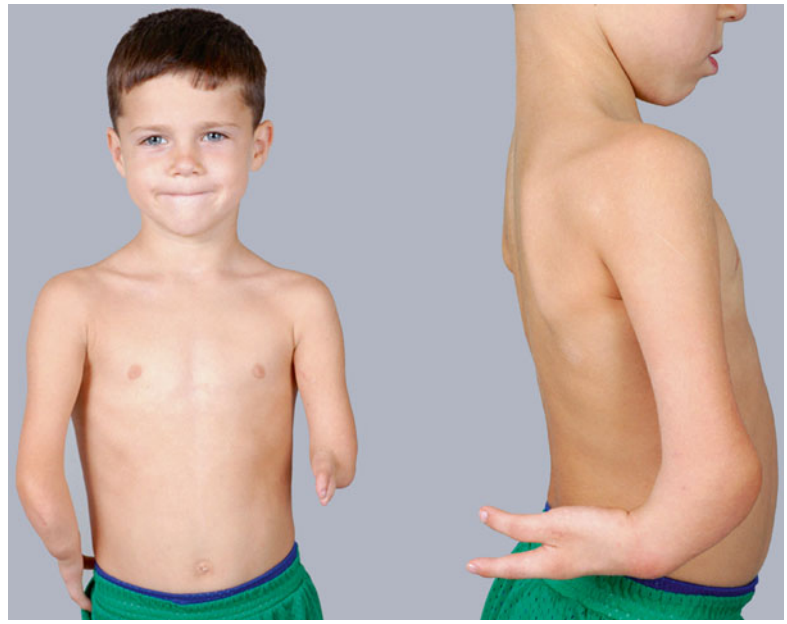

Fig. 7.19 5 year-old presents with bilateral radiohumeral synostosis associated with ulnar deficiency (Courtesy of Shriners Hospital for Children, Philadelphia)

"false hope" to the family, and perhaps to the surgeon, for subsequent movement. Advanced imaging studies are unnecessary. The elbow can be fixed in various positions. In children with profound ulnar deficiency, the elbow is usually in extension. In contrast, in children with a normal length ulna the elbow is typically in flexion [21].

The child with radioulnar synostosis does not present at a young age, but rather between the ages of 2 and 6 years with painless absence of forearm rotation (Fig. 7.20a). The delay in diagnosis is attributed to the fact that an infant does not use forearm rotation and that shoulder and wrist motion can compensate during activities associated with early childhood. Shoulder abduction compensates for deficient pronation and shoulder adduction compensates for deficient supination (Fig. 7.20b). Furthermore, children with deficient forearm rotation develop excessive wrist intercarpal rotation (up to 60° of rotation), which further compensates for lack of forearm rotation [22]. Therefore, a delay in presentation is typical until the complexities of daily activities amplify, such as catching a ball, self-feeding, or toileting. Common complaints from lack of pronation are problems with keyboarding and table top activities. Common problems from lack of supination are eating, washing one's face, catching a ball, and using a soap dispenser.

The physical examination of forearm motion must be assessed at the radial styloid and ulnar head (distal radioulnar joint) to avoid measuring wrist motion (Fig. 7.20c). This is especially important is children with disproportionate intercarpal rotation, which can lead to misdiagnosis. Anteroposterior and lateral elbow x-rays are routine and the synostosis is nearly always about the proximal radioulnar joint (PRUJ) (Fig. 7.20d). In a young child, the connection can initially be cartilaginous but eventually ossifies to a bony connection. A concomitant radial head dislocation may be present. Advanced imaging studies are unnecessary.

Management

Nonsurgical Treatment

Radiohumeral synostosis most commonly occurs in children with ulnar deficiency (type IV). The synostosis is often bilateral and associated with hand differences [3]. The child is often born with the hand facing backwards and against the flank, which severely impairs function. Treatment recommendations have varied from observation to corrective osteotomy to better position the limb in front of the body and into a more functional

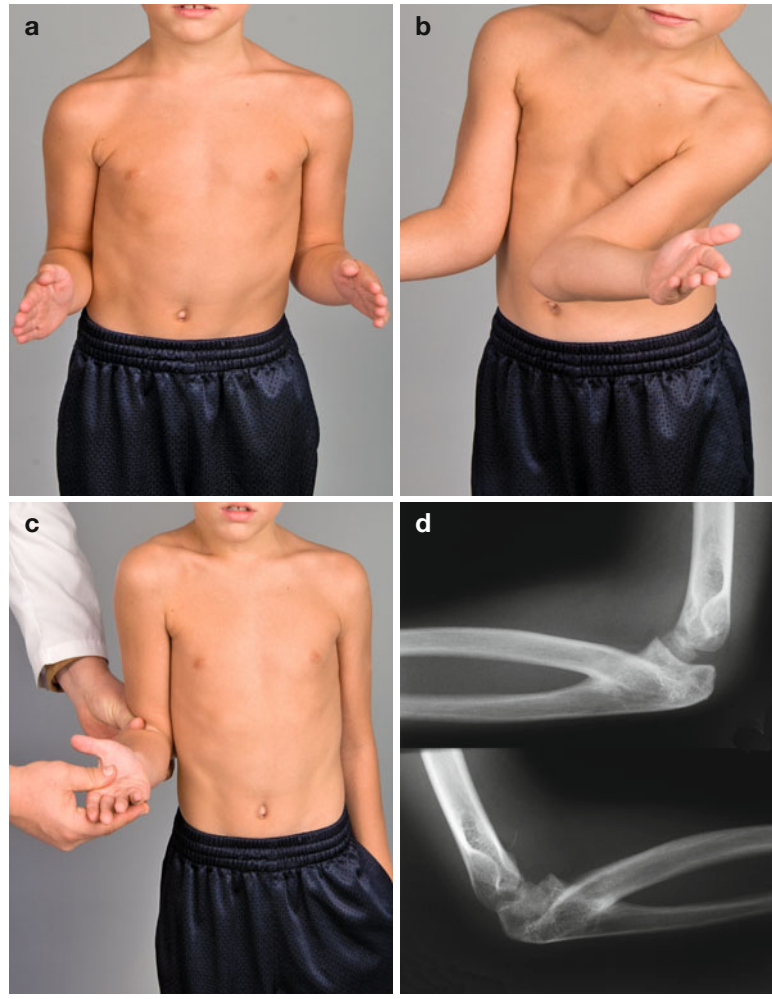

Fig. 7.20 7 year-old with bilateral proximal radioulnar joint synostosis (Courtesy of Shriners Hospital for Children, Philadelphia). (**a**) Right forearm fused in neutral and left fixed in slight pronation. (**b**) Shoulder adduction compensates for deficient supination. (**c**) Forearm motion must be assessed at the distal radioulnar joint and not the hand to avoid false interpretation of forearm movement. (**d**) X-rays confirm bilateral proximal radioulnar joint synostosis

position. In the past, I would surgically improve limb position via osteotomy through the fusion mass. The repositioning can be accomplished acutely or slowly via distraction techniques. Over time, I noticed that children born with their hand facing backward achieved similar repositioning as those children that underwent surgery (Fig. 7.21). In addition, their function is enhanced by their remarkable compensatory motions. The underlying basis for their correction is unclear, but may be related to extreme motion in the adjacent joints or changing bony torsion. Currently, I reserve osteotomy for recalcitrant cases that present with poor limb position despite time, growth, and development.

The limbs of children with radiohumeral synostoses are short due to loss of the growth plates about the elbow. Limb lengthening is a consideration to increase the child's workable reach space. However, these procedures are fraught with substantial complication rates and the risk/ benefit ratio needs to be measured. Furthermore, lengthening the extremity moves the hand farther from the child's face and may decrease the child's overall function. An occupational therapy evaluation is crucial to the decision making process to avoid doing harm.

In children with radioulnar synostosis, mild degrees of fixed pronation or supination are well tolerated by patients due to compensatory motion at the shoulder and/ or wrist joints [18, 22]. Therefore, surgery is not recommended as strategies that attempt to restore forearm motion are doomed to fail and universally unacceptable [23].

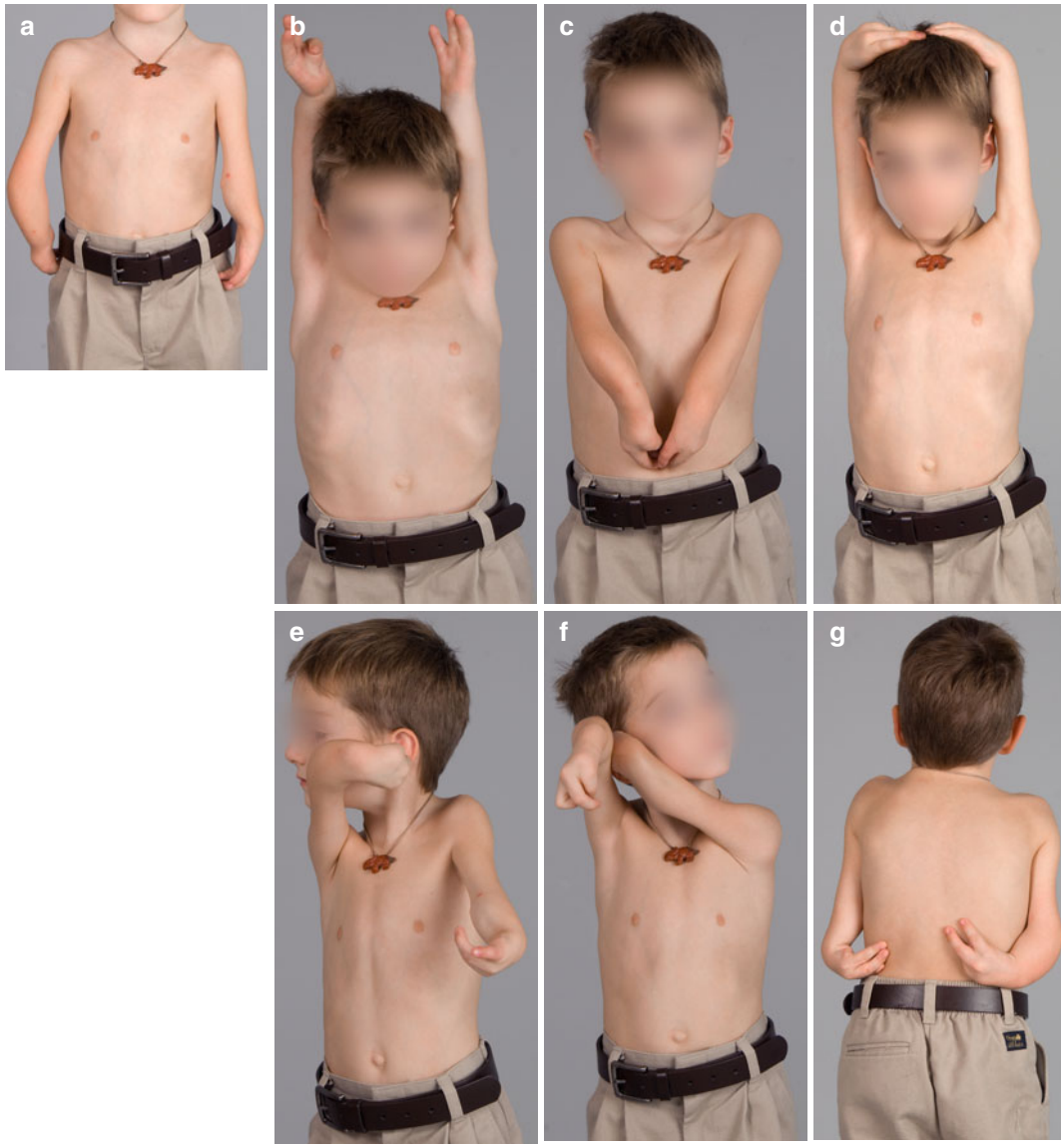

Fig. 7.21 6 year-old with untreated bilateral radiohumeral synostosis and ulnar deficiency (Courtesy of Shriners Hospital for Children, Philadelphia). (**a**) Resting posture with left hand facing backwards. (**b**) Overhead reach. (**c**) Midline activity. (**d**) Touching top of head for washing hair. (**e**) Touching left ear with right hand. (**f**) Touching left ear with right hand using left arm as assist. (**g**) Reaching back with both hands

Surgical Techniques

In children with radioulnar synostosis, extremes of pronation or supination cause functional difficulties for children especially as they become more independent and their activities become more complex. In the young child that presents with extreme rotation, early osteotomy is appropriate (Fig. 7.22). In the older child, when the complaints interfere with the child's daily activities, surgical intervention is warranted to place the limb in a more functional position.

Preoperative Evaluation

The optimal rotational position of the forearm is controversial as the best position varies with

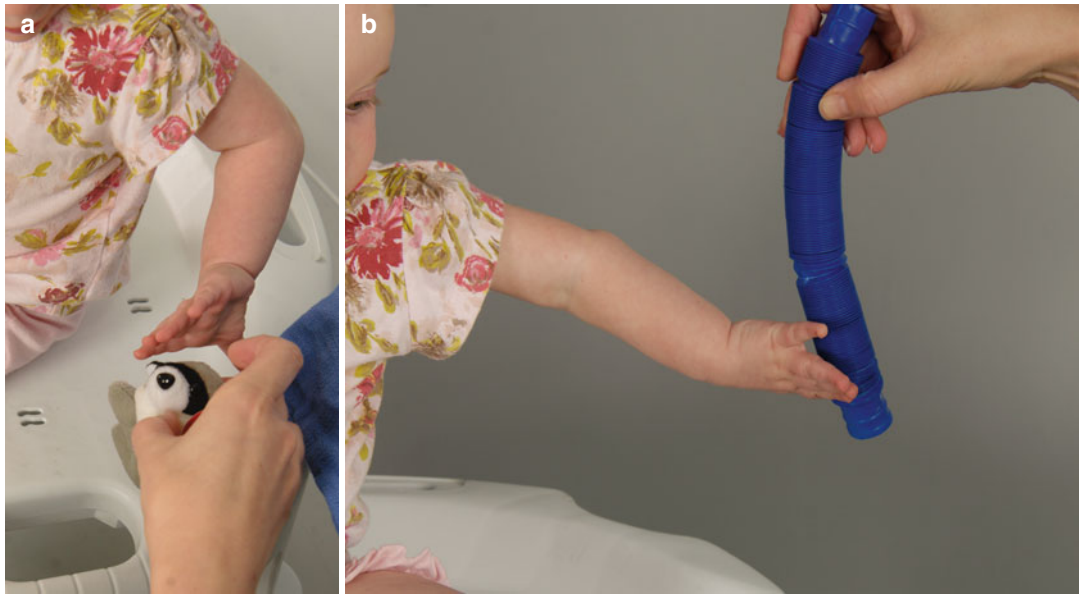

Fig. 7.22 1½ year-old female with bilateral radioulnar synostosis (Courtesy of Shriners Hospital for Children, Philadelphia). (**a**) Left arm extremely pronated. (**b**) Object acquisition with arm hyperpronated

the task. A pre-operative occupational therapy evaluation is helpful to determine the individual child's functional deficits and optimal forearm position. Bilateral recommendations are to place the dominant forearm in 10–20° of pronation and the non-dominant forearm in neutral rotation [24]. Unilateral recommendations are to place the forearm in 30° of pronation to provide the best function for keyboarding, writing, and working with small objects. The determination of optimal forearm positioning, however, should be based on the precise needs of the individual.

Surgical Technique- Osteotomy

The child is positioned supine and the arm is placed on a table. A pediatric tourniquet is placed high on the arm to allow access to the epicondylar axis for accurate measurement of forearm rotation. A longitudinal dorsal incision is made along the subcutaneous border of the ulna directly over the synostosis (Fig. 7.23). Sharp dissection is performed down to the synostosis between the extensor carpi ulnaris and the flexor carpi ulnaris. Reverse retractors or malleable retractors are placed around the synostosis.

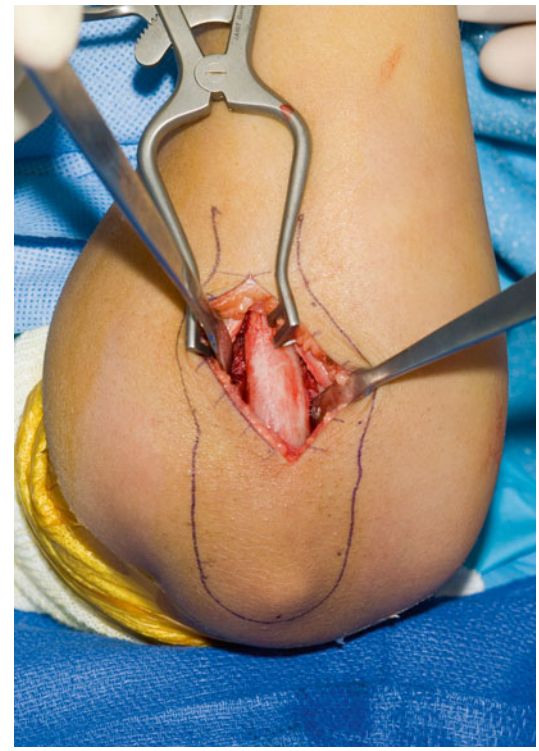

Fig. 7.23 Exposure of the synostosis along the subcutaneous border of the ulna between extensor carpi ulnaris and the flexor carpi ulnaris (Courtesy of Shriners Hospital for Children, Philadelphia)

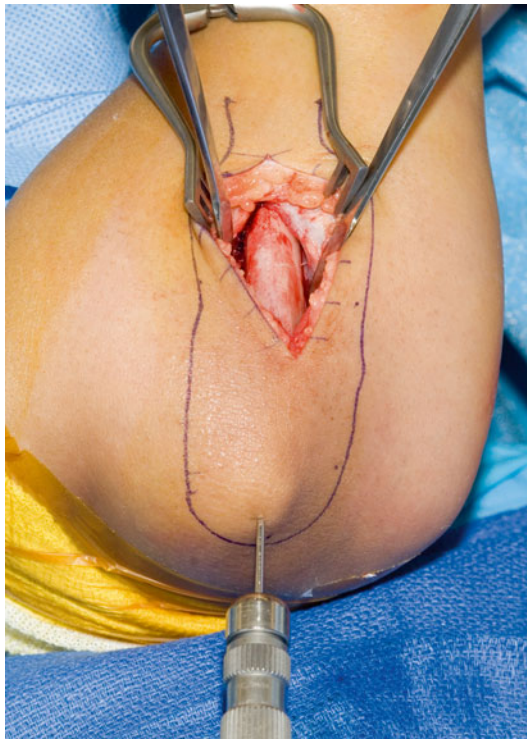

Fig. 7.24 Steinman pin inserted through the olecranon apophysis and into the intramedullary canal of the ulna (Courtesy of Shriners Hospital for Children, Philadelphia)

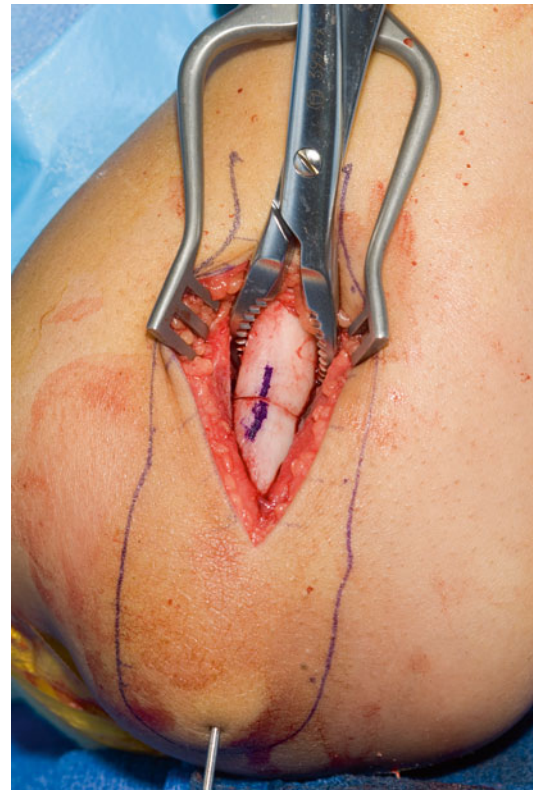

Fig. 7.25 Transverse osteotomy through the fusion mass (Courtesy of Shriners Hospital for Children, Philadelphia)

A 062-in. smooth Steinman pin is driven from a percutaneous position through the olecranon apophysis and down the intramedullary canal of the ulna (Fig. 7.24). The pin is positioned just proximal to the planned osteotomy through the synostosis. Mini-fluoroscopy is used to verify correct pin placement. The longitudinal axis of the ulna is marked prior to osteotomy (Fig. 7.25). This allows assessment of the rotational correction following osteotomy. An oscillating saw is used to cut a transverse osteotomy (Fig. 7.26). The osteotomy must be complete to allow rotation at the osteotomy site. The forearm is rotated into the predetermined position and the longitudinal wire is advanced across the osteotomy site. Once the position is satisfactory, an additional percutaneous oblique Kirschner wire is driven across the osteotomy for rotational control (Fig. 7.27). The pins are left in a percutaneous position. The subcutaneous tissue and skin is closed with absorbable suture. The arm is placed in a long arm splint with the elbow flexed to 90°. The child is admitted to the hospital overnight for elevation and neurovascular monitoring.

A cast is applied 2 weeks after surgery. At 6 weeks, the pins are removed in the office after x-rays substantiate bony union. Therapy is usually not required.

Outcome

Rotational osteotomy for proximal radioulnar synostosis is a reliable procedure to reposition the forearm. Patients perceive better limb function after surgery with regards to activities of daily living [22, 25]. Once the osteotomy site heals in the desired position, recurrence does not occur [26].

Complications

The reported complication rate following rotational osteotomy for proximal radioulnar synostosis is higher than one would expect [22, 24, 25]. Intra-operatively, the retractors placed

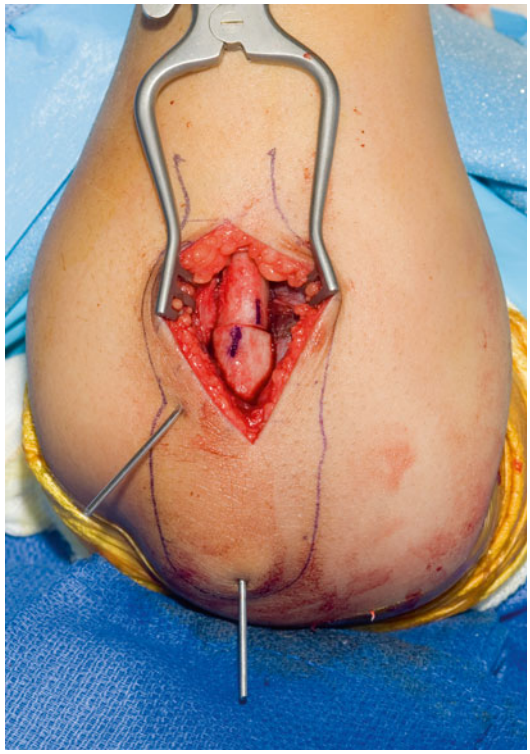

Fig. 7.26 The forearm is rotated and the longitudinal wire is advanced across the osteotomy site. An additional percutaneous oblique Kirschner wire is placed across the osteotomy for rotational control (Courtesy of Shriners Hospital for Children, Philadelphia)

around the synostosis may compress either the posterior interosseous and/or ulnar nerves resulting in a neurapraxia. Excessive forearm rotation, typically more than 85°, may lead to vascular compromise, nerve palsies, and compartment syndrome [25]. These are treated emergently by lessening the amount of rotational correction. In children greater than 10 years of age, the possibility of posterior interosseous injury increases as rotation is less tolerated [22, 24].

Pearls

Absence of skin creases is the hallmark of an underlying synostosis

Unilateral radioulnar synostosis in mild degrees of fixed pronation or supination is well tolerated by patients due to compensatory motion at the surrounding joints

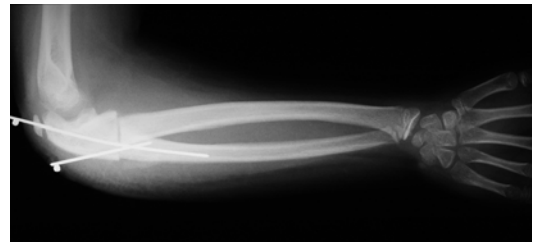

Fig. 7.27 X-rays after rotational osteotomy with good fixation (Courtesy of Shriners Hospital for Children, Philadelphia)

Consult an occupational therapist to help determine the optimum position of forearm for function when considering surgery

Avoid operations to restore motion to elbow/ forearm synostosis

Beware of potential neurovascular issues after rotational osteotomy for radioulnar synstosis

Metacarpal Synostosis

Incidence/Etiology

Metacarpal synostosis is a transverse bony fusion that most commonly involves the ring and small fingers. This anomaly occurs bilaterally in 60–80 % of the cases [27]. Metacarpal synostosis can occur as an isolated finding, but is also found in Apert syndrome, symbrachydactyly, and central deficiencies.

Evaluation (Presentation, Investigation and Treatment Options)

The child with metacarpal synostosis presents at a young age with the small finger usually hypoplastic and noticeably abducted from the palm (Fig. 7.28a). There is deficient active and passive flexion and extension. In the older child, the finger is an annoyance as it gets caught in pockets and close spaces.

Plain x-rays show fusion of the proximal ring and small metacarpal. The extent of the fusion is variable (Fig. 7.28b, c). The extent of the synostosis relates to the degree of small finger shortening and ulnar deviation [27]. At the distal end of the metacarpal, the small finger is postured into abduction. This position prevents the finger from being brought parallel to the rest of the hand.

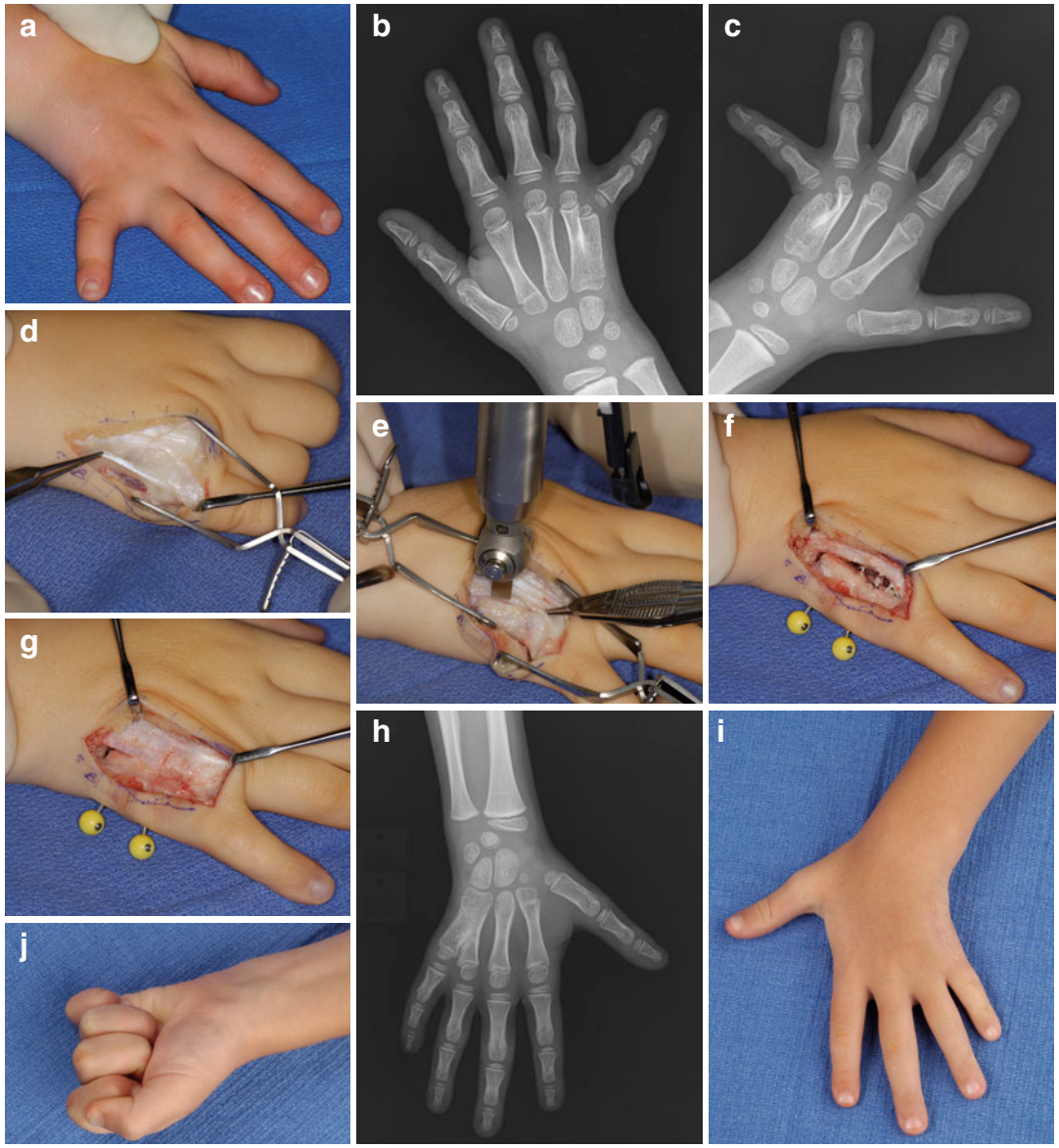

Fig. 7.28 4 year-old with bilateral ring/ small finger synostosis (Courtesy of Shriners Hospital for Children, Philadelphia). (**a**) Right hand with small finger abducted. (**b**) Right hand x-ray reveals extensive ring/ small finger synostosis. (**c**) Left hand x-ray with similar synostosis. (**d**) Dorsal approach and isolation of the extensor apparatus. (**e**) Synostosis divided with an oscillating saw. (**f**) The small finger metacarpal is pulled in a distal direction until satisfactory position has been obtained and percutaneous Kirschner wires are placed. (**g**) Extensor tendons are realigned over the metacarpal and marked improvement in small finger position. (**h**) Post-operative x-ray with realignment of metacarpal and small finger. (**i**) Clinical follow-up with improved alignment. (**j**) Good fist formation

Management

Nonsurgical Treatment

Mild abduction with good motion does not warrant surgical management. Observation is the rule with ongoing follow-up.

Surgical Techniques

The status of the small finger usually dictates the treatment. A vestigial small finger is best left alone or amputated. A functioning small finger deserves reconstruction. The goals of surgery

are to re-align and lengthen the small metacarpal [27, 28]. In addition, the extensor apparatus may require centralization and the radial collateral ligament may need reconstruction.

The child is positioned supine and the arm is placed on a table. A pediatric tourniquet is placed on the arm. After exsanguination, a dorsal approach is made over the metacarpal synostosis. The extensor apparatus is isolated. The abductor digit quinti is released from the extensor hood (Fig. 7.28d). The ring-small metacarpal synostosis is isolated along its entire length. The synostosis is divided with a knife or oscillating saw depending upon the age of the child (Fig. 7.28e). At the base of the small finger metacarpal a transverse cut is performed to prevent propagation into the carpometacarpal joint. The small finger metacarpal is separated from the ring finger using a lamina spreader and then pulled in a distal direction. Once satisfactory position has been obtained, percutaneous Kirschner wires are placed to maintain the position (Fig. 7.28f). In the young child, bone graft is unnecessary. Fat can be placed between the synostosis to lessen the chance of recurrence.

The extensor tendons are inspected for alignment and the collaterals tested for stability. Extensor tendon malignment requires centralization (Fig. 7.28g). Collateral ligament instability can be reconstructed with local tissue if available or a piece of extensor tendon fixed to the head of metacarpal and the base of proximal phalanx. Collateral ligament reconstruction, however, induces additional stiffness to an already inflexible joint.

Outcome

The surgical results are dependent upon the degree of synostosis and status of the small finger. X-rays demonstrate improved alignment of the small finger metacarpal (Fig. 7.28h). The hand appearance is improved and the small finger is less likely to become caught in pockets and tight spaces (Fig. 7.28i, j). Parents are usually satisfied with the outcome [27, 28].

Complications

The most common complication is that the hypoplastic small finger does not appear normal or function normally. Stiffness in the metacarpophalangeal joint is a common preoperative finding that persists after surgery (Fig. 7.29). Other uncommon complications are damage to the metacarpal physis and nonunion.

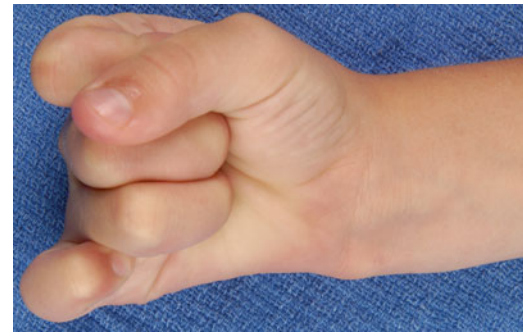

Fig. 7.29 Left hand with metacarpophalangeal joint stiffness after synostosis correction

Pearls

Delay surgery until the extent of the synostosis is evident on plain x-rays

Back cut the small metacarpal to prevent osteotomy propagation into the carpometacarpal joint.

Discuss metacarpophalangeal joint stiffness prior to surgery and explain this diminished motion will not improve.

Only reconstruct the collateral ligament(s) when necessary as this causes additional metacarpophalangeal joint stiffness.

Camptodactyly

Introduction

Camptodactyly is a painless flexion contracture of the proximal interphalangeal (PIP) joint. [29–31] Secondary metacarpophalangeal and distal interphalangeal (DIP) joint deformities may develop over time. Camptodactyly may be flexible or fixed (more common). The cause and treatment algorithm remains controversial.

Incidence/Etiology

Camptodactyly occurs in less than 1 % of the population and most cases are asymptomatic. Camptodactyly is bilateral two-thirds of the time with asymmetric involvement. The small finger is most commonly involved with the incidence decreasing toward the radial side of the hand [29–31]. Camptodactyly is usually sporadic in occurrence, although an inheritable form has been described that is an autosomal dominant trait with variable expressivity and incomplete penetrance [30, 32]. Camptodactyly can also occur in a variety of syndromes including craniocarpotarsal dystrophy (Freeman-Sheldon syndrome), congenital contractural arachnodactyly (Beal's syndrome), and orofaciodigital syndrome (Fig. 7.30) [3].

Etiology is variable and controversial. Nearly every structure about the PIP joint has been implicated as the primary cause or a contributing factor. The most prevalent anomalies affect the flexor digitorum superficialis (FDS) and intrinsic musculature [29, 30, 33, 34]. The FDS tendon and/ or muscle have been described as "tight" and unable to elongate during growth, which causes a PIP joint flexion contracture. The intrinsic lumbrical has been described as absent or having an abnormal insertion.

Evaluation (Presentation, Investigation and Treatment Options)

Camptodactyly can present shortly after birth, during infancy, or during the course of adolescence. The involved digits are examined for active and passive movements. Involvement of the radial digits warrants a genetic consultation. The degree of PIP joint contracture is assessed with the metacarpophalangeal joint in both extension and flexion. Increased PIP joint posture in extension implies FDS tendon tightness [29, 30, 33]. The ability of the patient to extend the PIP joint while holding the metacarpophalangeal joint in flexion is also assessed. The inability to extend the PIP joint infers attenuation or insufficiency of the extensor hood.

Plain x-rays of the involved fingers are routine. The lateral x-ray is the key to assess secondary changes about the PIP joint. Potential findings are a misshapen head of the proximal phalanx, flattening of the base of the middle phalanx, and volar subluxation. Secondary changes negatively impact surgical outcome [29, 33].

Management

Nonsurgical Treatment

Any contracture less than 30° does not warrant surgery [29, 30]. Since 80 % of patients with camptodactyly will progress, nap and night splinting is recommended [33]. The precise type and time of splinting is controversial. Static progressive splinting or serial casting is recommended for contractures greater than 30°. The goals are to prevent progression and to maintain bone/joint integrity.

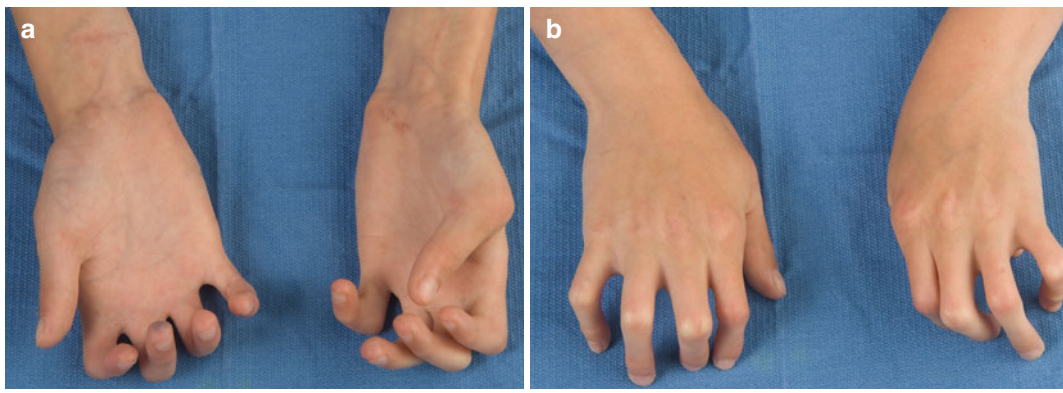

Fig. 7.30 7 year-old child with contractural arachnodactyly or Beal's syndrome and bilateral hand camptodactyly involving multiple digits. (**a**) Palmar view. (**b**) Dorsal view

Surgical Techniques

Surgery is reserved for a progressive or substantial deformity that has failed nonsurgical management and interferes with function. The PIP joint status decides whether a reconstruction can be performed or a salvage procedure is necessary.

Reconstruction

A PIP joint with minimal subluxation and secondary bony changes can be treated with reconstruction. A stepwise surgical approach is recommended with release of all offending structures [29, 30]. The skin incision various with the degree of contracture and the presence or absence of a ptyergium. Lesser contractures can be managed by Z-plasty, while larger contractures require a supplemental full thickness skin graft (Fig. 7.31a) [29, 30, 34].

Dissection is performed down to the flexor tendon sheath with release of any abnormal fascia. The flexor tendon sheath is opened via an L-shaped incision over the A3 pulley. Persistent tightness is uniform and subsequent release of the check-rein ligaments, flexor digitorum superficialis, and palmar plate is necessary (Fig. 7.31b). Instead of formal release of the palmar plate, the plate is fenestrated with a knife blade until adequate extension is obtained.

Any anomalous lumbrical muscle is excised [30, 34]. The decision to transfer the flexor digitorum superficialis tendon to the extensor apparatus is difficult [29, 30, 34]. The pros and cons of flexion versus extension need to be weighed. If transfer is selected, the transfer is passed through the lumbrical canal and sutured into the central

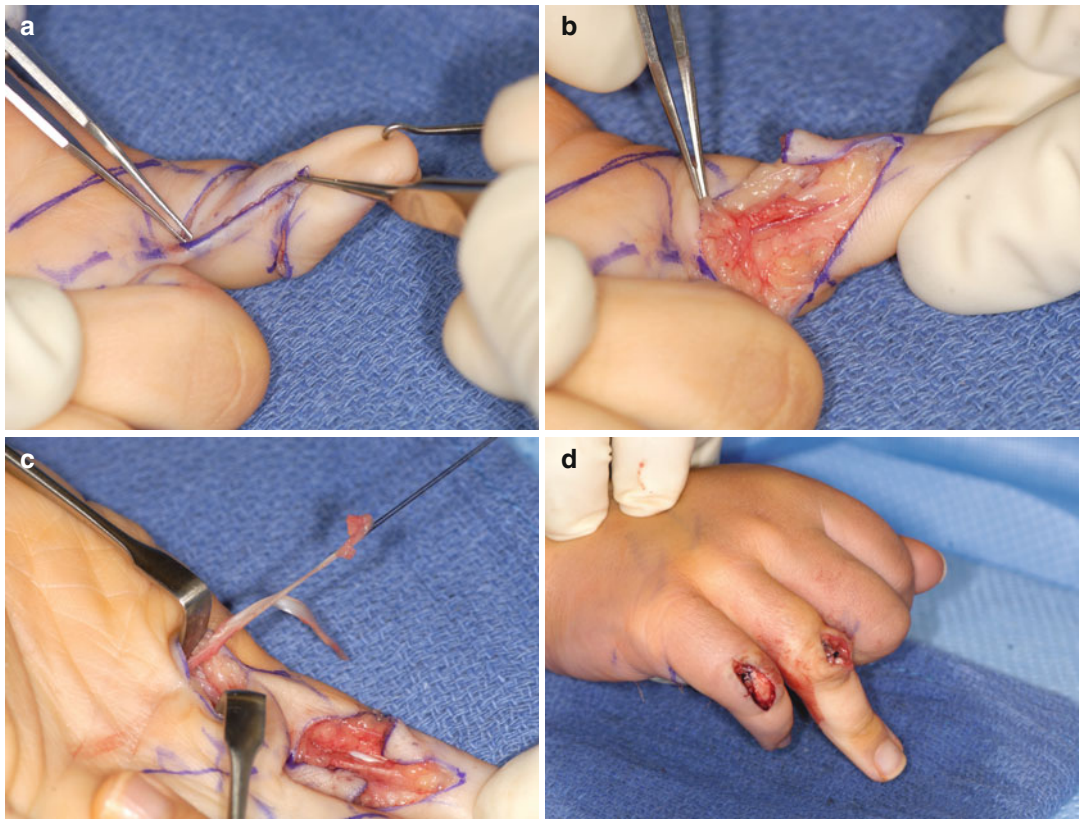

Fig. 7.31 8 year-old child with ring and small finger camptodactyly (Courtesy of Shriners Hospital for Children, Philadelphia). (**a**) Z-plasty along tight cord. (**b**) Isolation of underlying neurovascular bundle. (**c**) Flexor digitorum superficialis incised at A3 pulley. (**d**) Flexor digitorum superficialis transferred to extensor mechanism

slip/ extensor hood (Fig. 7.31c). The surgeon must avoid tensioning too tight, which would result in a swan-neck deformity and impairment in grasp. Following ample surgical release ± tendon transfer, a temporary percutaneous Kirschner wire can be placed across the PIP joint. Closure is performed with Z-plasty and/or skin graft (Fig. 7.31d).

Salvage Procedure

A PIP joint with marked volar subluxation and secondary bony changes is not a candidate for reconstruction. Bony re-alignment is the only method to lessen the excessive flexion. The PIP joint can be better positioned by a dorsal closing-wedge osteotomy of the proximal phalanx or a PIP joint fusion. Bony shortening is necessary to overcome the extreme volar tightness.

Outcome

The nonsurgical and surgical results are fairly disappointing. Rapidly increasing PIP joint contractures are often recalcitrant to splinting and require surgery before joint deformation. The inconsistent surgical findings amongst various reports confound universal statements regarding surgical outcome. The results tend to be unpredictable, although Smith [30] and Foucher [29] report better results with a systematic approach.

Complications

Surgical complications particular to camptodactyly include skin loss, neurovascular compromise, and stiffness. Skin loss occurs when flaps are sutures under tension, which must be avoided. In these cases, it is better to add supplemental skin graft than risk flap loss. Neurovascular compromise can be secondary to direct injury during dissection, but is more commonly related to excessive tension after release. In these cases, the digit must be repositioned in less extension to restore blood flow. Loss of motion is endemic after surgery. Insufficient extension is better tolerated than inadequate flexion.

Pearls

There are a myriad of potential causes for camptodactyly with the flexor digitorum superficialis and lumbrical musculature most implicated

Mild camptodactyly should be splinted with avoidance of surgery.

Surgical requires a stepwise approach to lessen the contracture and maximize outcome

Bony changes and PIP joint subluxation negate the possibility of reconstruction

The surgical complications rate is considerable

Thumb Camptodactyly

Thumb camptodactyly can be related to the "clasped thumb" (hypoplasia of the extensor pollicis brevis) or secondary to arthrogryposis. In general, the clasped thumb is less rigid and more responsive to non-operative management with thumb spica splinting. In contrast, the arthrogrypotic thumb camptodactyly is more rigid and less responsive to non-operative management.

The treatment of both types of "thumb-in-palm" deformity begins with stretching and splinting. Most of the clasped thumb variants do not require surgery. A persistent extension lag at the metacarpophalangeal joint can be treated with tendon transfer to augment thumb extension. The donor tendon is usually the extensor indici proprius transferred to the residual extensor pollicis brevis tendon or directly to the base of the proximal phalanx.

The arthrogrypotic "thumb-in-palm" deformity is an entirely different story. The thumb is often recalcitrant to non-operative measures and surgery is necessary to liberate the thumb out of the palm. The surgery must address the deficient volar skin across the thumb metacarpophalangeal joint and the rigidness of the metacarpophalangeal joint. The skin deficiency

can be overcome by a rotational flap from the radial side of the index or dorsum of the hand. In addition, full thickness skin graft can be used to cover the defect once the thumb in straightened. The metacarpophalangeal joint is usually treated by chondrodesis with preservation of the physis at the proximal phalanx. Ample metacarpal head is removed to position the joint in full extension without excessive tension across the neurovascular bundles. Fixation is accomplished with Kirshner wires that are removed after healing.

Clinodactyly

Introduction

Clinodactyly is an abnormal deviation in the radioulnar plane and most commonly involves radial deviation of the small finger DIP joint. [3]. The deformity is usually fixed and there is no intra-articular or periarticular swelling.

Incidence/Etiology

A DIP joint deviation less than 10° is so common, it may be considered normal [3, 35]. Clinodactyly affects somewhere 1.0 and 19.5 % of normal children [36]. Clinodactyly can be inherited via an autosomal dominant trait with variable expressivity and incomplete penetrance. Familial clinodactyly is usually not associated with systemic conditions. There are many genetic syndromes and chromosomal abnormalities that have clinodactyly as a physical finding, including Down syndrome (incidence between 35 and 79 %), Apert syndrome, Rubinstein-Taybi syndrome, diastrophic dwarfism and triphalangeal thumbs [35, 37].

Idiopathic clinodactyly is caused by an abnormal inclination of the middle phalanx articular surface adjacent to the DIP joint. [35, 37, 38] Abnormal deviation of a digit, however, can be caused by other aetiologies. A longitudinal orientation of the middle phalanx growth plate can alter the configuration of the phalanx and yield coronal deviation [37, 39, 40]. This finding is termed a longitudinal epiphyseal bracket, longitudinal bracketed diaphysis, or delta phalanx (Fig. 7.32).

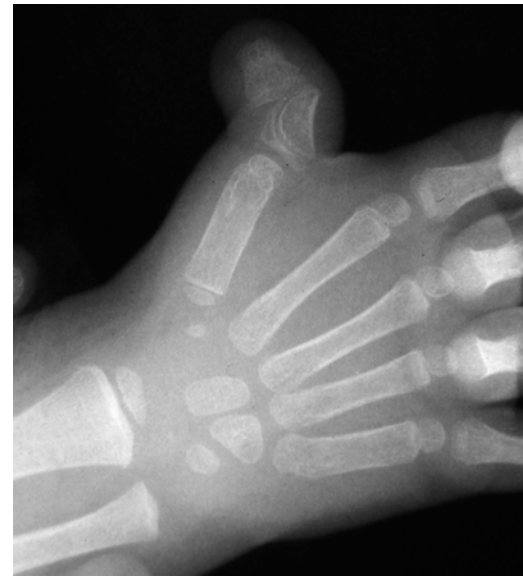

Fig. 7.32 3 year-old with Rubinstein-Taybi syndrome and thumb clinodactyly secondary to a longitudinal epiphyseal bracket of the proximal phalanx (Courtesy of Shriners Hospital for Children, Philadelphia)

Evaluation (Presentation, Investigation and Treatment Options)

The diagnosis of clinodactyly is straightforward. Idiopathic clinodactyly is usually noted shortly after birth or during infancy with a crooked small finger (Fig. 7.33a). The deviation is usually rigid. The anteroposterior x-ray exposes a middle phalanx articular surface that is inclined. The x-ray should be examined for a longitudinal epiphyseal bracket along the side of the phalanx (Fig. 7.33b) [3, 40]. This longitudinal bracket represents a functioning growth plate that retards longitudinal growth and may cause progressive angulation of the finger. The eventual shape of the phalanx depends upon the propensity of this abnormal growth plate to grow. Serial examinations and x-rays are necessary to determine its ultimate configuration [37, 39].

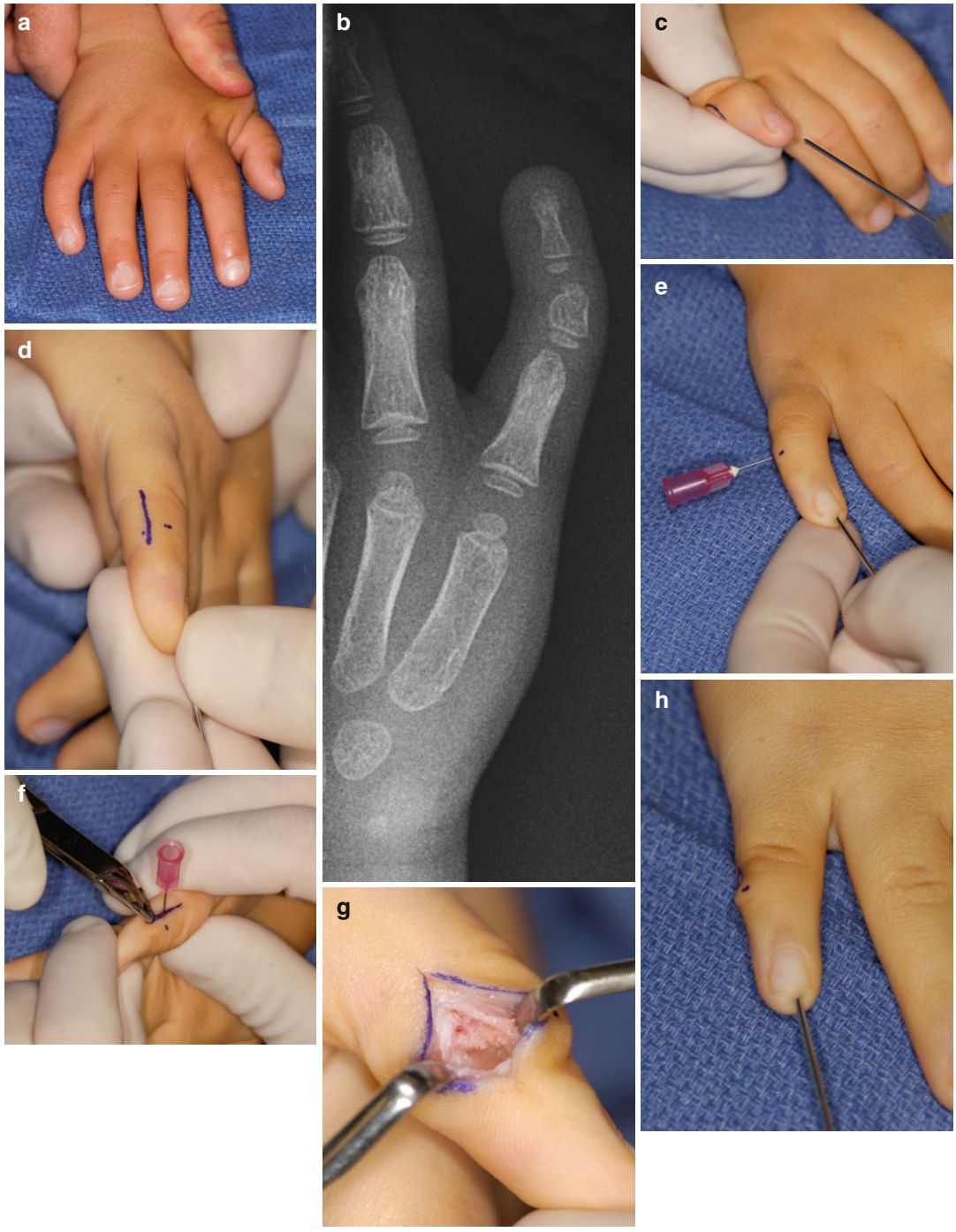

Fig. 7.33 4 year-old with bilateral small finger clinodactyly. (**a**) Clinical picture. (**b**) Anteroposterior x-ray shows an abnormal inclination of the middle. phalanx articular surface adjacent to the DIP joint. (**c**) A 0.045″ Kirschner wire is drilled from the fingertip across the DIP joint and into the middle phalanx. (**d**) Mid-lateral approach along the middle phalanx. (**e**) Physis is marked with a 25-gauge needle. (**f**) Removal of wedge with a small ronguer. (**g**) Close-up view of wedge removed. (**h**) The finger is manipulated and the longitudinal Kirschner wire advanced across the osteotomy site for fixation

Management

Nonsurgical Treatment

Idiopathic clinodactyly requires diagnosis and observation. Parents are often satisfied with the diagnosis and do not pursue additional treatment. Splinting is not recommended.

Surgical Techniques

Surgery is reserved for a substantial deformity that causes functional problems (e.g., digital overlap) or a progressive deformity attributed to a longitudinal epiphyseal bracket.

A variety of surgical techniques have been described to realign the digit. In idiopathic clinodactyly, a closing wedge osteotomy usually provides sufficient correction of the deformity [41]. The wedge is removed from the convex side of the middle phalanx. Our preferred method begins by drilling a 0.035 or 0.045″ Kirschner wire from the fingertip across the DIP joint and into the middle phalanx (Fig. 7.33c). The Kirschner wire is carefully positioned in the coronal and sagittal planes using mini-fluoroscopy. This wire defines the center of rotation of angulation (CORA) within the middle phalanx and the site for wedge osteotomy. The wire is partly withdrawn and a mid-lateral approach along the middle phalanx is performed (Fig. 7.33d). The physis is marked with a 25-gauge needle to avoid inadvertent injury (Fig. 7.33e) The extensor and flexor tendons are elevated and the wedge is removed with a small ronguer leaving the periosteum and radial cortex intact (Fig. 7.33f, g). The cortex is cracked via manipulation and the finger straightened. The longitudinal Kirschner wire is advanced across the osteotomy site to fixation (Fig. 7.33h). Fluoroscopy is used to check alignment of the digit and the position of the Kirschner wire.

Other osteotomy configuration have been described including opening and reverse wedge (resects a wedge from the convex side and inserts the segment into the concave side) configurations [37, 42]. These types of osteotomies are more cumbersome than a closing wedge and are seldom performed.

A prophylactic procedure has also been described to treat a longitudinal epiphyseal bracket that is causing progressive deformity [42–44]. Using a midlateral approach, the apex of the longitudinal epiphyseal bracket is exposed. The longitudinal portion of the bracket is excised and fat inserted to cover the ends of the divided physis. Over time, the digit slowly straightens as digital growth now occurs through the horizontal portions of the growth plate.

Outcome

The results of closing wedge osteotomy for idiopathic clinodactyly are uniformly good. This technique provides adequate correction of the deformity, improved hand function, and has high parental satisfaction [41]. The results of fat interposition into a longitudinal epiphyseal bracket appear better in children less than 6 years of age and with a trapezoidal shaped phalanx [43].

Complications

Complications related to wedge resection are infection, non-union, recurrence, joint or finger stiffness, and under- or over-correction. Fortunately, these complications are unusual. Idiopathic clinodactyly treated by wedge resection is a relatively straightforward operation. Wedge osteotomy could potentialy cause adhesions about the extensor or flexor tendons, although this is uncommon. The most common complication related to fat interposition is undercorrection, which necessitates an additional wedge osteotomy.

Pearls

- Idiopathic clinodactyly is attributed to an abnormal inclination of the distal articular surface of the middle phalanx
- Most clinodactyly does not require treatment
- Substantial angulation secondary to diopathic clinodactyly is best treated with a closing wedge osteotomy

Clinodactyly can also be caused by a longitudinal epiphyseal bracket across the middle phalanx

The behavior of the longitudinal epiphyseal bracket is variable and determined by serial examinations and x-rays

A longitudinal epiphyseal bracket that is causing progressive angulation is treated with bracket excision and fat interposition

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Polydactyly

8

Emiko Horii

Keywords
Congenital hand • Polydactyly • Duplicated thumb • Ulnar polydactyly

Introduction

Polydactyly is one of the most common congenital hand anomalies. Polydactyly that includes the thumb is referred to as radial polydactyly, whereas that involving the little finger is referred to as ulnar polydactyly. Central polydactyly, which is often associated with syndactyly, is now classified as abnormal induction of finger rays [29].

Radial Polydactyly

The primary reconstruction procedure for radial polydactyly varies according to the bifurcation pattern. Since the pathological anatomy of radial polydactyly has been clarified, reconstruction procedures have improved. However, since the duplicated thumb is inevitably hypoplastic, a secondary deformity may develop along with the child's growth. In order to meet the high expectations of parents and patients, the pros and cons of the various reconstruction procedures should be thoroughly discussed.

Background/Etiology

Incidence

Radial polydactyly is more commonly seen in an Asian populations, where 65 % of the patients are reportedly male. A family history is positive in 8–14 % [4, 6, 30, 33]. The incidence of bilateral involvement is reported as 8–16 %. Most cases are sporadic and unilateral. Approximately 20 % of patients have associated anomalies either generally or of an extremity, which is less frequent than in radial ray deficiency [6, 8, 23].

Etiology

Insults to the limb bud may occur by 8 weeks of gestation, when the differentiation of the thumb ray is completed. Experimental studies suggested that radial polydactyly develops as a result of a disorder of the interaction between the mesoderm and ectoderm in the pre-axial limb bud at the beginning of cell proliferation [27, 32].

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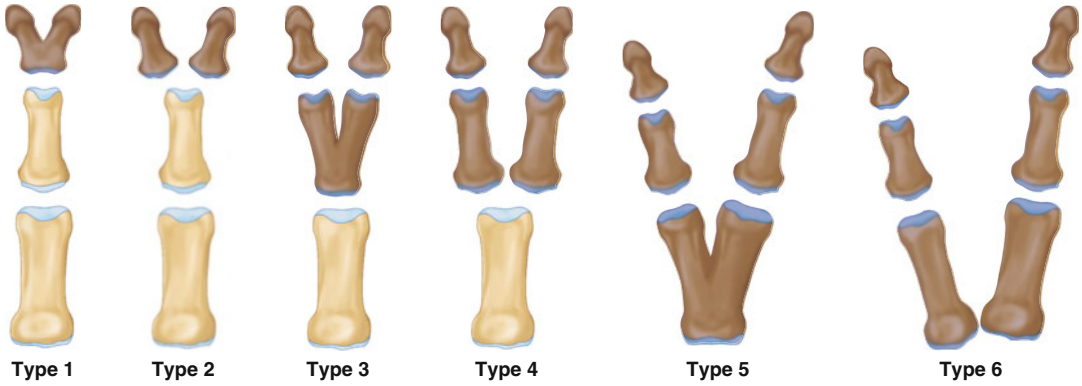


Fig. 8.1 Illustrations of Wassel's classification Type 1–6. *Type 1*: Bifid distal phalanx. *Type 2*: Complete duplication of the distal phalanx. *Type 3*: Bifid proximal phalanx. *Type 4*: Complete duplication of the proximal phalanx. *Type 5*: Bifid first metacarpal. *Type 6*: Complete duplication of the entire first digit

Classification

Radial polydactyly is commonly classified by Wassel's classification [35, 36], which divides it into seven types according to the branching level (Fig. 8.1). Type 4 is the most common, which involves approximately 45 % of cases, followed by Type 2.

Wassel's classification can however cause some confusion. First of all, the existence of the middle phalanx in either thumb puts the classification into type 7. However, the pre-operative radiograph, which is usually obtained on the immature infant, does not depict the epiphysial plate correctly. In addition, triphalangeal type polydactyly can appear at any level of duplication [14, 21, 37]. Therefore I feel that the Wassel type 7 sub-group is unhelpful in expressing the characteristics of radial polydactyly. The existence of three phalanges in the retained digit is only important accompanying information [38].

Secondly, Wassel's classification fails to provide sufficient details about the pathology of duplication. Type 4 polydactyly especially includes several important anatomical variables. Several reports have proposed a new classification for subdivisions with important implications for surgical management [3, 9, 13] although there is, as yet, no universal agreement about these subdivisions.

Thirdly, several types of radial polydactyly are difficult to classify, such as a 'radially deviated type' reported by Ogino [28], 'floating type', and polydactylies associated with other hand anomalies. Abnormal pathology differs in those cases, so reconstruction procedures and surgical outcomes also vary. It might be appropriate for these cases to be grouped as miscellaneous.

Preoperative Examination

Physical Examination

The key to the treatment of radial polydactyly is awareness of the pathology [5, 6, 23]. Preoperative patient examination is very important. The size of the thumb to be conserved is compared to that of the contra-lateral normal thumb. Symmetry of the pulp and the paronychium are assessed, and active and passive range of motion is measured on the thumb to be conserved. Excursion of the extensor pollicis longus (EPL) and flexor pollicis longus (FPL) can be estimated based on the IP joint motion and finger creases both dorsally and volarly.

Radiological Examination

The radiographic evaluation should be done on a carefully obtained postero-anterior view of the thumb. Branching level, the shape and size of each bone, and bone alignment are assessed on the radiograph in comparison to the contra-lateral thumb. Usually, the epiphyseal plate of the distal phalanx is not ossified before age 1. Early ossification might represent an abnormal epiphysis, which may cause an unstable or stiff IP joint.

Timing of Surgery

The timing of surgery depends on the physical condition of the patient and any co-morbid conditions, the maturation of bone, and branching type. When osteotomy or soft tissue augmentation is necessary, the surgery should be delayed. But, if it is delayed too long, the supernumerary component displaces the normal component. The surgery is usually completed before the age of 2. The author prefers performing the surgery at age 8–12 months. The best timing may be determined depending on the surgeon's experience [30].

Explanation to Parents

The family should be informed of the complexity of the surgery. It is important to emphasize the high re-operation ratio to correct deformities that may develop as the thumb grows [20, 25, 33]. Moreover, surgeries do not create a thumb identical to the contra-lateral thumb. When bilateral thumbs are compared side-by-side, a reconstructed thumb is usually visibly different, and has less motion.

Clinical Pearl

Wassel classification useful, but understanding of complex pathology more important for surgery planning

Surgery usually between age 8 months −2 years, older if thumb more complex

Secondary surgery common

Reconstructed thumb always smaller and less mobile than contra-lateral thumb

Stability more important than mobility

Surgical Techniques

In principle, the more hypoplastic and less functional thumb (usually the radial thumb) is removed. Simple ablation of the extra thumb is no longer acceptable [7]. Reconstructive surgery is always indicated for functional benefit and to improve the appearance of the reconstructed thumb. A deviation of the digital axis might be caused by crooked bones, tilt of articular surface, lax collateral ligaments, or abnormal insertions of tendons. These must be rebalanced at the initial surgery as much as possible. A fundamental error is to assume that minor deviations will correct themselves after resection of supernumerary digit. The aims of the initial surgery are: (1) to reconstruct a joint that is stable and mobile; (2) to align the joint surfaces perpendicular to the long axis, (3) to centralize the flexor and extensor tendons; and (4) to transfer the thenar muscles, and hence restore normal anatomy to the remaining thumb.

Details of surgical procedure for type 4 polydactyly is described first. Then several particular procedures for each type are described separately.

Type 4: Proximal Phalangeal Type

Since Wassel recommended avoiding the linear incision, a dorsal zigzag incision has been widely used [35]. However, this leaves a poor scar over the dorsal thumb. The author prefers the radial mid-lateral incision (Fig. 8.2b). In case the skin fillet from the radial thumb is necessary, the dorsal curved incision and the volar zigzag incisions are incorporated (Fig 8.3b). A small Z-plasty is added, if necessary. Usually there is a single artery for each thumb [18], so it is unnecessary to pay careful attention to the anatomy of the digital nerve and artery.

The EPL/FPL tendons usually show Y-shape branching, and the radial one is simply excised (Fig 8.2c). The tendon excursion should be examined. Eccentric tendon insertion, if such exists, causes deviation and restriction of IP joint motion, so insertion should be centralized. After the radial digit is excised, the ulnar digit is centralized. Although the metacarpal head is wide, shaving it is usually unnecessary. The radial collateral ligament can be reconstructed by a ligamentous/periosteal flap from the excised radial digit [22]. The thenar muscles, which usually insert into the radial digit, are detached, then

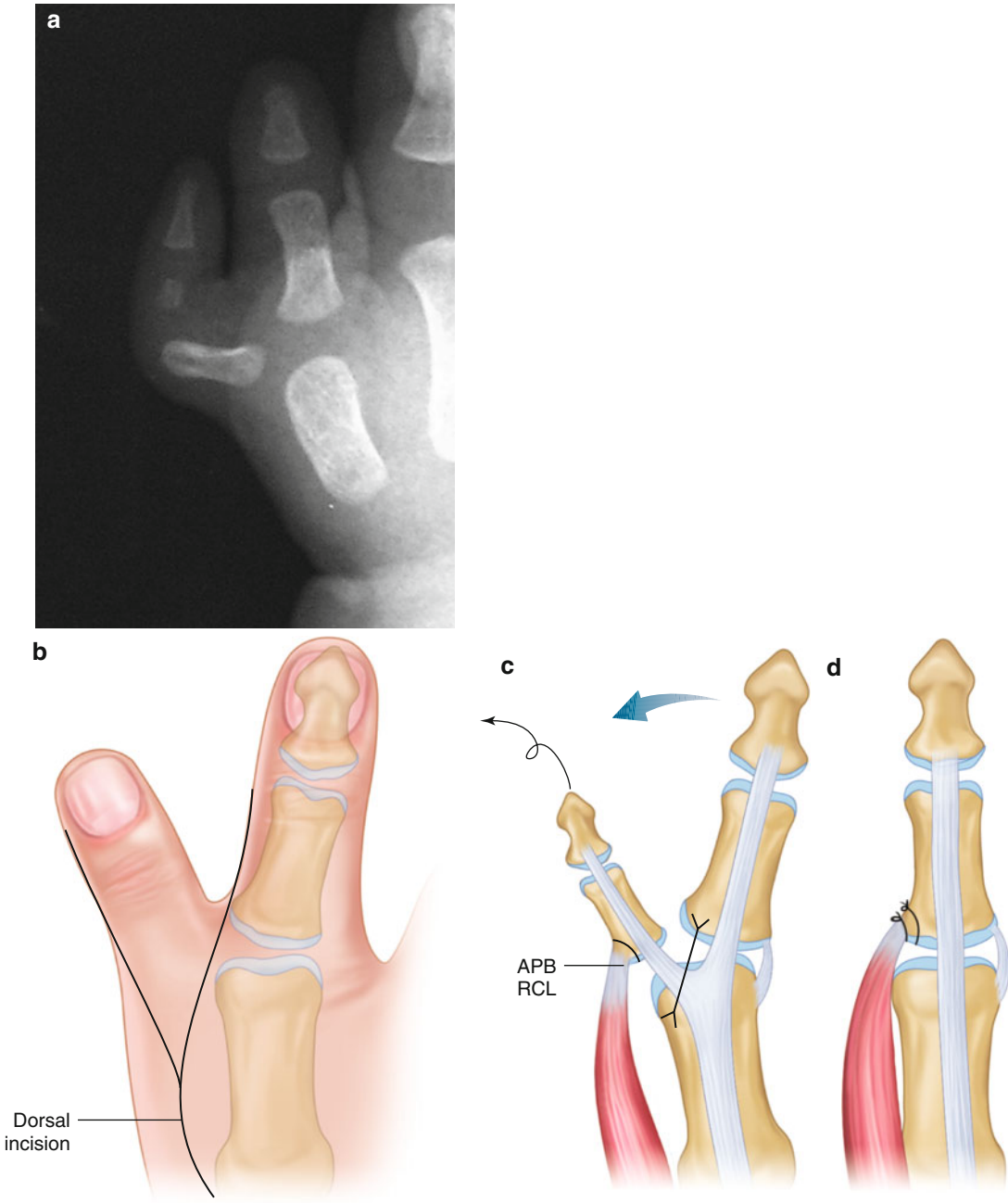


Fig. 8.2 Surgical technique for type 4 radial polydactyly in case the ulnar thumb is large. (**a**) PA view of radiograph. The ulnar digit is larger than the radial digit, and the axis of the ulnar digit is almost straight. In this particular case, the radial digit has a middle phalanx. (**b**) The radial curved incision is placed. (**c**) The radial EPL tendon branching at the MP joint level is excised, then the tendon excursion of the ulnar thumb is evaluated. The FPL tendon excursion is also examined; centralization of the FPL tendon is occasionally necessary. The MP joint is explored, and the radial collateral ligament with thenar muscle insertion is detached from the radial digit. (**d**) After the excision of the radial thumb, the ulnar thumb is centralized. The radial collateral ligament and thenar muscles are sutured to the radial side of the proximal phalanx

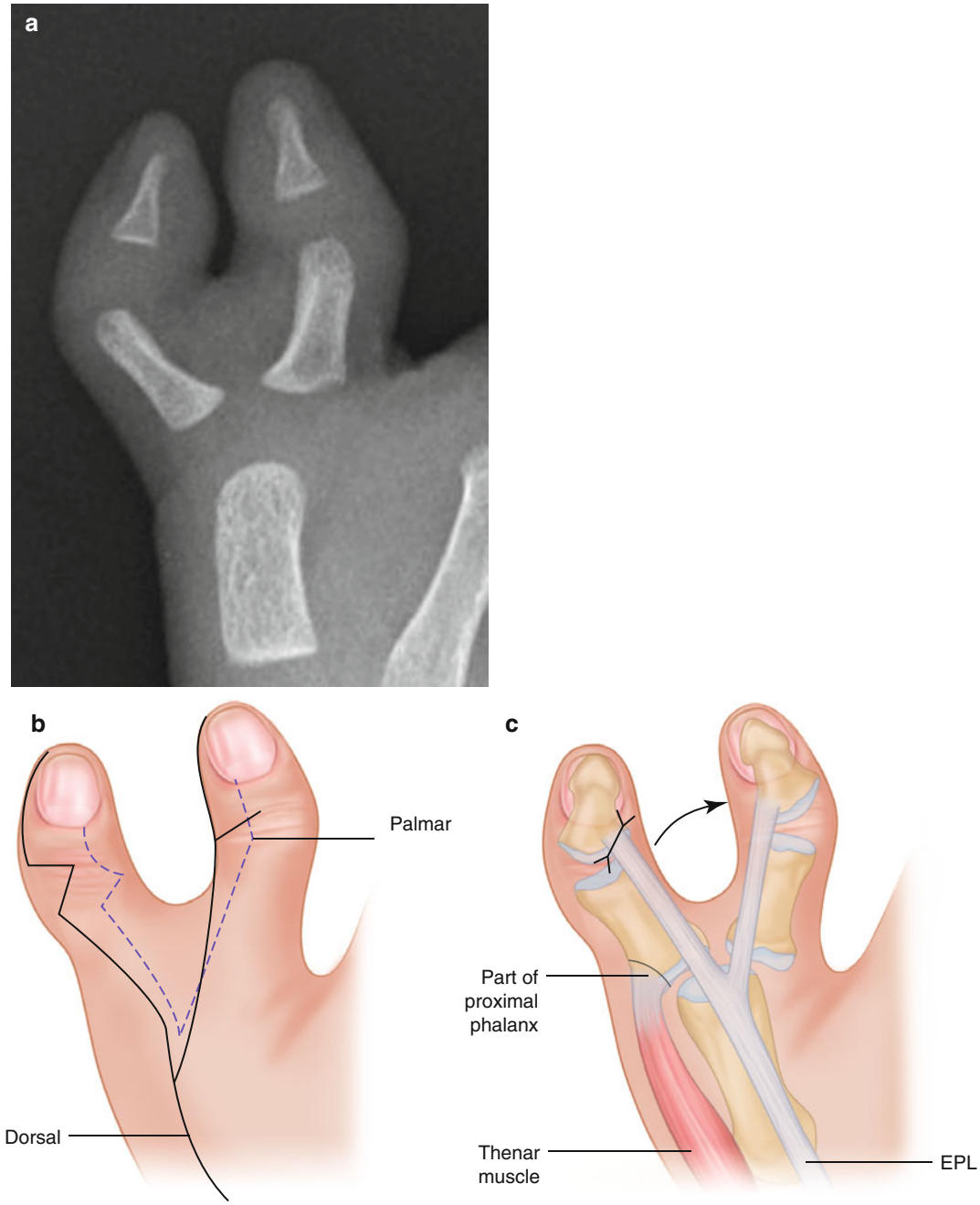
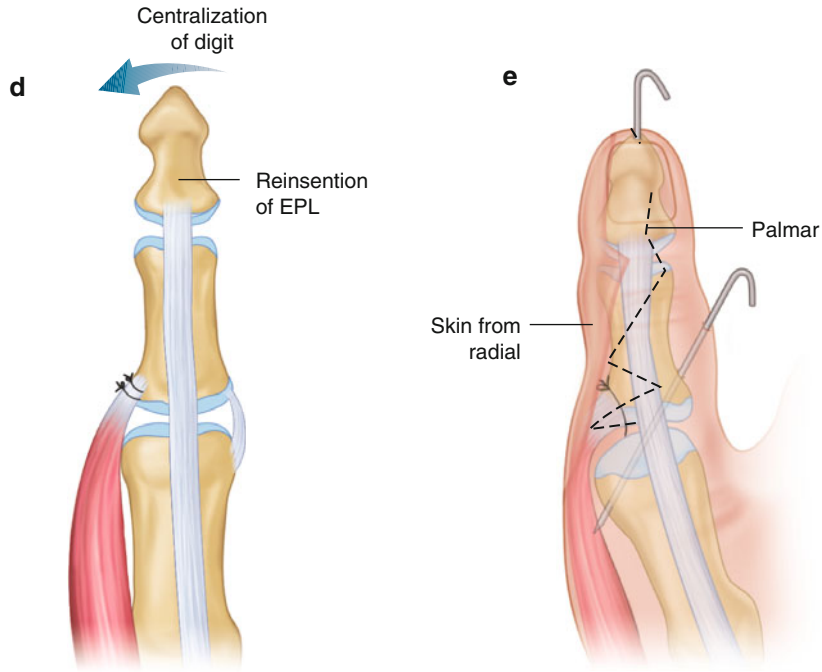


Fig. 8.3 Surgical technique for type 4 in case both digits are small. (**a**) Radiograph shows the proximal phalanges are divergent and both thumbs are small in size. (**b**) Skin incision. In order to maintain the girth of the ulnar digit, the fillet from the radial digit is preserved. A small zigzag incision or Z-plasty is added to the dorsal curved incision. (**c**) The EPL tendon shows V shape and is inserted into the lateral border of the distal phalanx. The EPL tendon of the radial digit is transferred to the ulnar thumb. The FPL tendon usually shares a common pulley in both thumbs. The pulley is cut and the radial FPL tendon is excised. The osseous/ligamentous flap from the base of the radial proximal phalanx is elevated with thenar muscle. (**d**) The radial digit is excised, then the ulnar digit is centralized. The osseous/ligamentous flap with the thenar muscle is transferred to the base of the proximal phalanx of the ulnar thumb in order to secure the MP joint stability. The insertion of EPL and FPL are centralized, then the IP joint is balanced. (**e**) The skin fillet from the radial digit supports the radial side of the reconstructed thumb. The IP and MP joints are temporally fixed by a k-wire

Fig. 8.3 (continued)



transferred to the base of the proximal phalanx of the ulnar digit or neck of the metacarpal.

Preoperatively, the thumb to be preserved occasionally shows lateral inclination. This should be corrected at the time of primary surgery. In these cases, in which the shafts of phalanges or metacarpals are obviously angulated, corrective osteotomy is considered. Type 4 polydactyly, however, seldom requires osteotomy. The majority of lateral bending deformities can be corrected by rebalancing the tendons and radial collateral ligament of the metacarpophalangeal joint. Failure to identify and correct these abnormalities is likely to result in abnormal dynamic action of the tendons which could lead to instability or angulation of the joints as the patient grows. Too much shaving of the metacarpal head, reduction osteotomy to reduce the width of bones, or inappropriate correction of bone axis might cause gross instability of the thumb.

Evenly Developed Type

When the thumbs are of nearly equal size, usually both thumbs are hypoplastic. There are several options for reconstruction; the first, excision of the radial digit and reconstruction of the ulnar digit, results in a smaller thumb. The second option is the Bilhaut procedure, which excises central parts of bone and soft tissues [2]. In the third option, the soft tissues from the excised thumb, are used to increase the size of the reconstructed thumb (Fig. 8.3) [1, 10].

The reconstructed thumb is usually smaller in comparison to the normal thumb. The appropriate size of the reconstructed thumb is unclear. Dobyns recommended having at least 80 % of the normal size [6]. Tonkin stated that when the nail width is less than 70 % of the contra-lateral side, nail fusion should be performed [34]. The Bilhaut technique might be applicable, if both nails are very small. There are two criticisms of the Bilhaut procedure: firstly joint stiffness and physeal growth disturbance are the major concern in terms of function. But, the diminished joint motion may be a lesser problem than joint instability. The second concern is nail deformity with a central ridge and broad thumb [22, 25, 35]. Meticulous nail fusion techniques have been reported, but this option is still unpredictable and requires a high learning curve [15, 34].

In order to resolve these problems, Baek et al recommended the extra-articular procedure in order to avoid joint stiffness and damages to physes [1]. The author prefers to reconstruct the ulnar digit by using soft tissue flaps from the radial digit [10, 26]. The bony mal-alignment is corrected by osteotomy at the proximal phalanx, if such exists. The joint stability is retained by a large osseous/ligamentous flap from the radial digit, and the tendons are also augmented by the radial tendon. The skin fillet from the radial digit supports the radial side of the reconstructed thumb. When the IP joint of the retained thumb is extremely unstable, primary chondrodesis is considered.

Wassel 1 and 2 (Distal Phalangeal Type)

Type 2 is the second common type of radial polydactyly. The basic surgical principle is similar to that of type 4. After the excision of the lesser radial digit, the retained digit is centralized, then the radial collateral ligament is secured by using a ligamentous/periosteal flap [22] (Fig. 8.4). Care should be taken not to perform too much shaving of the joint cartilage. Hence, in order to secure the joint stability, the IP joint is temporally fixed by a k-wire for 3 weeks. Osteotomy at the level of the proximal phalanx is usually unnecessary. The surgical scar around the nail is always noticeable, much attention should be paid to creating a symmetrical finger tip and paronychium, and avoiding scars.

Wassel 3

Type 3 is the most difficult type of polydactyly [19, 20]. Both digits are usually small and have inherited IP joint instability (Fig. 8.5). The Bilhaut technique is occasionally applied for a very small nail [15, 34]. The author prefers radial digit excision with augmentation by bone and soft tissue from the excised digit1 [4] (Fig. 8.5b). The EPL tendon is transferred from the radial digit to the ulnar one, and the FPL tendon is centralised if necessary. When the retained IP joint is extremely unstable with a hypoplastic EPL tendon, then primary IP joint chondrodesis is considered.

Wassel 5 and 6 (Metacarpal Type)

In type 5 and 6, the pathology is more complicated. The ulnar thumb is often in an adducted position, and bone mal-alignment exists preoperatively (Fig. 8.6). The thenar muscles are often hypoplastic, and the MP joint is unstable.

After excision of the radial digit, the stability and position of the retained thumb should be assessed intra-operatively. For correction of the adduction, the metacarpal is either osteotomized or transferred onto the base of the radial digit [31]. Deviation of bone axis and supination of the retained thumb are correctable at the osteotomy site. The first web contracture is released, if it exists, and Z-plasty is occasionally performed. For severe adduction contracture, Ogino recommended use of a rotation flap from the removed digit [31]. When the MP joint is very unstable, the reinforcement of the ulnar ligament is necessary. The thenar muscles detached from the radial digit are carefully transferred to the neck of the metacarpal instead of the base of the proximal phalanx.

Clinical Pearl

Radial digit usually planned for excision
Transfer of thenar muscles to retained digit and augment soft tissue and skin if needed
Reconstruct radial collateral ligaments with ligamentous/periosteal flap
Centralize tendons to retained digit; consider joint chondrodesis if tendons poor/absent
Osteotomy not usually necessary

Radially Deviated Polydactyly

Although it is rare, radially deviated polydactyly exists as reported by Ogino [28]. The pathology is not clear, but the proximal phalanx is usually short and deformed (Fig. 8.7), and the IP joint of the retained ulnar digit shows severe deviation. The reinforcement of the ulnar collateral ligament of IP joint and repositioning of tendons are not enough to correct the deformity and maintain

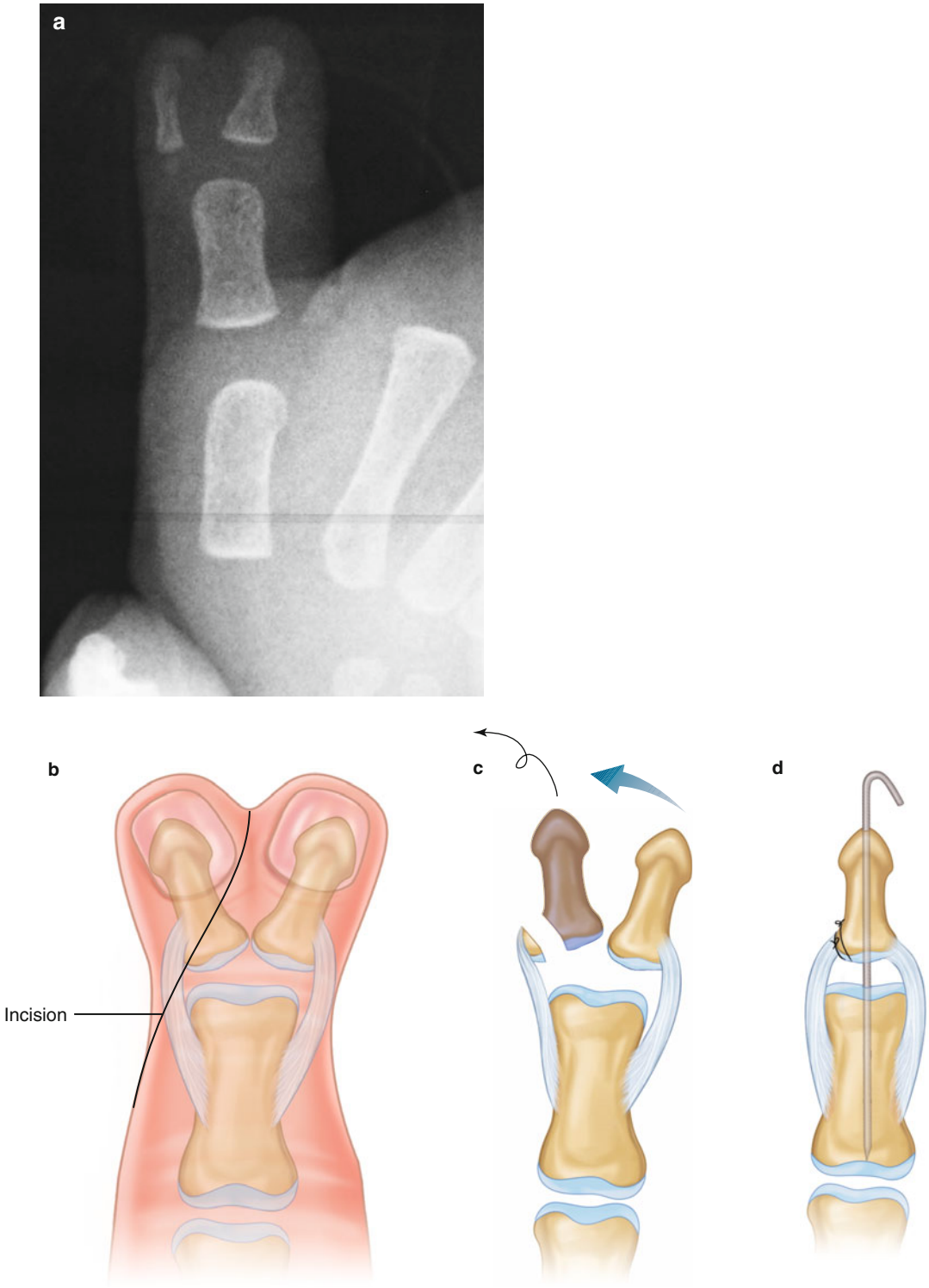


Fig. 8.4 Surgical technique for type 2 radial polydactyly. (**a**) Radiograph. (**b**) Skin incision. Care should be taken to created symmetric pulp and paronychium. (**c**) Excision of the radial digit. The radial collateral ligament with periosteal flap is detached from the radial digit. (**d**) The ulnar digit is centralized, and the ligamentous flap is secured to the radial base of the ulnar digit. A temporary K-wire fixation is performed

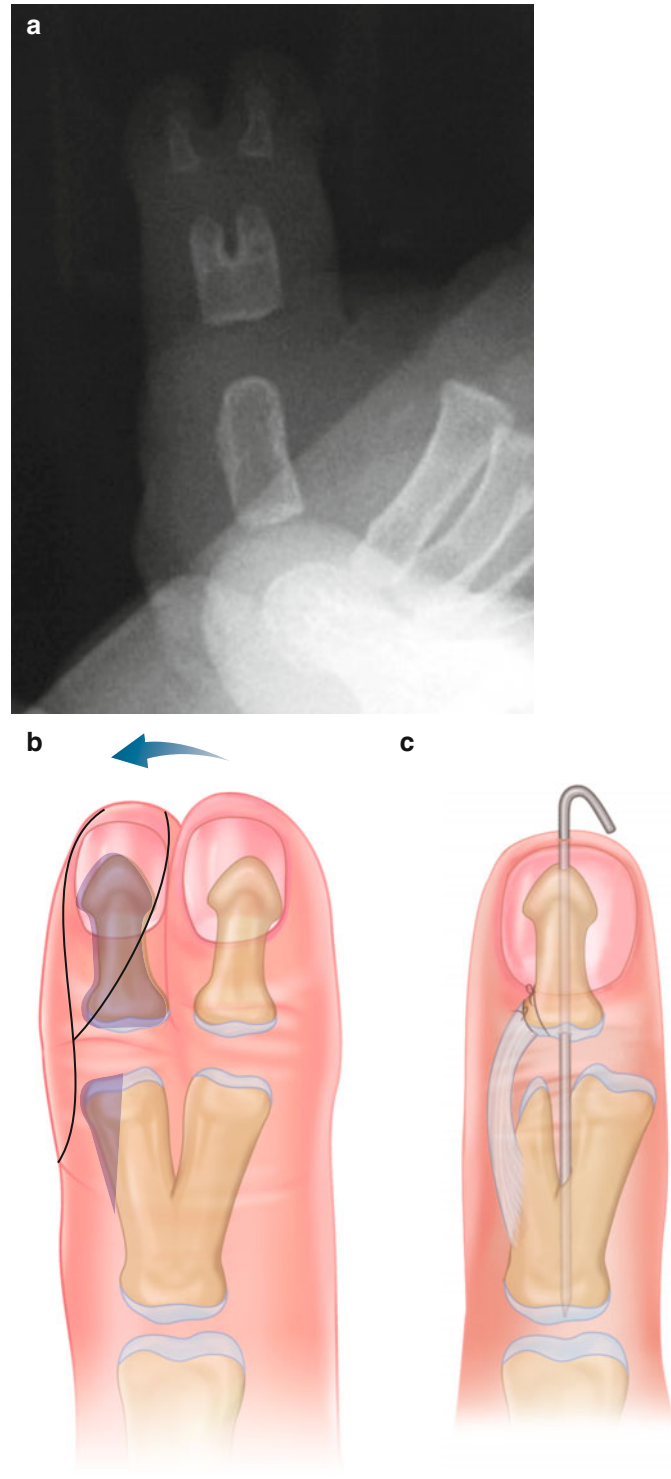


Fig. 8.5 Surgical technique for type 3 radial polydactyly. (**a**) Radiograph shows both digits are small and the IP joints are narrow. (**b**) The radial lateral incision is used to explore. The radial digit is partially excised, but the MP joint is left untouched. (**c**) Centralization of preserved distal phalanx is performed. Both the EPL and FPL tendons are also rebalanced, and the skin fillet from the radial digit supports the ulnar digit

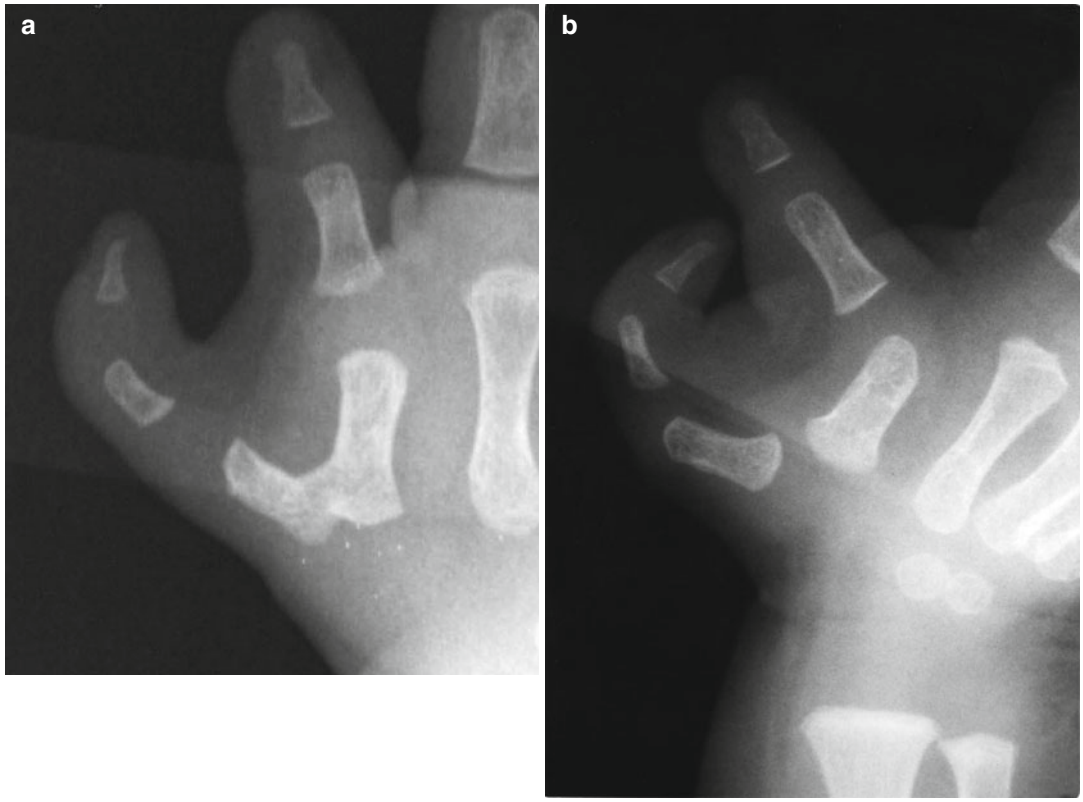


Fig. 8.6 Radiographs of type 5 and 6 radial polydactyly. The PA view is carefully examined to evaluate the bone alignment. (**a**). Type 5 polydactyly shows the ulnar digit has good alignment. (**b**) The ulnar digit in type 6 polydactyly shows adduction of the metacarpal and lateral bending at the MP joint

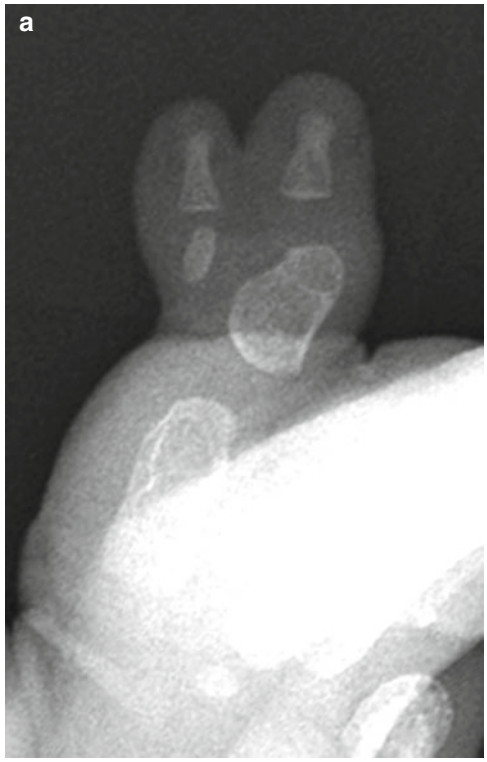
stability. In order to correct the digital axis, an open wedge osteotomy is necessary at the initial surgery, and then the bone harvested from the excised digit should be grafted. When the IP joint itself is narrow and unstable, primary chondrodesis might be necessary. The correction of the digital axis is maintained by k-wire fixation, which may be removed in 4–5 weeks.

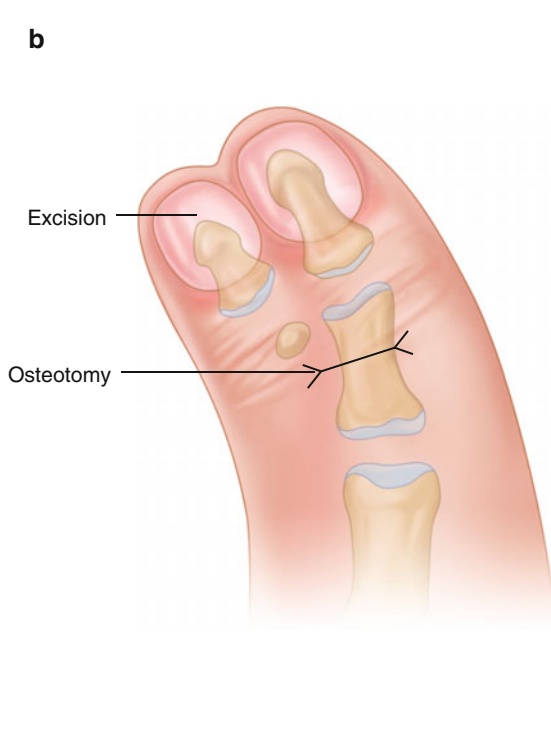
Other

Radial polydactyly, which associates with other anomalies, shows a different bifurcation pattern (Fig. 8.8). Occasionally the ulnar digit is hypoplastic, then the radial digit is preserved. Reconstructive surgery should be performed according to the given pathology.

Association of Triphalangeal Thumb

The triphalangeal type can be observed in any type of radial polydactyly. Existence of a middle phalanx is often not elucidated on the preoperative radiograph. Clues are the early appearance of the epiphysis or a too-wide space at the IP joint (Fig. 8.9). When there is any doubt of the existence of a middle phalanx in the retained thumb, surgery should be postponed until confirming the shape of the distal phalanx epiphysis. When the middle phalanx exists, it should be excised at the initial surgery. Care should be taken to repair the collateral ligament, and a temporary k-wire fixation is necessary after excision [10, 12]. When the distal phalangeal epiphysis is cone-shaped, it might cause stiffness or an unstable IP joint.





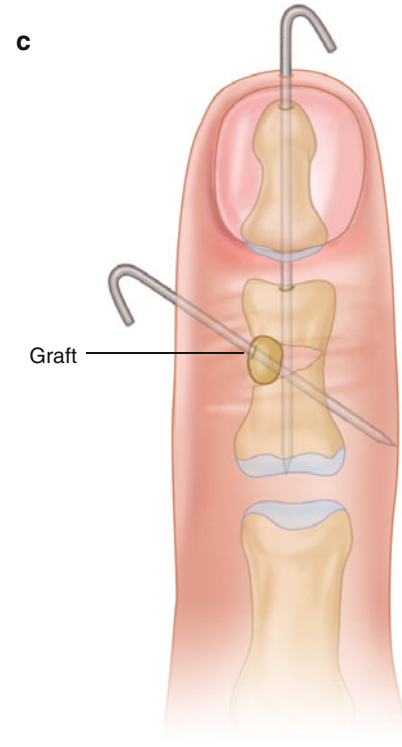


Fig. 8.7 Surgical technique for radially deviated polydactyly. (**a**) Radiograph; the proximal phalanx is deformed, and the IP joint of the ulnar thumb shows severe radial deviation. (**b**) The radial digit is excised, and open wedge osteotomy of the proximal phalanx is performed. (**c**) The bone chip from the excised digit is grafted, and fixed with k-wires

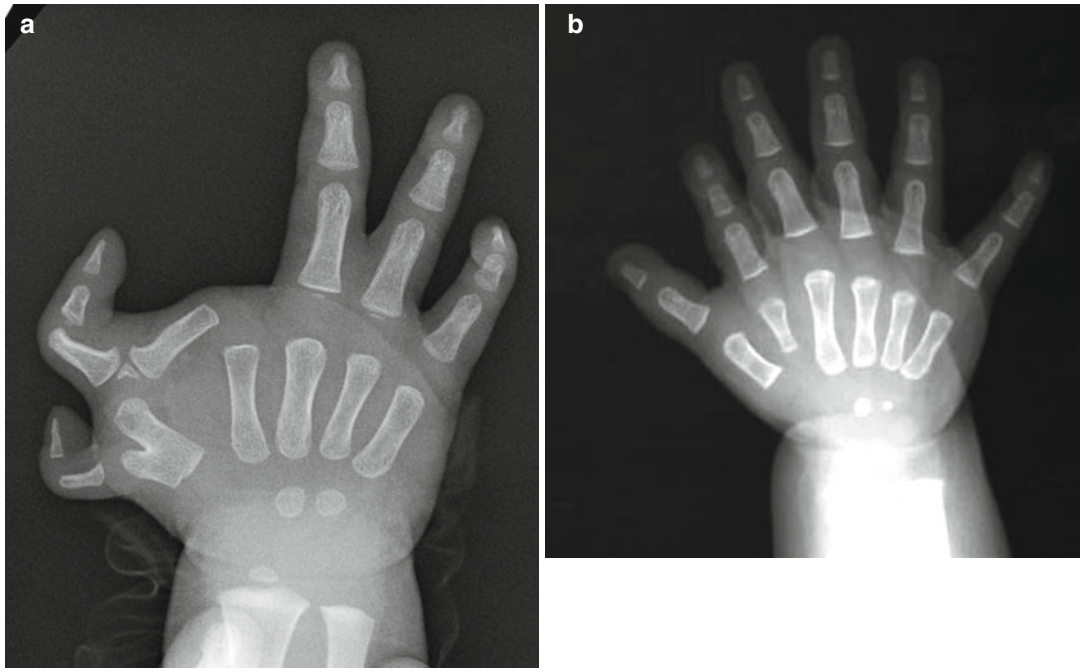


Fig. 8.8 Unclassified radial polydactyly. (**a**) The radial polydactyly is associated with the cleft hand. (**b**) This patient had bilateral radial polydactyly, and had a positive family history. The patient also had bilateral preaxial polydactyly in foot. The ulnar thumb is hypoplastic, then excised

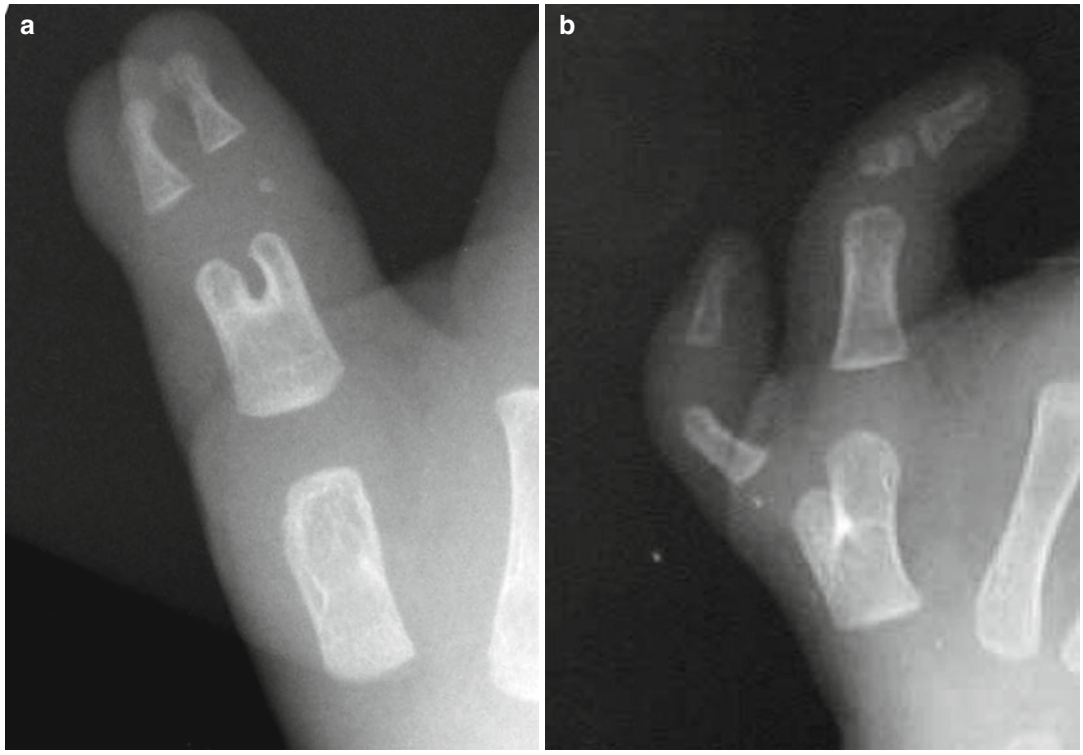


Fig. 8.9 Reconstruction of the triphalangeal thumb. (**a**) Type 3 radial polydactyly at 6 months of age. The ulnar thumb has a small ossification between the phalanges, which will turn out to be the middle phalanx. (**b**) Type 5 radial polydactyly has a middle phalanx in the ulnar digit

However, this cannot be resolved at the time of initial surgery. Stability of the IP joint should be carefully followed and either chondrodesis or osteotomy may be indicated later.

Postoperative Care

Place the patient's thumb under a bulky dressing for 3 weeks postoperatively. The author prefers using bandage fixation instead of a cast or splint. The dressing is changed at 3 weeks, then the temporary k-wires are removed at 3–4 weeks. Patients are allowed to freely use their hands after 4–5 weeks.

Evaluation

Tada's criteria are most often used for evaluation of the radial polydactyly. This evaluation system includes only objective assessments. Several evaluation systems have been proposed, including not only more detailed function but also the cosmetic evaluation [11, 13]. The author prefers the evaluation system proposed by the Japanese Society for Surgery of Hand [11], which is relatively easy to assess (Table 8.1).

Outcomes and Complications

The surgical outcomes vary with the degree and complexity of the thumb duplication. The reconstructed thumb is always smaller and has less motion than the normal side. Satisfactory results are readily obtainable in types I, II, and IV, and unsatisfactory results are prevalent in types III, V, and VI, and association with triphalangeal thumbs [20]. The majority of papers reported 80 % of the results were good (Table 8.2). However, Larsen [19] et al indicated that only 26 % of their cases had good results with an average of 22 years follow-up. They found that there was a higher incidence of instability with long-term follow-up, and explained the discrepancy from other studies indicating that stability decreases over time after ligament reconstruction. However, they could evaluate only 10 % of their cases.

Table 8.1 Assesement for radial polydactyly

Function			Point
Abnormal alignment	***IP joint***	<5°	2
		6–20	1
		20°<	0
	MP joint	<5°	2
		6–20	1
		20°<	0
Instability	***IP joint***	<10	2
		11–19	1
		20°<	0
	MP joint	<40	2
		41–59	1
		60°<	0
Acitve flexion	***IP+MP joint***	90<	2
		60<	1
		<60	0
Extension lag	***IP+MP joint***	0°	2
		<30	1
		30<	0
Palmar abduction	***MP+CM joint***	60<	2
		31~59	1
		<30	0
Cosmesis	**Size**	Acceptable	1
		Unacceptable	0
	Finger pulp/ nail	Acceptable	1
		Unacceptable	0
	Surgical scar	Acceptable	1
		Unacceptable	0
	Bulging	None	1
		Outstanding	0
Subjective assessments			
	Pain	None	1
		Painful	0
	Patients' satisfaction	Satisfactory	1
		Unsatisfactory	0
Total assessments			Points
		Excellent	20
		Good	17–19
		Fair	14–16
		Poor	0–13

By JSSH [11]

Reported reoperation rates are between 20 and 25 % [20, 24, 26]. Any offset in bone axis, persistent joint instability, or tendon imbalance

Table 8.2 Surgical outcomes in literature (evaluation criteria differs in each report)

Study	Avg. F-up (years)	Good	Fair	Poor
Tada et al. (1983) [33]	2.5	81 (%)	14	5
Cheng et al. (1984) [3]	3.6	86	9	5
Townsend et al. (1994) [35]	9	48	52	
Ogino et al. (1996) [30]	4	85	11	4
Horii et al. (1997)[a] [9]	8.1	72	20	8
Larsen et al. (2005) [19]	22	26	63	11

[a]Only type 4

will lead to problems over time. Angulation, joint instability, limited motion, and scar formation are common problems. There are two kinds of complications: one can be prevented if a surgeon knows the pathology well, while another is attributed to the hypoplasia of the thumb itself, in which case good outcomes are hard to obtain by any procedure.

Tada reported that one third of thumbs did not show any improvement by revision surgeries, and final results were good in 63.9 % of the cases [33]. Kawabata also reported that all of the cases improved after revision surgeries, but good results were obtained in 56 % [16].

Lateral Bending Deformity

Even though it is subtle, the radial deviation of the thumb becomes worse over time. It should be corrected as early as possible when the pathology of the bending moment becomes clear. The radial linear scar can be released by Z-plasty or a small rotation flap. Corrective osteotomy is effective when the bone axis is deviated. The dynamic bending force by tendons can be corrected by repositioning of tendon insertion. When the radial deviation occurs at either the MP or IP joint, which may then be associated with joint instability, then correction is not easy. Efficacy of an osteotomy, with reinforcement of the ligament, is limited. Arthrodesis is occasionally necessary.

On the other hand, if the ulnar bending of the thumb is present this will usually not progress, and surgical treatment is usually unnecessary. If it causes cosmetic problems, it might be corrected after bone maturation.

Instability

The pathology causing joint instability is not simple. Instability is usually attributed to subtle joint incongruity. Despite great effort at stabilizing and rebalancing the thumb at the initial surgery, inherit joint incongruity is difficult to stabilize [16, 17]. The effect of revision surgeries such as ligament reinforcement or tendon transfer, is limited. Persistent instability will cause a painful thumb with weak pinch or grasping function. If it becomes symptomatic, arthrodesis is the only way to solve the problem.

Restricted Motion

The reconstructed thumb usually has less motion in comparison to a normal thumb. However, restricted motion rarely causes functional limitations. The primary concern in the majority of patients is cosmesis [6]. Therefore, extension lag due to EPL insufficiency is one of the major complaints. In these cases, the EPL tendon is very thin and weak distal to the MP joint level. The efficacy of EPL reinforcement is limited. Tenolysis in order to increase motion is also not effective in the majority of cases.

Personal View

Since the pathological anatomy of radial polydactyly has been clarified, the incidence of severe residual deformity has decreased. But, patients' and parents' desire to have a normal thumb is still strong. In addition, when each component is very hypoplastic, reconstructed thumb is also hypoplastic in terms of function and cosmsis. Consequently, the incidence of revision surgery is still high. Good outcomes are difficult to obtain in cases having: (1) poor joint congruity in IP or MP joint; (2) hypoplastic EPL tendon; and (3) too small nail.

I prefer to reconstruct the radial polydactyly at 8–12 months, if it is a relatively simple duplication. But if the retained thumb seems very immature and needs additional osteotomy or soft tissue augmentation, I prefer to wait until

structures grow sufficiently. When reconstruction of the thumb is very complicated and hypoplastic, I seek to reconstruct a stable thumb with good alignment. Since the long-term result shows that joint instability develops with patient's growth, the reduction osteotomy to narrow the joint is seldom performed. Although the reconstructed thumb looks slightly wide for some time after surgery, cosmesis improves with growth. Maintaining good stability while the patients are growing is the key to long-term success in the reconstruction of radial polydactyly.

Ulnar Polydactyly

Ulnar polydactyly is often inherited via an autosomal dominant pattern with variable penetrance pattern, and is more common in Africans and African-Americans. It occasionally appears in syndromes, such as Ellis-van Creveld syndrome (Table 8.3). In comparison to radial polydactyly, this anomaly is often associated with other skeletal anomalies, such as syndactyly or polydactyly in feet.

Ulnar polydactyly is classified into type A (well developed) (Fig. 8.10) and type B (rudimentary) (Fig. 8.11). A prospective screening research study in California reported that a prevalence of type B is one in 531 live births, 76 % were bilateral, and 86 % had a family history [39].

Treatment

Ulnar polydactyly seldom causes hand dysfunction. A small post axial element in type B (Fig 8.11) can be safely removed by tying the base of the pedicle. The digit will become necrotic and fall off in a week although a residual bump is a common complication of this treatment. For treatment of type A, transfer of the abductor digiti minimi, ligamentous enforcement, or corrective osteotomy is occasionally necessary. Restricted motion or bending deformity in the remained little finger might occur in particular cases.

Table 8.3 Syndromes occasionally associated with ulnar polydactyly

Ellis-van Creveld syndrome
Rubinstein Taybi syndrome
Bardet-Biedl syndrome
Trisomy 13, 18, D-1 syndrome
Jeune Thoracic dystrophy
Short rib-polydactyly syndrome
Acrofacial dysostosis
Oral-facial-digital syndrome

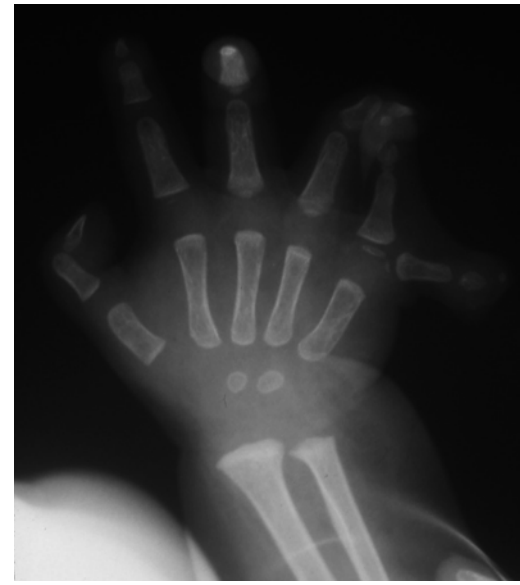


Fig 8.10 Type A ulnar polydactyly. The patient had had bilateral ulnar polydactyly

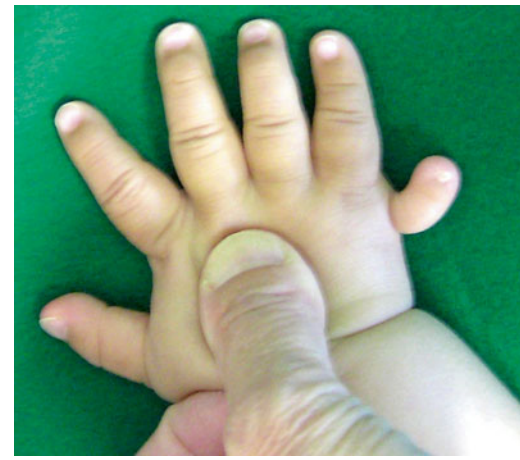


Fig 8.11 Type B ulnar polydactyly. This type of polydactyly is often ligated at birth. The functional loss is usually minimal

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Upper Limb Overgrowth

9

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Keywords

Overgrowth • Macrodactyly • Hemihypertrophy • Lymphoedema • Vascular malformation • Haemangioma • Macrodystrophia lipomatosa • Neurofibromatosis • Proteus syndrome • Amputation • Toe transfer

Overgrowth of the upper limb is a description of a category of conditions with the clinical finding in which the limb or a component of the limb develops in size beyond that expected for an individual's age and sex. This can be due to excessive growth of the anatomical structure or due to an abnormal accumulation of some substance. These groups of conditions are relatively rare and diverse in presentation and aetiology; so presenting the clinician with difficult challenges. Lister suggested that the condition diminishes even the giants of hand surgery [1].

Excessive growth in an anatomical structure can be due to an enlargement of specific tissue component within the limb such as vessels resulting in haemangioma or lymphangioma or fat resulting in lipoma. Alternatively they may arise as a consequence of enlargement of all or most of the tissue components in varying amounts. The overgrowth can present as diffuse overgrowth of the whole limb or of a segment of a limb or may be a localized as a tumour like growth.

The condition can be congenital or acquired. Acquired causes of overgrowth include tumour, acromegaly, lymphoedema and amyloidosis. For congenital conditions it may occur clinically in isolation or as part of a wider syndrome. Syndromes noted to be associated with overgrowth include Neurofibromatosis – type 1, Ollier's syndrome, Maffuci syndrome, Proteus syndrome or syndromes associated with complex vascular malformations such as Klippel Trenaunay syndrome or Parkes Weber Syndrome. Those congenital conditions occurring in isolation rarely have any hereditary association.

In this chapter we focus on congenital causes of overgrowth, highlighting the clinical pictures that occur and suggesting appropriate investigations. We examine the pathogenesis of the conditions and look at the treatment.

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Aetiology

The origin of overgrowth is multi-factorial. The cause of isolated overgrowth is unknown; a genetic link has not been established. Hereditary tendency to limb overgrowth and associated conditions is well recognized and has been described even in antiquity. Many of the Parthian Kings suffered from a skin disorder now thought to be neurofibromatosis. They were descendants of ancient Persian emperors, one of whom was Artexerxes (grandson of Darius the Great) also known as μακρόχειρ Macrocheir (Latin: 'Longimanus'), because his right hand was longer than his left. This suggests the earliest link between a hereditary neurofibromatosis and limb overgrowth [2, 3].

In isolated limb overgrowth abnormal nerve physiology and signaling during development appears to be a contributor to the condition. Recent studies propose a mechanism involving nerve regulated growth pattern of bone and soft tissue [4, 5]. Neural induction patterns demonstrated by animals regenerating limbs may have a role. Kumar et al. reported the discovery of newt Anterior Grading protein (nAG) that seems to be sufficient for induction of limb regeneration. nAG is found around the Schwann cells surrounding the injured nerve, where it induces the steps required for limb regeneration. It is possible that a similar pattern may be true for peripheral nerves that innervate the developing limb and that deregulation of these factors during the growth process may contribute to limb overgrowth [6].

The clinical distribution of isolated overgrowth along the median nerve distribution along with radiological and surgical observation of fatty infiltration of the nerve leads to the hypothesis that this may be a result of an abnormal nerve physiology [5, 7].

The dense innervation of bones also supports the neural control of bone metabolism and subsequent growth [8]. The connection between neural pathways with growth regulation may be one of the factors resulting in the clinical observation of reduced limb length following Obstetric Brachial Plexus Palsy however the cause of this is inevitably multifactorial with limb underuse also contributing to this finding [9]. Although the etiology still remains unclear some abnormal nerve physiology or nerve pathology appears to result in overgrowth.

Clinical Pearl

Some as yet undetermined factor in nerve physiology or pathology causes non-syndromic limb overgrowth

Other theories including vasomotor disturbances, abnormal nerve supply, endocrine aberrations, defective fetal position, lymphatic defects, autonomic nervous system defects and localized neurofibromatosis have been proposed but not confirmed [10].

Clinical Examination

Clinical examination should concentrate on a number of aspects.

Features of the Overgrowth Anatomy

Is the overgrowth confined to the digits or does it extend more proximally? Is there any nerve territory distribution? Median nerve related overgrowth of digits is probably the most common form of overgrowth [11]. Is the overgrowth uniform or does it have a nodular component? A nodular component would suggest neurofibromatosis or a lesion-related overgrowth such as a haemangioma. If it appears uniform then are all the components within the area enlarged or is one component more marked than others such as engorged veins or excess subcutaneous fat?

Features Suggesting the Presence of a Syndrome

Are there any other features suggestive of a syndromic abnormality such as a capillary haemangioma – port wine stain (Klippel Trenaunay Syndrome), neurofibromas with axillary freckling,

(neurofibromatosis) cerebroid thickening of palms and soles (Proteus Syndrome) or any family history of syndromes?

Loss of Function

This can be related to a number of consequences of the overgrowth. It can be a result of excess subcutaneous tissues mechanically restricting to range of movement. It can be as a consequence of the function of adjacent normal digits being obstructed by the overgrowth or a combination of both. Loss of function may be due to pain and stiffness secondary to associated early degenerative changes within the joint of affected digits. Swelling of soft tissue in the palm leads to flexor sheath thickening resulting in triggering of the affected digit and is demonstrated if motion is preserved [12].

The enlarged nerves (or fatty infiltration of nerves) is a component of some forms of overgrowth, leaving the nerves prone to mechanical compression neuropathies [13, 14] Mechanical neuropathy may occur at the carpal tunnel or cubital tunnel [15, 16]. Fatty infiltration of nerves may result in tenderness of the nerve in superficial anatomical sites such as the superficial branch of the radial nerve at the wrist.

Investigations

Investigations aims to identify the nature of the overgrowth (the components involved) and this will guide the clinician to diagnosis and also allow targeted treatment. Investigation should include:

Plain radiographs of the affected areas to identify any bony component to the overgrowth (Fig. 9.1). In haemangioma or lymphangioma there will be radiological signs of hypertrophy of the soft tissues without bony overgrowth. In Klippel Trenaunay Syndrome plain x-ray can show soft tissue and bone enlargement together with phleboliths within the vascular anomalies. Plain radiographs will also identify bony causes for the overgrowth such as Ollier's disease.

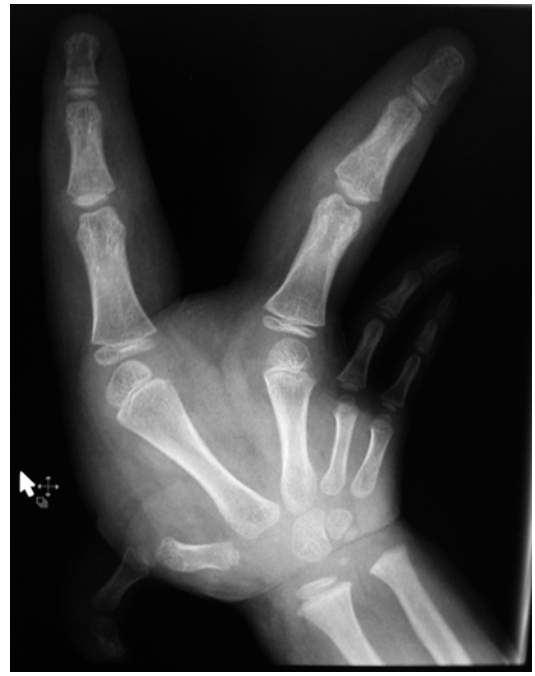

Fig. 9.1 Plain radiograph – a patient with hand overgrowth, both soft tissue and skeletal enlargement are noted from the film

Ultrasonography is useful, especially in children, because of its painless and harmless nature [17]. Ultrasound scan in utero can identify macrodactyly with three-dimensional examination offering even greater information on the foetal hand [18, 19]. This prenatal ultrasound assessment may provide clinicians important diagnostic information and warrant further investigation. Importantly parents are provided with prenatal diagnosis allowing time for psychological support and information prior to delivery [20]. Ultrasound combined with Doppler imaging is able to determine the flow rate of vascular lesions and differentiate one from another. Ultrasound scan can differentiate between haemangioma and arteriovenous malformation and other vascular malformations and is indicated if there is suspicion of an underlying vascular cause for the overgrowth (such as Klippel Trenaunay Syndrome) [21].

Magnetic Resonance Imaging (MRI) can characterise the type and extent of overgrowth without radiation exposure and helps to differentiate between most of the common diagnoses [17].

In neurofibromatosis T2 weighted MR images show high signal hyper intense neurofibromas, which will be situated close to the nerve [22].

In Klippel-Trenaunay-Weber syndrome limb hypertrophy, capillary malformations and arteriovenous fistulae are present and MRI identifies the presence and the extent of the vascular anomalies. These tend to be of high signal on T2 weighted images, although areas of low signal can be seen and represent haemosiderin deposition or areas of calcification [23].

In haemangioma T2 weighted MR imaging shows increased signal from the serpiginous vascular channels within the haemangiomas [22]. In lymphangiomatosis, the lymphangiomas are hyper intense to muscle on T1 weighted images and hyper intense to fat on T2 weighted images [24].

Classification

The terminology and descriptions of overgrowth in the upper limb have been varied and confusing, because overgrowth is as a consequence of diverse conditions. Even simple descriptive terms are used in a variety of fashions. The term macrodactyly (Greek: makros, large and dactylos, digit) is commonly used to describe overgrowth of digits in the upper limb yet Barsky described macrodactyly when used properly to describe an increase in the size of all the elements or structures of a digit but not the metacarpals [12]. In practice the degree and distribution of overgrowth of structures is rarely so simple. With this in mind any reference to macrodactyly should be followed by a very clear definition of the anatomy involved. There are a number of ways to classify overgrowth; we suggest that it may be worth classifying the condition in relation to the:

A. **Anatomy involved**
B. **Pattern of growth**
C. **Associated conditions**
D. **Tissue of origin**

Anatomy of Involved Parts

The extent to which the overgrowth involves the whole or part of the limb:

Overgrowth of Whole Limb

- As seen for example in
- Lymphoedema
- Hemihypertrophy
- Neurofibromatosis
- Combined complex vascular malformations
  - Klippel Trenaunay Syndrome
  - Parkes – Weber syndrome

Segmental Overgrowth of Limb (Excluding Hand)

- Diffuse – as seen in
  - Neurofibromatosis
  - Lymphoedema
  - Combined complex vascular malformations
- Localised – Tumours
  - Haemangioma
  - Enchondroma

Overgrowth of Hand

- Diffuse – as seen in
  - Macrodystrophia lipomatosa
  - Proteus Syndrome
  - Neurofibromatosis
- Localised – Tumours such as
  - Haemangioma
  - Enchondroma
  - Lipoma

Pattern of Growth [12, 25]

- Progressive- enlarged digit that grows proportionately faster than the adjacent digits
- Static – enlarged digits at birth which then continue to grow in proportion to the adjacent digits

Associated Conditions

Overgrowth can occur in isolation or as part of a syndrome [26].

This classification presented by Temtamy and McKusick in 1978 distinguishes overgrowth according to its presentation as an isolated anomaly or associated with another syndrome [27].

Isolated Overgrowth: overgrowth may be present at birth and can be static or progressive in nature.

Syndromal Associations: Overgrowth can present as part of a broader anomaly. The patient should be examined systemically.

The following syndromes may present with overgrowth as a component feature; proteus syndrome [28], Klippel-trenauney syndrome [29], Ollier's disease and Neurofibromatosis [30]. Neurofibromatosis has been associated with macrodactyly since the early reports of overgrowth became available [30].

Congenital Partial Gigantism may involve both metacarpals and metatarsals. It is the most common type of hypertrophy and has been further divided into segmental, crossed and hemihypertrophy [27]. The soft tissues are always involved however the long bones may show an increase in radial growth only [31].

Tissue of Origin

There are two classifications that refer to tissue of origin and histological type of the overgrowth. Flatts classification and Evans Classification. These classifications provide us with greater clinical intuition regards management.

Flatts Classification: Flatt classified Macrodactyly, one form of overgrowth in the upper limb. This classification is based on the histological disease process involved in macrodactyly. It was first presented by in Kelikian 1974 [32]. This was further modified by Flatt and is the most commonly used classification system in modern literature with regards to macrodactyly [33].

Type 1 – lipofibromatous hamartomas (fat)
Type 2 – neurofibromatosis (nerve)
Type 3 – hyperostosis (bone and soft tissue with no neural association)
This has been supplemented with the addition of type 4
Type 4 – hemi hypertrophy (intrinsic muscular hypertrophy)

Type I

This is the most common type of macrodactyly [11] (0.5 % of all hand anomalies), almost all cases are unilateral [11], male to female ratio is 3:2 and can be associated with other congenital hand anomalies in 10 % of cases [32, 33].

Type II

This is an autosomal dominant inherited trait, the reported incidence is 1 in 2,500–3,300 [27]. Macrodactyly in cases of neurofibromatosis are often bilateral, enlargement of the skeleton in the hand leads to reduced mobility. Other skeletal abnormalities like scoliosis and kyphosis and enchondromas may also be present [34].

Type III

Hyperostotic macrodactyly is a rare form with no hereditary influence. It is characterised by the severe limitation in mobility of joint due to the presence of osteochondral masses. Type III enlargement tends to follow median nerve distribution however the nerves are histologically noted to be normal [35].

Type IV

The aetiolgy of type IV is unknown and is also rare as compared to the latter forms of macrodactyly [35]. The enlarged digits are usually circumferentially enlarged and are accompanied with flexion contractures, deviation of digits and thenar and hypothenar muscle hypertrophy [34] (Table 9.1).

Isolated Presentation and Associated Conditions

Overgrowth in the upper limb most commonly presents as isolated overgrowth of the digits and palm usually in the form of macrodystrophia lipomatosa.

Macrodystrophia Lipomatosa

Macrodystrophia lipomatosa is a rare congenital disorder characterised by progressive overgrowth of the mesenchymal elements in the limb particularly the fibro adipose component. Feriz first described the condition in 1925 but only described

Table 9.1 Classification described by Evans et al, giving a structured representation between causes of overgrowth on the basis of tissue of origin and pathological entity

Histological component	Pathological entity	Conditions
Lymphatic	Primary lymphoedema (Milroy disease)	
	Lymphangioma	
Vascular	Vascular tumors	Haemangioma
	Slow flow vascular malformations	Venous malformations
		Lymphatic malformations
	Fast flow vascular malformations	Arteriovenous malformations
	Combined complex vascular malformations	Klippel Trenaunay syndrome
		Parkes-Weber syndrome
Soft tissue and nerve		Macrodystrophia lipomatosa
		Neurofibromatosis (type 1)
Bone	Hyperostosis	Ollier disease
		Maffucci disease

involvement of the lower limb and Golding extended the term to include upper limb involvement [36, 37]. The condition is not inherited but the underlying aetiology is poorly understood. However hypotheses include irregularities in the foetal circulation, lipomatous degeneration and abnormalities to growth-inhibiting factors [38, 39].

The condition is often noted at birth or in the neonatal period and it can affect upper or lower limbs. It is found more commonly in the lower limb where the overgrowth often occurs in the distribution of the plantar nerve; occasionally it can affect the entire limb. In the upper limb the overgrowth is usually within the distribution of the median nerve (Fig. 9.2). The condition is unilateral, painless and clinically the involved hand is noted to have volar enlargement in comparison to the dorsum, fitting with a characteristic fibro-adipose overgrowth pattern. This will also result in the tips of digits being proportionately larger. Deviation of the digit is also common and is thought to be a result of disproportional involvement of the digital nerve, the most involved border of the digit growing faster therefore resulting in deviation (Fig. 9.3) Macrodystrophia lipomatosa can be either static or progressive as described earlier. The static type is the most common [24].

The main concern for clinicians will be the impact of the overgrowth on function with mechanical obstruction as well as deforming forces on adjacent digits/tissues. The aesthetic appearance will also raise concerns regarding social acceptance and integration. In adolescence secondary joint disease may develop [40]. Plain radiographs may help to differentiate between macrodystrophia lipomatosa and other forms of overgrowth with the phalanges characteristically appearing broad and the distal ends splayed with a mushroom appearance [41]. MRI demonstrates an excess of fibro-fatty tissue around the affected digits particularly on the palmar surface. The fat component demonstrates a high signal on T1 and T2 weighted sequences and low signal on hit suppression sequences. The fibrous strands are demonstrated as low signal linear strands on TI weighted sequences. The fatty tissue may also be seen to infiltrate the adjacent muscles. Bony

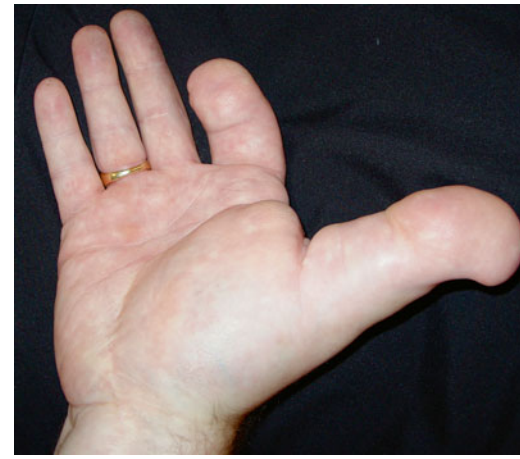

Fig. 9.2 Overgrowth in the distribution of the median nerve

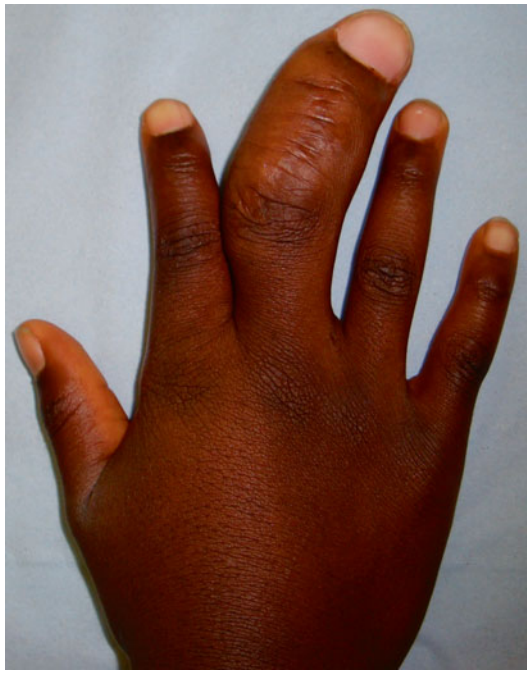

Fig. 9.3 Distal part of a digit is more commonly affected than that proximally

abnormalities such as cortical thickening and secondary degenerative changes are also identified with MRI, fibrous thickening of a nerve may also be seen [42].

Neurofibromatosis Type 1

Neurofibromatosis type 1 is an inherited neuroectodermal condition resulting in the formation of tumours around nerves along with other pathological features. It is the result of decreased production of the protein neurofibromin, which has tumor suppressor function [43].

Frederich von Recklinghausen first described the condition in a monograph in 1882, he identified that the skin tumours were derived from peripheral nerves [44]. Although the condition is inherited in an autosomal dominant manner with 100 % penetrance approximately 30 % of new cases arise spontaneously through gene mutation and are the most commonly occurring sporadic mutation in humans.

A diagnosis of Neurofibromatosis type 1 according to the National Consensus Developmental Conference Statement, is based on two or more of the following criteria

- Six or more café-au-lait spots or hyperpigmented macules greater than or equal to 5 mm in diameter in children younger than 10 years and to 15 mm in adults
- Axillary or inguinal freckles
- Two or more typical neurofibromas or one plexiform neurofibroma
- Optic nerve glioma
- Two or more iris hamartomas, Lisch nodules, (often identified only through slit-lamp examination)
- Sphenoid dysplasia or typical long-bone abnormalities such as pseudarthrosis
- First-degree relative with Neurofibromatosis type 1

Digital, hand or limb overgrowth are well-recognised sequelae of Neurofibromatosis type 1. The overgrowth tends to be predominately fibro fatty and localised around particular nerve distribution similar to macrodystrophia lipomatosa but osteochondral masses may be noted around the epiphyses of phalanges and metacarpals [45] (Fig. 9.4).

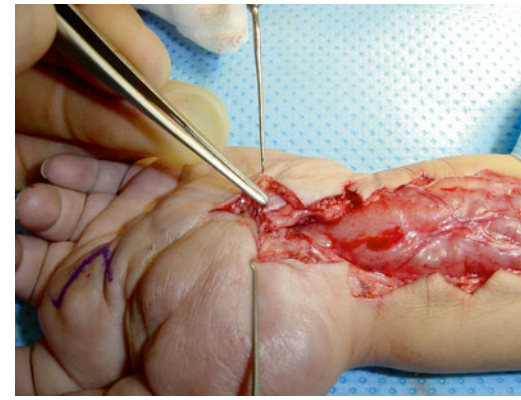

Fig. 9.4 Neurofibromatosis – both fibro fatty overgrowth in the thenar eminence and palm as well as nerve abnormality are noted in a patient with neurofibromatosis

Proteus Syndrome

Proteus syndrome is named after the Greek sea god Proteus. Homer's Odyssey refers to Proteus as the 'Old Man of the Sea' and herdsman of sea beasts. It is said that he had the power of

prophecy and also had the ability to change his form at will. Wiedemann first described the condition in 1983 [46]. It is a rare hamartomatous disorder characterised by multiple sites of overgrowth that can involve overgrowth of skin, bone, muscle, adipose tissue, blood and lymphatic vessels. Structures at multiple sites may contribute with varying degrees of overgrowth. Clinical presentation is extremely varied and includes some or all of the following: macrodactyly, hemi hypertrophy, subcutaneous masses, exostosis, cerebroid thickening of palms and soles and linear skin lesions. The condition can result in significant disfigurement. Once thought to have neurofibromatosis, Joseph Merrick (also known as "the elephant man") is now thought by some to have had Proteus syndrome [47].

Proteus syndrome arises from a sporadic mutation in the AKT1 gene and this arises during the early stages of development, so that not all cells within the developing foetus will have the mutation present [48]. The AKT1 gene helps regulate cell growth and division (proliferation) and cell death. Increased cell proliferation in various tissues and organs leads to the abnormal growth characteristic of Proteus syndrome.

The affected child is usually born without any obvious abnormalities and clinical manifestations appear in the first year. The clinical findings may include

- Neurological abnormalities as a consequence of mass effect on the brain resulting in learning difficulties, seizures and visual loss.
- Musculoskeletal abnormalities with the limbs and spine commonly affected. This results in a loss of function secondary to scoliosis and macrodactyly as well as symptoms of muscle and joint pain from the deforming action of masses on the skeletal growth (Fig. 9.5).
- Skin and subcutaneous tissue changes include lipomas, epidermal naevi, and connective tissue naevi with cerebriform thickening of the soles of the feet, which are pathognomonic, and vascular malformations.
- Abnormal appearance: other than the abnormal swellings the affected individuals may also have distinctive facial features such as a long face, lateral canthi that point downward (down-slanting palpebral fissures), a low nasal bridge with wide nostrils and an open-mouth expression. For reasons that are unclear, affected people with neurological symptoms are more likely to have distinctive facial features than those without neurological symptoms.
- Premature death from deep vein thrombosis and pulmonary embolus secondary to vascular malformation associated with the condition [49].

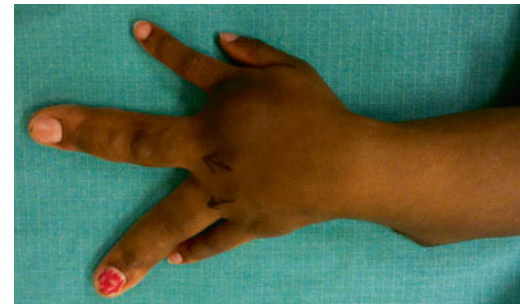

Fig. 9.5 Proteus Syndrome – macrodactyly in a patient with proteus syndrome

The mainstay of treatment for Proteus syndrome includes early identification of serious medical problems and the use of prophylactic and symptomatic treatment.

Vascular Malformations

Vascular malformations contribute a large proportion of overgrowth. These can result in localised overgrowth as a consequence of isolated haemangioma or lymphatic malformation. However more diffuse overgrowth may occur in the presence of venous malformations that cross tissue planes and invade more than one or all tissues in the extremities (skin, tendon, muscle, nerve, fat, or bone) [50]. Diffuse overgrowth is also a feature of many combined complex vascular malformations where skeletal overgrowth as well as soft tissue overgrowth is noted. Such malformations represent a wide array of lesions the most common of which are Klippel Trenaunay and Park Weber syndrome. These conditions display a triad of features including capillary malformation (port

wine stain), venous malformation with large varicosities and overgrowth.

Klippel Trenaunay Syndrome

Although the exact cause of Klippel-Trenaunay syndrome is unknown a number of theories have been postulated. These include intrauterine damage to the sympathetic ganglia or a mesodermal defect during foetal development leading to dilated microscopic vascular anastomoses [51, 52]. This leads to deep vein abnormalities with resultant obstruction of venous flow resulting in venous hypertension, the development of varices and limb hypertrophy [53].

Most cases are sporadic, although a few cases in the literature report an autosomal dominant pattern of inheritance [54].

The condition has no sex predilection and is noted at birth or soon afterwards. It generally affects a single extremity and the lower limb is involved in 95 % of cases.

A majority of patients demonstrate all three signs of the clinical syndrome: capillary malformations, varicose veins and overgrowth with both skeletal and soft tissues involved [55]. The capillary malformation presents first and is often noted on the lateral aspect of the limb. It may be limited to the skin or extend deeper to subcutaneous tissues, including muscle and bone. Varicosities may be extensive and may affect the superficial, deep and perforating venous systems. Bony and soft tissue overgrowth is the third sign. This may be appreciated at birth and can progress thereafter.

Prognosis is good and surgical intervention is only required to address limb discrepancy [55].

Parkes Weber Syndrome

Other than the triad of capillary and venous malformation with overgrowth, Parkes Weber syndrome is also characterized by arteriovenous fistulas, which are present from birth [56].

Some cases of Parkes Weber syndrome result from mutations in the RASA1 gene. The RASA1 gene controls several important cell functions, including cell proliferation and differentiation and appears to be essential for the normal development of the vascular system. When the condition is caused by RASA1 gene mutations, affected individuals usually have multiple capillary malformations [57]. Those patients who do not have multiple capillary malformations are unlikely to have the mutations and in these cases, the cause of the condition is unknown.

When Parkes Weber syndrome is caused by mutations in the RASA1 gene the condition has an autosomal dominant pattern of inheritance. Most cases of Parkes Weber syndrome are sporadic.

The clinical signs of overgrowth usually present in infancy and continue up to epiphyseal fusion unlike Klippel Trenaunay where overgrowth becomes static in the early teens. The arteriovenous fistulae are usually multiple in natures. It is a consequence of the fistulae that the condition has a poor prognosis and the condition can be associated with life-threatening complications including abnormal bleeding and heart failure.

Ollier's Disease

Enchondromata are benign cartilaginous tumors, which develop in close proximity to the epiphyseal growth plate cartilage [58]. Ollier's disease, also termed enchondromatosis is a rare condition characterized by multiple enchondromata. Ollier's disease is estimated to occur with a prevalence of 1/100,000. The condition occurs sporadically with no inheritance. The disease is not apparent at birth and clinical manifestations will develop in infancy and thereafter. Ollier's disease is characterized by the clinical findings of asymmetric and often unilateral distribution of cartilage lesions that can be extremely variable (in terms of size, number, location, evolution of enchondromata, age of onset).

Clinical problems caused by enchondromas include deformities and limb-length discrepancy. Enlarged digits are as a consequence of localised growths on the digit, which also result in delayed

ossification of the epiphyseal growth plate with resultant overgrowth.

Patients with Ollier's diseases may also develop pathological fractures through the skeletal tumours. The tumours do also present a risk of malignant transformation of enchondromas into chondrosarcomas therefore surveillance should be incorporated into the treatment.

Maffucci Syndrome

Maffucci syndrome is a rare sporadic disease characterised by vascular and skeletal tumours. The tumours primarily occur in hands, feet and long bones with clinical manifestations developing in the first decade of life. Like Ollier's disease it is also characterised by multiple enchondroma however unlike Ollier's the distribution is bilateral and the clinical picture also includes multiple haemangiomas. The tumours are asymmetrical and bilateral. Other than haemangiomas, soft tissue manifestations include the presence of lymphangioma and phlebectasia. Haemangiomas are most frequently located in the dermis and subcutaneous fat adjacent to areas of enchondromatosis. Intracranial tumours have also been reported in Maffuci syndrome therefore screening with MRI may be prudent. Complications include spontaneous fracture and sarcomatous degeneration of enchondroma [59]. Benign and malignant degeneration of haemangioma and lymphangioma can also occur.

Treatment should include careful surveillance for malignant degeneration of both skeletal and non-skeletal tumours.

Clinical Pearl

In a phenomenological entity such as overgrowth many diverse aetiologies make classification confusing. In practice the surgeons needs to determine whether there is a systemic condition (proteus, neurofibromatosis,) with their own prognoses, and whether the condition is progressive (vascular malformations, tumours etc) this will aid prognosis and guide decision making about surgery

Management

Overgrowth can lead to quite severe deformity resulting in a functional and aesthetic impact on the patient. There is also the significant psychological impact related to social stigmata of an obviously visible deformity. The family may seek treatment during childhood or the patient may present in later life with secondary consequences of overgrowth, which include compression neuropathy and degenerative joint changes.

The psychological effect of the anomaly on the patient and in most cases families of the children involved is important. As with all disfigurement in childhood the help of a clinical psychologist is valuable to support the family and with time help the child with the psychosocial consequences, they also assist in the family's attitudes to treatment. Overgrowth is a difficult condition to treat and aesthetic outcomes are nearly never normal and may include amputation and a clinical psychologist may help provide parents and child with a realistic view of the outcomes [10].

In view of the diverse presentation and aetiology, treatment is individualised. The interventions available for surgical treatment include volume and length reduction, arresting growth, correction of deviation and amputation. In many a combination of techniques and multiple procedures at different stages of growth will be needed to achieve the best possible outcome. Hardwicke reported that static disease required significantly fewer operations than progressive disease and patients with progressive disease should be made aware of the need for repeated treatments [60]. Choice of techniques and timing is influenced by a number of factors including age at presentation, rate of growth, size in comparison to expected normal adult finger size for the child (using the same sex parent as a guide to predict expected size), digits affected, the type extent and severity of the deformity and willingness to undergo numerous procedures. The options available include:

Arresting Growth

This is an important concept in the treatment and timing is critical. Once the size of digit/limb has reached its predicted adult size, the

epiphyseal growth plate must be destroyed in each bone involved (middle phalanx epiphysis may be spared to prevent PIPJ stiffness) producing epiphysiodesis. In some cases of overgrowth this may be the only surgical procedure required [55]. In the digits this is achieved via mid lateral incisions. Alternative techniques include ligating arteries and nerve stripping or resection although in the senior author's experience resecting the affected nerve does not arrest growth as might be expected [61].

Volume Reduction

A majority of cases have excess subcutaneous tissues. In the upper arm and forearm this can be addressed by a number of techniques as follows:

Wedge excision – this can be performed in a single excision or in a serial fashion with excision of excess skin and subcutaneous tissue down to fascia with closure.

Excision and skin grafted – in case of lymphedema or severe neurofibromatosis excess and abnormal skin and subcutaneous tissue can be excised and the resultant defect reconstructed by means of a split thickness skin graft. Excision of subcutaneous tissue by raising skin flaps reduced the need for grafting and was proposed by Homans and is performed in a serial fashion in order to maintain skin viability. For pure lymphoedema a number of modifications to this approach includes retaining dermal flaps as described by Thompson, which attempts to address the deformity and also aid with lymphatic drainage, this technique is rarely used in the upper limb [62]. Liposuction has become a popular method in addressing excess subcutaneous tissue particularly in lymphedema.

Techniques for volume reduction of the digits are dependent on the distribution of the excess tissue. For volar reduction alone access to the area can be performed by means of Brunner incisions and performed in one stage. If both volar and dorsal tissues require excision then midlateral incision may be used. Any planning of incisions must take into account the possibility of future surgery including skeletal reduction and amputation. The radial and ulnar side of each digit should be debulked in two separate procedures three months apart. When midlateral incisions are used the skin flaps are raised up to the midline both on the dorsal and volar surface of the digit. Regardless of incision the neurovascular bundles need to be dissected from the subcutaneous fat, the digital nerve can be quite tortuous within this tissue and in many cases the nerve itself can be significantly infiltrated with fat. The excess subcutaneous tissue and redundant skin is then excised and remaining skin closed.

Bony narrowing can be performed by means of longitudinal osteotomy. However this impacts on important structural soft tissue components of the finger needed for function such as the flexor sheath and ligamentous structure around the joints resulting in joint stiffness.

Length Reduction

This is achieved by skeletal shortening and a number of ways are suggested. During this stage one should consider the degree of angulations of the digit as this can be addressed as part of the skeletal shortening procedures.

Terminalisation to shorten the digit to its appropriate size is the easiest but least sophisticated method of shortening. More elegant techniques to shorten the digit, whilst preserving the cosmetic nail unit, have been described by Barsky and Tsuge. Barsky's technique involves removing the distal portion of the middle phalanx and proximal portion of the distal phalanx, thereby reducing the length of the finger whilst preserving the nail which was transposed on a on a volar skin pedicle [12]. Tsuge's technique also preserves the nail by overlapping the dorsal portion of the distal phalanx with the volar portion of the middle phalanx, in this technique the nail is preserved on a dorsal skin pedicle [61]. Bunching of the soft tissues in these cases will need to be addressed at a second stage and flexor tendon lengthening as a consequence of bone shortening needs to be addressed in cases where more than 3 cm of shortening has been performed. Uemera modified the above process on a case of macrodactyly affecting a toe by transposing the nail as an island on a neurovascular pedicle as opposed to a skin pedicle therefore negating the need for

a second procedure to excise the redundant skin that had been used as a pedicle [63]. Fatemi replicated the same process on digits on the hand [64]. Nail reduction may have to be considered in conjunction with transposition. However this always requires a compromise of either removing a central portion of nail preserving the eponychial flaps but resulting in a central ridge or excising a lateral portion of nail and sacrificing one eponychial fold.

Transverse osteotomy with square bone resection can be performed in each phalanx and in cases with deviation trapezoid bone resection should improve both deformity and length excess. Combination of epiphyseal resection combined with wedge reduction can be utilised to address deformity and arrest growth [61].

Clinical Pearl

Amputation is often a good early treatment for macrodactyly and courageous informed decision making will spare the child the multiple surgeries and medicalization that can all too often result in an unsightly stiff and excluded digit

Amputation of a digit must always be considered as a potential first line treatment and not as a fall back procedure. In overgrowth it is the most successful procedure. It is essential that patients who would benefit from amputation are identified early as in the correct patient it will eliminate the need for multiple procedures and a rapid return to function and normal living, which is essential particularly in the developing child. Border digits can be resected by fashioning a ray amputation with central digits amputated at mid metacarpal level with transposition of an adjacent border digit [65]. Digital amputations can be followed by micro vascular transfer of digit to the amputated area to achieve function; thumb amputation should however be avoided if possible unless adequate reconstruction is available [66–68].

Clinical Pearl

Toe to hand transfer has been very successful in our practice as a reconstruction for amputated fingers and thumbs in their distal portion and should be performed at the same time as the amputation

Summary

Overgrowth of the upper limb presents the clinician with a variety of clinical challenges, the aetiology of the condition is complex and multiple and terminology within medical literature is often confusing.

The clinician must addresses the patient's and the family's complaints in a holistic and individualised manner taking into account deformity and lack of function but also addressing psychological concerns for both patient and family. Secondary changes of joint degeneration, nerve compression and malignant transformation must be incorporated into one's long-term management plan.

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Undergrowth

10

Ruth Lester

Keywords

Undergrowth • Polands • Kirners • Brachymetacarpia • Brachydactyly • Phalangeal transfer • Toe-to-hand, symbrachydactyly, clinodactyly, symphalangism • Distraction osteogenesis

Introduction

Because of the enormous range of congenital anomalies, classification of all congenital anomalies of the upper limb is difficult. In particular, undergrowth/hypoplasia can be classified in many different ways. Developmental biology is an advancing field and significant progress has been made in identifying the complex molecular pathways, which orchestrate limb development. The pathology of these pathways is becoming better understood and thus has an impact on the classification of hand and upper limb anomalies [1].

The group of Undergrowth/hypoplasia in Swansons classification [2] can be interpreted in many different ways. Undergrowth as identified by Swanson can now be considered a malformation of the proximal-distal outgrowth (transverse arrest) or a failure of the hand-plate in an unspecified axis.

According to Swanson's classification, hypoplasia of whole digits, or parts of digits and/or hands and/or limbs is present in many categories of deformity.

There is also significant confusion in the literature over nomenclature. I am therefore going to group together certain clinically relevant presentations accepting that there are often multiple different presentations within one patient.

The groups to consider will be undergrowth of the whole limb with symbrachydactyly (short joined fingers), brachydactyly (short fingers), brachymetacarpia (short metacarpals). I will also include Kirners syndrome and clinodactyly of the little finger [3].

Clinical Presentations

1. Symbrachydactyly (Fig. 10.1) is most commonly associated with Polands syndrome and consists of brachydactyly, cutaneous syndactyly and

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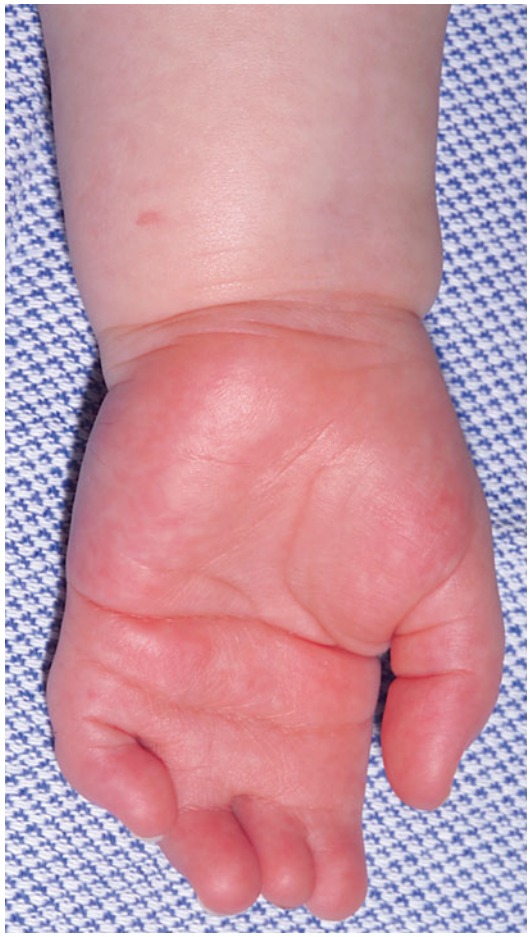


Fig. 10.1 Classical symbrachydactyly of Polands Syndrome

global hypoplasia of the hand [4]. The undergrowth of the hand in Polands syndrome can be very severe leading to an absence of the fingers althogether.

Poland's Syndrome The cause of Poland syndrome (Fig. 10.2) is unknown. However, an interruption of the embryonic blood supply to the arteries that lie under the collarbone (subclavian arteries) at about the 46th day of embryonic development is the prevailing theory [5]

2. **Brachydactyly** ("short fingers") is a general term that refers to disproportionately short fingers and toes.

   Brachydactyly can occur either as an isolated malformation or as a part of a complex malformation syndrome. Brachydactyly may therefore, also be accompanied by other hand malformations, such as syndactyly, polydactyly, reduction defects, or symphalangism.

   There are basically four types of short fingers:

   (a) small or absent phalanges (brachyphalangia) (Fig. 10.3)
   (b) missing digits leading to a central cleft (Fig. 10.4)
   (c) monodactylous – i.e. a single main digit only (Fig. 10.3a)
   (d) No fingers/nubbins – i.e. transverse arrest (Fig. 10.3b)

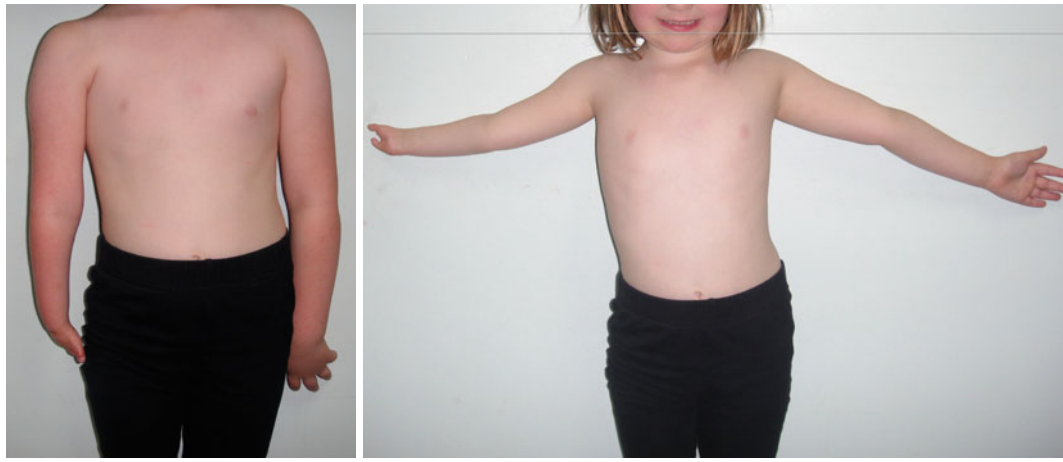


Fig. 10.2 Polands syndrome. Hypoplastic right forearm and absent sternal head of pec major

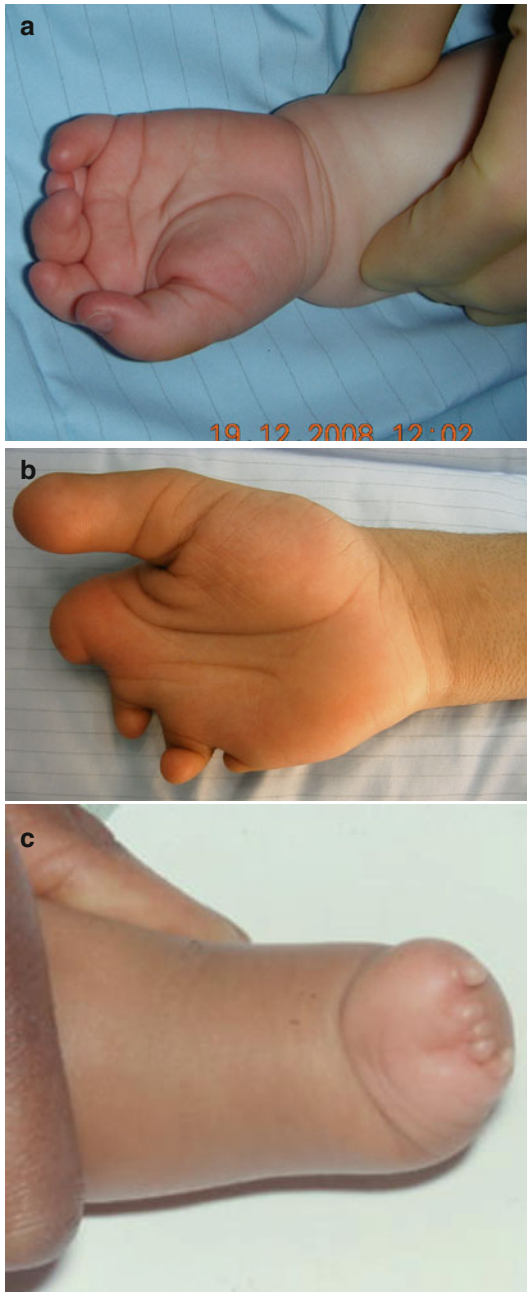


Fig. 10.3 (**a**) Brachyphalangia: Short floppy digits with normal hand. (**b**) Brachydactyly – Monodactylous type. (**c**) No fingers – i.e. Transverse arrest

In isolated brachydactyly, subtle changes elsewhere may be present. The various types of isolated brachydactyly are rare, except for types A3 and D. Some forms also result in short stature.

For the majority of isolated brachydactylies and some syndromic forms of brachydactyly, the causative gene defect has been identified. In isolated brachydactyly, the inheritance is mostly autosomal dominant with variable expressivity and penetrance [6]. (Table 10.1 with Fig)

3. **Brachymetacarpia** (Fig. 10.5) is a condition manifesting a shortened metacarpal that is caused by early closure of the epiphyseal plate and is believed to arise idiopathically.
4. **Kirner's deformity** (Fig. 10.6) affects the distal phalanx of the little finger leading to a progressive volar and radial angulation with time caused by an abnormal growth of the distal phalanx. It has been associated with Cornelia de Lange, Turner's and Downs syndrome. It is often misdiagnosed as an old fracture or acutely as a mallet injury, but, on close questioning, no history of significant injury can be obtained. It does not lead to any functional disturbance but can cause considerable anxiety from a cosmetic perspective.
5. **Clinodactyly** (Fig. 10.7) of the little finger describes a curvature towards the ring finger, caused by an abnormally shaped epiphysis of the middle phalanx. It is commonly an isolated finding, often familial, but can be associated with many syndromes such as Russell-Silver, Feingold and Downs Syndrome. Once again it is usually of cosmetic significance only

Presentation, Investigation and Treatment Options

During the initial consultation, a full history including family and prenatal history should be obtained.

A detailed clinical examination of the whole child including upper and lower limbs, feet and toes as well as hands, needs to be undertaken. In particular an observation of both upper limbs including the chest wall to look for inequality in shape and in nipple size and position enables confirmation of Polands syndrome.

Early x rays of the hands in the neonatal period may not be helpful for surgical planning

because of the cartilaginous nature of the bones at this time. However Xrays may be of value for the diagnoses of syndromes associated with hand anomalies and are often requested by the geneticists who should be available for consultation with the families at an early stage.

Surgical Principles

Because of the varied presentation of conditions in this category, each surgical plan needs to be individually worked out, after ongoing observation of the baby/child and their patterns of hand use. This usually requires more than one outpatient visit.

The surgical plan for functional improvement should be based on the following principles:

The minimum requirements for a functioning hand are two opposing digits. Even if tip-to-tip pinch cannot be achieved, a simple side-to-side pinch will be useful.

1. A simple pinch/grasp can be achieved by:
   (a) Deepening of a cleft and/or
   (b) Separation of a syndactyly, with or without:
   (c) Lengthening of digits.
2. Stabilising 'floppy/collapsible' short digits can be achieved with:
   (a) Free toe phalangeal transfer (Figs. 10.8, 10.9) or
   (b) Transfer of extra bone from within the hand, either:
      (i) Vascularised or
      (ii) Free intercalated bone graft

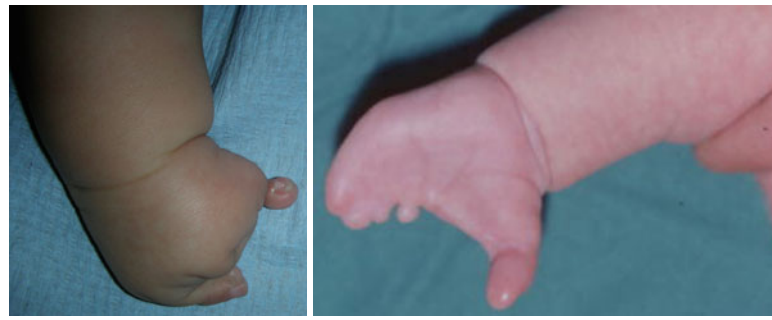


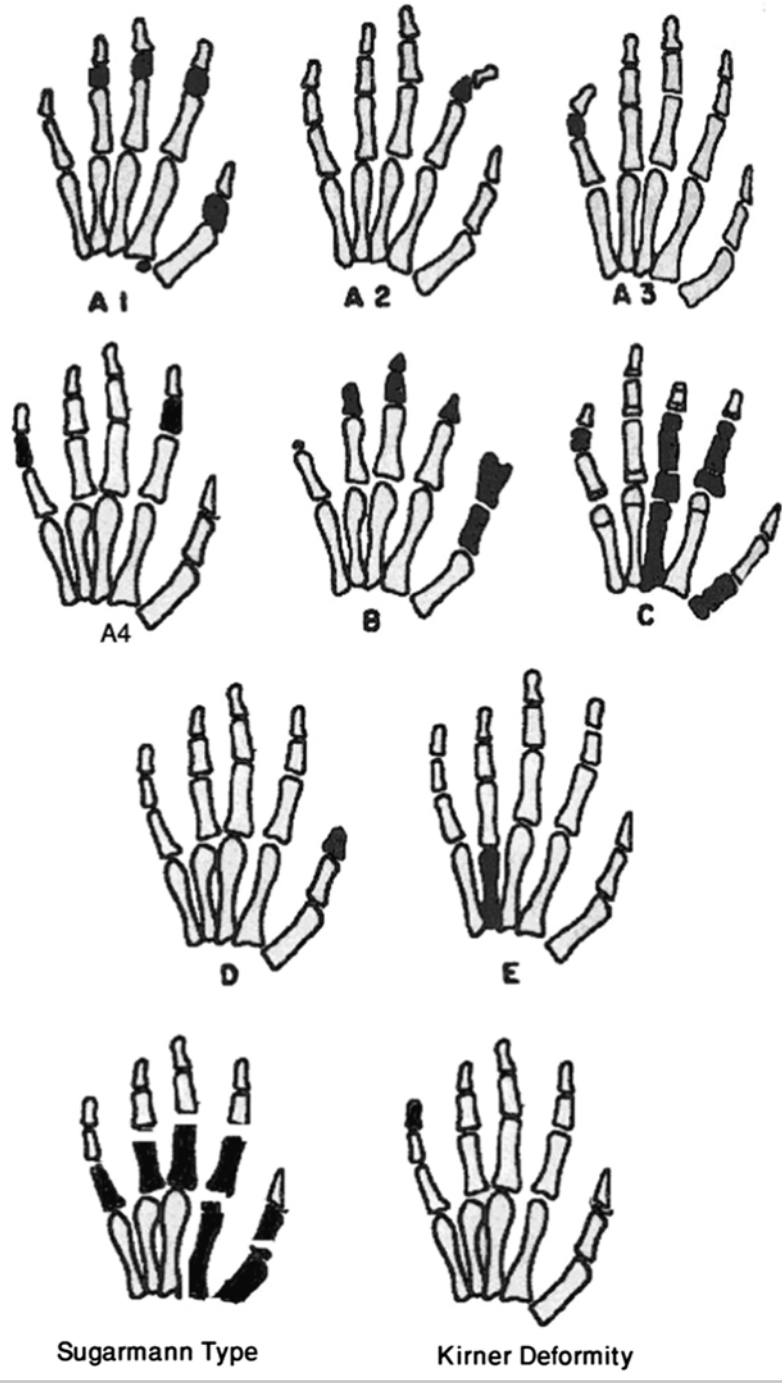
Fig. 10.4 2 examples of 'Atypical cleft hand' or Symbrachydactyly

Table 10.1 Types of isolated brachydactyly

Name	Synonyms	OMIM
Brachydactyly type A		
Brachydactyly type A1 (BDA1)	Farabee type brachydactyly	112500
Brachydactyly type A2 (BDA2)	Mohr-Wriedt type brachydactyly	112600
Brachydactyly type A3 (BDA3)	Brachymesophalangy V, Brachydactyly-Clinodactyly	112700
Brachydactyly type A4 (BDA4)	Brachymesophalangy II and V, Temtamy type brachydactyly	112800
Brachydactyly type A5 (BDA5)	Absent middle phalanges of digits 2–5 with nail dysplasia	112900
Brachydactyly type B (BDB)		113000
Brachydactyly type C (BDC)	Brachydactyly with hyperphalangism, Haws type brachydactyly	113100
Brachydactyly type D (BDD)	Stub thumb	113200
Brachydactyly type E (BDE)		113300
Brachymetatarsus IV	Metatarsus IV, short, Toes, fourth, short	113475
Sugarman brachydactyly	Brachydactyly with major proximal phalangeal shortening	272150
Kirner deformity	Dystelephalangy	128000

Table 10.1 (continued)

Name	Synonyms	OMIM



Reprinted with permission from Temtamy and Aglan [7]

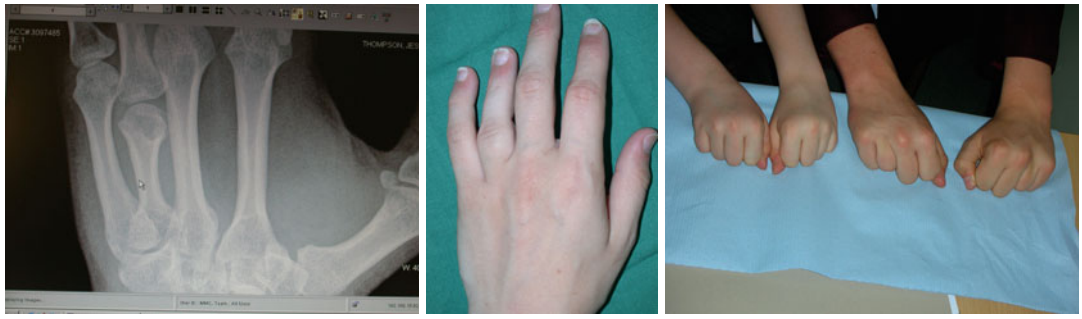


Fig. 10.5 Brachymetacarpia: Note the appearance of the short metacarpal visible on flexion of the fingers

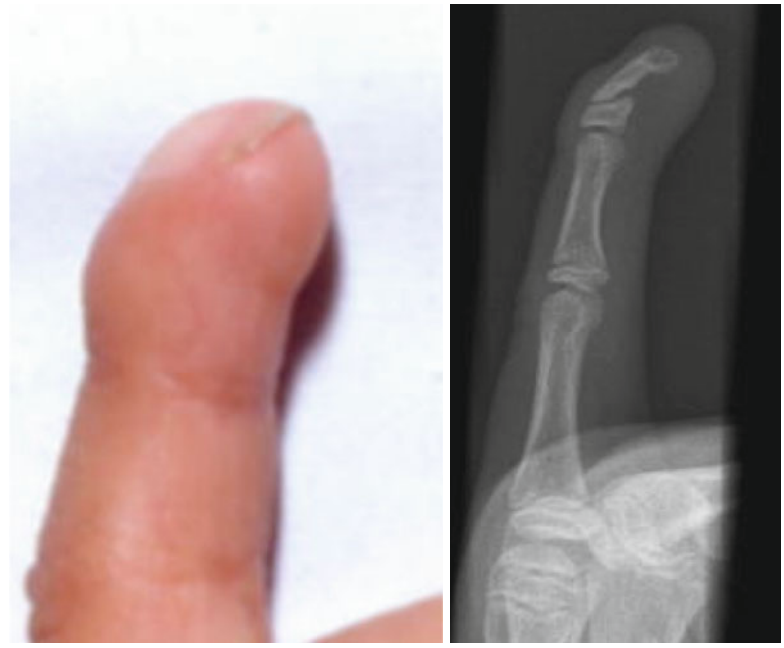


Fig. 10.6 Kirners deformity (Reprinted with permission from Temtamy and Aglan [7])

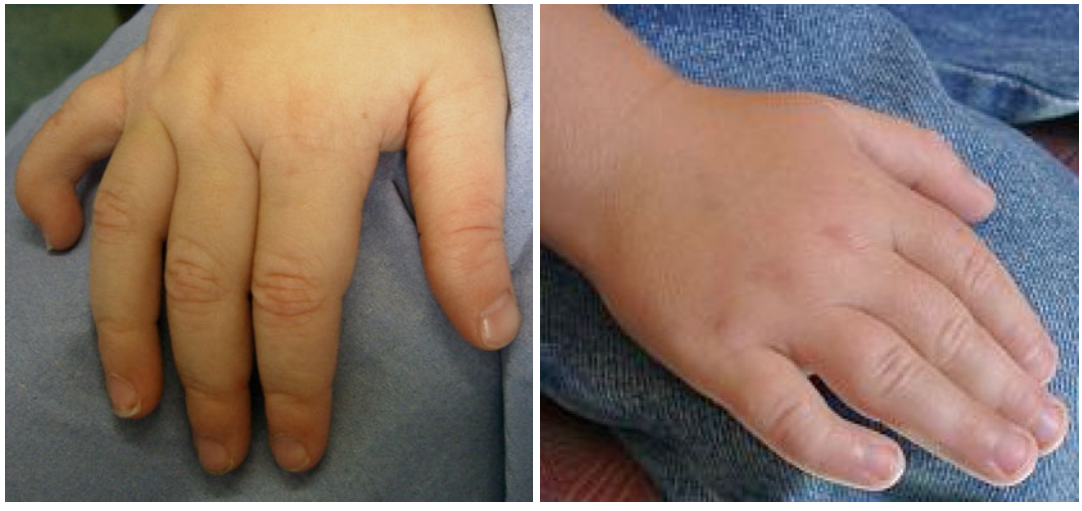


Fig. 10.7 Clinodactyly

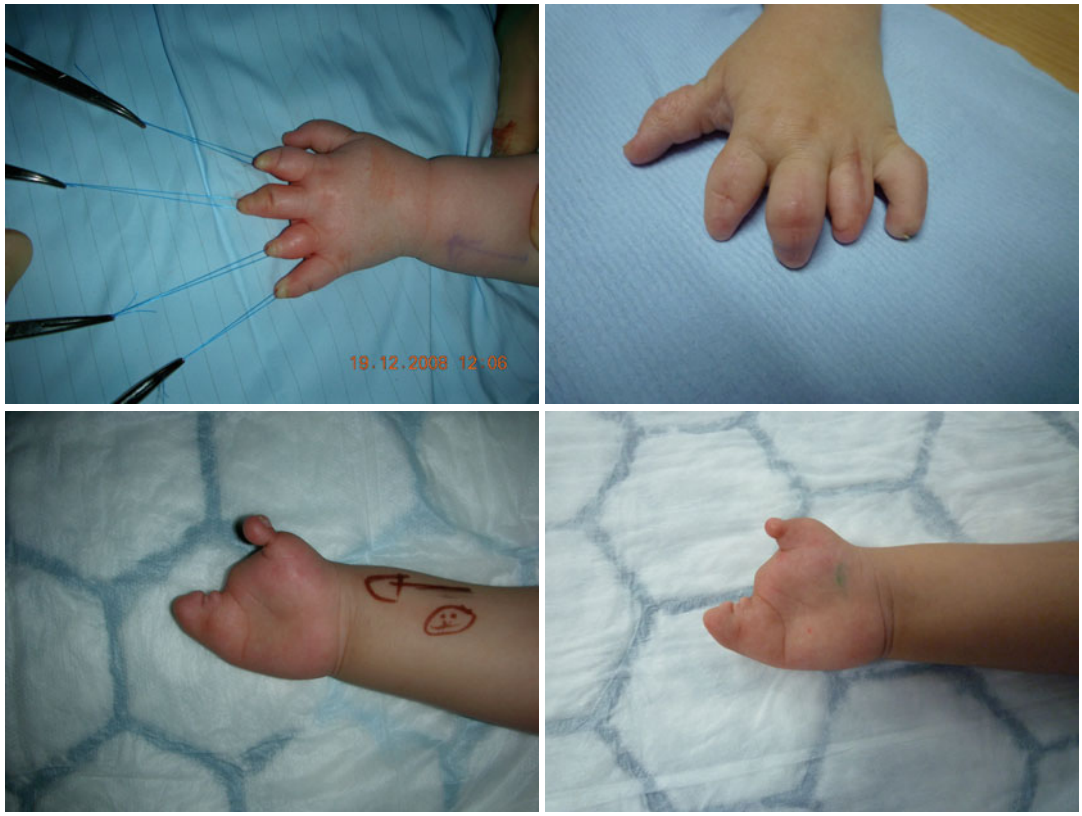


Fig. 10.8 Pre and post op phalangeal bone transfers for short floppy digits

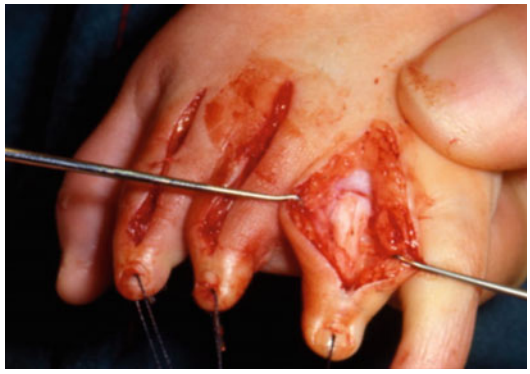


Fig. 10.9 Intra operative insertion of phalangeal bone grafts

3. The use of toe transfers in congenital hand surgery is limited but may be appropriate to provide an opposable digit to a single digit already present as long as the intrinsic muscles are present.

   Two toe transfers working opposite each other will only offer the possibility of a pinch grip if the intrinsic muscles are present to at least one recipient site.

   An additional toe transfer to provide a tripod pinch is feasible but leaves a large defect in the foot.
4. Approximately 1.5 cms. of additional length can be achieved with the use of bone distraction techniques (Fig. 10.10).

Surgery for cosmetic reasons only as in clinodactyly of the little finger and Kirner's deformity can usually be left until the child requests treatment.

Timing of Surgery

Deepening of a thumb web is best performed as early as possible i.e. at around 6 months of age. A useful early grasp is seen in the

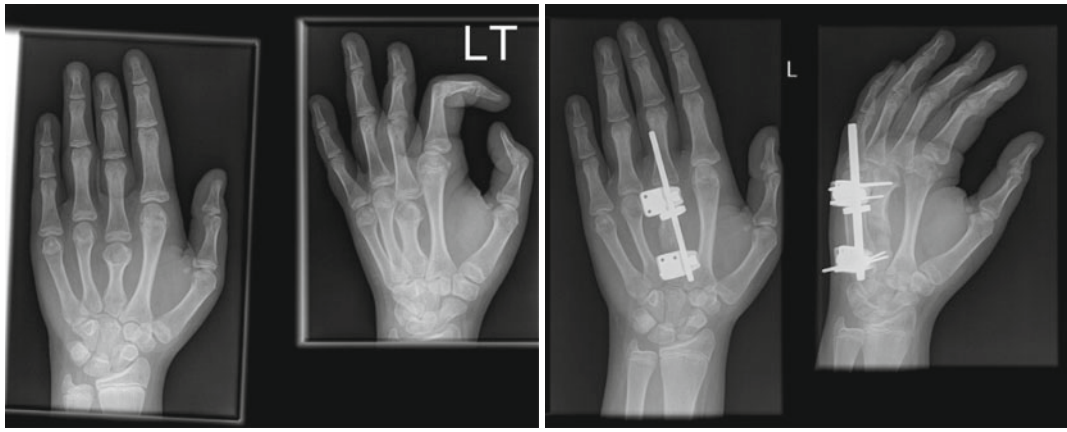


Fig. 10.10 External lengthening device for brachymetacarpia pre and post op

development of the child at around 4–5 months and therefore creating this opportunity is ideally timed as early as possible. The baby needs to be well enough to withstand anaesthesia of an hour or more.

Twelve to eighteen months of age is generally considered an optimal age for carrying out most reconstructive surgical techniques in children. At this age, most other congenital problems in the child, which may have an implication for anaesthesia, will have been identified. The child also is very distractible and therefore copes well with dressing changes and other necessary interventions.

Transfer of phalangeal bone grafts is considered best carried out before the age of 15 months as it has been shown that the growth of the transplanted phalanx is greater the earlier the transfer occurs [8]. However, in some circumstances, I have proceeded at a later age (even as old as 9 or 10 year) with no technical problems and a reasonable outcome. If the transfer is carried out at an older age, the transplanted phalanx has had more opportunity to grow in its normal position.

Vascularised toe transfers are best carried out after the age of 3 years old when the child is more co-operative. It is also valuable to have a period of time, observing the way the child using his/her hand and for ongoing discussions with the parents.

Distraction osteogenesis techniques require sufficient bone thickness to allow a screw fixation using a minimum of 2×1.2 mm screws. These screws will be under tension as the elongation process continues. The bone size is unlikely to be sufficient for this process until the child reaches 7–8 years old.

Opening, **closing or reverse wedge osteotomies to straighten a curved digit** for cosmetic reasons can be delayed until the child reaches skeletal maturity.

A severe clinodactyly of the little finger may be improved by early division of the abnormal epiphyseal bracket either with the insertion of a bone graft or fat graft, although care must be taken to avoid further damage to the epiphysis with subsequent progression of the deformity.

Techniques

Deepening of Thumb Web

This can be achieved in a variety of ways:

(a) Transfer of flap from radial side of index finger with or without full thickness skin graft (from antecubital fossa or lateral groin)
(b) Double opposing Z-plasty with advancement flap – i.e. ‘Jumping man procedure’
(c) Any standard syndactyly separation technique
(d) Occasionally removal of small residual bones from within the hand may help deepen the cleft and thus effectively lengthen the digits.
(e) Using these segments of bones either vascularised or free to lengthen remaining digits by ‘intercalation’ or ‘on top’ can be helpful

Transfer of Phalangeal Bone Grafts

(a) Any 'collapsible' digit is suitable
(b) Dorsal 'Y'-shaped incision over recipient digit
(c) Use 3rd and/or 4th proximal or middle phalanges from toes
(d) Include periosteum
(e) The following methods of Reconstruction of the bony defect are described:
  (i) suturing flexor to extensor tendon
  (ii) inserting bone graft from ilium [9]
  (iii) Reversing a section of distal metatarsal
  (iv) Taking a longitudinally resected half of the phalanx
(f) Shape the graft to fit in recipient site
(g) K wire is not usually necessary

Vascularised Toe Transfers

(a) If radial and/or ulna intrinsics are present in the recipient hand, a single or double vascularised 2nd toe and/or partial great toe transfer can establish a pinch grip. The transferred digit only has extrinsic movement and therefore a contraindication to this surgery would be the absence of a good CMC joint or inability to 'cup' the hand using the recipient intrinsic muscles.
(b) Further surgery to stabilise an existing digit by arthrodeses or osteotomies with or without bone grafts may be necessary.
(c) This type of surgery should only be carried out in specialised centres where access to a microvascular surgical team is available 24 h a day.
(d) Various descriptions of the technique are available [10].

External Bone Distraction Techniques [11]

(a) External and internal distractors are available
(b) Approximately 1.5 cms of lengthening can be achieved
(c) Distraction at a rate of 0.25 mms per day is the maximum achievable and the child needs to be reviewed very regularly.
(d) Common complications including pin track infections and failure of the regenerate bone.
(e) The distractor needs to be left in place after distraction for the same length of time as the distraction period

Osteotomies to Straighten Digits

The following types of osteotomies can be used to straighten a radio – ulnar curvature in a bone. Careful assessment of the x-ray to decide which bone is leading to the curvature must be made

(a) Reverse wedge osteotomy should maintain length
(b) Opening wedge osteotomy plus interpositional bone grafts should increase length
(c) Closing wedge osteotomy will reduce length

Multiple volar opening wedges can be used to straighten the volar curve associated with Kirner's deformity of the distal phalanx without the need for a bone graft

Outcomes

The use of validated scoring assessments for children are difficult to achieve in a busy working practice.

In association with the individualised surgical plan and the variety of clinical presentations there are often too many variables both in the condition and in the surgery for such assessments to be meaningful.

Annotating discussions with parents and the child and repeated observations of the child's activities can give a lot of useful information around expected outcomes of surgery as well as the results of a surgical procedure carried out a few years before.

Parents will often report that the child can do a particular task more easily. Most children will be able to do most activities whatever the deformity.

Realistic outcomes for improvement of cosmetic issues needs to be thoroughly discussed with the parents and the older child.

Complications

This type of reconstructive surgery carries significant risk in addition to the potential complications of infection, wound healing problems and scarring.

Each surgical plan needs to be discussed with the parents and older child and a frank explanation of outcome goals is crucial prior to embarking on a reconstructive challenge.

Further surgery may often be necessary to complete the goals but the timing and type of surgery cannot always be predicted.

Conclusions

Undergrowth of a hand, limb or digits presents unique cosmetic and functional problems, which need to be individually assessed. An individualised surgical treatment plan should be constructed after observation and discussion with parents and colleagues. Various possible techniques and appropriate surgical timings have been presented with some guidance on technique and expected outcomes.

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Constriction Ring Syndrome

11

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Keywords

Constriction ring syndrome • Amniotic band syndrome • Constriction bands • Acrosyndactyly • Auto-amputation of digits in utero

Introduction/Nomenclature

Constriction ring syndrome is a rare condition of unknown etiology. Numerous names have been used to describe this entity in the past including annular band syndrome, constriction band syndrome, Streeter dysplasia, Torpin dysplasia, amniotic band disruption sequence, acrosyndactyly, fenestrated syndactyly and congenital amputation syndrome [1]. The syndrome is characterized by bands of ranging severity, always present at birth, that can affect any part of the body with a particular predilection for the upper or lower extremities [2].

The reported incidence in the literature varies from 1 in 1,200 to 1 in 15,000 live births depending on the population sampled and the diagnostic criteria used [2, 3]. When found in conjunction with significant limb involvement its incidence has been recorded as low as 0.19 per 10,000 [4]. No genetic inheritance pattern has been identified but some environmental associations have been postulated. These have included possible links with abnormal gestational histories [5] and oligohydramnios [6].

Aetiology

The aetiology of constriction ring syndrome has been debated for centuries. Hippocrates is attributed with the first reference to a syndrome of encircling fetal membranes that resulted in band formation and amputation of extremities [7]. In 1832 Montgomery gave an account of 'complete ligatures about the arms and 2/3 of the leg' of an infant. He described a theory of intra-uterine threads which can entwine themselves around a developing fetus [8]. Since these accounts three different theories to the aetiology

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of constriction ring syndrome have emerged – the intrinsic theory, the extrinsic theory and that of intra-uterine trauma.

Intrinsic Theory

The intrinsic theory was put forward by Streeter, the director of embryology at the Carnegie Institute in 1930 [9, 10]. In contrast to the explanation offered by Montgomery, Streeter theorized the ring constrictions were localized areas of imperfectly formed tissue due to 'defective areas of germ plasm' [2, 9]. On histological examination of the tissue areas of degeneration were found that appeared to arise from the normal tissue at the base of the rings suggesting a primary defect of mesenchymal origin [2]. The validity of this theory is supported by the clinical presentation of external constriction rings alongside internal visceral abnormalities, such as diaphragmatic defects and ectopic gallbladders.

Extrinsic Theory

This alternative theory, first suggested by Hippocrates was re-introduced by Torpin in 1965. He postulated that spontaneous rupture of or damage to the amnion during the first and early second trimester can result in the formation of amniotic strands [11]. These mesodermic strands become entangled with the foetus and in particular the extremities within the sac. In addition resultant transient oligohydramnios caused by a loss of amniotic fluid can lead to abnormal pressure on developing limb buds resulting in deformities such as congenital talipes equinovarus (CTEV) [12]. This theory is supported by a growing body of evidence. A higher rate of entrapment has been noted in the longer central fingers and corresponding longer great toe [13]. Fibrous bands have been discovered at sites of constriction rings at birth [10, 14–16]. Documented evidence exists of the delivery of amputated foetal parts [17]. Furthermore reports exist of amputated parts such as digits becoming ectopically engrafted on the body of the foetus [18, 19]. Bands have also been seen to cause neural dysfunction distal to the site of the stricture, in some cases with vascular compromise, implying an extrinsic pressure effect [20, 21]. Serial ultrasonographic and doppler studies have also demonstrated evidence of strangulation leading to reduced blood supply and ultimate resorption of a limb subjected to amniotic band formation [22]. All of these cases add weight to Torpin's extrinsic hypothesis.

Intra-Uterine Trauma Theory

This third theory was put forward by Kino in 1975 [23]. Using a rat model Kino demonstrated that excessive uterine contraction, induced by amniocentesis, could result in haemorrhage from the marginal sinuses of digital rays during the first trimester. This intra-uterine traumatic disruption of digital formation was seen to produce a phenotypical appearance of constriction band syndrome in the form of fenestrated syndactyly – acrosyndactyly.

It is now generally accepted that in view of the varied presentation of the condition and the evidence supporting each of the above theories that the aetiology of this disorder may vary from case to case.

Classification

A number of classification systems have been proposed for constriction ring syndrome. The most widely used of these is that described by Patterson – [2], a clinically useful system of four different groups which reflect a range of severity (See Table 11.1).

A later classification put forward by Hall divided the continuum of severity caused by constriction rings into three broad clinical categories – mild, moderate and severe (See Table 11.2). Severity was graded according to depth of tissue involved and disability in terms of the presence of lymphoedema and distal amputation [24]. Hall did however emphasise the continuous spectrum of pathology that exists across these three categories.

Table 11.1 Patterson classification of constriction ring syndrome

Type	Description
1.	Simple ring-constrictions
2.	Ring-constrictions accompanied by deformity of the distal part with or without lymphoedema
3.	Ring-constrictions accompanied by fusions of distal parts ranging from mild to severe acrosyndactyly
4.	Intra-uterine amputations

With permission from Ref. [2]

Table 11.2 Hall classification of congenital ring constrictions

Type	Extent of involvement	Disability
Mild	Partial thickness, subcutaneous tissue	No disability
Moderate	Full thickness, subcutaneous tissue	Lymphoedema
Severe	Deep tissue, can include bone	Intra-uterine amputation

With permission from [24]

On the whole distal tissue necrosis as a result of constriction rings tends to be completed and healed during development of the foetus *in utero*. In some cases a picture of delayed healing can be seen after birth – leading to worsening constriction, progressive vascular compromise and subsequent tissue loss [25]. Salvage of distal parts is possible during this phase through decompression and staged reconstruction. The classification systems of Patterson and Hall do not refer to this 'intermediate stage'. Therefore Weinzweig in 1994 put forward a revised classification incorporating this dynamic stage of lymphatico-venous/arterial compromise [25] (Table 11.3).

Table 11.3 Weinzweig Classification of the "Intermediate" Stage of Constriction Ring Syndrome

Type	Description
1.	Mild constriction (no lymphoedema)
2.	Moderate constriction with distal deformity, syndactyly, or discontinuous neurovascular or musculotendinous structures without vascular compromise **A**. Without lymphoedema **B**. With lymphoedema
3.	Severe constriction with progressive lymphaticovenous or arterial compromise **A**. Without soft tissue loss **B**. With soft tissue loss
4.	Intra-uterine amputation

With permission from [25]

Associations

The percentage incidence of associated anomalies varies from 40 to 80 % [1, 3, 10]. These anomalies can be extremely variable and together are often referred to as the 'amniotic band disruption sequence [26]. Almost all associated defects involve the musculoskeletal system with an equal distribution in the upper and lower extremities [1] (Table 11.4). Of these congenital tapiles equinovarus (CTEV) is the most common [5, 10]. The parallel presentation of CTEV and constriction rings has been attributed to increased intra-uterine pressure on the foetus as a result of oligohydramnios [6]. Other limb anomalies appear relatively unrelated such as radial and ulnar dysplasia [27].

In addition to musculoskeletal anomalies a wide range of other associated malformations have been reported (Table 11.4). In a similar way to CTEV, some of these anomalies such as cleft palate as part of Pierre Robin sequence, appear closely linked to the effects of oligohydramnios [28]. Similarly there is a strong association between limb ring constrictions and rare craniofacial clefts again supporting the idea of a common causality [26]. The majority of these facial clefts occur in similar para-median axes and can be extremely disfiguring [26]. The common association between these specific craniofacial clefts and constriction limb anomalies is termed the ADAM complex – amniotic deformity, adhesions, mutilations [29]. Other such syndromic entities involving constriction rings include Adams Oliver syndrome where there is a co-occurrence of paramedian scalp aplasia and acral limb defects and 'limb–body wall complex' in which there is an association with facial clefting, thoracoshisis (a congenital defect in the chest wall that may result in evisceration of mediastinal organs) or abdominoschisis (a congenital defect in the anterior abdominal wall

Table 11.4 Upper and lower limb musculoskeletal anomalies

Associated with constriction ring syndrome
Acrosyndactyly
Syndactyly
Amputations
Lymphoedema
Club foot
Pseudoathrosis of the tibia and fibula
Ulnar dysplasia
Radial dysplasia
Other associated anomalies
Anencephaly
Rare paramedian craniofacial clefts
Ocular and orbital defects
Encephalocoeles
Choanal atresia
Cleft palate as part of Pierre Robin Sequence
Ectrodactyly–ectodermal dysplasia–cleft syndrome
Ear deformations
Abdominal wall defects including abdominoschisis
Thoracic wall defects including thoracoschisis
Scoliosis and kyphosis
Omphaloceole
Absent or dysplastic kidneys
Diaphragmatic abnormalities
Imperforate anus
Cutis aplasia of the scalp
Anaemia
Congenital heart defects

with herniation of abdominal contents) and exencephaly/encephalocoele (a congenital defect in the cranium permitting herniation of intra-cranial tissue) [30, 31] .

Differential Diagnosis

Conditions that are commonly confused with constriction ring syndrome include symbracyhydactyly, vasculocutaneous catastrophe of the new born and transverse growth arrest [32].

Symbrachydactyly refers to a wide spectrum of anomalies ranging from short central digits to complete digital aplasia [33]. Cases are usually unilateral and unlike in constriction ring syndrome the entire hand is usually hypoplastic [34]. In addition the associated syndactyly is usually of the simple rather than acrosyndactylic type. Characteristic features include vestigial digits with nail remnants which helps distinguish this entity from transverse growth arrest [33]. Unlike constriction ring syndrome the disorder can be familial and has been associated with Poland syndrome – a condition whose key diagnostic feature is a congenital absence of the sternocostal portion of the pectoralis major muscle [35, 36].

Vasculocutaneous catastrophe of the new born or congenital Volkmanns ischaemic contracture is a rare condition that can present as an extensive necrotic plaque involving the upper extremity that is often circumferential from the wrist to the elbow [37]. Its aetiology is related to a vascular insult that leads to muscle and nerve injury [38]. At the time of birth cutaneous involvement in the form of bullae, ulceration and necrotic eschar can be seen. This pattern of extensive skin involvement helps distinguish this condition from constriction ring syndrome [37].

Transverse growth arrest of the upper limb represents a spectrum of failure of formation that can present at any level from the shoulder to the phalanges [39, 40]. As both transverse arrest and constriction ring syndrome can present as limb, hand or digital amputations differentiating the two conditions can sometimes be difficult. Distinguishing features include the pattern of limb involvement. In transverse deficiency bilateral cases are much rarer. The most common site of growth failure is amputation at the proximal third of the forearm. Unlike constriction ring syndrome the proximal upper limb in transverse failures of formation tends to be hypoplastic. Furthermore associated features such as acrosyndactyly are not seen in transverse arrest. However in isolated cases of limb amputation the only discernable difference between the two conditions may be the features of the amputation stump. The amputations seen in transverse arrest are often of the disarticulation type and rudimentary digital appendages or nubbins are often found at the end of the limb. Whereas the amputations seen in constriction ring syndrome are more tapering [40].

Clinical Presentation

No two cases of congenital ring syndrome are identical. The deformities are always present at the time of birth and prenatal ultrasonographic techniques enable early diagnosis [22, 41]. More than one limb is usually affected and associated musculoskeletal anomalies are commonly present [2, 7, 40] (Table 11.4). There is a predilection for the more distal extremities – in particular the longer central three digits and longer great toe [13] (Fig. 11.1a). In line with an extrinsic aetiology, involvement of the thumb is rare due to its flexed adducted posture during intra-uterine development making entrapment more difficult. More proximal rings do occur but with much less frequency. However it is rare for only one ring to be present as an isolated abnormality [42]. The severity of the deformity can vary dramatically even between affected limbs of the same individual. The depth of the groove can range from mild indentations with minimal involvement of subcutaneous tissue to deep deficiency with associated disruption of lymphatic channels, nerves and blood vessels [7, 20, 21]. Generally the rings tend to be deeper on the dorsal surface of affected extremities. Deep volar involvement can be associated with tendon involvement and joint contractures [43]. Skeletal involvement is rare because in these cases auto-amputation tends to have already occurred [1]. Amputated parts are usually resorbed during foetal development but can be delivered separately or even found ectopically engrafted on the body [17–19]. Amputations can occur at any level and stumps tend to be tapered (Fig. 11.2) [40]. Sensation tends to be normal and despite the presence of rings and amputations function is generally good as proximally all soft tissue and skeletal anatomy remains unaffected [14].

Most constriction rings have reached their final form *in utero*, occasionally rings and amputation stumps appear still ulcerated and bleeding after birth. Resultant delayed healing can produce further constriction and associated distal swelling, lymphoedema and vascular compromise (Fig. 11.3) [25]. Unimpaired distal swelling may require expeditious decompression to prevent permanent neurological dysfunction and in worse cases distal necrosis. A degree of distal vascular impairment will be manifest in severely affected digits regardless of treatment. This produces a temperature gradient across defects and associated cold intolerance which may be evident throughout patients' lives [10, 42].

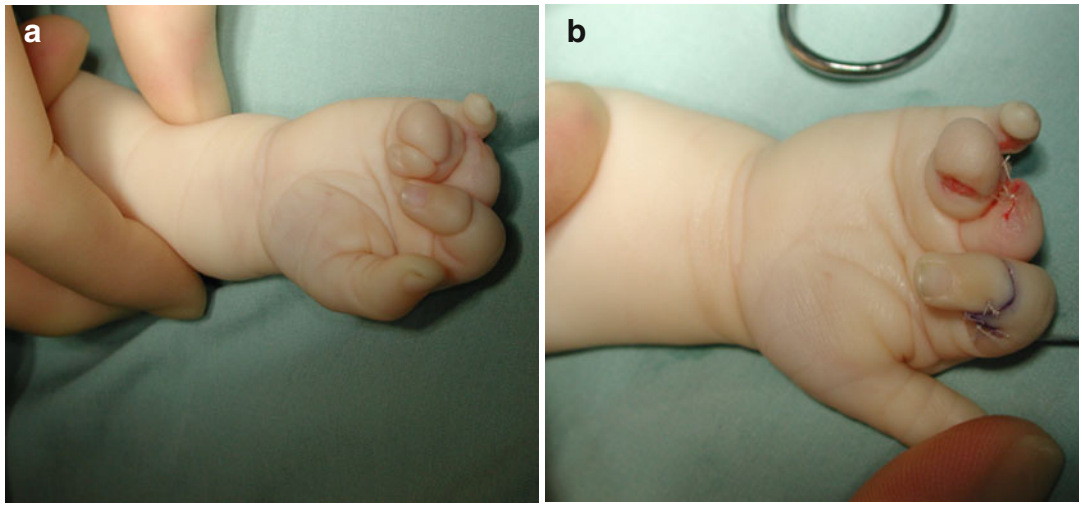

Fig. 11.1 Right hand showing constriction rings affecting the index, middle and ring fingers. (**a**) Pre-operative image showing a circumferential ring affecting the distal index finger and further constrictions producing acrosyndactyly of the middle and ring fingers. (**b**) Intra-operative image showing a one stage circumferential release of the index finger with Z plasties placed in the mid-lateral axes and separation of the middle and ring acrosydactyly

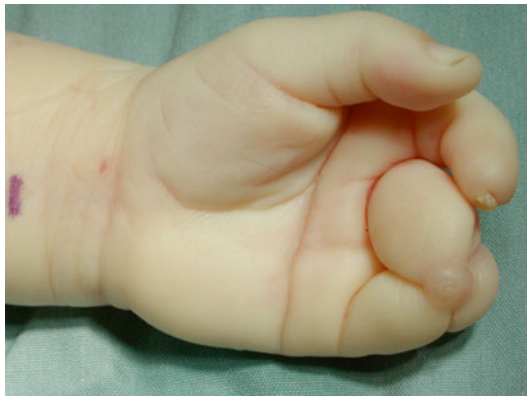

Fig. 11.2 Left hand constriction rings that have resulted in tapered amputations of distal index and middle digital extremities

Fig. 11.3 Gross distal swelling of a thumb affected by a tight constriction ring at its base. The thumb requires expeditious decompression to avoid vascular compromise and subsequent tissue necrosis. Note additional areas of delayed healing at the tips of the central three digits. This represents the delayed intermediate stage as detailed by Weinzweig [25]

Fusion of distal parts is also common in cases of constriction ring syndrome in the form of acrosyndactyly [2]. Distal fusions can be either simple or complex with bony fusions present between adjacent digits. Proximal fenestrations tend to lie characteristically distal to the level of the commissure. Multiple digital coalition can occur with a fibrous union at the site of constriction. Distally the cluster of digital tips have the appearance of a 'bunch of grapes' in which the index finger tip is usually most volar (Fig. 11.4) [42]. Probing of proximal fenestrations can assist pairing of proximal and distal digital segments. Neurovascular supply to these distal parts can be

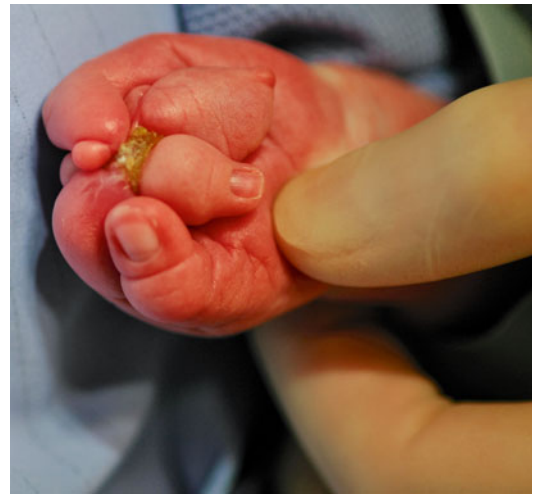

Fig. 11.4 Acrosyndactyly affecting the right hand of a neonate. The cluster of digital tips distal to the fibrous union at the site of constriction have the appearance of a 'bunch of grapes' in which the index finger tip is usually most volar [42]

unpredictable particularly if the ring constriction involves two digits which are crossing over. In these cases separation must be carried out judiciously in stages [1].

Treatment

Timing

Treatment must be individualized to the patient in such a hugely variable condition. Prenatal imaging using ultrasonography allows for early diagnosis, monitoring of progression and planning of treatment. In some cases spontaneous resolution of strangulating amniotic bands has been recorded without any long-term sequelae [44]. In other cases *in utero* lysis of amniotic bands has successfully prevented critical limb ischemia [45]. Throughout pregnancy it is possible to assess limbs for vascular compromise using doppler assessment [46]. The need for prenatal surgical intervention must be weighed against the risks of compromising foetal viability, initiating premature labour or causing maternal injury [1, 47].

In the neonatal period the main treatment that may be required is the division of small soft tissue

bridges between digits and the thumb to ensure unrestricted growth [1]. This procedure can be effectively carried out under local anaesthetic.

Clinical Pearl
Release of small distal soft tissue bridges (formed as a result of acrosyndactly) can be done under local anaesthetic in the neonate (<3 months)

In some instances it may be possible to see fibrous bands encircling the fingers and toes of newborns. These can be simply removed or unwound to release the tethered digits [10]. Occasionally ring constrictions can produce severe distal oedema and vascular compromise particularly in cases of delayed healing in the postnatal period (Fig. 11.3). These cases represent an emergency situation which requires expeditious decompression in the early neonatal period in order to prevent impending tissue ischaemia [25]. Hemi-circumferential decompression can yield a dramatic improvement in the status of the distal extremity. Once the decompression has had time to take effect resultant redundant skin and the remaining half of the ring can be dealt with as a delayed second stage.

Clinical Pearl
Severe distal oedema and venous congestion requires expeditious decompression. This can be achieved through a hemi-circumferential dorsal release of the tight constriction ring.

Similarly in the absence of critical compression the debulking and reconstruction of lymphoedematous swelling can be effectively carried out during infancy. At this time the tissue is swollen with interstitial fluid and not as affected by scarring, hence facilitating surgical excision [1].

Separation of syndactylies and correction of constriction rings, as described below, are best undertaken within the first year of life. Bilateral procedures and those treating upper and lower limbs simultaneously are possible but best performed before the child is ambulatory. More complex procedures such as toe to hand transfers, skeletal lengthening and digital transpositions can be delayed to optimize patient and family compliance and the size of surgical anatomy [1].

Single Versus Staged Surgery

For digits that are circumferentially involved the traditional approach has been a staged release and reconstruction of no more than 50 % of the ring at one time to avoid vascular embarrassment to the distal segment [42, 48, 49]. More recently however authors have advocated routine circumferential band release [14, 24, 50–52]. Generally it is now well accepted that a single stage circumferential release is safe but that staged corrections are more appropriate for deep rings or those associated with pronounced distal swelling [42].

Clinical Pearl
It is now considered safe to perform circumferential surgical release unless rings are very deep or the distal limb compromised

Techniques for Correction of Constriction Rings

Traditional methods of correction involved excision of grooves, mobilization of adipose tissue and re-approximation of skin using Z-plasty or W-plasty based techniques [42, 49]. However resultant scars have a tendency to indent with time and occasional partial loss of flaps and resultant delayed healing can result in a conspicuous depressed saw tooth appearance [14].

Clinical Pearl
To ensure complete release and an optimal post-operative contour excision of all the atrophic scar tissue along the side walls (up to the shoulders) of a

constriction ring is essential. Gentle compression of one marked side against its opposing unmarked partner guides correct incision placement on the shoulders of the ring.

The ideal correction of a deep constriction ring should address the tight scar at the base of the indentation, atrophic tissue along the side walls and indurated fatty tissue within the distal swollen segment [1]. A method described by Upton in 1991 ensures complete excision of the band of constricting scar tissue and long-term contour improvement. It also incorporates Z-plasty corrections to further minimize recurrence but places them in less conspicuous mid-lateral locations. Transverse incisions are first marked on the either side of the depressed area on the limb or digit. These are marked on the shoulders flanking the constriction to ensure complete removal of all scarred tissue from within the ring (Fig. 11.5) Gentle compression of one marked side against its opposing unmarked partner guides correct incision placement. All scarred tissue is then excised from within the depth of the ring preserving large dorsal veins, volar neurovascular structures and flexor tendons. Initially all native skin on either side is preserved. Occasionally dorsal veins and extensor tendons are absent but prominent venae comitantes are found volarly. The remaining tissue on either side of the excision is then elevated in two planes. The superficial layer of skin and some superficial fat is separated first. The deeper layer of areolar tissue is then raised off the extensor tendon dorsally and flexor sheath volarly. Excess fatty tissue, usually most apparent distally, can then be excised and the remaining deeper tissue can be redraped to fill the annular contour defect. In this way final closure of the layers can be staggered to minimize the effects of vertical scar contracture. Constriction on the volar surface tends to require less debulking. Excess tissue on either side of the digit is then excised and Z-plasties are carried out ensuring transverse limbs of the correction are placed along mid-lateral axes (Fig. 11.1b). In designing these Z-plasties it is recommended that the dorsal skin triangular flap be mobilized into place first before the committing to incision of the less mobile volar skin. Deeper rings with significant distal swelling can be approached using this technique but in a staged manner [1, 14].

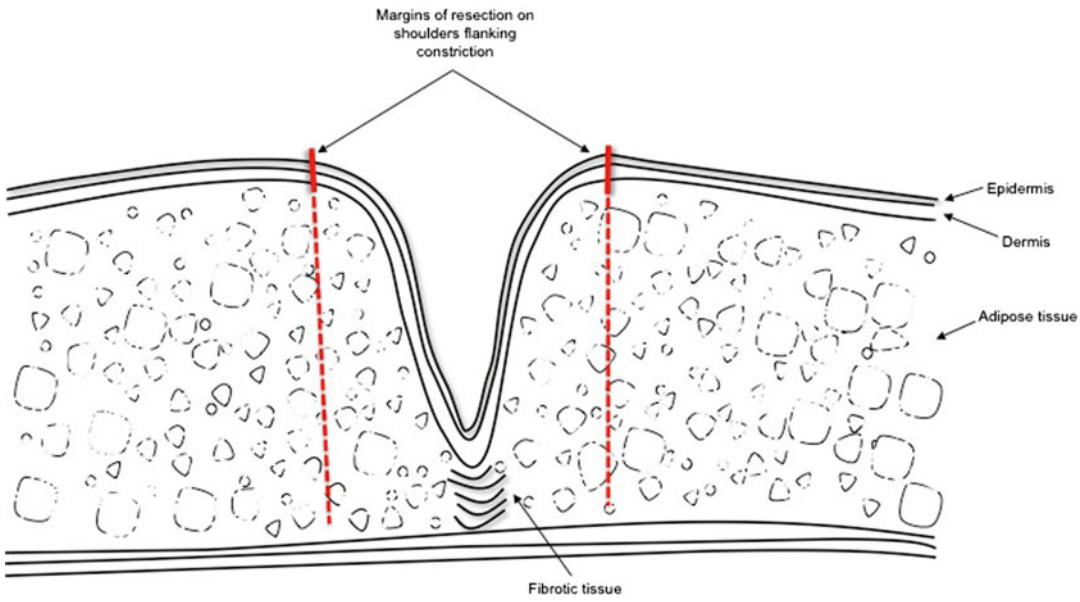

Fig. 11.5 Schematic demonstrating excision margins for correction of ring constriction. Transverse incisions are made on the shoulders flanking the constriction to ensure complete removal of all scarred tissue from within the ring

Clinical Pearl

Once scarred tissue within a ring has been excised, elevation of the remaining tissue on either side can be carried out in two layers – superficial skin and subcutaneous fat and a deeper layer of areolar fat. The deeper layer can then be redraped to fill the annular defect (left after excision of the ring) and a staggered closure of the two layers will reduce chances of the scar re-tethering in the future.

Treatment of Acrosyndactyly

Acrosyndactyly is the characteristic form of digital fusion seen in constriction ring syndrome [2]. The distal syndactyly is usually comprised of scar tissue alone. Multiple digits can be joined together in this manner giving a peaked appearance to the hand. Proximal epithelial-lined fenestrations penetrate from what can appear as dorsal dimples through to the volar surface [10, 42]. Often an initial early release is possible and can be achieved by following the fenestrations in a straight line distal release. This early maneuver can be invaluable produce a surprising increase in digital length.

In terms of web space reconstruction it is important to recognize that sinuses and fenestration do not reflect the correct position of the proximal commissure. In addition as the tissue in these windows is often macerated and of poor quality it is best not incorporated into local flap web space reconstruction.

Clinical Pearl

Macerated skin within the fenestrations of an acrosyndactyly should ideally not be included in local flap web reconstruction as it is of poor quality and likely to breakdown increasing chances of web creep during the healing process.

It is however possible to de-epithelialise discarded skin and use it to improve contour deformities [1]. Some authors recommend initial excision of fenestrations combined with advancement of normal full thickness tissue across the defects to promote venous and lymphatic return from distal swelling. This is then followed by a formal separation of the syndactylised digits as a second stage [1]. Separation of the syndactyly follows the general principles outlined by Flatt [42].

In more complex cases the acrosyndactyly can involve digits that cross over one another. Remnants of digital tips starved of vasculature can protrude like grape-like nubbins from the constricture [42]. In these cases distal neurovasculature can be abnormal and a staged approach is recommended avoiding dissection of both sides of a digit at the same time. Probing of fenestrations can help identify which tip belongs to which proximal digit. Careful proximal identification of blood vessels and nerves is recommended to aid distal dissection.

Clinical Pearl

Placement of probes into proximal fenestrations can help identify which tip belongs to which digits in convoluted distal acrosyndactylies.

Separation of multiple digit acrosyndactyly can also be complicated by the hypoplastic nature of the distal segments involved. In some cases of combined acrosyndactyly and digital amputation, separation may result in thin short digits which do not function as well as the original coalition. In these cases increased functionality must be the primary objective and separation is not helpful.

Treatment of Digital Aplasia or Short Fingers

The components of a basic functional hand are a mobile digit on the radial side, a well lined pliable broad web space with at least one other digit or post on the ulnar side to enable rudimentary pinch and grasp [42]. In most cases of constriction ring syndrome the thumb is usually preserved as it is held protected in an adducted and

flexed position in the palm during foetal development [13]. However transverse amputations of the thumb do occur. Constriction ring amputations of the thumb and digits are ideal candidates for vascularised toe to hand transfers as proximal anatomy including the neurovasculature is normal [53–55].

Clinical Pearl

Constriction ring amputations of the thumb and multiple digits are ideal candidates for vascularised toe to hand transfers as the proximal of tendon and neurovasculature anatomy is usually normal.

These techniques can dramatically improve hand function through the reconstruction of a sensate mobile thumb or digits which will show a degree of commensurate growth with the child [56–58]. Extremely positive long-term results have been seen into adolescense and adulthood in terms of function, appearance, donor site and psychological and social well-being [58]. However this approach can occasionally be limited by a lack of useable toes in the context of constriction ring syndrome.

An alternative approach is the use of distraction lengthening [59, 60]. This method is very effective at restoring a functional prehensile unit but can be very involved process that requires a great deal of patient and family commitment. Indications for surgery must be carefully considered [61]. This method appears to be more effective when used at the metacarpal and forearm levels as the distracted tissue is surrounded by more soft tissue with greater vascularity [1]. Different approaches have been described. Some authors prefer to distract to the desired length and wait until the intercalated gap fills with bone through a process of consolidation [60]. Others feel the consolidation phase is best bypassed as complication rates with such techniques are directly proportional to the length of time the external distraction device is *in situ*. Therefore alternative approaches have been adopted using intercalary bone grafts to fill distracted gaps [62] [1]. Distraction at the phalangeal level is possible but is extremely challenging due to problems of poor skeletal stock and soft tissue coverage.

Clinical Pearl

Distraction lengthening at the metacarpal level produces more predictable results to that at the phalangeal level. This is due in part to a better surrounding soft tissue envelope.

In cases of index finger or thumb insufficiency in the context of constriction ring syndrome, a third treatment option is possible. Transposition of a redundant digital tip based on its neurovascular pedicle as an ‘on-top’ plasty including its proximal phalanx can lengthen digits improving length for prehension [63]. Transposition of a short non-functioning index finger in this way can also broaden a first web space. This technique can be combined with distraction lengthening to further maximize length of the transposed skeletal segment [1].

Complications

Early complications associated with surgical correction of constriction ring syndrome include infection, haematoma, flap necrosis and distal vascular compromise. Care must be taken in view of the abnormal vasculature architecture and significant soft tissue induration and lymphoedema that may be present. Careful planning, meticulous surgical technique and adequate post-operative immobilizations are key to success. Long-term problems of sensory loss are not usually detectable until the child is older. Problems associated with skeletal manipulation and lengthening are more common in cases of poor bone stock and insufficient soft tissue coverage. Children and parents must be counseled appropriately and understand that with growth as with other congenital conditions further interventions may be required to preserve optimal function.

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Congenital Anomalies of the Hand

12

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Keywords

Congenital anomalies • Hand malformations • Antenatal ultrasound screening • Hand embryology • Ossification centres of hand

Introduction

This chapter will cover the embryological development of the upper limb, in particular the salient congenital hand malformations but mainly focusing on skeletal abnormalities. The diagnostic role and merits of antenatal sonographic evaluation and postnatal imaging will also be discussed.

Prevalence

The prevalence of upper limb anomalies in new born babies is around 0.2 % [1]. In one Australian study, a high proportion (46 %) of those affected had another non-hand congenital anomaly, 51 % bilateral hand anomalies, and 17 % suffered from multiple different hand anomalies. Congenital upper limb anomalies were shown to be more common in boys; preterm, post-term, multiple births and older mothers. Ethnicity, sidedness and survival were not significant variables.

A large Swedish population study of infants born after IVF (n > 9,000) has indicated increased risk of foetal abnormality in babies resulting from assisted conception (risk ratio 1.47) but without specified increased risk of limb anomaly [2]. The overall excess risk could be explained by increased maternal age and other maternal characteristics (such as >4 years involuntary childlessness).

Classification

There is an international classification [3] for congenital hand anomalies based on extension of an earlier classification system proposed by Swanson [4]. Congenital hand malformations are divided into the following seven groups:

I. Failure of formation
II. Failure of differentiation
III. Duplication
IV. Overgrowth

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V. Undergrowth
VI. Amniotic band syndrome/constriction bands
VII. Generalized skeletal syndromes

Any remaining entities are grouped as "unclassifiable" for example carpal fusion and Madelung's deformity.

In one series, the most common anomalies were failures of differentiation (35 %), duplications (33 %), and failures of formation (15 %) [1]

Recent studies have clarified the roles of different molecules and their interactive pathways. This knowledge of developmental biology has provided further insight into the aetiology of many limb malformations. Based on this knowledge, Oberg et al have advocated an updated scheme for the above classification [5].

Embryology

It is important to understand that the conventional embryological description is based on the conceptual age, this is to be distinguished from the gestational or menstrual age used in the clinical setting. The gestational or menstrual age is defined as being from the first day of the last menstrual period and is therefore the conceptual age plus 2 weeks.

The embryological sequence of upper limb development is summarised in Table 12.1. The most critical period for the development of upper limb anomalies is from 24 to 36 days, when the influence of teratogens is most potent [6]. The first sign of an upper limb bud are shown in the fourth week of embryological life (approx. Day 26).

Clinical Pearl

Embryological description is based on conceptual age which is gestational age minus 2 weeks

Most critical period for development of upper limb anomalies is from 24 to 36 days

The upper limb develops from the somatic mesoderm that forms the mesenchyme and is, in turn, covered by ectoderm. The ectodermal cells multiply and condense to form a thickening on the ventromedial border of the distal limb bud called the Apical Ectodermal Ridge (AER). A layer of proliferating mesenchymal cells underneath AER is called the Progress Zone (PZ) and both are vital in inducing limb growth and differentiation [5, 7] (Fig. 12.1).

Embryological knowledge has evolved from purely anatomical understanding to encompass genetic expression and chemical signalling that controls progression and differentiation [5] (Fig. 12.2).

Table 12.1 Overview of upper limb and hand embryology [6]

Menstrual age	Conceptual age	Anatomical development arm	Anatomical development hand
6th week days 40–41	4th week days 26–27		Limb buds appear – lower before upper by 1–2 days
7th week	5th week	Mesenchymal skeleton forms followed by chondrofication centres	Mesenchymal skeleton forms followed by chondrofication centres
End 8th week	End 6th week	Limb skeleton (cartilage) is complete	Digital rays formed from mesenchyme to form hand
9th week	7th week	Ossification starts	
End 10th week	End 8th week		Separate fingers have formed
10 weeks	8 weeks	Elbows face caudally	

Clinically the *menstrual age* is used, this should be used here also to ensure understanding
The sixth week = 5^{+1} weeks – 6^{+0} weeks GA
Thus the eighth week refers to an embryo that is 7+ weeks GA
Critical period of development of the upper limb (and thus that which is most vulnerable to teratogens) is between days 24–42 (menstrual days 38–56, 5^{+3} – 8 weeks GA)

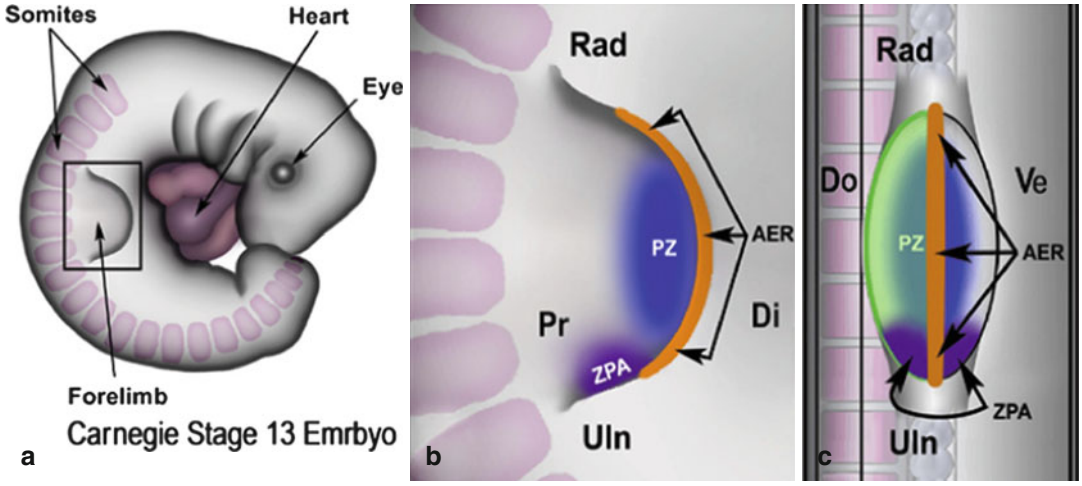

Fig. 12.1 Limb bud coordinate axes and signaling centers. (**a**) The forelimb (*boxed region*) of a Carnegie stage 13 embryo depicting the 3 coordinate axes—each with their own signaling center: the apical ectodermal ridge (*AER—orange*) coordinating proximal-distal (*Pr-Di*) outgrowth and differentiation; radial-ulnar asymmetry is controlled by the zone of polarizing activity (*ZPA—purple*). Dorsal-ventral (*Do-Ve*) asymmetry is regulated by dorsal ectoderm (*green*); within the progress zone (PZ—blue) the fate of mesodermal cells is determined by these signaling centers. The axes and signaling centers are shown in different orientations: (**b**) dorsal view and C lateral, end-on view. Figure and legend reprinted with permission from Oberg et al [5]

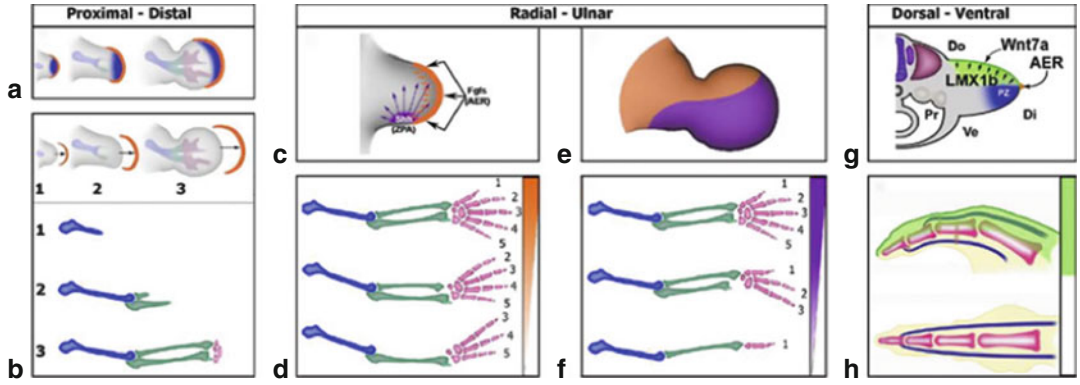

Fig. 12.2 Disruption of axis formation/differentiation. (**a**–**d**) Impact of signaling centers on normal development. (**a**) Fibroblast growth factors (*FGFs*) from the AER (*orange*) initiate and maintain limb outgrowth. Formation of skeletal elements progresses from proximal to distal, shown in 3 progressive stages that depict arm, forearm, and hand formation. (**b**) During limb outgrowth, SHH emanating from the ZPA (*purple*) establishes an "ulnarizing" gradient (*purple arrows*) and maintains posterior (ulnar) proliferation. In addition, FGFs (*orange arrows*) secreted from the AER promote proliferation in the underlying progress zone contributing to limb volume and width. (**c**) Relative contributions of SHH (*purple*) and FGFs (*orange*) to skeletal elements. (**d**) Dorsal–ventral section of an embryo through the forelimb, illustrating the regulation of LMX1B (*light green*) by WNT7A (*green*). (**e–h**) Failure of axis formation/differentiation. (**e**) Abrogation of AER/FGF function, as depicted in the top panel, leads to transverse defects that correspond to the time of disruption. (**f**) Progressive reduction of FGF function leads to loss of radial structures. (**g**) Loss of SHH function leads to loss of ulnar-associated structures. (**h**) Disruption of LMX1B is associated with loss of dorsalization (Figure and legend reprinted with permission from Oberg et al. [5])

In the 5th week, mesenchymal condensations called blastemas start to form cartilage anlage (framework of hyaline cartilage that is precursor of future bone development) of the arm bones. This process extends proximal to distal and by day 41, digital rays begin to delineate in the hand starting from the border rays. Ossification starts in the diaphyseal primary ossification centres in the seventh week. Two common muscle masses also form during 7th week, starting as condensations of cells derived from dermatomyotomes. AER begins to fragment and notched digital rays are formed by day 46. Webbed digits separate out by the end of the 8th week as a result of apoptosis within digital interspaces. Throughout the foetal period (8 weeks onwards), the muscles and neurovascular structures continue to differentiate and develop.

Clinical Pearl

Complete differentiation into recognisable upper limb is achieved by the end of the eighth week of conceptual age

Muscle and neurovascular differentiation continues throughout foetal life

Prenatal diagnosis

During the course of his day-to-day practice an upper limb surgeon will encounter congenital abnormalities amongst which will be cases not identified before birth. It may be that the parents & child, and indeed the surgeon himself, will ask why this was not seen on the ultrasound scan done during the pregnancy. This section aims to provide an understanding of the sensitivity of prenatal ultrasound in the detection of upper limb abnormalities under two separate sets of conditions. First, the conditions under which most prenatal sonograms are performed as a screening examination carried out by a sonographer to a protocol in a fixed time slot and, second, ideal conditions when the examination is performed by an experienced and highly trained sonologist with little constraint on his/her time.

Clinical ultrasound practice varies between countries; in the UK there is an NHS-run prenatal screening programme defined by the National Screening Committee (NSC) and subject to annual audit [8]. The thrust of the NSC programme for the detection of structural abnormalities is in the second trimester; a scan is also performed in the first trimester to date the pregnancy and screen for chromosomal abnormalities, especially trisomy 21. North America run a similar programme to the UK but with State variations, the scans are performed to a standard set by the American Institute of Ultrasound in Medicine (AIUM). The practice throughout non-UK Europe and the rest of the world is variable; those countries that offer prenatal screening are encouraged to adhere to guidelines and standards published by their own national professional bodies. Internationally accepted guidelines are published by professional bodies such as the World Federation of Ultrasound in Medicine and Biology (WFUMB) and the International Society of Ultrasound in Obstetrics and Gynecology (ISUOG).

Clinical Pearl

Ultrasound screening programme for foetal anomalies in UK recommends:

Dating scan – between 11+0 and 14+1 weeks

Anomaly scan – between 18+0 and 20+6 weeks

Given the huge range of structural anomalies that may be detected, their varied prognoses and the difficulty in attempting to look for all anomalies due to constraints imposed by limited resources (including skill and time), the NSC has recommended that the second trimester scan targets lethal chromosomal anomalies (trisomies 13 & 18) and nine specific structural anomalies (see Table 12.2). The abnormalities have been selected as they all have a poor prognosis being either lethal, life-limiting or seriously disabling. The only skeletal abnormality listed is that of lethal skeletal dysplasia with a target detection

Table 12.2 The 11 auditable conditions and detection rates

Conditions	Detection rate (%)
Anencephaly	98
Open spina bifida	90
Cleft lip	75
Diaphragmatic hernia	60
Gastroschisis	98
Exomphalos	80
Serious cardiac abnormalities	50
Bilateral renal agenesis	84
Lethal skeletal dysplasia	60
Edwards' syndrome (Trisomy 18)	95
Patau's syndrome (Trisomy 13)	95
Down Syndrome (trisomy 21)	90[a]

The NHS Foetal Anomaly Screening Programme has agreed that only 11 conditions may be detected by an ultrasound scan and these have limited detection rates, as derived from a number of major reference sources
[a]90 % detection rate for a screen positive rate (SPR) of 2 % for combined screening

rate of 60 %. This represents a shift in emphasis of the screening second trimester scan and is explained to the women who are required to 'opt in' to this ultrasound screening programme. The following is clearly stated in the guidance document [8]:

> It is important that both women and health professionals appreciate that the scan is a screening test and because of that it has limitations. Inevitably some conditions will be missed or misidentified. Women should receive comprehensible information before the scan and if a woman chooses to decline the screening test then this must be respected.

The screening second trimester scan includes a review of the limbs. The sonographer is required to measure one femur as part of the biometric foetal assessment and to check that there are four limbs. The sonographer identifying metacarpals and metatarsals in each of two upper and two lower limbs achieves the latter requirement. It must be emphasized that this is the minimum accepted standard for review of the limbs. The sonographer is NOT required to (a) count the digits or (b) check that all three long bones are present in each of the four limbs in order to complete the second trimester screening scan.

Clinical Pearl

Limb assessment on the Anomaly Scan in second trimester includes:
- Femur length (FL) measurement in one lower limb
- Confirmation of four limbs (e.g. by identification of metacarpals and metatarsals in each limb)

Trained, competent and experienced sonographers will make additional observations during the screening scan relating to posture and movement. They will also make a subjective assessment of the presence and relative proportions of the long bones of the limbs, their modelling and ossification thus permitting the identification or suspicion of phocomelia, postural deformities, arthrogryposis, non-lethal skeletal dysplasias, genetic syndromes (eg Meckel Gruber which can be associated with short curved femora) and radial ray anomalies to name a few. Therefore whilst there is the potential for prenatal diagnosis of many skeletal abnormalities their identification is neither a requirement nor expected of the screening programme. This is important when considering why skeletal abnormalities are 'missed' on prenatal screening ultrasound.

Following the identification or suspicion of a skeletal abnormality by the sonographer during the screening scan the parents are referred to a specialist Unit for a further scan by a Consultant (usually an Obstetrician in Foeto-Maternal Medicine but possibly a Radiologist) with higher specialist training in foetal ultrasound, invasive testing and counselling. This constitutes the second set of conditions indicated in the introduction. At this scan a comprehensive survey of the skeletal system is performed in great detail covering structure (both bone and soft tissue), movement and posture together with a full survey of the remainder of the foetus, placenta and liquor. This comprehensive survey is important as many foetuses will have abnormalities in other body systems leading to the suspicion of either a chromosomal or syn-

dromal abnormality [9]. From this scan information the Specialist constructs a diagnosis (or differential) and prognosis and explains this to the parents together with options for further investigation and options for the pregnancy, including termination if appropriate and within that country's laws on termination of pregnancy. The prenatal diagnostician now has two further diagnostic tools at his/her disposal, 3-D ultrasound (3DUS) and in utero magnetic resonance imaging iuMRI. The former, whilst not shown objectively to improve diagnosis per se, can be helpful in defining the position and posture of the arm and hand and is helpful in explaining the appearance of the defect to the parents [9]. iuMRI is of limited added value to ultrasound in the evaluation of upper limb defects and is not routinely used.

We shall now look at what detail may be observed outwith the context of a screening examination given specialised scanning skills, appropriate equipment and sufficient time. Such information will be helpful in informing the timing and utility of ultrasound in a pregnancy subsequent to the index case. First we need to refresh our knowledge of the embryology of the limbs (Table 12.1) as an understanding of the normal sequence and timing of limb development is essential before being able to make a diagnosis of abnormality [6, 7]. The ability of ultrasound to image the upper limb is influenced by a number of factors:

1. The **equipment** used must have the spatial resolution to permit visualization of the detail of the bone and soft tissues.
2. The **operator** (the sonographer or medical consultant) must have higher training and expertise to obtain the images.
3. The **woman** herself as, if she is obese, then image quality is compromised.
4. The **abnormality** as some abnormalities are more readily imaged and recognizable than others and the presence of multiple abnormalities in a case further raises the likelihood of detection 1 [9].
5. The **gestation** at which the scan is performed with the best imaging window for comprehensive system review being between 16 and 24 weeks' gestation with the peak at 20–22 weeks.

At the time of the first trimester scan (between 11+0 and 14+1 weeks' gestation) the upper limb has a complete cartilaginous skeleton and ossification has commenced. The hand and separate digits would have developed (Table 12.1; Fig. 12.2). It is therefore possible, under ideal conditions, to look for limb reduction abnormalities or for complete radial aplasia with associated hand anomaly in a previously affected family before 15 weeks' gestation. A retrospective study of 100 consecutive cases of first trimester scans in a high risk population gave sensitivity of 60 % in the detection of upper limb anomalies [10]. It would not be possible, other than in exceptional cases, to reliably count digits or to look for conditions such as syndactyly or clinodactyly at a first trimester scan. At the second trimester scan it should be possible to reliably image the upper limb in its entirety with detailed analysis of the bony skeleton (Figs. 12.3), the soft tissues, the digits, limb movement and posture. The two most likely deterrents to achieving this level of resolution are low volume or absent liquor and maternal obesity. In a series of 100 consecutive cases (GA 13–33 weeks), only 12 cases were isolated (i.e. sonographically diagnosed abnormality confined to the upper limb) and there was a strong link between upper limb abnormalities diagnosed prenatally (Figs. 12.4, 12.5, and 12.6) with both aneuploidy (28 %, majority trisomy 18) and non-chromosomal syndromes (67 %) [9].

Clinical Pearl

88 % of foetuses with upper limb anomalies have associated structural or chromosomal anomalies

Most common associations are aneuploidy/trisomy and non-chromosomal syndromes

Many of these families will be offered (and frequently accept) termination of the pregnancy while others will have increased risk of foetal death in utero and perinatal death. Consequently,

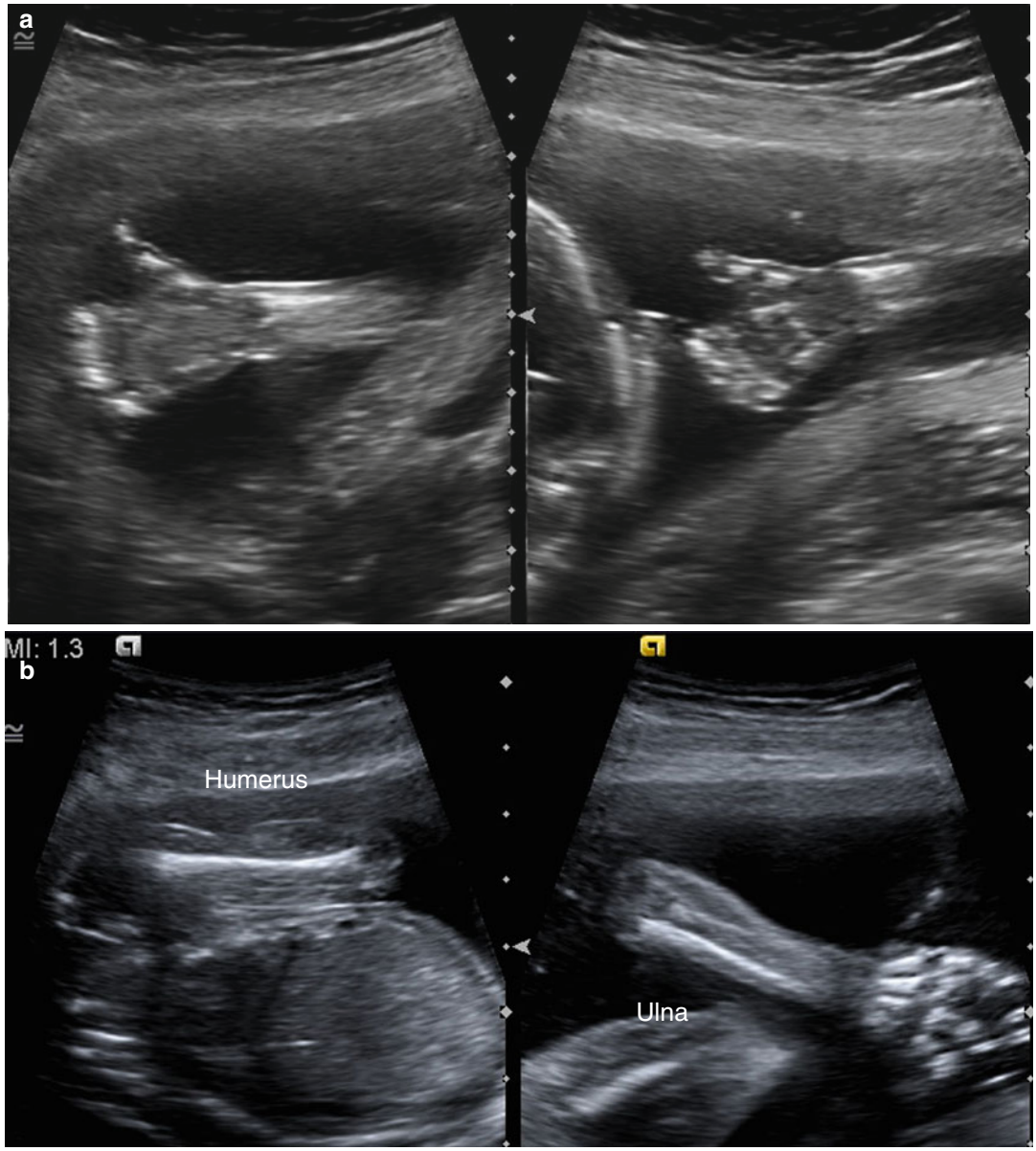

Fig. 12.3 Prenatal sonogram at 20 weeks shows normal appearance of distal forearm bones and hand bones including metacarpals/phalanges (**a**). Humerus, ulna and proximal radius are also well visualised (**b**)

the cases seen post-natally represent the 'tip of the iceberg' of all upper limb abnormalities seen in the pregnant population.

In summary, it is hoped that this section has enhanced the understanding of the prenatal screening programme in existence in UK and the wider world and also of the role of ultrasound in screening any subsequent pregnancy for the detection of recurrent abnormality.

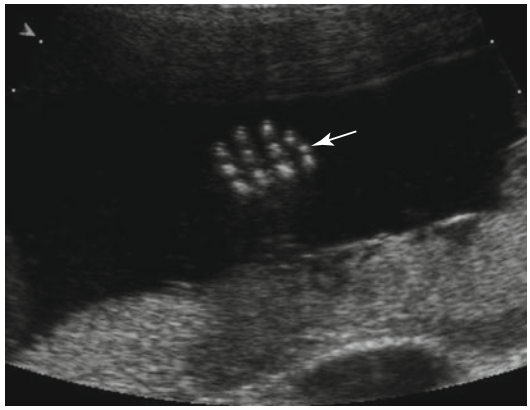

Fig. 12.4 Anomaly scan at 20 weeks gestation shows an incurved little finger in keeping with clinodactyly

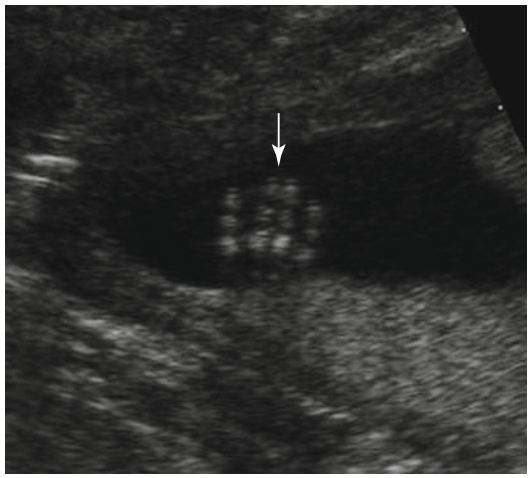

Fig. 12.5 Anomaly scan in another foetus reveals closely apposed third and fourth digits representing central soft tissue syndactyly

Post-natal Imaging Work Up

X-ray evaluation alongside physical examination suffices in most cases of congenital hand anomalies.

Ultrasound and MRI are helpful in the evaluation of un-ossified cartilage anlage and associated musculo-tendinous structures in cases such as polydactyly and transverse deficiency.

A significant number of foetuses with hand anomalies have other syndromic features and many are terminated. Such foetuses are assessed by post-mortem whole body x-rays (foetogram) using anterior-posterior and lateral projections. Separate views of hands and feet may be required if they are inadequately imaged on whole body views.

Table 12.3 shows timing of appearance of different primary and secondary ossification centres in wrist and hand.

Important Congenital Skeletal Hand Anomalies

The spectrum of congenital hand malformations is wide. In association with syndromes, the commonest hand anomalies are polydactyly and syndactyly. Polydactyly is usually preaxial or postaxial but occasionally may be central. Syndactyly can be due to either bony or soft tissues fusion at different levels. Other anomalies which are less severe and often recognised later on include delta phalanx (often resulting in clinodactyly) or isolated short metacarpals. Failure of formation – longitudinal with radial/ulnar dysplasia (previously club-hand) is a more severe spectrum of abnormalities involving either radial or ulnar side of the hand and forearm.

Syndactlyly

Syndactyly represents either bony or soft tissue fusion of multiple fingers. It is often associated with other congenital anomalies and as part of several syndromes e.g trisomy (13, 18 or 21), Apert's syndrome, Poland's syndrome, cranio-facial syndromes (e.g. Mobius – more often missing digits). Delta phalynx is often associated with acrosyndactyly

Depending on severity and extent, syndactyly can be detected on antenatal sonography (Fig. 12.5).

Polydactyly

Polydactyly is one of the commonest congenital hand anomalies. It is usually pre-axial or post-axial in location but can occasionally be central. Post-axial polydactyly is more common than the other two varieties (Fig. 12.7). When genetically determined, it is usually autosomal recessive in inheritance pattern and associated with a syndrome. It may represent a simple soft tissue growth (Stelling type 1) through to a well developed digit (type 3). Syndromes associated with polydactyly include short rib-

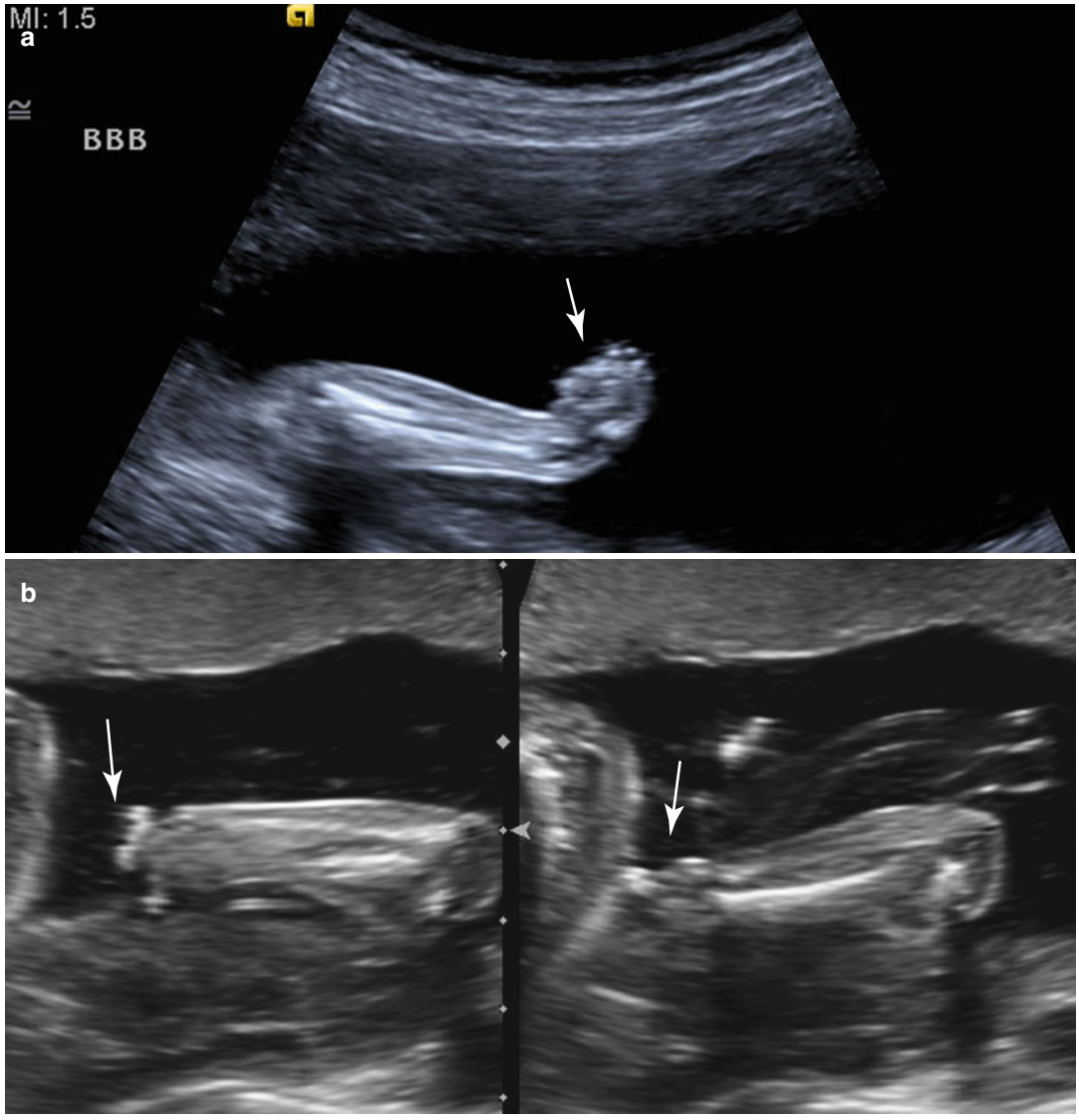

Fig. 12.6 Prenatal sonograms at 20 weeks gestation showing two examples of transverse deficiency at two different levels, mid hand (**a**) and wrist (**b**)

polydactyly syndromes (SRPS) type 1 and 2, Carpenter's syndrome, Ellis –van Creveld syndrome, Meckel-Gruber syndrome, asphyxiating thoracic dystrophy, chondroectodermal dysplasia and otopalatal digital syndrome. A variation of polydactyly includes the triphalangeal thumb.

Triphalangeal Thumb

Triphalangeal thumb (TPT) represents a long finger-like thumb with three phalanges and is regarded as a subtype of preaxial polydactyly. It can occur as a sporadic disorder, but is more often seen as a dominant familial trait [11]. It is often associated with a degree of radial deviation and frequently a longer than normal first metacarpal, hence metacarpal shortening is one effective way of dealing with the abnormal thumb length.

Triphalangeal thumb may be associated with delta phalanx, polysyndactyly and other phalangeal abnormalities of the fingers e.g uniphalangeal and biphalangeal fingers. (Fig. 12.8).

Table 12.3 Timing of appearance of different ossification centres of the hand

Ossification centre	Time of appearance
Metacarpals – primary	9th intrauterine week
Phalanges – primary	9th intrauterine week
Metacarpals – secondary[a]	2 years
Phalanges – secondary	2 years
Capitate[b]	1 year
Hamate	2 years
Triquetral	3 years
Lunate	4 years
Scaphoid	5 years
Trapezium	6 years
Trapezoid	7 years
Pisiform	10–12 years

[a]All metacarpals have secondary ossification centres in their heads except the first metacarpal which has a basal secondary ossification
[b]Carpal bones ossifications (except pisiform) appear in anticlockwise fashion, roughly one centre appears every year starting from the age of 1

Failure of Formation – Longitudinal with Radial or Ulnar Dysplasia

Failure of formation-longitudinal can be divided into radial or ulnar varieties depending on which side fails to develop. The radial variety is associated with variable hypoplasia of the radial ray including thumb (Fig. 12.9). It is often divided into types 1–4 with type 1 being the mild variety and type 4 representing the most severe degree of hypoplasia.

Classified among longitudinal deficiencies, the radial/ulnar ray anomalies comprise of a spectrum varying from partial to complete deficiency of radius and thumb or ulna and medial rays. An animal study has suggested that radial deficiency and ulnar deficiency are caused by a common teratogenic mechanism in humans. However, the critical period for radial deficiency is one day later than that for ulnar deficiency in rats, which probably contributes to the greater frequency of radial deficiency than ulnar deficiency in clinical cases [12].

Important syndromes associated with these anomalies include Holt-Oram, Cornelia de Lange, TAR (thrombocytopaenia- absent radius), Trisomy 18, VACTERL association and Fanconi's anemia. In one study 50 % of patients with radial ray anomalies were syndromic [13]. Unilateral radial aplasia has also been described in 22q11 deletion [14].

Clinodactyly

Clinodactyly refers to incurving of a finger, usually the little finger. It is a relatively common anomaly both in association with syndromes and as an isolated entity (Fig. 12.10). Sometimes it is associated with delta phalanx [15]. It can also be an acquired anomaly due to either trauma or infection.

Delta Phalanx

A delta phalanx is an abnormal phalanx of trapezoid or triangular shape due to an abnormal "C" or "J" shaped epiphysis, hence it is also called "longitudinally bracketed epiphysis" (Fig. 12.11). It has a strong familial association and usually occurs in association with other anomalies such as polydactyly, syndactyly, clinodactyly (usually middle phalanx), triphalangeal thumb (usually proximal phalanx), cleft hand and hypoplastic hand. Triangular deformity of the bone of the index finger usually is associated with Apert's syndrome. The syndromes commonly associated with delta phalanx include Apert's, Poland's and Holt-Oram syndromes [16].

Intercarpal Fusion

Occurs as normal variation in about 0.1 % population and is usually asymptomatic. When isolated it involves carpal bones of the same row but syndromic variety usually affects bones in different carpal rows. Triquetro-lunate fusion is probably the commonest (Fig. 12.12). Syndromes commonly associated with carpal fusion include Ellis-van Creveld, arthrogryposis, diastrophic dwarfism and Turner's. Achondroplasia can also be an association (Fig. 12.13).

Failure of Differentiation – Central Deficiency (Previously Ectrodactyly or Lobster-Claw Hand)

Central deficiency is a rare condition characterized by absence of digits, ranging from single absence of a finger to the split hand or foot deformity [17].

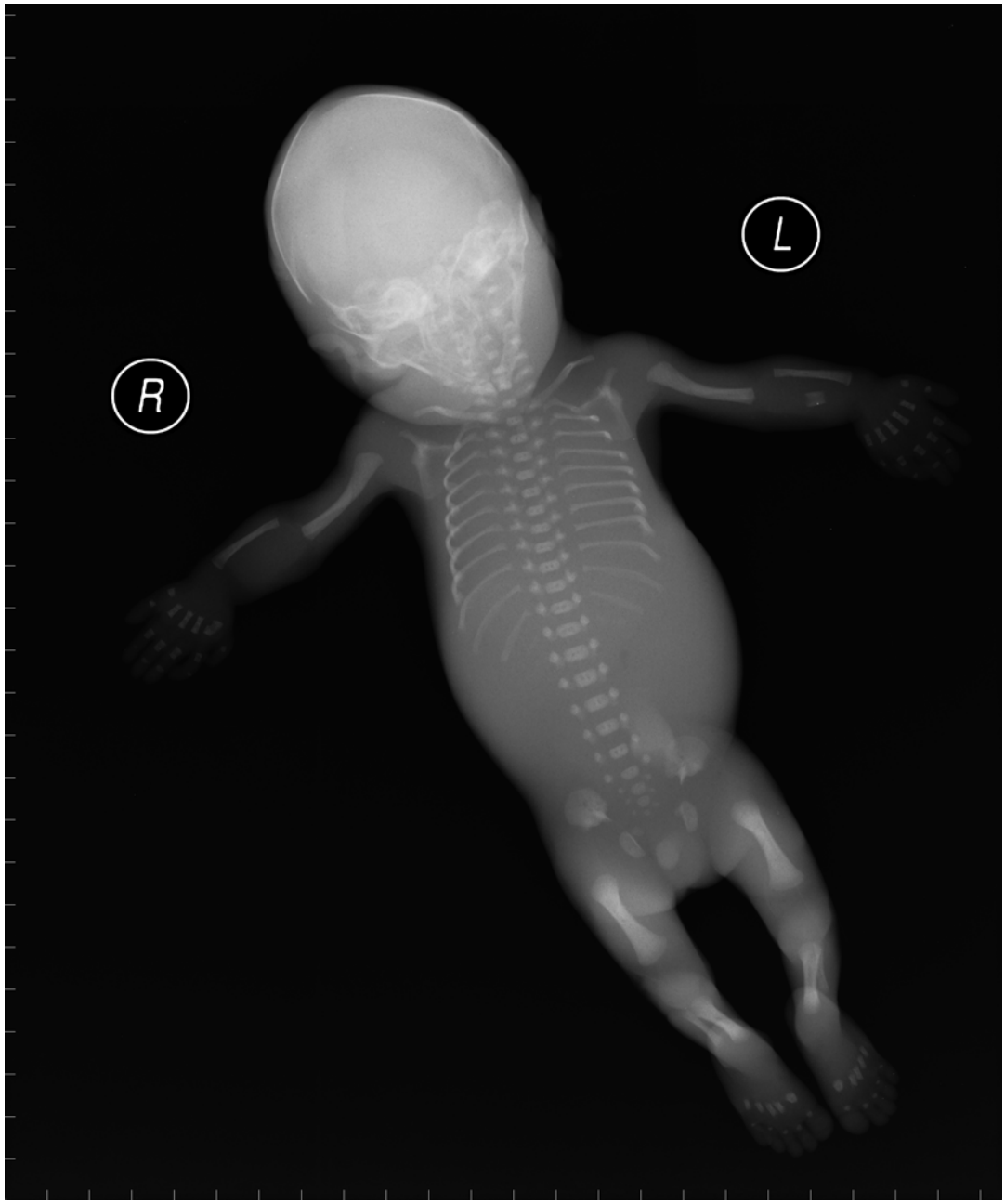

Fig. 12.7 Foetogram of a terminated foetus shows multiple skeletal anomalies including bilateral short upper and lower limbs, curved humeri, short radii, ulnar ossific deficiency and post-axial polydactyly

Ectrodactyly (Fig. 12.14) is one of the three cardinal features of EEC syndrome (ectrodactyly, ectodermal dysplasia and cleft lip with or without cleft palate). It usually occurs as an autosomal-dominant trait or less commonly in a sporadic form [18].

Transverse Deficiency and Congenital Constriction Band Syndrome

Both terms have been used loosely and interchangeably and have certain overlapping and distinctive features. Both represent a form of

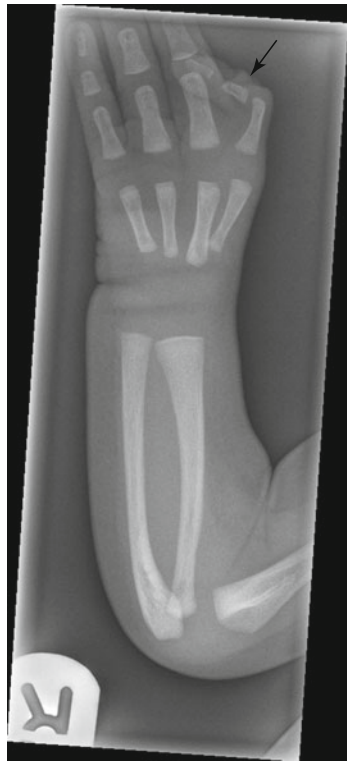

Fig. 12.8 Triphalangeal thumb in a young infant also showing oligodactyly and camptodactyly

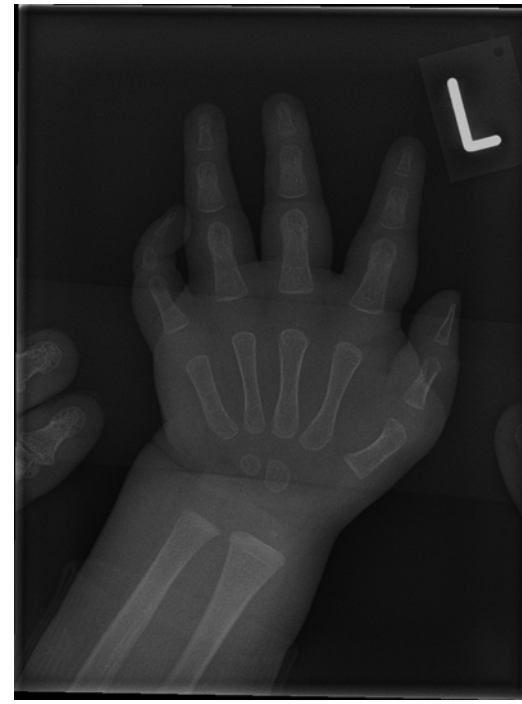

Fig. 12.10 Hand x-ray of 1.5 years old showing incurved little finger in keeping with clinodactyly. Note associated dysplastic middle phalanx of the little finger

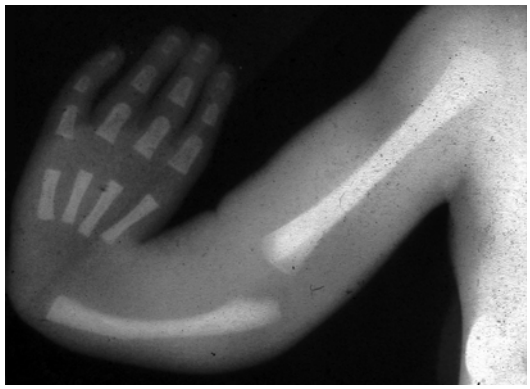

Fig. 12.9 Forearm x-ray in a neonate - An example of longitudinal deficiency with thumb and radial aplasia

amputation of a distal limb, previously thought to be due to amniotic bands (Fig. 12.15).

Ogino et al showed that the classical transverse deficiency was unilateral in their cohort, associated with absence of pectoral muscle and the level of amputation was more proximal than seen in congenital constriction band syndrome [19].

Brachymetacarpalia/Hypoplastic Metacarpal(s)

Metacarpal hypoplasia usually involves 4th and/or 5th metacarpals. This finding can be an isolated anomaly (Fig. 12.16) but is usually familial or syndromic. Common syndromes associated with it include Turner's, Klinefelter's and Kallman as well as metabolic conditions such as pseudo-hypoparathyroidism and pseudo-pseudo-hypoparathyroidism.

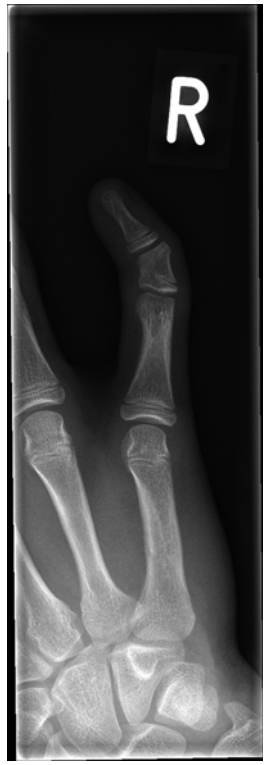

Fig. 12.11 14 years old with a delta middle phalanx of the little finger and associated clinodactyly

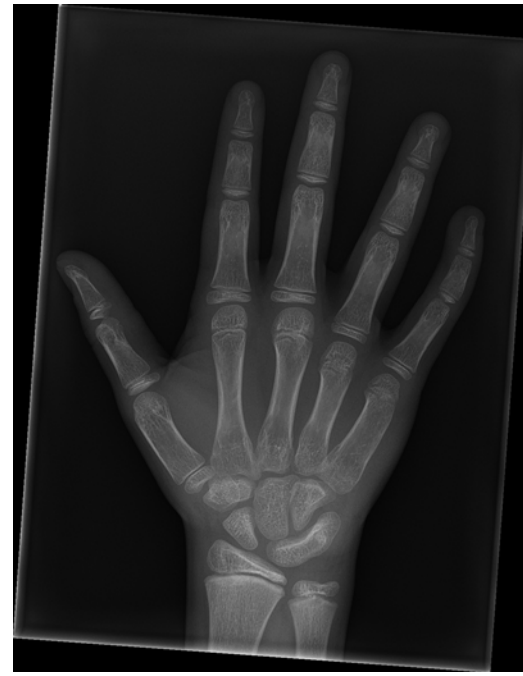

Fig. 12.12 Hand x-ray of a 10 year old child demonstrates an isolated carpal fusion between lunate and triquetral bones

Madelung and Reverse Madelung deformities

Madelung's deformity of the wrist results from disturbance of distal radial epiphyseal growth plate, mainly involving its ulnar and volar portions. It is characterised by dorsal and lateral bowing of the radius, increased ulnar and volar slanting of the distal radial articular surface and relative overgrowth (positive ulnar variance) and dorsal subluxation of the distal ulna. Radio-ulnar-carpal joint shows a V-shaped configuration on x-ray with proximal invagination of the carpal bones and lunate forming its apex. Reverse Madelung's deformity refers to the wrist abnormality similar to Madelung's but in association with an abnormally short ulna (Fig. 12.17).

Madelung's deformity is bilateral in two third of cases and majority of cases are congenital

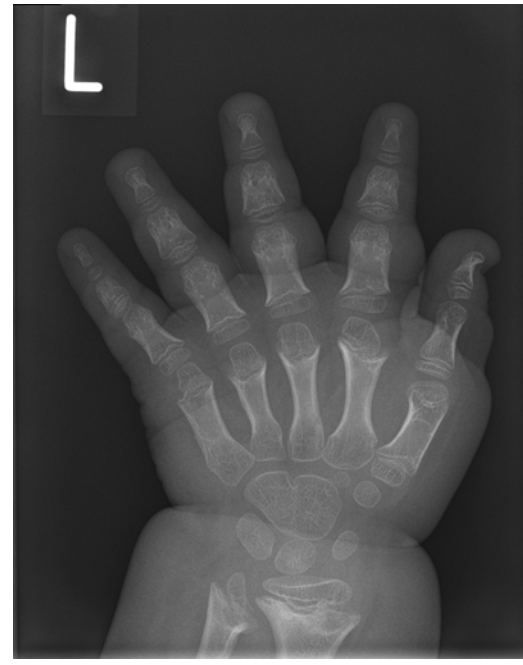

Fig. 12.13 Fused capitate and hamate in an 8 years old achondroplastic child. Please note brachydactyly, divergent fingers, coarse trabeculae and irregular metaphyses including of the distal radius and ulna

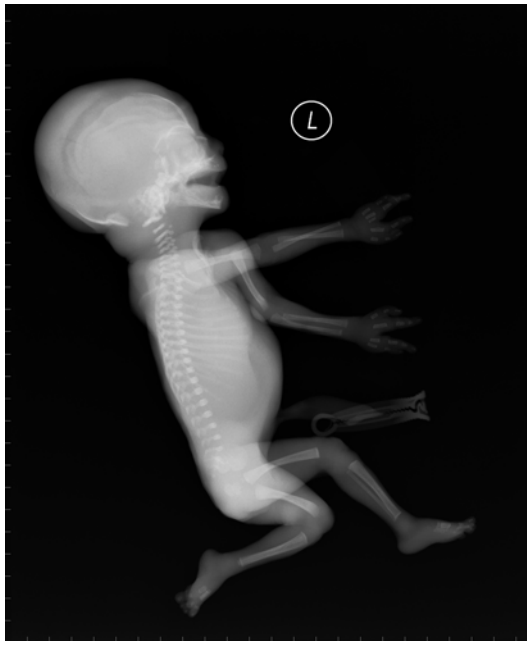

Fig. 12.14 Lateral x-ray of a terminated foetus shows central deficiency/ectrodactyly of both hands with associated oligodactyly and syndactyly

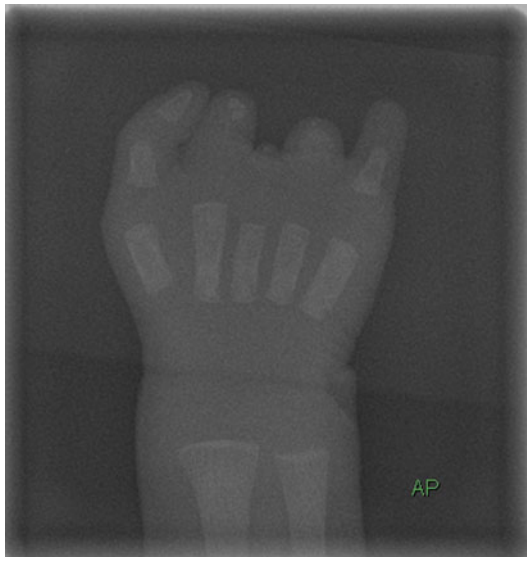

Fig. 12.15 Hand x-ray of a newborn baby showing irregular bony and soft tissue deficiency of all 4 fingers due to amniotic band syndrome

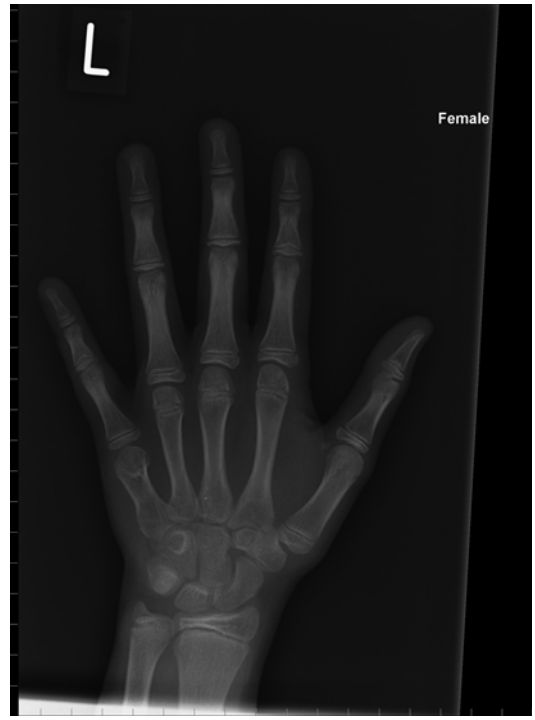

Fig. 12.16 Hand radiograph of a 12 years old girl with shortened 5th metacarpal of a familial nature

when it is usually inherited as an autosomal dominant trait. Acquired causes include trauma and infection. The congenital form usually presents during childhood growth between the ages of 8 and 14 years. Presenting symptoms include wrist pain, deformity and limited function or range of motion. Females are four times more likely to be affected than males.

Different chromosomal syndromes and skeletal dysplasia are linked with Madelung's deformity including Turner syndrome, Leri-Weill dyschondrosteosis, nail-patella syndrome, diaphyseal aclasis, achondroplasia and Ollier disease. Madelung's deformity can also be associated with some types of Mucopolysaccharidosis.

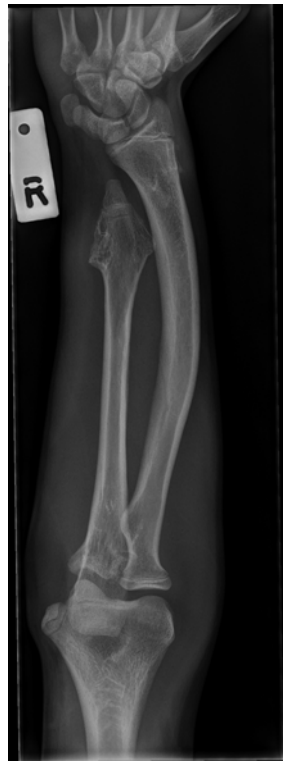

Fig. 12.17 Antero-posterior radiograph of forearm of a 15 year old with Diaphyseal Aclasis showing a Reverse Madelung's deformity. Ulna is foreshortened with an "*arrow-head*" shaped distal end, radius is laterally curved and the ulnar side of its distal epiphysis is hypoplastic. This latter feature results in proximal subluxation and v-shaped crowding of proximal carpal bones. Note multiple small osteochondromas in relation to the distal radial and ulnar metaphyses

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Part III

Surgical Techniques

Wrist Arthroscopy – Diagnosis and Therapeutic

13

A.R.E. Keogh and G.I. Bain

Keywords

Wrist • Arthroscopy • Biopsy • Infection • Ganglia • Synovectomy • Triangular fibrocartilage complex • TFCC • Distal radioulnar joint • Stabilisation • Carpal instability • Contracture • Capsular release • Scaphoid • Limited wrist fusion • Kienböck's disease • Distal radius fractures

Introduction

Professor Kenji Takagi was the first to report on the use of large joint arthroscopy in 1920 [1]. Whilst early attempts were hampered by cumbersome instrumentation, developments over the following 50 years would allow small joint instrumentation. In 1979, Chen reported on the development of a technique for wrist arthroscopy [2]. In 1986 Roth et al. [3] presented an "Instructional Course Lecture" on wrist arthroscopy at the American Academy of Orthopaedic Surgeons meeting, which brought it into the mainstream of orthopaedic surgery. Since then, wrist arthroscopy has continued to evolve as an important diagnostic and therapeutic tool. Wrist arthroscopy now has many indications and these continue to be extended as the principles of open surgical procedures are applied to the arthroscope (Table 13.1).

Whilst its earlier application was mainly as a diagnostic tool, wrist arthroscopy has become a valued therapeutic tool. There has been a continued evolution of therapeutic advances and even complex reconstructive procedures are being performed arthroscopically. Adaptation of open surgery to the arthroscope, with the development of arthroscopic tools such as wands, graspers, cannulas, and anchors, has led to an explosion of arthroscopic procedures. Innovative surgeons willing to push the limits of arthroscopic skills and joint access have pioneered what is an expanding field. We herein outline current applications and raise awareness of future directions in the field of wrist arthroscopy.

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Table 13.1 Indications for wrist arthroscopy

	Soft tissue	Bone
Diagnostic	Wrist pain of unknown origin	Assessment of instability
	Synovial biopsy	Staging of Kienbock's disease
"Ectomy" procedures	Bacterial sampling, lavage	Distal pole of scaphoid
	Dorsal and volar ganglia	Distal ulna (wafer procedure)
	Ligament debridement	Hamate
	Synovectomy	Lunate
	TFCC tears	Proximal row
		Carpectomy
	Articular cartilage lesions	STT debridement
		Ulnar styloid
Tissue shrinkage	Radiofrequency capsule or ligament shrinkage	
Surgical release	Volar capsular release	
	Dorsal capsular release	
Repair procedures	Lunotriquetral instability	Distal radius fractures
	Scapholunate instability	Scaphoid fractures
	TFCC repair	
Reconstructive procedures	Scapholunate ligament Reconstruction	Bone graft to scaphoid nonunion
	Distal radioulnar joint stabilization	Limited wrist fusion

Reproduced from Bain et al. [7] courtesy of Elsevier

Anatomy

Fundamental to wrist arthroscopy is a knowledge of the dorsal and volar wrist anatomy. Tendons, nerves and arteries are all at risk when establishing portals. A table of the portals and the structures at risk is displayed above (Table 13.2).

Wrist Arthroscopy Technique and Portals: The Box Concept

Arthroscopic examination of the wrist typically involves visualisation of the radiocarpal and midcarpal joints. The traditional dorsal radiocarpal portals include the workhorse visualisation portal in the 3–4 interval and the 4-5 and 6R working portals (Fig. 13.1).Accessory portals include the 6U, STT and 1/2 portals. Radial and ulnar midcarpal portals are used as visualisation and working portals. With the more recent description of the radial and ulnar volar radiocarpalportals [4–6] (Fig. 13.2), it is possible to have viewing and working portals that completely encircle the wrist. This enables the arthroscopic surgeon to view and instrument from all sides of the wrist, which we refer to as the box concept [7] (Fig. 13.3). Other portals have been described for other specific applications. Distal Radioulnar Joint (DRUJ) portals may be used for assessment of the DRUJ and the triangular fibrocartilage complex (TFCC). The scapho-trapezio-trapezpoidal (STT) portal lies just ulnar to the Extensor Policis Longus (EPL) tendon and allows visualisation and debridement of the STT joint.

Our Surgical Technique

General or regional anesthesia is the norm. The patient lies supine on the operating table, with the shoulder aligned with the table edge. A tourniquet is placed above the elbow, but not inflated. The shoulder is abducted to around 80° and the forearm lies on a hand table. A weight of 8 lbs is suspended by a padded strap over the tourniquet and this will later provide countertraction. The limb is prepared and draped and the forearm is suspended vertically, by use of finger traps, from an articulating arm attached to the operating table. The limb is exsanguinated and the tourniquet inflated. A 2.5-

Table 13.2 Wrist arthroscopy portals in common use, with structures at risk

	Structure at risk		
Portal	Tendon	Nerve	Artery
1/2	EPB, ECRL	Superficial branch radial N.	Radial artery
3/4	EPL, ECRB, EDC		
4/5	EDC, EDM		
6 Radial (6R)	EDM, ECU		
6 Ulnar (6U)	ECU, FCU	Dorsal branch ulnar N.	
Midcarpal ulnar (MCU)	EDC, EDM	Dorsal branch ulnar N.	
Midcarpal radial (MCR)	ECRB, EDC		
STT (Scaphotrapeziotrapezoid)	EPL, ECRL	Superficial branch radial N.	Radial artery
Volar ulnar (VU)	FCU	Ulnar N.	Ulnar artery
Volar radial (VR)	FCR		Radial artery

EPB extensor pollicis brevis, *ECRL* extensor carpi radialis longus, *EPL* extensor pollicis longus, *ECRB* extensor carpi radialis brevis, *EDC* extensor digitorum communis, *EDM* extensor digiti minimi, *ECU* extensor carpi ulnaris, *FCU* flexor carpi ulnaris, *FCR* flexor carpi radialis)

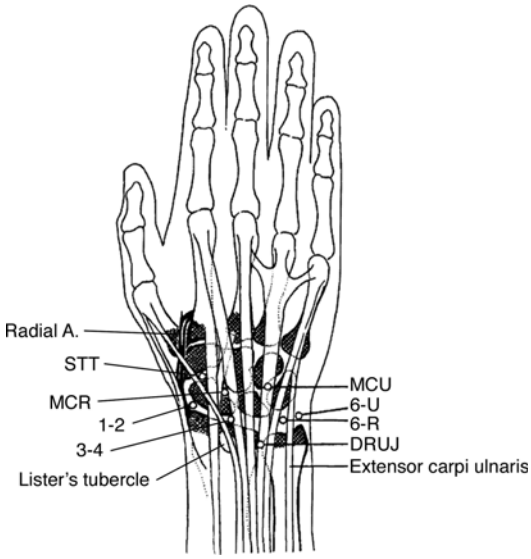

Fig. 13.1 Dorsal wrist portals. Portals are named according to anatomical landmarks. *DRUJ* distal radioulnar joint (Adapted from Bain et al. [113])

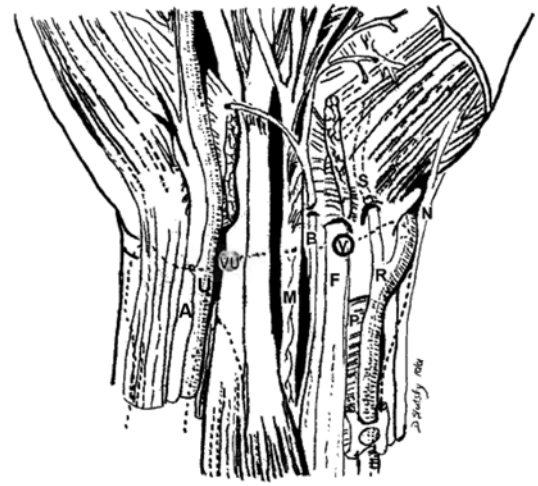

Fig. 13.2 Volar aspect of the right wrist with relative positions of the volar ulnar and volar radial portal shown. *N* superficial radial nerve, *R* radial artery, *S* superficial palmar branch of the radial artery, *P* pronator quadratus, *F* flexor carpi radialis, *V* volar radial portal, *B* palmar cutaneous branch of the median nerve, *M* median nerve, *FDS* flexor digitorum sublimis, *VU* volar ulnar portal, *A* ulnar artery, *U* ulnar nerve (Reproduced from Slutsky [5, 6]. Reprint permission provided by Elsevier)

mm arthroscope is used with a 30° viewing angle. A short bridge arthroscope (lever arm of 100 mm) allows for greater control. The hand can envelop the arthroscope shaft and the tips of the fingers abut the patients skin to enable fine control of the arthroscope tip in the joint space. The wrist is examined and landmarks are palpated (radial styloid, Lister's tubercle, ulnar styloid, triquetrohamate joint). We recommend that the anatomy and portals be drawn in until the surgeon is experienced in portal placement. Using the tip of the thumb, the 3–4 interval is established in the soft spot 1 cm distal to Lister's tubercle. An 18-gauge needle is inserted first and angled 10° volar to the horizontal plane to be parallel to the joint surface, thereby decreasing the risk of articular damage. The wrist can then be distended with normal saline solution or local anaesthetic. A vertical stab incision is

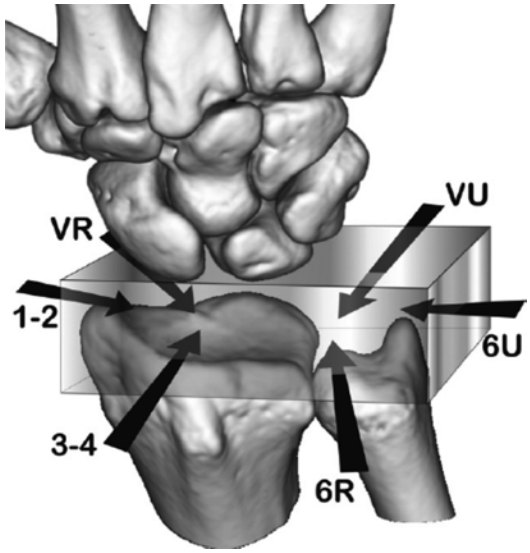

Fig. 13.3 The box concept – the radiocarpal joint can be accessed circumferentially with the arthroscope much like looking into a box. With the use of two dorsal (3/4 and 6R), two palmar (VU and VR), one radial (1/2) and one ulnar (6U) portals it is possible to visualize and instrument all parts of the radiocarpal joint (Reproduced from Bain et al. [7]. Reprint permission provided by Elsevier)

made in the skin with a no. 15 scalpel blade, the soft tissues are spread with an artery forcep and then these are used to gently enter the radiocarpal joint. Once the return of injected fluid is seen, the forceps are held open in order to dilate the path through the joint capsule. A blunt trocar is then introduced into the joint. Inflow (lactated Ringer's solution) is gravity-fed (a pump is not required). Subsequent portals are made by use of an outside-in technique, where a needle is introduced in to the joint under vision. The 6R portal is the commonly used radiocarpal portal. It is identified by the use of transillumination and the introduction of the needle radial to the extensor carpi ulnaris (ECU) tendon and distal to the triangular fibrocartilage complex (TFCC). If required, a mini-open approach is used for the 1–2 and 6U portals because of the proximity of the cutaneous nerves. An inside-out technique, with an exchange rod for the volar radial portal, is preferred [5]. Our preferred method for entering the midcarpal joint is to palpate the triquetrohamate joint, a palpable dorsal ridge one centimeter distal to the ulnar styloid and introduce other portals using an outside-in technique. The radial midcarpal portal is in the soft spot, 1 cm distal to the 3–4 portal. The transillumination technique can be used to guide the placement of the radial midcarpal portal.

Pearls for Arthroscopy Technique

Set up all equipment before making incision.

Identify the soft spot between EPL, ECRB and EDC index for the 3/4 portal.

Hold thumb tip in one position while establishing portals; do not take thumb away.

Insert needle into the joint and infiltrate.

Open the ulnar midcarpal portal before the radial midcarpal portal, identifying it by the dorsal ridge of the triquetrohamate joint.

Indications for Arthroscopy

The indications for wrist arthroscopy continue to expand (Table 13.1). In the following pages we will summarize the present indications for wrist arthroscopy, identify areas of recent progress and speculate on the future.

Soft Tissues

Diagnostic Arthroscopy

Wrist arthroscopy is becoming an increasingly popular procedure. However whilst it is perceived as relatively innocuous, its reliability is yet to be proven. Some claim that it represents the gold standard of investigative tools [8]. However, Sennwald [9] believes that it is not without complication and may overdiagnose or indeed underdiagnose. There is also a recognized learning curve associated with its use.

Stanley and Saffar [10] consider wrist arthroscopy appropriate in three situations: for diagnostic, clinical and therapeutic reasons. When used for diagnostic purposes it is useful for patients presenting with pain, instability, weakness or stiff-

ness, where investigations have been inconclusive. Magnetic Resonance Imaging (MRI) historically had poor sensitivity for scapholunate and triangular fibrocartilage complex tears (TFCC) [11] and, consequently, arthroscopy gained acceptance as a more reliable diagnostic tool. However, with the development of newer MRI technology, the sensitivity and specificity has improved. Indeed, when used in concert with arthrography, MRI has been shown to detect lesions not readily identifiable at arthroscopy [12], a criticism levelled at arthroscopy by Sennwald [9].

We must also be ever mindful of the ability of arthroscopy to diagnose when there is no clinical accompaniment. It has been shown using arthrography, that there is a high incidence of changes in the contralateral asymptomatic wrist [13] and that the correlation between arthroscopic findings and pain in the symptomatic wrist is poor [14, 15].

The diagnostic capabilities of wrist arthroscopy were reported almost 20 years ago in a study which investigated the arthroscopic appearances of post traumatic wrist pain in 30 patients [16]. Time to arthroscopy from injury varied from 4 to 50 months. In 70 % of patients (21 patients) there were intra-articular findings that correlated with the clinical findings. Five of those patients had ligament injuries with frank or potential wrist instability. Arthroscopy allowed direct inspection and assessment of the extent of the injury and revealed any associated injuries. Dynamic manoeuvres also allowed the diagnosis of segmental instabilities.

In spite of Sennwald's scepticism, arthroscopy continues to be performed regularly for diagnostic purposes and it has many proponents. As surgeons and radiologists improve their diagnostic skills, and as there is an improving awareness of wrist pathologies, this area will be in a continuing state of flux.

Regardless of the potential for newer imaging techniques to define the extent of the injury, there is nothing that compares with probing and manipulating injured tissues in order to gauge the likelihood of effecting tissue repair. Another major advantage of wrist arthroscopy is the ability to assess the wrist dynamically, which may reveal underlying dynamic instability patterns.

Synovial Biopsy

Arthroscopic synovial biopsy is a relatively simple procedure. Utilising a viewing and working portal, material can be obtained with rongeurs and sent for histological examination, culture and sensitivity tests. Synovectomy can be performed at the same time in an attempt to improve symptoms.

Wrist arthroscopy has been used for the diagnosis and therapy of gout of the wrist. Wilczynski et al. [17] reported on a cohort of seven such patients. Having failed non- operative treatment, patients underwent arthroscopy, at which time synovial biopsies were taken. They reported a significant risk of scapholunate ligament attenuation within gout affected wrists, the most common pattern of arthritic change corresponding to a SLAC wrist (Scapholunate Advanced Collapse). Additionally, they noted that when sampling synovium from a gouty wrist, the tissue should be sent to the laboratory fresh. Formalin dissolves the crystals of monosodium urate giving false negative results. This problem is not encountered with Calcium pyrophosphate deposition (CPPD) disease as the formalin does not dissolve the crystals.

Infection

Wrist arthroscopy is a simple way of sampling wrist synovial fluid, taking synovial biopsies and lavaging the joint in cases of suspected bacterial infection. It is usual to place an inflow viewing portal and an outflow portal. Both the radiocarpal and midcarpal joints are inspected and associated pathologies such as concomitant pseudogout, gout or ligament injuries can be diagnosed. The state of the carpal cartilage is also documented. The advantage of wrist arthroscopy is that the small portals allow earlier return to function when compared to an open approach [18].

Sammer and Shin have previously compared the results of open and arthroscopic treatment of septic arthritis of the wrist [19]. In a retrospective study over a period of 11 years, they treated septic arthritis of the wrist arthroscopically in 21 wrists and open in 19 wrists. Fewer patients in

the arthroscopic group required repeat operation and, if it was required, it was more usual for the repeat procedure to be performed open. There was no significant difference in the number of operations required between groups, but the average hospital length was shorter for patients undergoing the arthroscopic procedure. As it was a retrospective study, the authors acknowledge that there may have been a selection bias and patients with more severe infections may have been preferentially chosen to undergo open procedures.

Dorsal and Volar Ganglia

Dorsal and volar ganglia are two of the most common tumors of the wrist and can involve the radiocarpal and the midcarpal joints [20]. They are frequently asymptomatic. However, some patients develop persistent symptoms, unresponsive to traditional means. Other methods of treatment, such as closed rupture or ganglion aspiration, are associated with high recurrence rates [21–23].

When conservative means fail then operative treatment can consist of open or arthroscopic surgery. Arthroscopic surgery offers reduced postoperative pain, earlier return to function and smaller incisions. Similar or better rates of recurrence have been reported [24, 25].

Author's Preferred Technique

Arthroscopic excision of a dorsal ganglia requires the use of the 3/4 and 6R portals. The 6R portal in this situation is used as the viewing portal towards the scapholunate ligament. The 3/4 portal is then used as the working portal through which an arthroscopic shaver is placed. The axilla of the dorsal capsule is then freed from the dorsal scapholunate region, taking with it the ganglia. The stalk is identified at the scapholunate interval and this area is debrided to leave a fenestration. It has been reported by some authors [26] that formal identification of the ECRB and EPL tendons is routinely performed with a dorsal capsulectomy.

In a recent prospective and randomized study by Kang et al. [26], the rates of recurrence and ongoing pain between arthroscopic and open dorsal ganglion excision were compared. There were 41 patients in the arthroscopic group and 31 patients in the open group. At 12 months, outcomes were similar between groups with differences for ganglion recurrence and ongoing pain not achieving significance. At 12 months, 11 % of the arthroscopic group and 9 % of the open group reported a recurrence. There was one superficial radial nerve neuropraxia in the arthroscopic group.

In a report on the 5 year follow up of dorsal and volar radiocarpal and midcarpal ganglia, Rocchi et al. [20] presented the results of a series of 47 patients undergoing arthroscopic excision. They reported 16 cases of dorsal radiocarpal ganglia. Only a proportion had an identifiable stalk. They used the 3/4 portal as a working portal and switched between the 4/5 and 6R portals. Resection continued until there was a 2 cm gap in the dorsal capsule and the extensors were commonly seen. In half of their dorsal wrist ganglion cases, the ganglion stalk was found in the midcarpal space, or indeed straddling the radiocarpal and midcarpal spaces. They found the dorsal midcarpal ganglia more difficult to treat than the radiocarpal ganglia because of the smaller dimension of the joint.

Radiocarpal volar ganglia were resected using a 3/4 portal for a resector and the 4/5 portal for viewing. The ganglion was identified by ballotment of the volar capsule, most often being seen through the junction between the radioscaphocapitate and the long radiolunate ligament. Resection of the volar capsule between these ligaments decompressed the ganglion and was continued until a 1 cm defect in the capsule was created. They recommended not advancing the resector palmarly as the radial artery is at risk and the Flexor Pollicis Longus tendon can also often be seen. Midcarpal volar ganglia were found in four patients. After examination of the radiocarpal joint and finding only minimal bulging in the usual position for a volar ganglion, the authors examined the midcarpal joint. Using the midcarpal radial portal, the ganglion base was found in the region of the scapho-trapezial-trapezoid joint. A 1/2 portal was then used to resect the ganglion base with a 2.0 mm end cutting shaver.

The resection was extremely difficult because of the limited space, in spite of finger traps, being applied to the thumb to improve distraction.

At 15 months follow up, Rocchi et al. [20] reported 39 out of 47 excellent results. There were two axonotmeses of the superficial branch of the radial nerve and one case of extensive hematoma. Injury to a branch of the radial artery occurred in one case, requiring an open approach to the volar STT ganglion and the vessel. There were two recurrences, one a radiocarpal volar ganglion, the other a midcarpal dorsal ganglion.

Pearls for Arthoscopic Ganglion Excision

Dorsal Radiocarpal:

Use the 6R portal as a viewing portal

Identify the recess of the dorsal capsule and the ganglia

Debride using a shaver through the 3/4 portal

Volar Radiocarpal:

Volar ganglia can be identified in the radiocarpal joint between the radioscaphocapitate and the long radiolunate ligament.

Resect the edges of these ligaments to marsupialize the opening of the ganglion.

Release the tourniquet after resection of the ganglion stalk to ensure patency of the radial artery

Intraossesous Ganglia

Intraosseous ganglia of the carpus are a rare cause of disabling wrist pain. The most commonly reported site is in the lunate, but intraosseous ganglia of the scaphoid have been reported [27]. The pathogenesis of these lesions remains unclear [28], although its etiology is thought to be similar to that for soft tissue ganglions. Treatment, if required, consists of either open or arthroscopic techniques.

Author's Preferred Technique

The senior author (GIB) has previously published a technique for the arthroscopically assisted debridement and bone grafting of intraossesous ganglia of the lunate [29]. A preoperative CT scan is imperative for identifying the location of the cyst in the lunate, as some are predominantly volar and hence require a volar portal. A standard wrist arthroscopy is performed using 3/4 and 6R portals. A 3.5 mm drill is advanced through a trocar placed in the 3/4 portal. Fluoroscopy is performed to confirm positioning prior to drilling. The contents of the ganglia is sampled with a curette and sent for histology. A 3.0 mm motorised resector is inserted into the mouth of the ganglia. Bone graft from the distal radius is inserted with a rod, through the trocar placed within the ganglia.

Pearls for Arthroscopic Assisted Resection of Intraosseous Ganglion of the Lunate

CT is mandatory to identify the location of the lesion (dorsal or volar)

3/4 working portal for a dorsal intraosseous ganglion, volar radial portal for volar ganglion

Trocar and 3.5 mm drill are essential

Bone grafting of the defect is recommended

After a minimum of 1 year follow up, all patients had a significant improvement in pain scores, grip strength, range of motion and function [29]. Postoperative radiographs revealed trabeculation within the grafted lunate, reflecting incorporation of the bone graft. The authors felt that this was a safe procedure for advanced wrist arthroscopists providing a quick recovery and likely return to full employment.

Synovectomy

Surgical synovectomy has been utilized for over 100 years for the treatment of arthritic joints. With the advent of arthroscopy its application has become more accepted. However, the longer term outcomes for wrist synovitis are contested. Regardless, in the short to medium term there is improvement in pain and function [30, 31].

In rheumatoid arthritis, in spite of the recent advances in pharmacological treatment, there may, on occasion, be a requirement for wrist synovectomy. Some authors report that those suffering from severe rheumatoid arthritis have not been shown to benefit from surgery [32] whilst others believe that arthroscopic debridement should be considered for all grades of arthritis [30]. Arthroscopic debridement is not appropriate if a large open wrist debridement is anticipated, but it can be combined with other procedures, such as carpal tunnel decompression, tenosynovectomy and fusion/joint replacement procedures.

Anecdotal evidence for the arthroscopic debridement of wrists of patients with systemic lupus erythematosus, psoriatic, and postinfectious monoarthropathy has been presented [33]. Similarly, pseudogout can be treated with arthroscopic synovectomy for transient relief. Chondral defects associated with osteoarthritis will be discussed in a later section, but if synovitis is the principal concern then athroscopic synovectomy may be applicable. Debridement of synovitis following trauma is possible, although it is often combined with division of adhesions and capsular release.

Outcomes of arthroscopic synovectomy for rheumatoid patients have been studied [30, 32]. Park et al. [30] analysed the results in 18 patients with rheumatoid arthritis. Patients were included if they had not responded to systemic treatment for 6 months and excluded if they had extensor tenosynovitis or severe DRUJ involvement. Standard 3/4 and 4/5 portals were used, but 1/2 and 6U portals were helpful in visualising the dorsal capsule. When the TFCC was degenerate, admission to the DRUJ was possible and separate DRUJ portals were occasionally used. Outcomes, including pain and satisfaction, were assessed using visual analogue scales, in addition to flexion extension range.

All patients, other than two, experienced a reduction in pain (VAS 8.6–4.4) at final follow up (29 months). Mean satisfaction score was 6.3. The flexion extension arc increased from 81 to 92°.

The authors reported that the DRUJ could be accessed arthroscopically, but not completely and when severe radiographic changes were apparent, then an open approach should be combined with an arthroplasty procedure (ie. interpositional, excisional, replacement). They also recommended that patients with severe radiographic changes could be treated with arthroscopic synovectomy, as they also reported good relief of symptoms. They suggested that patients with severe degenerative changes should undergo arthroscopic debridement prior to fusion procedures.

TFCC Lesions

The triangular fibrocartilage complex (TFCC) is positioned between the distal ulna and the ulnar side of the carpus. It has dual functions: both stabilising the distal end of the ulna in relation to the radius and also transmitting load from the carpus to the ulna [34]. TFCC tears are a common source of pain following trauma and also as a consequence of degeneration [35]. The peripheral 25 % of the TFCC is vascularised [36] and may be amenable to repair, similar to the meniscus of the knee. Wrist arthroscopy has become the gold standard for the diagnosis and treatment of these lesions [37, 38]. The key advantage of arthroscopic treatment is that associated lesions can be assessed and attended to at the same sitting.

Palmer [39] has divided TFCC lesions into Type 1 (traumatic) and Type 2 (degenerative), there being further subdivisions of each (Table 13.3).

Traumatic (type 1) tears of the TFCC are classified according to their location. Central tears (Type 1A) are managed with arthroscopic debridement as the potential for healing of the central portion is poor [40]. Viewing via a ¾ portal whilst placing a 3.5 mm oscillating resector through the 4/5 or 6R portal allows good access to these tears. Synovitis and loose fibrocartilage can be resected, with the majority of patients experiencing good relief of symptoms [41, 42]. In patients who have a central tear associated with positive ulnar variance, a wafer procedure or an ulnar shortening should be combined with debridement.

Table 13.3 TFCC injuries – classification and recommended management

Tear classification	Description	Author's recommended management
Traumatic		
1A	Tear in horizontal or central portion of disk. Often with an unstable flap.	Initial Splinting ± Steroid Injection. Arthroscopic debridement of central torn portion.
1B	Tear from distal ulna insertion ± ulnar styloid fracture.	Arthroscopic repair. Inside-out technique. ± ECU sheath open repair.
1C	Tear with ulnocarpal ligaments disrupted. (Ulnolunate and Ulnotriquetral ligaments).	Arthroscopic augmented repair using a mini open approach ± FCU augmentation.
1D	Tear from insertion at radius	Debridement of torn portion or reattachment to sigmoid Notch.
Degenerative		
2A	TFCC wear but no perforation	Diagnostic Arthroscopy followed by open diaphyseal ulna shortening.
2B	TFCC wear but no perforation Chondromalacia of lunate or ulnar head	
2C	Central perforation of TFCC Chondromalacia of lunate or ulnar head	Arthroscopic TFCC debridement plus Arthroscopic Wafer procedure or Open diaphyseal ulna shortening.
2D	Central perforation of TFCC Chondromalacia of lunate or ulnar head Perforation of LT ligament	
2E	Central perforation of TFCC Perforation of LT ligament Ulnocarpal arthritis	

Reproduced from Bain et al. [7] with permission from Elsevier
Abbreviations: *TFCC* triangular fibrocartilage complex, *FCU* flexor carpi ulnaris, *ECU* extensor carpi ulnaris, *LT* lunotriquetral ligament

Peripheral tears of the TFCC (type 1B) are a difficult diagnostic problem and may be associated with instability of the distal radioulnar joint. Patients can complain of ulnar sided wrist pain following either a fall on the outstretched hand or a twisting injury. There is associated giving way of the wrist, with twisting and painful clicking. Resisted supination and pronation may reproduce the symptoms. Our pre-operative assessment includes ballotment of the distal ulna with comparison to the opposite side. Imaging, including plain X-rays and MRI, can reveal dorsal or palmar ulnar subluxation, or a TFCC tear. We find the 3 Tesla MRI scanners will clearly demonstrate the TFCC foveal insertion and adjacent structures. At arthroscopy, loss of the normal "trampoline" effect, felt as an increased sponginess or softness of the TFCC [43] indicates a peripheral tear. Additionally, distraction of the TFCC from the foveal attachment can be performed (the "hook test") [44]. If repairable, these tears can be managed either with an outside-in, or an inside-out repair technique [42, 45]. Other, newer, techniques involve the use of suture welding, mini-open with arthroscopic assistance approaches and meniscal arrows [46–48].

It is the senior author's (GIB) experience that some patients present acutely with pain, but without instability and imaging reveals an associated deep surface TFCC tear. These patients are treated with the inside out Tuohy needle technique. The majority of patients with DRUJ instability have a deep surface tear of the TFCC, commonly not observable at arthroscopy of the radiocarpal joint. For patients with a deep surface TFCC tear and instability, who have a delayed presentation, we perform our preferred technique of peripheral TFCC repair using a tendon graft (see below). If there is a complex tear of the TFCC (ie central and deep) and there is associated instability, we perform a reconstruction using a technique similar to that reported by Brian Adams [49] (see section on DRUJ reconstruction).

Author's Preferred Technique for Peripheral TFCC Tear Repair

For simple, deep surface, acute TFCC tears, without instability, we prefer the inside out repair technique popularized by Poehling [50]. Using a 20-gauge Tuohy needle and a 2–0 PDS suture (Fig. 13.4), the suture is passed to the radial side of the TFCC tear and then out through an ulnar wound. A peripheral TFCC tear may also, on occasion, be associated with subluxation of the nearby ECU tendon. In these situations an open reconstruction would be combined with the TFCC repair.

For those patients with a confirmed deep surface TFCC tear and instability, we prefer our technique of tendon reconstruction, with either palmarislongus (PL) or flexor carpi radialis (FCR) tendons. Arthroscopic assistance is used, but this approach is primarily performed in an open fashion. We prefer to use a tendon graft in those patients with a delayed presentation, as we are not confident that we can achieve good fixation between the undersurface of the TFCC and the foveal attachment by simple repair techniques. We have been happier using the tendon graft, which can incorporate into the TFCC and the ulnar tunnel. We use a dorsal s-shaped incision centred over the 5th extensor compartment. The dorsal capsule of the DRUJ is reflected, respecting the dorsal radioulnar ligament, TFCC and extensor carpi ulnaris (ECU) subsheath. A view is obtained inferior and superior to the TFCC. An ulnar tunnel is drilled with a cannulated 3.5 mm drill, under fluoroscopic control, from the medial border of the ulna to the fovea. Tendon graft is harvested and the two ends are sutured, using a braided absorbale 2–0 suture, to give purchase on the separate tendon ends. A Spectrum II (ConmedLinvatec, Largo, Florida) suture passer is passed through the ulnar tunnel and then through the volar/ulnar TFCC. A suture shuttle is then passed and withdrawn out of the joint. The tendon suture is passed through the TFCC and retrieved. A second pass is made through the dorsal/ulnar TFCC, with the suture passer and the suture shuttle and tendon suture retrieved out the ulnar tunnel, creating a mattress configuration. The tendon is then advanced through the TFCC and back out through the

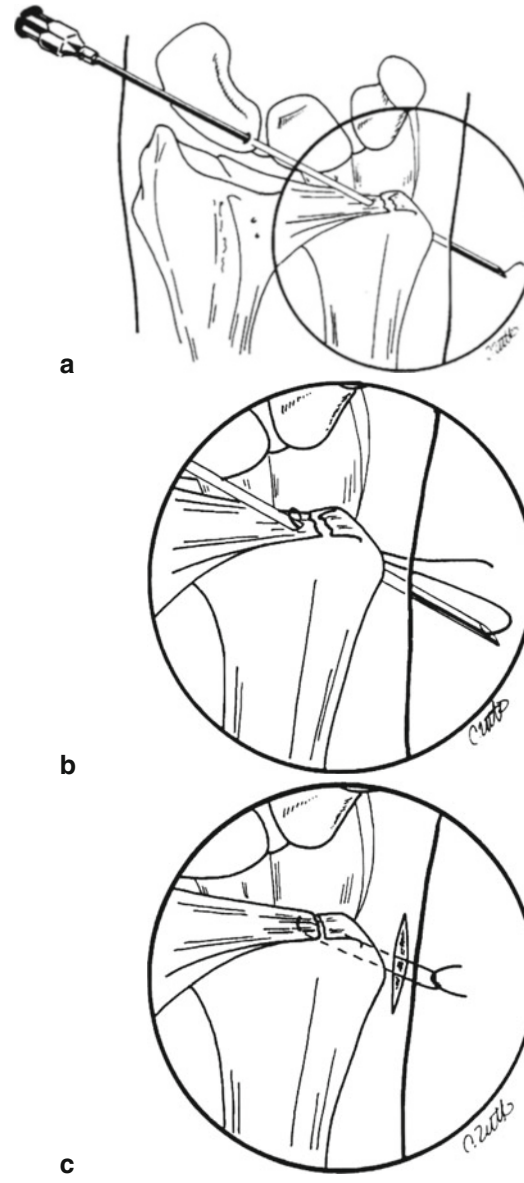

Fig. 13.4 (**a**) A 20-gauge Tuohy needle is inserted into the wrist joint through the 3/4 portal and is advanced through the ulnar edge of the TFCC, capsule, and skin. A 2–0 PDS suture is fed through the needle, and the free end is held with an artery clip. (**b**) The needle tip is drawn back into the wrist and is again advanced through the ulnar edge of the TFCC. The suture remains in the lumen of the needle and is passed through the TFCC a second time to create a mattress suture. The needle is again passed out through skin. The end of the suture from the second pass is then withdrawn from the needle. (**c**) The two ends of the suture are tied over the capsule after making a small incision, protecting cutaneous nerves (Reproduced from Bain et al. [113])

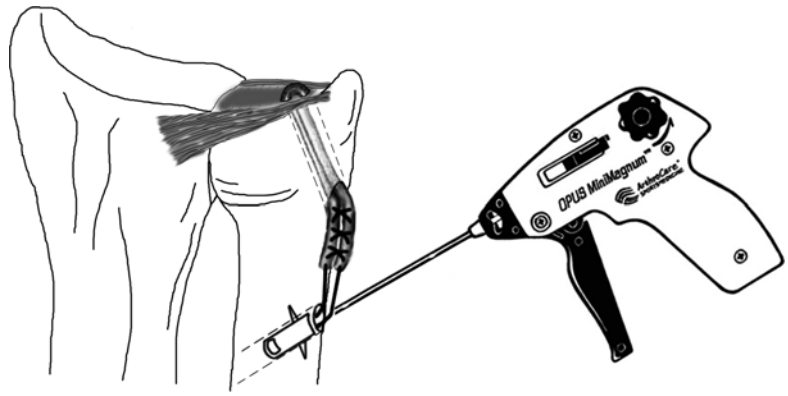

Fig. 13.5 Method of DRUJ stabilization using tendon graft and the Opus Labrafix (Arthrocare Corporation, Austin, Texas) device. This technique is useful in the setting of DRUJ instability with an intact central portion of the TFCC

ulnar tunnel, leaving two ends of the tendon at the entry to the ulnar tunnel. These two tendon ends are then sutured using a Bunnell configuration and secured to the ulna proximally using a Labrafix (Arthrocare Corporation, Austin, Texas) tensionable anchor (Fig. 13.5).

The uncommon Type 1C lesions involve disruption of the TFCC from the ulnocarpal ligament complex. Arthroscopic assessment demonstrates laxity of the ulnocarpal ligaments and, as the adjacent ligaments are torn, the pisiform may be unusually easy to see. Repair of these lesions, in a similar fashion to the type 1B lesions, has been undertaken successfully by Estrella et al. [34]. Radial sided tears (Type 1D) have a poorer chance of healing, due to the relative avascularity of the radial side of the TFCC. Debridement usually results in a satisfactory result, particularly as these ligaments are not essential for stabilisation of the distal radioulnar joint [51]. Suture repair techniques have been described, with two-thirds of patients having good to excellent results [52].

In the Palmer classification, type 2 TFCC lesions describe the degenerative pattern of TFCC tears. These tears are commonly asymptomatic. They are subdivided, based on the severity of degeneration and involvement of the underlying ulna head and carpus. The degenerative changes are caused by chronic overload of the ulnocarpal joint and there is often associated positive ulnar variance. Continued ulnocarpal overload leads to further degenerative changes on the ulnar side of the wrist, with consequent changes in the lunate, triquetrum and the ulnar head. There can also be lunotriquetral ligament attenuation.

Generally, arthroscopic treatment includes debridement of synovitis and chondral lesions, in addition to the degenerate TFCC. Debridement of the TFCC should not advance beyond a 2 mm peripheral rim [53] which represents the stabilising dorsal and palmar radioulnar ligaments.

Associated ulnar positivity is often the driving force of these degenerative lesions and disimpaction is therefore recommended. This can be performed either as an open diaphyseal shortening, or as an arthroscopic wafer procedure. The wafer procedure is performed by first debriding the central TFCC, through which the ulnar head can be seen. The ulnar head is then resected to achieve 1.5 mm of negative ulnar variance and this can be checked with fluoroscopy [54]. If 1.5 mm is not possible with the wafer procedure, then ulnar diaphyseal shortening is preferable. The full circumference of the ulnar head can be removed by pronating and supinating the forearm [55, 56]. Pain relief has been reported in 78 % of patients using an open procedure [56] and good to excellent results have been achieved in up to 75 % of patients undergoing arthroscopic management of type 2 tears [57].

The senior author (GIB) has found the wafer procedure less reliable and prefers the ulnar shortening procedure, except in low demand patients and those patients who are adverse to an osteotomy.

DRUJ Ligament Reconstruction

When the TFCC is degenerate and not repairable and there is associated distal radioulnar

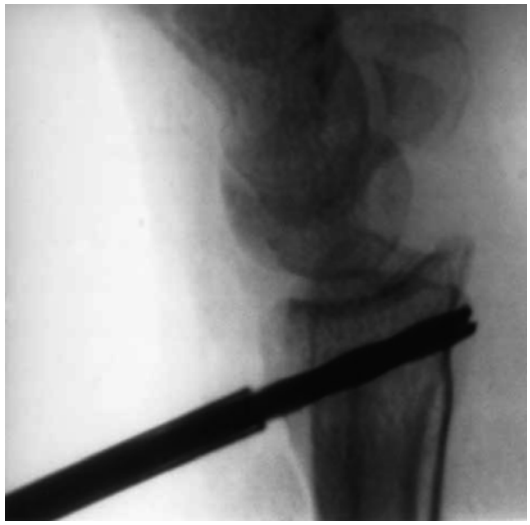

Fig. 13.6 A fluoroscopic image of a cannulated drill passing from dorsal to volar in a malunited distal radius (Reproduced from Adams and Berger [49]. Reprint permission obtained from Elsevier)

joint instability, then reconstruction of the distal radioulnar joint is required. This has been reported as an arthroscopic assisted technique, allowing the graft to be passed through the ulnocarpal joint, enhancing the stabilisation of the ulnar carpus [44].

Author's Preferred Technique for DRUJ Reconstruction

Our technique is similar to that described by Atzei [44]. A standard wrist arthroscopy is performed and the TFCC integrity is assessed with the trampoline and the hook test referenced previously. The TFCC remnant is debrided. We use: (1) a curved dorsoulnar incision, linking to the 4/5 portal and protecting dorsal sensory branches of the ulnar nerve and (2) a palmar ulnar incision, identifying the ulnar neurovascular bundle proximally. Palmarly the pronator quadratus is retracted proximally, or sometimes incised to expose the palmar corner of the distal radius. The palmaris is harvested through a small transverse incision at the distal wrist crease and a similar sized incision proximally. A 4.0 mm cannulated drill is driven over a wire, from palmar to dorsal, placed under fluoroscopic control (Fig. 13.6). This drill hole is intended to pass from the distal and ulnar-most part of the palmar and dorsal surfaces of the distal radius, whilst avoiding penetration or fracture into the joint. The same cannulated drill is drilled over a guidewire to the ulnar fovea under x ray control. The wire can be placed in the centre of the remnant TFCC/fovea under arthroscopic and fluoroscopic control. Atzei [44] uses a small joint compass, but we have no experience with this device. Using a combination of Keith needles and Linvatec Spectrum suture passers the graft is threaded through the distal radius, through the palmar capsule and 4/5 portal, then out through the ulnar tunnel. The two limbs of the graft are sutured together, using a Bunnell suture and then attached to an ArthrocareLabrafix (Arthrocare Corporation, Austin, Texas) anchor. A Labrafix drill is drilled into the distal ulna and the anchor is carefully placed under fluoroscopic control in the medullary canal of the distal ulna. Tension is then applied, carefully gauging stability of the distal radioulnar joint and supination/pronation movements, as tension is wound onto the anchor winding mechanism. Once sufficient tension is achieved, the anchor is locked off and the suture ends cut (Fig. 13.7).

Articular Defects

Articular defects are amenable to arthroscopic debridement [58]. Numerous sites are accessible by the chondrotome, including the STT joint, the radiocarpal joint and the midcarpal joints. Loose chondral flaps are debrided back to a stable base and limited synovectomy and or capsular release may improve the patient's symptoms.

Arthroscopic treatment of the STT arthritis has been reported by Ashwood et al. [29]. In a prospective series of ten patients, favourable outcomes were achieved including medium term relief of pain and good patient satisfaction. Wrist motion was somewhat improved with this procedure, particularly flexion/extension. Associated lesions, such as midcarpal arthritic changes, could be assessed and debrided as necessary. Regarding the safe placement of the STT portal, it is recommended that a 1.5 cm incision be used, gently dissecting the soft tissues and then pen-

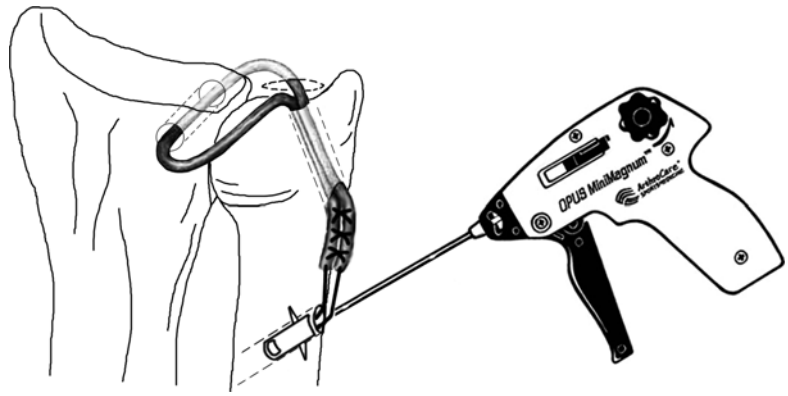

Fig. 13.7 Modification of the Adams technique of DRUJ reconstruction with tendon graft using the Opus Labrafix (Arthrocare Corporation, Austin, Texas) anchor. This technique is useful in the setting of DRUJ instability where there is inadequate TFCC tissue for repair

etrating the joint capsule of the STT joint. This helps to avoid the radial artery in the snuffbox and the superficial radial nerves which course nearby.

Following the recognition of the anatomical variation patterns of the lunate and its articulation in some patients with the hamate [59], it was shown that some cases of ulnar sided wrist pain were attributable to hamato-lunate impingement and so treatment has focussed upon arthroscopic excision of the proximal pole of the hamate. Patients in a case series of four reported good pain relief from arthroscopic debridement [60]. Further reports of similar patients revealed good to excellent outcomes in 18 of 23, following arthroscopic resection of the proximal hamate [61].

Other arthroscopic studies of the ulnar side of the wrist have revealed a similar distribution of lunate morphology to those of the cadaveric studies of Viegas et al. [59]. Contrary to their findings however, in a retrospective review of a younger patient group of 78 patients with ulnar sided wrist pain, it was found that not only type 2, but also type 1 lunates were associated with chondral lesions of the proximal aspect of the hamate [62]. They also described lesions of the helicoidal surface of the hamate, the capitohamate junction and the distal facet of the lunate. The differences between their findings and those of Viegas et al. [59] were attributed to the age difference of the patient group and also patient selection (ulnar sided wrist pain in this group vs a random sample in the Viegas et al. group).

Articular lesions of the wrist have also, on occasion, been treated with the microfracture technique popularised by Steadman et al. [63], although medium to long term results are not available and hence the outcomes are uncertain [64].

Carpal Instability

Acquired instability patterns of the wrist follow either trauma or degeneration and represent a spectrum of disease. There can be minor sprains with only minimal symptoms which can last up to 3 days [65]. Further injury can lead to the disruption of blood vessels with partial injury to the intrinsic and extrinsic ligaments and, where healing is expected without treatment, the so called "three week injury". Further injury results in the "three month injury," where there is complete rupture of intrinsic ligaments and secondary stabilizers of the carpus. The latter is the focus of this discussion.

Arthroscopy should now be considered the gold standard for diagnosing carpal instability patterns. It is a dynamic study which allows the assessment of: (1) joint spaces whilst performing provocative manoeuvres, (2) the ligaments responsible for the instability pattern and the viability of repair and (3) associated lesions such as chondral defects, and enables therapeutic measures such as arthroscopic repair. Additionally, a fluid arthrogram can be performed by infiltrating the midcarpal joint and observing spilling through to the radiocarpal joint, representive of an interosseous ligament disruption. Fluoroscopy can also be performed concomitantly.

Table 13.4 Geissler's [65] arthroscopic classification of interosseous carpal instability

Grade	Description
I	Attenuation/haemorrhage of interosseous ligament as seen from the radiocarpal joint. No incongruency of carpal alignment in the midcarpal space.
II	Attentuation/haemorrhage of interosseous ligament as seen from the radiocarpal joint. Incongruency/step-off as seen from the midcarpal space. A slight gap (less than width of a probe) between the carpal bone may be present.
III	Incongruency/step-off of carpal alignment is seen in the radiocarpal and midcarpal space. The probe may be passed through the gap between the carpal bones.
IV	Incronguency/step-off of carpal alignment is seen in the radiocarpal and midcarpal space. Gross instability with manipulation is noted. A 2.7 mm arthroscope may be passed through the gap between the carpal bones (so-called "drive-through lesion).

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Grading of the instability pattern can be performed arthroscopically for intrinsic ligament disruption. The Geissler classification scheme (Table 13.4) can be applied to both scapholunate and lunotriquetral instability patterns [66]. From the midcarpal joint the degree of separation of the scapholunate interval can be assessed using an arthroscopic probe. Additionally, the presence of a step between the two bones suggests instability.

Scapholunate instability is the most common pattern of wrist instability [67] and can lead to progressive degeneration of the wrist [68]. New methods of arthroscopic scapholunate ligament reconstructions are being developed currently [69] whilst other arthroscopic assisted repairs and reconstructions have been described [70].

Debridement Alone

For tears of the Scapholunate interosseous ligament, good results have been obtained with simple debridement alone. Weiss et al. [71] have reported on this technique in a series of 43 wrists. There was complete resolution or improvement of symptoms in 66 % of complete SLIO ligament tears. Similarly, excellent results have been reported with partial tears of the SLIO ligament [72].

Debridement and Temporary Stabilisation

Although controversial, some authors recommend debridement and percutaneous pinning of the scapholunate interval for treatment of the acute dynamic scapholunate ligament injury (<3 months) [73] and for the patient with dynamic chronic (>3 months) scapholunate ligament injury although the results are described as suboptimal [74].

Thermal Shrinkage

Similarly controversial, pundits of arthroscopic thermal capsular shrinkage tout its relative innocuousness and ease. Good results have been achieved with this technique in the lower grades of scapholunate instability (grade I and II) at short term follow up [75, 76]. However, long term studies have not yet been performed to confirm its efficacy.

Repair/Reconstruction

A technique for all arthroscopic repair of the scapholunate interosseous ligament has not yet been published in the English literature. A case report of the repair of a lunate fracture with the scapholunate ligament attached, provided a good outcome for one patient [77]. The fragment was fixed with screws and the patient was allowed to mobilise from day five post-operatively. Arthroscopic assisted techniques have been employed for scapholunate reconstruction procedures [70], but the results remain unpublished. Mathoulin et al. [69] have recently published their results of a series of patients undergoing an arthroscopic dorsal capsulodesis procedure. There was excellent pain relief and good rates of return to high levels of activity. More work is required to confirm the long term success of these encouraging early results.

Reduction and Association of the Scapho-Lunate Ligament (RASL Procedure)

The RASL procedure has been proposed as a satisfactory treatment for the early stages of scapholunate instability [78]. It was recognized that failed attempts at scapholunate fusion for scapholunate instability could, in some cases, provide acceptable results, leading Rosenwasser [79] to describe his technique for creating a stable pseudarthrosis, or "neoligament," between the scaphoid and lunate by debriding the interval and then placing a screw across it. Lipton et al. [80] published their results for this procedure, reporting excellent rates of return to work, almost normal dynamic and static radiographic parameters, with only few complications (one screw removal and one conversion to limited wrist arthrodesis). Aviles et al. [78] reported encouraging early results however long term results were not available.

Bony and other associated ligament injuries

Wrist arthroscopy has revealed the extent of soft tissue injuries associated with distal radius and scaphoid fractures. Richards et al. [81] reported on injuries of the soft tissues in a cohort of 118 patients with a mixture of intra and extra-articular distal radial fractures. Up to 21.5 % of patients also had injuries of the scapholunate interosseous ligament, while combined scapholunate and lunotriquetral ligament injuries were seen in up to 13.3 % of extra-articular fractures of the distal radius. Similarly, wrist arthroscopy has been useful in the assessment of combined scaphoid fractures and interosseous ligament injuries [82]. In this study of 41 wrist arthroscopies, in patients with either non-displaced or displaced scaphoid fractures, 29 patients (71 %) had scapholunte injuries. Ten (24 %) of these associated scapholunate injuries were Geissler grade 4, representing a complete scapholunate ligament tear. There was no association between the grade of the scapholunate injury and the displacement of the scaphoid fracture. Further work is required in this area, particularly in regard to treatment of the associated ligamentous lesions found in association with scaphoid fractures.

Lunotriquetral instability arises from injuries to the intrinsic and extrinsic stabilizers of the lunotriquetral interval, as a consequence of hyperextension or twisting injuries of the wrist. The stronger volar and the dorsal interosseous ligament are the primary stabilizers, with the ulnolunate, ulnotriquetral, dorsal radiotriquetral and scaphotriquetral ligaments providing secondary support. Acute injuries can be treated arthroscopically [83], by simply plicating the volar-ulnar sided ligaments. The technique involves confirmation of the instability, with examination of the radiocarpal and midcarpal joints. Gapping, or loss of co-linearity of the lunate and the triquetrum, suggests instability. Through a 6U portal, a spinal needle is passed just volar to the ulnocarpal ligaments and enters the wrist joint, just radial to the ulnolunate ligament (Fig. 13.8). A 2–0

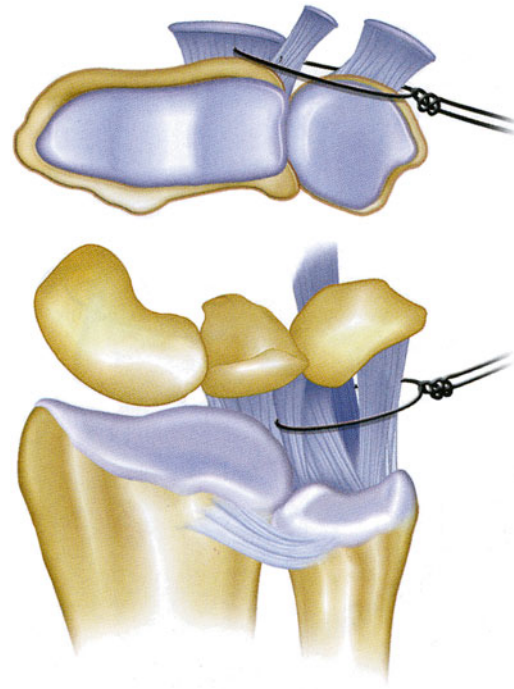

Fig. 13.8 The technique for stabilization of the lunotriquetral interval with plication of the volar ulnar sided ligaments (Reproduced from Moskal in *AANA advanced arthroscopy*: *the elbow and wrist*. Reprint permission obtained from Elsevier)

PDS suture is passed through the needle and is retrieved out of the 6U portal. A second suture is passed 5 mm distal to the first. A final suture is placed through the volar capsule and TFCC for later tying over the ulnar capsule. The arthroscope is moved to the midcarpal portal. Traction is placed on the sutures and the lunotriquetral interval is closed with the aid of a probe. Two Krischner wires are then placed across the luontriquetral interval and the reduction is checked on X-ray. Finally, the sutures are tied over the ulnar capsule through a small incision, avoiding the dorsal branch of the ulnar nerve.

A retrospective review of 21 patients undergoing this procedure at a mean of 2.5 years revealed good to excellent results in 19 [83]. One patient experienced ulnar neuritis while three others had persistent ECU tenderness. The senior author (GIB) warns of the close proximity of the ulnar nerve and that this must be respected. Ulnar nerve injury as a consequence of this technique would be devastating.

Arthroscopic Release of Contracture

Wrist stiffness is the most common complication following wrist trauma or surgery. It is a multifactorial condition and some cases may remain refractory to conservative treatment. Causes can be grouped into either extra-capsular, capsular or intra-articular [84]. In cases that are caused primarily by arthrofibrosis and who have failed conservative treatment, arthroscopic surgical release is our preferred treatment option. Arthroscopic release of the volar and dorsal wrist capsule has been shown to have a low complication rate with good patient outcomes. Poor outcomes will result if the patient has intra-articular degenerative changes, or if the major pathology is extra-capsular.

Luchetti has previously reported good results of arthroscopic debridement of the stiff wrist following distal radial fractures [85]. In a study of 22 patients, routine radiocarpal arthroscopy was performed, augmented by accessory portals as required. Fibrotic adhesions were removed with a resector or diathermy probe. Either the volar or the dorsal radiocarpal ligaments were resected from the edge of the distal radius. It is not recommended that both the dorsal and volar ligaments be resected as ulnocarpal translation may result. Articular steps were levelled with a chondrotome and tears of the TFCC or lunotriquetral ligament were debrided. If supination and pronation remained problematic following radiocarpal and midcarpal debridement, then the distal radioulnar joint was entered and adhesions gently detached from the articular surfaces of the ulnar head, the sigmoid facets of the radius and the undersurface of the TFCC. At mean follow up of 28 months, those with osteochondral injuries or an articular step off had the least improvement in ROM and had the most pain after surgery. In all patients, pain was either significantly improved or absent following surgery. There was also statistically significant improvement in wrist movement and grip strength.

Author's Preferred Method

Release of either the volar or dorsal wrist capsule is chosen dependent upon the clinical indication, whether it be restriction of extension or flexion. Release of the volar capsule is performed through the 3/4 and 6R portals, with the use of a diathermy probe. Care must be taken to avoid injury to the underlying tendons, nerves and arteries. The radioscaphocapitate ligament should be preserved in order to prevent ulnar translocation of the wrist.

Release of the dorsal capsule is likewise performed with the use of the 3/4 and 6R portals, but with the addition of a volar viewing portal (either ulnar or radial). A nylon tape is passed deep to the tendons between the dorsal portals. A resector or basket forceps is then passed into the dorsal portals whilst maintaining traction on the nylon tape, thereby avoiding the risk of tendon injury when resecting the dorsal capsule.

Pearls for Arthroscopic Dorsal Capsular Release

Nylon tape is passed under the extensor tendons using an artery clip (in through the 3/4 portal, out the 6R portal)

View the dorsal capsule through the volar radial portal
Traction is placed on the nylon tape
Debridement of dorsal capsule is performed using arthroscopic basket forceps
Structures at risk are the extensor tendons

Bone

Scaphoid Fixation

Scaphoid fractures are common amongst the young active population. Undisplaced scaphoid fractures are traditionally managed in a cast for 8–12 weeks until union occurs. Displaced scaphoid fractures are typically managed with reduction and internal fixation. This can be performed either in an open manner, percutaneously, or with the assistance of arthroscopy. The concept of arthroscopic assisted scaphoid fixation was introduced by Terry Whipple in the early nineties [86]. Arthroscopic assistance allows direct visualisation of the reduction (particularly rotation) and also allows assessment of associated injuries, such as scapholunate dissociation [82] and TFCC tears. Waist fractures are best visualised from the midcarpal joint while proximal pole fractures are best visualised from the radiocarpal joint.

Author's Preferred Method of Arthroscopic Assisted Acute Scaphoid Fracture Fixation (Geissler Method)

The line of the scaphoid is marked on the skin with the assistance of a Kwire and the fluoroscopy machine. Arthroscopy is performed in the usual manner, creating radiocarpal and midcarpal portals. The ulnar midcarpal portal gives the best visualisation of the reduction of a waist fracture. An arthroscope placed in the radial midcarpal portal can displace the fracture. In order to reduce the fracture, K-wires are placed in the proximal and distal fragments. A small stab incision is made distal to the STT joint. The volar lip of the trapezium is excised with a rongeur, to provide a good line of access to the scaphoid. Once reduction is achieved, K-wires (one down the central axis and another anti-rotation wire) are advanced along the pre-assessed planes and the position of the wire and the fracture is checked with fluoroscopy. A cannulated drill and cannulated screw of choice are then used to internally fix the scaphoid fracture.

Fractures of the proximal pole can also be fixed arthroscopic assisted and percutaneously via a dorsal approach [87]. A 3/4 and 6R radiocarpal portals are required. The arthroscope is transferred to the 6R portal and, after flexing the wrist 30°, a14 gauge needle is advanced into the 3/4 portal. The tip of the needle is advanced into the proximal aspect of the scaphoid, adjacent to the scapholunate ligament and the position is checked on fluorscopy. A guidewire is passed through the needle and the reduction is assessed on fluoroscopy and arthroscopy (midcarpal ulnar portal). If the reduction is unsatisfactory, the guidewire can be driven out volarly, but kept in the distal fracture fragment. K-wires can once again be used to manipulate the fracture fragments, with the reduction viewed either from the radiocarpal or midcarpal portals. Once happy with the reduction, the guidwire is advanced into the proximal pole and out through the dorsal portal again. After checking the position on fluoroscopy, the length of the screw is chosen. A cannulated drill is passed and the screw inserted. The advantage of this method over other methods of dorsal percutaneous fixation is the avoidance of hyperflexion of the scaphoid and the possible production of a humpback deformity.

Slade et al. have reported on the use of arthroscopy of the wrist in the assessment and management of scaphoid fibrous non-unions [88]. Fifteen patients with undisplaced scaphoid fractures and either fibrous union or non-union were treated. Where the cartilage envelope was considered intact on arthroscopy, fractures were treated with internal fixation only, without the use of bone graft. All fractures healed at 14 weeks and there were no complications.

"Ectomy" Procedures

In addition to the aforementioned resection of the proximal pole of the hamate for impingement on the type 2 lunate, arthroscopic resection can be used elsewhere in the carpus.

Radial styloidectomy has been used for the treatment of radial stylocarpal pain since 1948 [89]. Its utility is mainly for carpal instability patterns, which lead to abnormal movements between the distal scaphoid and the styloid, such as SLAC and SNAC (Scaphoid Nonunion Advanced Collapse) wrists and post STT fusion. It may also be used to improve range of motion, following proximal row carpectomy and four corner fusion. Performing radial styloidectomy as an arthroscopic procedure affords advantages of assessment of the remainder of the carpus and also monitoring the extent of resection with particular regard to the radioscaphocapitate (RSC) ligament. Resection beyond this ligament and its adjacent radiolunate ligament can lead to ulnocarpal translocation. The current recommendation is that between 3 and 4 mm may be resected safely [90]. Arthroscopic styloidectomy is performed through 3/4 portal and a 1/2 portal. A 3.5 mm burr placed through the 1/2 portal gives a good idea of the extent of resection required. Reliable pain relief is achieved with this procedure, although improvement in range of motion is less reliable [91].

Arthroscopic excision of the proximal pole of the scaphoid for Preisser's disease has also been reported in a case study [92]. Ruch et al reported on arthroscopic radial styloidectomy and distal scaphoid excision for AVN of the proximal pole of scaphoid with non union [93]. In spite of a progression of carpal collapse, at 2 years all three patients reported an improvement in movement and satisfaction with the procedure, similar to others using an open procedure [79].

Proximal Row Carpectomy (PRC) is a salvage procedure useful for Kienbock's disease and wrist instability patterns such as SLAC and SNAC. It has several advantages over four-corner fusion, including the ability to mobilise the wrist early and bony union is not required. PRC has been performed as an entirely arthroscopic procedure with good results [94] and has advantages over the usual open procedure. Immobilisation is not required, as there is no dorsal capsule incision to heal and it is anticipated that there will be less post-operative pain and scarring. The risk of post-operative wrist instability (ulnocarpal translation) is reduced as the wrist capsule remains intact.

In their non-comparative study of 17 patients undergoing arthroscopic PRC, Weiss et al. [94] reported good post-operative flexion extension arc and grip strength. Patients were either very satisfied or satisfied, whilst 13 out of 16 returned to their previous employment. The technique involves a midcarpal arthroscopy, with the viewing portal being the midcarpal ulnar portal and the working portal, the midcarpal radial portal. The resection is initiated by a burr on the proximal scaphoid from the radial midcarpal portal. Meticulous care is taken to avoid injury to the capitate. The distal pole of the scaphoid is removed through an STT portal. Portals are then switched and the resection continues for the lunate and then the triquetrum. Consideration is given to a radial styloidectomy, if impingement occurs on fluoroscopy.

Ulnocarpal impaction syndrome is caused by abutment of the distal end of the ulna against the lunate and triquetrum. It is typically related to positive ulnar variance, either static or dynamic. Patients with ulnar positive variance, who have TFCC lesions and who don't respond to conservative treatment, are candidates for the arthroscopic wafer procedure as was described in a previous section of this chapter. Meftahet al. [95] have reported on the outcomes of the arthroscopic wafer procedure (average 2.3 mm resection). Studying 26 patients, they found that the presence of MRI changes, consistent with ulnar impaction syndrome (cystic changes, sclerosis and oedema of the proximal lunate) were a good predictor of outcome. Twenty-two patients reported good or excellent pain relief.

Ulnar stylocarpal impaction is an uncommon cause of ulnar sided wrist pain. It is usually seen in patients who have a longer than normal ulnar styloid of 3–6 mm [96]. Open excision for impaction has been described previously [97]. However

Bain et al. [98] perceived benefits in arthroscopic excision. The TFCC could be viewed directly whilst the resection was carried out, hence avoiding the possibility of resecting too much of the ulnar styloid provoking instability of the distal radioulnar joint.

Limited Wrist Fusions

PC Ho [99] has reported the only series in the English literature of cases of 12 partial wrist fusions utilising all arthroscopic techniques. Fixation was achieved with either K wires or cannulated screw devices. Nine of the cases went on to union, two had a stable fibrous union, and another proceeded to non-union at an average follow up of 70 months. Pain and range of motion was reported to have improved in all cases.

Kienbock's Disease

The Lichtman radiological classification system of Kienbock's disease has traditionally been used for assessment and management. This system has been shown to be unreliable [100] and X-ray findings often do not correlate with the arthroscopic findings [101]. Bain and Begg [102] have developed a useful arthroscopic classification scheme, based upon the number of functioning articular surfaces and the presence of a coronal lunate fracture (Fig. 13.9). It was their observation that the pattern of degenerative changes proceeded from the proximal surface of the lunate, to involve the lunate fossa of the radius, followed by the distal end of the lunate and then finally the proximal capitate was involved. In a separate category, the coronal fracture of the lunate creates two non-functional surfaces. This "joint based approach" to Kienbock's can guide the reconstructive approach based on the number of non-functional surfaces.

This grading system has a logical progression and gives the surgeon direction in the management of the condition. Grade 0 may require a joint levelling procedure or lunate re-vascularisation [102]. Arthroscopic forage procedures have been described [70], but efficacy is unproven. Grade 1 can be managed either with proximal row carpectomy or radioscapholunate fusion. Grade 2a can be managed with radioscapholunate fusion, while 2b requires proximal row carpectomy. Grades 3 and 4 would typically be treated with salvage procedures, such as wrist arthrodesis or wrist arthroplasty. Arthroscopic debridement alone for late stage Kienbock's disease has been investigated and has shown promising results at 19 months follow up [103], although long term outcomes are unpublished. The debridement involved removing all synovitis and loose chondral flaps back to a bleeding base.

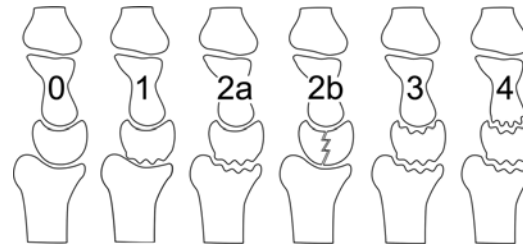

Fig. 13.9 Grade 0 Kienbock's disease represents the early stages of symptoms and changes on MRI (low signal on T1 weighted imaging and high signal on T2 weighted imaging). At arthroscopy the joint surfaces are normal. Grade 1 involves the proximal lunate surface. Grade 2a reveals degenerative changes in the lunate fossa as well as on the proximal lunate. Grade 2b is the coronal fracture of the lunate, a poor prognostic indicator. Grade 3 has involvement of the distal lunate surface and grade 4 has pan-lunate changes (Reproduced from Bain et al. [7]. Reprint permission obtained from Elsevier)

Assessment of Intra-articular Distal Radius Fractures

The aim of treatment for intra-articular distal radial fractures is bony union enabling pain free movement of the wrist joint. Better radiographic or clinical outcomes have been associated with reduction of articular steps to within 1 [104] or 2 [105] mm. Others [106, 107], however, have not shown correlation between articular gaps or steps and clinical outcomes. Wrist arthroscopy has been shown to better assess the articular step than fluoroscopy [108] and on that premise, some advocate wrist arthroscopy as a necessary adjunct to fixation of intra-articular distal radial fractures

[109]. There are other advantages to the use of wrist arthroscopy for the assessment of distal radius fractures, including the ability to assess chondral injuries, assess associated ligament lesions and prevent intra-articular placement of screws. Further research is required to determine whether the treatment of associated ligament injuries affects outcomes. Disadvantages of combining wrist arthroscopy with distal radial fracture fixation include increased expense and use of resources, longer operative time and the requirement for a new skill set.

Ruchet al. [110] undertook a prospective non-randomised study of arthroscopic assisted versus fluoroscopic assisted fixation of intra-articular distal radius fractures. Fifteen patients in each group underwent fixation, with an external fixator combined with Kirschner wires. Seven peripheral TFCC tears identified at arthroscopy were repaired with an outside in technique. Range of motion at a minimum 12 month follow up was significantly improved in the arthroscopic group. Radiographic parameters and DASH scores were not significantly different between groups.

In a similar but randomised, prospective study, Varitimidis et al. [111] examined the clinical outcomes following fluoroscopic versus fluoroscopic with arthroscopic assisted treatment of intra articular fractures of the distal radius. All patients underwent external fixation and percutaneous pinning. Associated ligamentous and chondral lesions found at arthroscopy were attended to at the time of fixation, either with closed or open repair. At 12 month follow up there was a significantly improved range of motion and at 24 months an improved Modified Mayo Wrist score in the arthroscopically assisted group. They suggested that attending to associated injuries at the time of the fixation was necessary, as their clinical outcomes were either similar or improved.

Author's Preferred Technique

With the advent of fixed and variable angle volar plating, we now rarely use the traditional techniques of external fixation and K-wires for intra-articular distal radial fractures. The majority of patients are managed without arthroscopy. However, we use arthroscopy for the young, high demand patient with an intra-articular fracture, where there is a suspicion of associated ligament injury and in those cases in which there is concern that the screws may penetrate the joint.

A combination of open and arthroscopic surgery is used. We use the standard FCR approach to the volar distal radius. The fracture is provisionally reduced under fluoroscopic control and the plate applied to the volar surface of the radius. A cortical screw is inserted into the sliding hole of the plate. K-wires are inserted into the main articular fragments allowing manipulation. The limb is suspended in a traction tower and the arthroscope is placed in the ¾ portal. Switching the arthroscope between the ¾ and 6R or a volar portal can be used to assess articular steps or gaps and allows manipulation of the articular fragments with the K- wires or a probe. Temporary fixation can be achieved by advancing the fragment wires through the opposite cortex and then the distal locking screws are placed to achieve angular stable fixation.

Associated ligamentous lesions are treated at the time of fixation of the radial fractures. Peripheral tears of the triangular fibrocartilage are repaired as per the technique described earlier in this chapter. Lower grade (grade II Geissler) tears of the scapholunate and lunotriquetral ligaments can be percutaneously pinned after joystick manipulation, to a reduced position. For higher grade tears (grade III and IV Geissler) the author uses a dorsal open repair with tensionable suture anchors, as we have found the percutaneous techniques unrewarding.

Post Operative

Following wrist arthroscopy, the wrist is typically put into a crepe bandage for simple excision or debridement procedures. Where more extensive surgery, such as a ligament reconstruction has occurred, the arm is put into a plaster splint. The operated wrist is elevated. The hand is kept elevated and dry until the post-operative clinic visit, usually in 4–7 days. Analgesia is prescribed and leaking dressings are reinforced. Hand therapy is usually begun on the first day,

including long flexor exercises and elbow and shoulder exercises.

Complications

Wrist arthroscopy is considered a low risk procedure. Complications do occur however, including tendon injury from inappropriate portal placement, septic arthritis and wound infections, skin slough from finger traps, nerve injury and chronic regional pain syndrome. One must have a thorough understanding of the anatomy over the dorsal and palmar wrist, so that arthroscopic portals are inserted with minimal risk to the adjacent structures. Portals which are used uncommonly, such as the 1/2 and the 6 U portals, present the greatest risk. Longer skin incisions and careful subcutaneous dissection is required, in order to preserve the sensory nerves and the radial artery.

The Future of Wrist Arthroscopy

We have seen that wrist arthroscopists have been willing to challenge the limits of their abilities in their attempts to apply open techniques to the arthroscope. The purported benefits apply to all arthroscopic procedures, reduced wound problems, improved range of motion and faster recovery. With this in mind, areas under development include the treatment of scaphoid nonunions, scapholunate ligament reconstruction and wrist fusions. Slade and Dodds [112] have reported clinical success with their arthroscopic treatment of scaphoid non-unions. They emphasize that the surgical principles of non-union surgery remain: maintenance of blood supply, nonunion debridement, fracture reduction, bone grafting, and rigid internal stabilization. Arthroscopic scapholunate ligament reconstruction [70] has been proposed, but no clinical data has been published as yet. Arthroscopic scapholunate ligament capsuloplasty has been reported in a limited series [69]. PC Ho has pioneered arthroscopic limited wrist fusion techniques with success [99]. These are all areas which will require further investigation of their clinical efficacy over the coming years.

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PIPJ Replacement

14

Jonathan Hobby

Keywords
Arthritis • Arthroplasty • Proximal interphalangeal joint • Surgical technique • Surgical outcomes • Total joint replacement • Silastic arthroplasty • Osteoarthritis • Inflammatory arthritis • Treatment • Hand

Introduction

Mobile proximal interphalangeal joints (PIPJ) are necessary for normal hand function; both inflammatory and degenerative arthritis can affect the PIPJ leading to pain, stiffness, deformity and loss of function. The aims of arthroplasty are to relieve pain, provide a functional range of movement and reduce deformity. The proximal interphalangeal joints (PIPJ) in the hand have long been a challenge in arthroplasty surgery. There has recently been a surge in the commercial interest in small joint replacement in the hand, with a number of new implants reaching the market. Inevitably, few of these have an established track record in clinical use. This is a controversial area, as past experience of small joint arthroplasty has been almost universally disappointing and concerns have been raised regarding the long term outcomes of the new generation of implants. The results of PIP joint arthroplasty have not yet matched the success of large joint replacements [1].

A predictable and durable arthroplasty of the PIP joint remains an unsolved problem in the care of the osteoarthritic hand [2].

Background

The PIPJ is an important link of the kinetic chain of finger joint movement, as it provides about 40 % of the total active range of movement in the finger. It is functionally important for grasping smaller objects and handling irregularly shaped objects. The functional range of motion of the PIPJ has been described as between 30° extension to 85° flexion [3]. If the metacarpophalangeal joints are intact, an extension deficit is functionally better tolerated than a lack of flexion.

The kinematics of the PIPJ are supposedly simple. It is a bicondylar diarthrodial joint, with a single fixed centre of rotation at the insertion of the collateral ligaments. It is as near to a hinge as any other joint in the body, with one degree

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of freedom. The stability of the joint is primarily due to the bicondylar joint surface and the collateral ligaments. Other structures contribute significantly to stability, including the volar plate and the accessory collateral ligaments, the lateral bands of the extensor apparatus, the transverse and oblique retinaculae, and the digital fascia. The normal range of movement is 0–115°. There is very little lateral deviation or rotation throughout the range of movement in the normal PIPJ joint, but there are some anatomical variations in the bony anatomy between digits which may mean the uncomplicated hinge concept is an over-simplification [4, 5].

The importance of maintaining PIPJ motion has long been recognised. Burman reported the use of a vitallium cap for PIPJ arthroplasty in 1940 [6]. In 1959 Brannon & Klein published the results of the first total joint replacement, using a hinged prosthesis [7]. Two years later Flatt reported a device with two intramedullary prongs to increase the rotational stability of the fixation in bone [8]. There were some encouraging early results, but none of the initial implants achieved satisfactory long-term results [9].

Indications for Anatomical PIPJ Replacements

Inflammatory arthritis commonly affects the PIPJs. However, the effect of these arthritides on the soft tissues and the implications of this on joint stability has led some to question whether surface replacement arthroplasty is appropriate [9]. The increasing use of disease modifying drugs has reduced the need for reconstructive surgery in the rheumatoid hand. The soft tissues in these patients are better preserved and, in my experience, arthroplasty is a reasonable option. The primary indication for Anatomical PIPJ arthroplasty is osteoarthritis. The increasing demands of an ageing population, to maintain hand function has increased the interest in PIPJ replacement for osteoarthritis. These are a higher demand patient group, in whom silicone arthroplasty is less durable. Arthroplasty can also be considered following trauma, but the results have in general been disappointing, particularly in younger high demand patients.

Adequate soft tissues are required to maintain stability following surgery; the collateral ligaments and volar plate are particularly important. Fixed deformities or severe bone loss may also compromise the results of surgery. Contraindications to surgery include active infection, and loss of extensor or flexor tendon function.

Clinical Pearl

Patient selection: adequate bone stock and sufficient soft tissue integrity to allow a stable reconstruction in a well motivated patient with realistic expectations

Presentation, Investigation and Treatment Options

Patients present with pain, stiffness and deformity. Digits may deviate in the coronal plane, leading to overlap in flexion with marked functional impairment. In some cases the finger may be excluded from hand function entirely. Patients may also be concerned about the cosmesis of the finger.

Clinical assessment should include a thorough history, including age, occupation and leisure pursuits; pain and use of analgesia; functional restrictions; previous hand injuries and infection; previous treatments. The whole hand should be examined and the PIPJ assessed for deformity, tenderness, stability and both active and passive range of motion. The MCPJ and DIPJ should also be assessed, as they will have a significant impact on the final functional result. Standard AP and lateral radiographs should be obtained.

Treatment options include analgesia and hand therapy, corticosteroid injections can afford lasting pain relief and, if combined with a gentle manipulation under local anaesthetic, they can significantly improve symptoms and hand function, delaying the need for arthroplasty. The surgical alternatives include arthrodesis, silicone arthroplasty, hinged designs and anatomic joint replacement.

Arthrodesis

Arthrodesis affords reliable pain relief and the results are maintained in the long term. The procedure is technically simple and fixation can be achieved with k-wires, wire loops or plates. The preferred position varies by finger. The index should be arthrodesed near full extension to allow tip and tripod pinch. The ring and small finger in mid flexion to allow grasp. Although reliable, arthrodesis imposes significant functional restrictions.

Silastic Implants

The Swanson silicon implant developed in 1962 has been the most widely used PIP joint prosthesis and has the longest documented follow up [10]. Good results have been reported in 60–90 % of patients at 5 years follow-up [11–13]. These studies report consistent pain relief and an average range of movement of approximately 40° [3]. However, the Swanson prosthesis has recognised complications and deficiencies; in particular prosthetic fracture and silicone synovitis [14, 15].

The Swanson implant has been reported to provide pain relief in 49/70 (70 %), a mean ROM 31° (17–48), a revision rate of 13 % (9/70), with implant deformation in 18/70 (26 %) and implant fracture in 11/70 (16 %) at a mean follow-up of 6 years (3–20 years) [10]. The outcomes deteriorated significantly after 10 years. It is not recommended for use in the index finger, due to problems with instability and is probably best restricted to use in older patients. Despite these limitations the Swanson implant remains the standard to which any new design should be compared. There are a number of other designs of silicone implant, including the Neuflex and Avanta, which perform similarly.

An implant consisting of a flexible silastic spacer connected to titanium stems has been developed in Sweden. At 8 year follow-up 47/50 stems had osseointegrated, but 17/25 (68 %) of the silicone spacers had fractured [16].

Silastic arthroplasty remains a simple reliable procedure, which is well suited to the ring and middle finger in older low demand patients, but it has a restricted range of motion and limited life span which makes it a less attractive option for younger high demand patients

Hinged Implants

Most of the early designs for PIPJ were based upon a simple hinge. These designs can closely mimic normal joint kinematics, and have advantages in-terms of stability. However, the level of constraint places huge stresses at the hinge of the implant and at the interface between the implant and the bone. The long-term results have been universally disappointing.

A recent hinged design is the LPM prosthesis, which is a two component, semi-constrained hinge joint (Van Straten Medical, Netherlands). The implants are manufactured from cobalt-chrome alloy, with a titanium porous coating (proximal 1/3rd). The proximal (female) component has a concave cylinder with a central socket. The distal (male) component is convex and fits into the proximal component socket. The articular surfaces have a ceramic titanium-niobium coating, to minimise initial wear. The bearing is designed to allow some abduction and adduction of the interphalangeal joint, which is gradually limited as the finger is flexed. This is designed to mimic the stability of the healthy PIPJ in flexion.

There were some early reports of encouraging short-term results, but none were published [17, 18]. Anecdotal reports of early failure, coupled with the first published series of results, led the Research and Audit Committee of the British Society for Surgery of the Hand to review the outcomes of the LPM proximal interphalangeal joint replacement in the United Kingdom in 2008 [19, 20]. Outcomes were retrieved for 164/257 Implants that had been sold in the UK at that time; 80/164 (49 %) were showing clinical and radiological signs of failure, and 47/164 (29 %) were known to have been revised. Massive osteolysis was a common finding with aseptic loosening

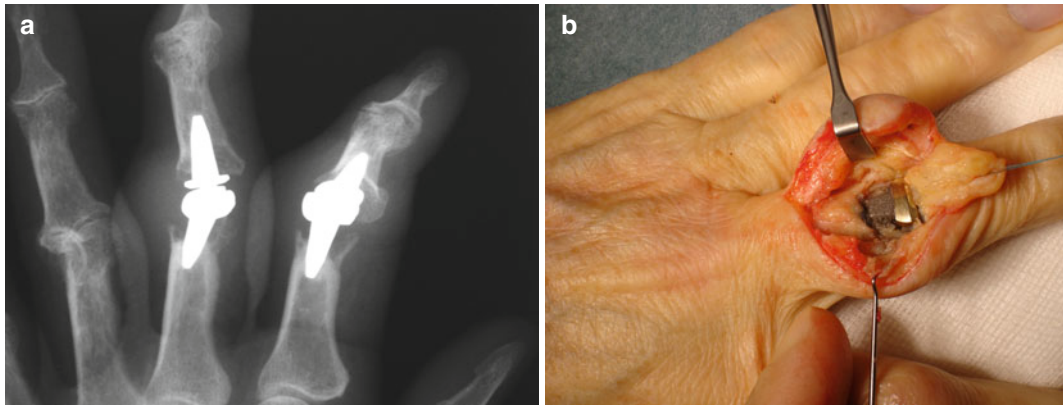

Fig. 14.1 (**a**, **b**) Radiographs and operative photographs showing osteolysis and metallosis in failed LPM implants

of many implants. The majority of failures had started showing signs of failure within 18–24 months. There were reports from surgeons that the function of many of the surviving LPM implants deteriorates with time, with progressive stiffness and osteolysis. This audit highlights that, in spite of the reports of good early results from some early users of the LPM, many of these implants started to fail after 1–2 years.

At revision surgery there was usually a dark stained membrane with severe osteolysis (Fig. 14.1a, b). Despite the osteolysis many implants remained well fixed. Surgical specimens submitted for histological examination have revealed a chronic inflammatory membrane with collections of macrophages containing dark particles (which are presumably metallic debris). The cause of failure seems likely to have been a hypersensitivity reaction to metallic debris, akin to ALVAL recently described in metal on metal hip replacements. LPM Implants have been successfully revised with Swanson implants, arthrodesis and surface replacements. Some have also been revised to larger LPM implants with or without cement. A number of fingers have required amputation. The authors recommended that the LPM implant should not be used and that all patients who have had this prosthesis inserted should be kept under regular clinical and radiographic review. The early failure of the LPM illustrates the importance of collecting outcome data for new implants.

Surgical Technique and Approaches

Dorsal Approach

The dorsal approach is probably the easiest and most forgiving approach to the PIPJ. A straight or curved dorsal incision is made, and thick skin flaps are raised from the extensor mechanism. A straight incision can be converted to a z-plasty if the dorsal skin is tight. Branches of the dorsal veins are coagulated. There are three options for dealing with the extensor mechanism. A distal flap based upon the central slip can be raised (Chamay). This has the potential advantages of leaving the insertion of the central slip intact and it allows the balance of the extensor mechanism to be adjusted during closure. However, access may be limited by large dorsal osteophytes. The simplest approach is to split the central slip and elevate the insertion subperiosteally; adequate exposure can usually be obtained without completely elevating the insertion. Dorsal osteophytes can then be trimmed. An approach between the central slip and lateral band has been described; this has the advantage of leaving the central slip insertion intact and affords sufficient exposure to perform a silastic replacement, but is probably insufficient for anatomical replacements.

Adhesions of the extensor mechanism can be mobilised with a blunt dissector. The capsule is opened and a synovectomy performed. The origins of the collateral ligaments can be released

sub-periosteally from the head of the middle phalanx. When severe arthritic changes are present, collateral release may be required to allow joint flexion, but if anatomical joint replacement is intended, great care must be taken to preserve the integrity of the collateral ligaments and their distal insertion into the base of the middle phalanx, as they are essential for joint stability.

The main disadvantage of the dorsal approach is the risk of stiffness due to adhesions of the extensor mechanism. The tendons of the extensor mechanism have a small excursion relative to the flexors and, if adhesions form, they can be extremely difficult to overcome. Most series of anatomical replacements report some secondary surgery for stiffness, which usually involves tenolysis, but unfortunately this rarely produces a satisfactory improvement in the range of motion. Good surgical technique is important, dissection of the extensor mechanism should be minimised and the tissues must be handled sympathetically and closed accurately. Early motion should be instituted under hand therapy supervision. Patient selection is also important, as patients who are unable to co-operate with post-operative rehabilitation are unlikely to achieve satisfactory outcomes.

The medullary canals of the proximal and middle phalanges are opened with a sharp awl. Accurate implant positioning is dependent upon identifying the centre of the medullary canal and I would strongly recommend using an image intensifier to assist with positioning of the entry point for reamers, and to check the position and size of broaches. The head of the proximal phalanx is resected with a powered saw. Care should be taken to preserve the insertions of the collateral ligaments. The bone cuts should be perpendicular to the long axis of the bone; some implant systems have jigs to assist with orientation of the bone cuts. The amount of bone resected is crucial to accurate soft tissue balancing, as there is limited opportunity for soft tissue releases in the PIPJ. If too little bone is removed, the joint will be overstuffed, the balance of the extensor mechanism will be altered and the postoperative range of motion is likely to be restricted. If too much bone is resected, joint instability is more likely and the joint may hyperextend leading to a swan-neck deformity which greatly increases the risk of implant loosening.

The bone is then prepared to accept the implant stems. Most implants have broaches to allow accurate reaming of the medullary cavity. Implant sizing is important. An appropriately sized implant will allow a satisfactory press fit within the bone, too small and the risk of loosening/subsidence and implant migration is increased. If too large an implant is used there is a risk of fracture/cortical perforation during reaming. If the implant is too small the joint may be lax and unstable, if too large then the joint is likely to be "overstuffed" with a risk of stiffness. Some implants allow a mismatch between the size of the proximal and distal components; others require the same size to be used for both, in which case the implant size is usually determined by the distal component.

When preparing the base of the middle phalanx there are often large marginal osteophytes; there may be bone loss leading to angular deformities of the joint, which require correction. A powered saw can be used, but I prefer to use a burr to create a flat surface to seat the implant. I have found that it is difficult to preserve the bony footprint for the insertion of the central slip of the extensor tendon when using a saw, and the use of a saw also risks disrupting the insertion of the collateral ligaments and volar plate, which can lead to instability. I prefer to use broaches to prepare the medullary cavity, as this creates an accurate press fit and I believe that there is a risk of thermal osteonecrosis leading to implant loosening if the medullary cavity is prepared with a high-speed burr. If the press fit obtained after reaming is unsatisfactory, impaction of bone graft can be used to improve fixation. Implant sizing and positioning are vital to the stability and kinematics of the joint. The centre of rotation and joint line should be reproduced as accurately as possible. For this reason I would recommend the use of an image intensifier at each stage of the procedure.

A side to side repair of the extensor tendon is performed. The central slip can be reattached to the middle phalanx with trans-osseous sutures,

but great care must be taken to avoid altering the balance of the extensor tendon. Very satisfactory results have been reported for a central slip splitting approach, without reinserting the tendon into the base of the proximal phalanx [21].

Volar Approach

The volar approach is technically demanding, but has the potential advantage of leaving the extensor mechanism undisturbed. A Volar Brunner type incision is made. The neurovascular bundles are protected and a thick skin flap raised. The flexor sheath is exposed and Clelland's ligaments are released to allow the neurovascular bundles to be mobilised. The Flexor tendon sheath is opened, preserving the A2 and A4 pulleys and the flexor tendons are retracted. The Volar plate is released from the base of the middle phalanx and a proximally based "trap-door" type flap is raised. This exposes the joint, which can then be hyper-extended to allow preparation of the bone surfaces. Particular care must be taken during closure to repair the volar plate preventing swan neck deformity post-operatively.

The volar approach does have some potential advantages, particularly in facilitating early rehabilitation, without the need to protect a tendon repair and may offer a better range of motion. However, it is technically more demanding that the dorsal approach and preparation of the bone surfaces is more difficult.

Lateral Approach

The lateral approach has been advocated for both silicone and pyrocarbon implants. A mid-lateral incision of approx. 4 cm is made on the radial side of the digit. The neurovascular bundle (including the dorsal branch) is identified, mobilised and protected. The transverse retinacular ligament is divided and the lateral band of the extensor tendon is retracted to expose the collateral ligament. The collateral ligament can either be released subperiosteally from its origin from the head of the middle phalanx and the accessory collateral released; or the collateral can be split in line with it's fibres and divided as a z. The volar plate and central slip should be preserved. The joint can be hinged open. Osteophytes are removed and bone cuts made. Prior to closure tranosseous drill holes are made to allow the ligament to be reattached with a non-absorbable suture. Post-operatively, the finger is protected with a resting splint and/or neighbour strapping.

Good results have been reported with the lateral approach. The lateral approach leaves the flexor and extensor mechanisms undisturbed. However, there is a risk of varus/valgus instability, due to failure of the ligament repair and orientation of the bone cuts can be difficult until experience is gained with the approach. Most of the implant jigs are designed for a dorsal approach.

Clinical Pearl

Dorsal approach, splitting the central slip, is the easiest approach

Preserve collateral ligaments for joint stability

Use image intensifier to allow accurate implant placement and sizing

Precise bone resection and accurate broaching

Soft tissue balance for stability and ROM

Meticulous extensor repair – do not over-tighten central slip

Early supervised mobilisation

Anatomical Designs

In an attempt to overcome the problems associated with silastic arthroplasty, other designs have been developed using different materials. The Ascension, Avanta-SRA and Finsbury are all anatomical PIPJ total joint replacements with press fit components designed for uncemented implantation (Figs. 14.2, 14.3 and 14.4). The Avanta-SRA has a cobalt chrome proximal component and a titanium distal component with an UHMWPE bearing surface [22]. The Finsbury is a similar design, with an HA coating to pro-

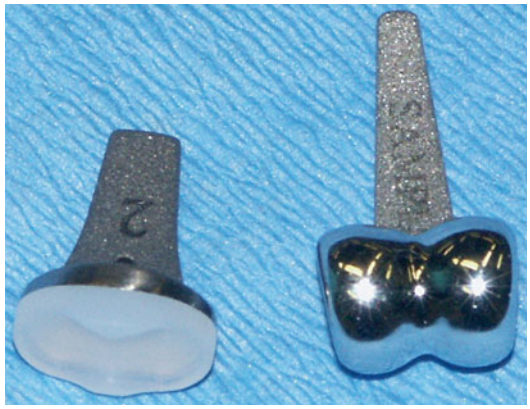

Fig. 14.2 The Avanta SRA PIPJ replacement

Fig. 14.3 The Ascension Pyrocarbon PIPJ replacement

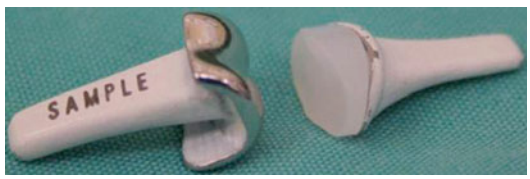

Fig. 14.4 The Finsbury PIPJ replacement

mote osseointegration and a rotating hinge to reduce stresses at the bone implant interface. The Ascension implant (Bravo et al. 2007) is made of a graphite substrate coated with pyrocarbon [23]. All three prostheses aim to provide a stable joint with a functional range of movement and therefore retain PIP joint function.

The current generation of prostheses are designed as uncemented implants, relying on an accurate pressfit for initial stability and bone ingrowth for long term fixation. Most have metal stems with a porous coating and some are coated with hydroxyappatite to encourage bone ingrowth. Cement has fallen out of favour due to the difficulties posed by revision of failed implants and concerns regarding the effects of heat generated during polymerisation at implantation, upon the small bones of the finger. However, significant problems with early implant loosening have been reported with uncemented Avanta SRA implants, and reports suggest that the mid-term results are better when cement is used [24, 25]. If an implant does truly osseointegrate, then revision of a failed implant can present a major challenge; as it can be extremely difficult to remove a well fixed stem. Early reports suggested that pyrocarbon implants would osseointegrate, but subsequent animal studies and clinical experience has shown that few if any of the pyrocarbon implants truly integrate [26, 27]. In clinical use, some initial migration is common, affecting 40–50 % of implants in some series, which has caused considerable concern [28]. However, most implants seem to stabilise after 12–18 months.

Linschied et al. reported acceptable mid-term results with the Avanta-SRA implant [22]. Other authors have reported problems with implant loosening when the Avanta-SRA is used without cement, with radiographic loosening in 30–33 %, and revision rates of 17–26 % at 3 years [24, 25]. Both have reported much better results when the Avanta-SRA is implanted with cement. My experience suggests that it may be possible to salvage loose implants with bone cement, but I am reluctant to use cement for primary PIPJ surgery.

The Ascension has also shown radiographic signs of loosening, but it appears to subside and then stabilise in most cases. Implant migration is well recognised with this prostheses [23, 28]. The Mayo Clinic group, who designed the prosthesis, has reported "secondary stabilisation," but this is controversial [23]. Similar short and medium term results have been reported by many other authors (Table 14.1). Sweets and Stern (2011) have expressed reservations over the longer-term results of the Ascension, due to implant migration and progressive stiffness [30]. Problems have been reported when the Ascension implant is used for post traumatic arthritis [35].

The Moje implant is an anatomical design made of a ceramic. Satisfactory early results were reported by Pettersson et al. (2006) for a series of 20 implants, with 12 months follow-up [36]. However, at mid-term follow-up Weseman et al. (2008), reported that although 80 % of patients reported pain relief, the average range

Table 14.1 Showing pooled results for the Avanta SRA and Ascension Pyrocarbon implants, including unpublished results from Hobby, Watts and Herren, all cause re-operation rates include revision and amputations, pain scores are VAS out of 10

	FU (years)	Joints	Migration		Dislocation		Reoperation		Revision		Amputation	ROM	Pain
Ascension													
Bravo et al. (2007) [23]	3	50	20	40 %	2	4 %	15	30 %	6	12 %	2	47	1
Herren et al. (2007) FESSH	4	17	9	53 %	0	0 %	1	6 %	1	6 %	0	39	1.5
Wijk et al. (2010) [29]	2	50			0	0 %	7	14 %	6	12 %	0	52	0.4
Watts et al. (2012) [40]	5	97					22	23 %	13	13 %			
Hobby et al. (2010, unpublished)	4	17	6	35 %	2	12 %	4	24 %	2	12 %	0	65	3
Sweets and Stern (2011) [30]	5	31	15	48 %	5	16 %	6	19 %	5	16 %	0	31	
Hutt et al. (2012) [31]	6	18	10	56 %	0	0 %	4	22 %	2	11 %	1	45	0
McGuire et al. (2012) [32]	2	57	17	30 %	0	0 %	16	28 %	5	9 %	0	66	1.6
Mashhadi et al. (2012) [33]	4	24	0	0 %	1	4 %	4	17 %	0	0 %	0	46	0.9
Total	**3.7**	**361**	**77**	**36 %**	**10**	**4 %**	**79**	**22 %**	**40**	**11 %**	**3**	**46**	1.1
Cemented Avanta													
Linscheid et al. (1997) [22]	4	*63*	*0*	0 %	5	8 %	12	19 %	*2*	3 %	2	*47*	
Johnstone et al. (2008) [25]	6	*24*	*1*	4 %	0	0 %	5	21 %	*2*	8 %	0	49	1
Jennings and Livingstone (2008) [24]	3	*45*	*2*	4 %	0	0 %	2	4 %	*2*	4 %	0	56	
Total	**4.3**	***132***	*3*	**2 %**	**5**	**4 %**	**19**	**14 %**	**6**	**5 %**	2	50	
Uncemented Avanta													
Johnstone et al. (2008) [25]	4	19	13	68 %	0	0 %	5	26 %	5	26 %	0	55	1
Jennings and Livingstone (2008) [24]	3	41	16	39 %	0	0 %	16	39 %	16	39 %	0	56	
Luther et al. (2009) [34]	2	24	4	17 %	0	0 %	14	58 %	4	17 %	0	50	3
Hobby et al. (2010, unpublished)	4	21	6	29 %	2	10 %	7	33 %	4	19 %	0	50	3
Total	**3.2 years**	**105**	**39**	**37 %**	**2**	**2 %**	42	**40 %**	29	**28 %**	0	53	2.4

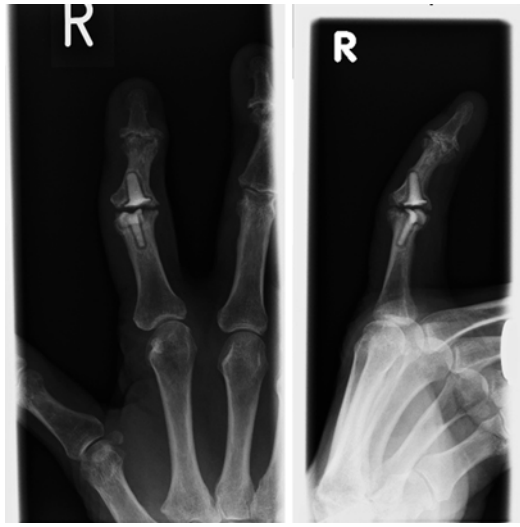

Fig. 14.5 Migration of an Ascension implant

of motion was less than 20° and re-operation in 9/21 (43 %) implants; They concluded that "The Moje ceramic implant shows neither the required performance, nor the ideal attributes needed for an adequate PIP joint replacement" [37].

Anatomical PIPJ replacements remain a controversial procedure. There have been many reports of deteriorating results with time; with implant migration (Fig. 14.5), progressive stiffness and instability. The surgery is technically demanding and patient selection is important, but I have found that the majority of patients are satisfied with the outcomes. The Royal College of Surgeons of England have highlighted a clinician's duty to "audit the outcomes of new procedures" and "review their progress within an appropriate peer group" (RCS 2002) [38]. The National Institute for Health and Clinical Excellence (NICE) published guidelines on the use metacarpophalangeal and interphalangeal joint replacement in February 2005 [39]. It has recommended that: "the results of the procedure are monitored" (NICE/IPG 110). It is cleat that surgeons engaging in implant surgery have a responsibility to monitor the outcomes and share their results, good or bad.

I believe the mid-term results with the Ascension implant to be acceptable and believe this gives better functional results than arthrodesis of the PIPJ. I no longer use the Avanta-SRA implant, as I feel that the revision rate for this implant is unacceptably high; it is higher than that found for the Swanson implant, which has been reported to have a revision rate of 13 % at 6 years [10]. I have also found that the greater bulk of the distal prosthesis in the Avanta-SRA system has caused me difficulties with preservation of the collateral ligament insertions and soft tissue balance. I believe that soft tissue balance is key to this procedure. The collateral ligaments should be preserved to maintain joint stability. Soft tissue balance is also key in achieving a satisfactory range of motion, stiffness is common if the joint is too tight. I have also experienced problems when the extensor tendon balance is not restored correctly. I have found PIPJ stiffness to be a problem if the central slip is advanced during closure.

Complications and Salvage

Deep infection is a serious complication, which is rare (>1 %) following PIPJ replacement. As with all implant surgery, meticulous aseptic technique should be observed and peri-operative antibiotic prophylaxis is indicated. If a deep infection occurs early after surgery it may be possible to salvage an implant with an open debridement and a prolonged (6-week) course of appropriate antibiotics. For most deep infections the implant will need to be removed. It is crucial to identify the infecting organism: all antibiotics should be stopped for at least 48 h prior to surgery and deep tissue specimens should obtained for microbiological culture at the time of implant removal. A thorough debridement should be performed with copious irrigation. An antibiotic loaded bone cement spacer can be inserted and a prolonged course of antibiotics should be given before revision to another prosthesis or arthrodesis.

Early dislocations can often be managed with a closed manipulation and splinting. If there is gross instability, implant loosening or malposition, revision of the implant may be necessary.

Stiffness is a relatively common complication of surgery. Careful soft tissue balancing and early

supervised post-operative mobilisation is essential. Most authors report disappointing results from secondary surgery to improve range of motion. The post-operative range of motion will, to a large degree, depend upon the degree of stiffness prior to surgery.

Deformity; Swan neck and Boutonnière deformities have been reported following surgery. Preserving the volar plate and appropriate soft tissue balancing is crucial. It is probably unwise to perform arthroplasty in patients with a pre-operative swan neck deformity. The balance of the extensor mechanism must be maintained and, if the central slip is reattached, particular attention should be paid to avoid over tightening the tendon.

Failed implants, can at times, be revised to another anatomical implant, often a larger sized implant is required to obtain a satisfactory press-fit and stability. If not, then bone cement can be used. Otherwise, a silastic implant can usually be inserted. Arthrodesis is an alternative, which may be more appropriate in the index finger, where joint stability is probably more important than range of motion. A structural bone graft is likely to be required, but joint fusion can usually be achieved.

Problems of implant loosening and migration have already been discussed.

To date there have been few if any reports of problems due to wear of the bearing surfaces.

Conclusions

Anatomic PIPJ arthroplasty has a disappointing track record, but the new generation of implants have achieved better mid-term results. The surgery is technically demanding and factors leading to poor outcomes have been identified; these include component malposition, excessive bone resection and inadequate soft tissue balancing. The procedure should be considered in younger, higher demand patients, but they should be fully advised of the risks and uncertainty regarding long-term outcomes. Surgeons using these implants have a duty to collect and report the outcomes of their patients. Silastic arthroplasty remains a simple reliable procedure, which is well suited to the ring and middle finger in older low demand patients, but it has a restricted range of motion and limited life span, which makes it a less attractive option for younger high demand patients

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MCP Arthroplasty

15

Arnold-Peter C. Weiss

Keywords
Finger • Rheumatoid • Osteoarthritis • Implant • Arthroplasty • Metacarpophalangeal • Joint • Replacement • Silicone • Arthritis

Arthritis of the metacarpophalangeal (MCP) joint can be a debilitating condition, with patient complaints of substantial pain, stiffness, and progressive deformity [1, 2]. This condition is seen most frequently in patients with rheumatoid arthritis, although it can also occur in primary or post-traumatic osteoarthritis (OA) [1–5].

The MCP joint functions as a complex hinge joint, important in gripping and pinching activities. The normal range of motion is 0°–90° flexion, with a slight amount of radial and ulnar deviation. The joint is reliant upon its surrounding soft tissue structures for stability, including the volar plate, extensor mechanism/sagittal bands and radial and ulnar collateral ligaments [2].

In rheumatoid arthritis patients, inflammation within the synovium of the joint is seen often in a symmetric fashion. The subluxation pattern is typically radially at the wrist joint and ulnarly at the MCP joint. With disease progression, the proximal phalanx will subluxate in a volar direction on the metacarpal head and the intrinsic musculature will contract [1–3]. Patients with primary osteoarthritis of the MCP joint generally complain of pain at the MCP joint, exacerbated by activities of gripping and pinching. Clinically, there may be soft tissue swelling adjacent to the joint itself, although there is no obvious deformity.

Diagnosis of MCP arthritis can be made clinically, but is often confirmed with plain radiographs. In primary osteoarthritis, plain radiographs identify joint space narrowing, subchondral sclerosis and osteophyte formation. In the rheumatoid patient, along with the normal OA finding, radiographs will identify erosions, subluxation, ankylosis, and joint malalignment [6].

MCP arthroplasty is performed to reduce pain, improve motion and function and correct deformity [1–11]. Silicone MCP implants were the first developed in the 1960's and are still considered to be the gold standard [3]. The original Swanson implant was an intramedullary-stemmed one-piece design, that provided both immediate stability and intrinsic flexibility [12].

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Fig. 15.1 A pyrolytic carbon implant

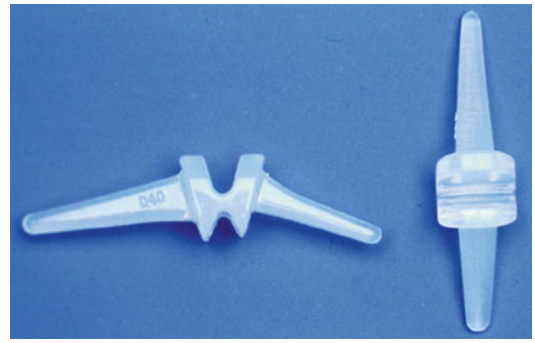

Fig. 15.2 A NeuFlex silicone implant with an anatomically neutral angulation

The classic silicone implant was created as a straight implant which provided stability, however, for the majority of patients, limited flexion. If patients were able to achieve full flexion, the stresses on the implant were high and the risk of fracture increased [3]. Newer generation implants have a lower risk of fracture than silicone, but require adequate bone stock and adjacent soft tissue support for stability [1, 2, 13, 14]. This makes primary osteoarthritis and post-traumatic arthritis amenable to the newer generation, however, its use is limited in the rheumatoid patient where bone loss and significant deformity are more common [14]. The biomechanical properties of the pyrolytic carbon implant makes it an appealing choice, as it has an elastic modulus similar to cortical bone and shows excellent bone-implant incorporation (Fig. 15.1) [15, 16].

There are multiple silicone MCP arthroplasty options currently available, including the Swanson (Wright Medical, Memphis, TN), Integra Lifesciences (Plainsboro, NJ), Small Bone Innovations (SBI, , Morrisville, PA) and Neuflex (DePuy Orthopaedics, Warsaw, IN) [4]. The Neuflex implant was the first having a 30° pre-bend, mimicking the anatomic neutral position of the MCP joint [3, 4]. Weiss et al. compared the mechanical properties of these three implants and found the Neuflex implant to most closely match the center of rotation, tendon excursion and moment arm, when compared to a native MCP joint (Fig. 15.2) [4]. The SBI and Integra implants mimicked the pre-bent hinge as well as the overall design features of the NeuFlex implant.

Indications

MCP arthroplasty should be considered in patients with advanced MCP arthritis, who have failed a 3–6 month course of conservative therapy. In the osteoarthritis (OA) group, this advanced OA generally consists of pain with loss of joint space, but reasonably preserved range of motion. In the rheumatoid patient population, the inflammatory arthritis manifests as painful deformity with destruction of the joint space and a fixed deformity [1, 2].

In the rheumatoid patient, problems at the MCP joint may only be “one of many” and, as such, the decision to undertake surgery is often by committee, including other health professionals. Other factors to be considered prior to surgery include nutritional status, risk of infection, use of DMARDs and the presence of cervical spine instability [1, 2]. The incidence of unstable cervical spine joints is nearly one-third in the rheumatoid patient, with atlantoaxial subluxation being the most common [2]. The cervical spine must be adequately assessed prior to considering endotracheal intubation [2].

Contraindications

Significant bone loss and severe deformity at the MCP joint, most frequently secondary to inflammatory arthritis, may be a contraindication to MCP arthroplasty, depending on sur-

geons comfort level. Flexor or extensor tendon rupture can result from rheumatoid arthritis and should be managed prior to MCP arthroplasty, to allow maximum range of motion results. Complicating factors of rheumatoid arthritis, such as cervical spine disease and infection risk, may pose challenges in the peri-operative management of these patients, but are generally not considered contraindications. Other relative contraindications include vasculitis and poor skin condition [1, 2].

Surgical Technique for Silicone Arthroplasty

The surgical incisions vary, depending on whether or not the patient has rheumatoid or osteoarthritis. In the osteoarthritic patient, a longitudinal 5-cm dorsal incision is made over the affected MCP joint. In the rheumatoid patient, most often requiring multiple arthroplasties simultaneously, a 5 cm incision is made between the index and middle finger MCP joints and another between the ring and small finger MCP joints (Fig. 15.3). Each of these incisions provides access to the adjacent MCP joints. Alternatively, a transverse incision across all four MCP joints can be made, to obtain exposure. Careful soft tissue dissection is performed through the subcutaneous tissue to the central extensor tendon. In primary osteoarthritis, without significant deformity, the extensor tendon is incised longitudinally, which exposes the underlying MCP joint (Figs. 15.4a–c). In rheumatoid patients with ulnar deviation at the MCP joint, the tight ulnar-sided extensor hood is incised longitudinally, which allows radial subluxation of the extensor tendon to expose the joint surface. A transverse cut of the metacarpal head is performed at the distal-most aspect of the metacarpal flare and the metacarpal head is excised (Figs. 15.5a, b). The level of this cut is very important, in order to allow appropriate seating of the implant without impingement. Using a rongeur, osteophytes are removed from the base of the proximal phalanx, as a proximal phalanx resection is rarely necessary. If the proximal phalanx has been chronically subluxed, there can be some erosion of the base. In cases of significant erosion, resection of the proximal phalanx base can be performed, to provide a perpendicular base for implant insertion. Evaluation of flexion and extension gaps after bony resection is critical to identify and release any soft tissue constraints. A specially designed awl is placed in the intramedullary canal of the proximal phalanx and the metacarpal (Fig. 15.6). Sequential broaching is performed for appropriate implant sizing (Fig. 15.7). The goal of broaching is to insert the largest broach possible, that allows complete seating of the flanged stop. A trial implant is then inserted, corresponding to the size of the largest broach used. The MCP joint is then taken through a range of motion to assess stability and fit. If full extension or full flexion is unable to be obtained, then either soft-tissue release or further bony resection should be performed, so that full range of motion can be obtained. If significant volar soft-tissue tightness is present, release of the volar plate and intrinsic tendons can be performed to assist in balancing flexion and extension. The definitive implant is then placed (Fig. 15.8). For the index finger, radial-sided drills holes are

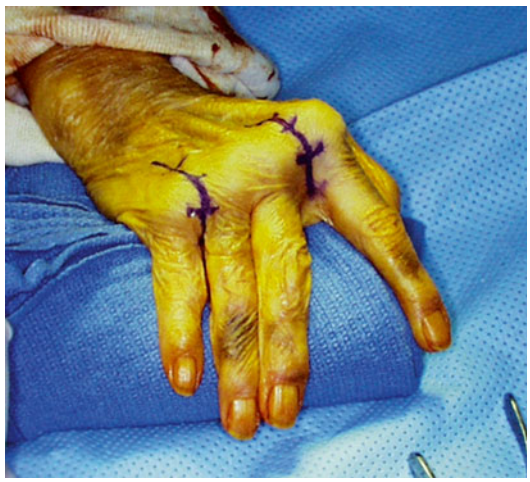

Fig. 15.3 Two separate longitudinal incisions made in a rheumatoid patient undergoing MCP arthroplasty of all four joints

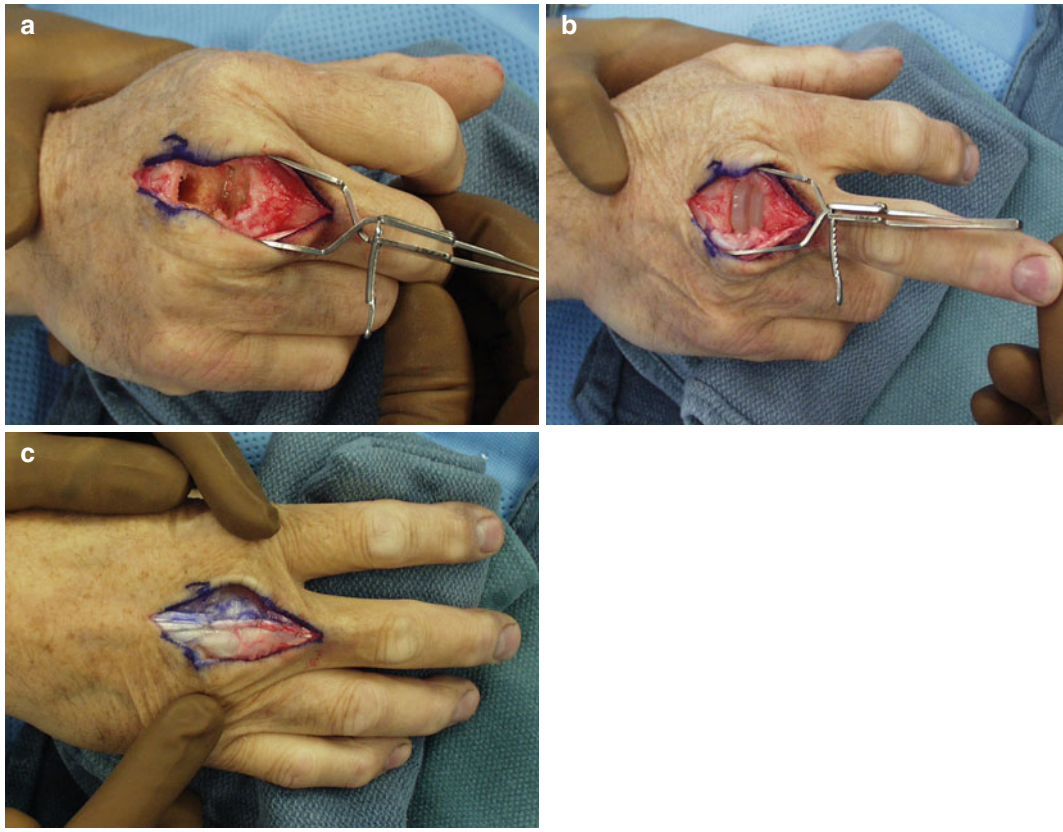

Fig. 15.4 An osteoarthritic patient following longitudinal incision through the extensor tendon and subsequent resection of the metacarpal head and broaching (**a**). A NeuFlex implant in place (**b**). Closure of the extensor tendon (**c**)

created in the distal metacarpal shaft to allow reconstruction of the collateral ligament. This is not generally required for the remaining digits. The wound is copiously irrigated and the definitive MCP implant is placed. The joint is again assessed for stability and range of motion. In rheumatoid patients, the extensor tendon is relocated to midline and a radial, sagittal extensor hood imbrication is performed, to maintain appropriate tendon position. In primary osteoarthritis patients, the split extensor tendon requires repair. Hemostasis is obtained and the skin is closed. The fingers should rest in a relatively straight position, with significant radial or ulnar deviation. A light, bulky dressing with a volar splint is applied, maintaining the MCP joint at 30° of flexion. Rehabilitation is started at 2 weeks, with active and active-assisted range of motion and passive motion is started at 4 weeks (Figs. 15.9a, b).

Summary Technical Tips

Release the tight ulnar sagittal hood, exposing the MCP joint

Excise the MCP head at the distal metacarpal flare

Ronguer osteophytes and the proximal phalanx base

Broach to as large implant size as possible

Trial implant followed by definitive implant; check motion

Imbricate (reef) radial sagittal hood to centralize extensor

Tricks for Problems

Tight volar structures = release entire volar plate

Index finger too unstable = reconstruct RCL through drill holes
Tight with implant in = resect more metacarpal first
Significant dorsal P1 erosion = use saw to resect P1 base

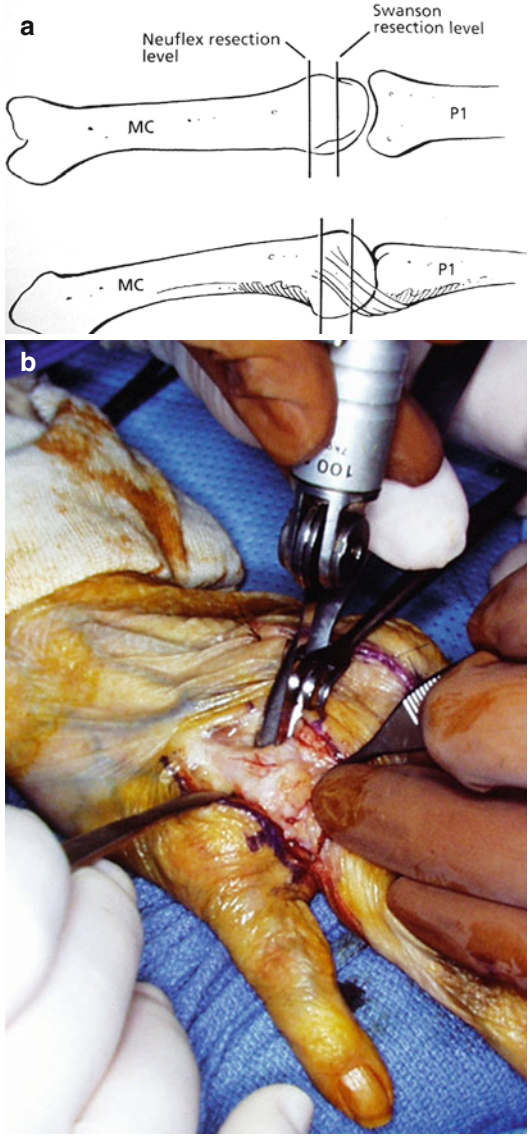

Fig. 15.5 The resection level for silicone based implants (**a**). A sagittal saw is used to resect the metacarpal head at the distal flare of the metacarpal (**b**)

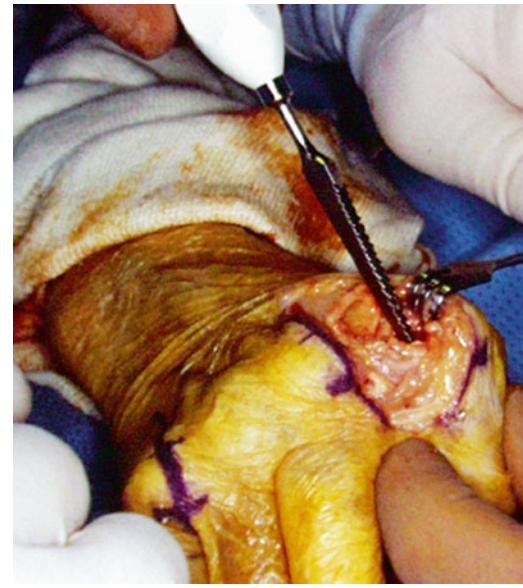

Fig. 15.6 An awl is used to prepare the canals

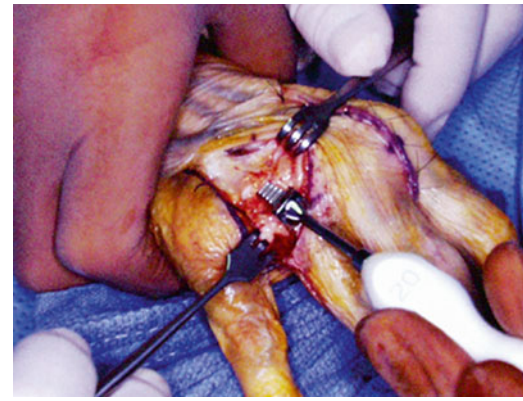

Fig. 15.7 Broaching is undertaken to the maximum size possible

Results

Surgical intervention of MCP arthritis has shown to be beneficial for patients with rheumatoid arthritis, in comparison with non-operative management. Chung et al. performed a prospective study, comparing MCP arthroplasty to non-operative care. At 1-year follow-up, the surgical group had significant improvements in function, aesthetics, ADLs and satisfaction, whilst the conservative management group had no improvement or deterioration in function [17].

The results for MCP arthroplasty have consistently shown good early function, with gradual and progressive loss of motion and recurrence of deformity at long-term follow-up. Chung et al. reported on 1-year follow-up of silicone MCP arthroplasty and noted a significantly improved range of motion, ulnar drift and subjective outcomes [18]. Weiss and Strickland reported on 50 consecutive patients (92 % rheumatoid) who underwent Neuflex MCP arthroplasty at 14 months and found improved range of motion and deformity, with excellent patient satisfaction [3, 19]. In a study of MCP arthroplasty using the Neuflex prosthesis in patients with primary osteoarthritis, at 4-year follow-up, patients had a high rate of pain relief and satisfaction [20]. In a similar study, Rettig et al. reported good pain relief and function at 40-month follow-up for silastic MCP arthroplasty in an idiopathic osteoarthritis patient population [21].

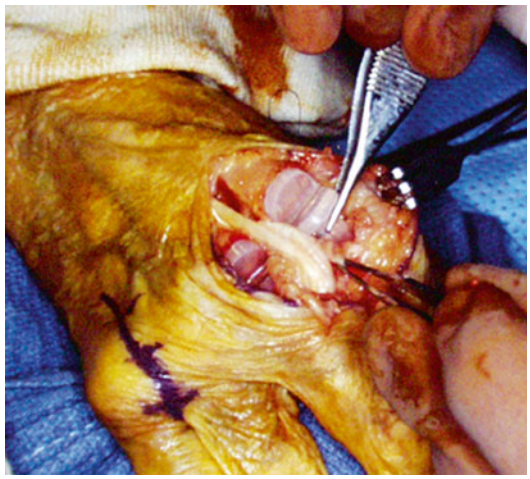

Fig. 15.8 Definitive implants are placed following successful trial implant check

In looking at long-term follow-up, Goldfarb et al. reported on 208 rheumatoid patients, who underwent four-digit MCP arthroplasty, at an average of 14-year follow-up. These patients had immediate improvement in range of motion and ulnar drift post-operatively, which recurred to some degree at long-term follow-up, with 38 % expressing satisfaction with hand function and only 27 % described pain-free function. 63 % of these silicone implants were fractured at the time of evaluation [22]. A similar study by Trail et al. reviewed the records of 381 patients with

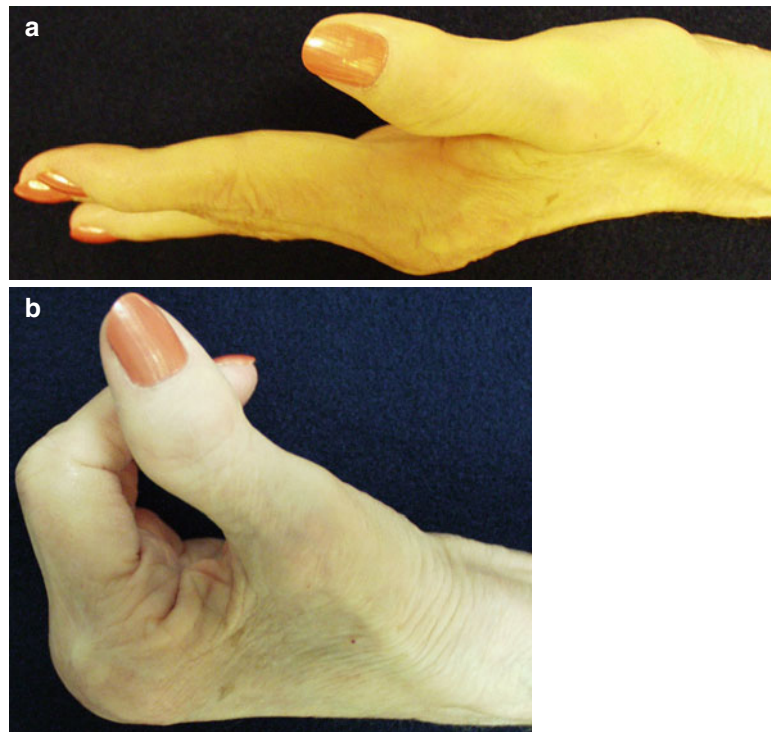

Fig. 15.9 A 3 month follow-up range of motion in a typical MCP arthroplasty demonstrating extension (**a**) and flexion (**b**)

rheumatoid arthritis, who had undergone silastic MCP arthroplasty for a total of 1,336 implants. With revision surgery as the end-point, they found an 83 % survivorship at 10 years and a 63 % survivorship at 17 years [23]. However, at the 17-year mark, approximately two-thirds of the remaining implants were fractured in an asymptomatic patient. Only 2.9 % of the revision surgeries were performed for fracture. This group identified soft-tissue balancing, crossed intrinsic transfers and management of wrist deformity prior to MCP arthroplasty as the keys to preventing revision surgery [23]. The value of crossed intrinsic transfer, to prevent recurrent ulnar subluxation after MCP arthroplasty, has been further evaluated in the literature. Clark et al. found that the addition of a crossed intrinsic transfer to the Swanson arthroplasty, decreased the recurrence of ulnar drift at 50-month follow-up (6° vs 14°, $p=0.01$) [24].

In comparing the various silastic implants, Escott et al. compared the Swanson implant with the Neuflex and found that at 12 months both groups showed improvement in range of motion and ulnar drift. The Neuflex group achieved a greater post-operative arc of motion, while the Swanson implant group self-reported better function and aesthetics [25]. As previously mentioned, in a biomechanical study, Weiss et al. identified that the Neuflex implant most closely mimicked the motion of the native MCP joint [4]. Hussein et al. performed biomechanical testing, comparing the Swanson and Neuflex implants. This group found less implant toggling within the intramedullary canal and lower stress at the implant hinge in the Neuflex implant. The Swanson implant did require less force to achieve full extension than the Neuflex [26]. Another comparative study, looking at the Swanson and Avanta/Sutter silastic implants at 5-year follow-up, found the Avanta to have greater range of motion, but with a higher fracture rate [27].

With the vast majority of rheumatoid disease at the MCP joint involving all digits, Chung et al. compared the outcome of each digit 1 year after silastic arthroplasty. The authors identified that the ulnar digits (ring and small) had better MCP arc of motion, less extension lag and similar ulnar drift to the radial digits (index and middle) [28].

Results of the new generation MCP implants are encouraging, primarily in primary and post-traumatic osteoarthritis. Parker et al. reported on 142 consecutive MCP arthroplasties, noting significant improvement in pain, range of motion and function at an average of 14-month follow-up for patients with osteoarthritis. Patients with rheumatoid arthritis showed improvement, but to a lesser degree and the authors noted that the pyrolytic carbon arthroplasty is a good option for inflammatory arthritis with good bone stock and preserved soft tissues [14]. Cook et al. reported on 151 pyrolytic carbon MCP arthroplasties in 53 patients and found that 18 implants required revision for subluxation, dislocation, or soft tissue imbalance, with 71 implants (26 patients) available for long-term follow-up (11.7 years). In this long-term follow-up group there was a 13° improvement in range of motion and no evidence of bony resorption [15].

Complications

The most frequently seen complication of the silicone MCP arthoplasty is fracture. Bass et al. reported a 20 % fracture rate, at an average of 27-month follow-up with the Sutter silicone MCP arthroplasty [29]. Interestingly, in this study there was no correlation between implant fracture and patient satisfaction. Kirschembaum et al. reported a 10 % fracture rate for the Swanson implant at 8.5 year follow-up, while Blair et al. reported a 21 % fracture rate at 4.5 year follow-up of the same implant [30, 31]. Despite the high rate of fracture noted in multiple studies, generally less than 5 % require revision surgery [12, 13, 23, 32]. Other, less frequently occurring complications, include silicone synovitis (<1 %), loosening (<1 %) and infection (<1 %) [2].

The complications of pyrolytic carbon MCP arthroplasty include; subluxation and dislocation, infection and periprosthetic erosion. The risk of fracture is much lower than in its silicone counterpart. Parker et al. reported a “minor”

complication rate of 6 % and a "major" complication rate of 9 % with this procedure [14]. In a study in primates by Cook et al., pyrolytic carbon arthroplasty showed no evidence of wear debris, or inflammatory joint reaction [15, 16].

Revision surgery following MCP arthroplasty most frequently occurs secondary to recurrent ulnar subluxation, pain and decreasing functional capacity [32]. Revision surgery for fractured implants occurs infrequently, because patients are rarely symptomatic. This is thought to be due to the fact that the implant functions as a spacer, rather than an articulating prosthesis [2]. Burgess et al. reported good pain relief, but generally poor objective outcomes in 62 revised MCP silastic implants at 5-year follow-up. Pre-operative examination compared to follow-up examination found no significant improvement in flexion, with a slight improvement in ulnar drift. There was a 34 % fracture rate in the revision implants. However, patients were generally pleased with their outcome. The authors concluded that the difficulty in revision surgery is management of the incompetent soft tissues [32]. Revision surgery, following unconstrained pyrolytic carbon MCP arthroplasty, occurs secondary to dislocation, subluxation, soft-tissue imbalance and implant loosening. Cook et al. reported a 12 % revision rate at long-term follow-up [15]. Fracture of these implants is rare, but has been reported [15, 33]. With the majority of the revisions occurring for instability, most are converted to a constrained silastic implant.

Conclusion

Metacarpophalangeal arthritis is a common hand problem, most frequently seen in patients suffering from inflammatory arthritis. This disease process can result in significant pain, loss of function and deformity. Non-operative management, including anti-inflammatory medications, splinting and corticosteroid injections, can be effective early in the arthritis process. The use of disease-modifying anti-rheumatic drugs has provided great benefit for patients with rheumatoid arthritis and MCP arthritis. Surgical intervention is considered in patients whom have failed conservative measures and consists of arthrodesis in the thumb and arthoplasty for the remaining four digits. Arthroplasty preserves motion of the MCP joints, important for functional hand motion in the index, middle, ring and small fingers. A variety of silicone and pyrolytic carbon implants are available for arthroplasty, with silicone being the "gold standard" in treatment of MCP arthritic disease. Arthroplasty has been shown to improve motion, pain and deformity, with reasonable survivorship in both primary osteoarthritis and inflammatory arthritis.

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Total and Partial Wrist Implant Arthroplasty

16

Brian D. Adams

Keywords
Wrist arthroplasty • Total wrist arthroplasty • Wrist replacement

Introduction

The wrist is a common site for arthritis, with 80 % of patients with rheumatoid arthritis (RA) complaining of functional impairment due to wrist pain. Osteoarthritis and post-traumatic arthritis are also common causes for symptomatic wrist pain, particularly in patients with severe scapholunate advanced collapse (SLAC), scaphoid non-union advanced collapse (SNAC) and secondary radiocarpal arthritis after severe distal radius fracture. Total wrist arthroplasty is attractive to clinicians allowing patients the same benefits provided by arthroplasty of the hip, knee and shoulder joints. A motion-preserving alternative to wrist arthrodesis is of particular importance when treating patients who are debilitated by arthritis affecting multiple joints. Basic activities of daily living, such as perineal care, fastening buttons, combing hair and writing are made easier if some wrist motion is preserved [1, 2], and thus, patients with rheumatoid arthritis typically prefer arthroplasty over arthrodesis to enhance the performance of daily activities [3–5]. Patients with post-traumatic or degenerative osteoarthritis may also select arthroplasty over arthrodesis to better perform specific vocational and other activities.

A recent cost-utility analysis compared non-surgical management, total wrist arthroplasty and wrist arthrodesis [6]. Hand surgeons, rheumatologists and patients with rheumatoid arthritis were given a time trade off/utility survey to assess preferences. Results showed that surgical management was preferred to non-surgical management and was deemed cost effective by quality adjusted life years (QALY). Arthroplasty had a slightly higher cost compared to fusion, but not so high as to make it cost prohibitive. Another study by the same group did a utility analysis of rheumatologists and hand surgeons on the treatment of wrist arthritis in rheumatoid patients [7]. The results showed that surgeons and rheumatologists agreed that arthroplasty was the better procedure in expected gain of QALY, compared to fusion. Murphy et al. compared 24 patients with a wrist arthrodesis and 27 with total wrist

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arthroplasty and found no difference in overall outcome scores; arthroplasty patients, however, tended to have greater ease with personal hygiene and selected other tasks and had slightly higher satisfaction overall [8].

Despite these potential advantages, wrist arthroplasty has not become a widely accepted option by most surgeons, primarily because arthrodesis is simpler to perform and it is more predictable, with fewer long term complications. The natural wrist is a highly complex joint, with multiple articulations which present substantial design challenges when developing implants. Although current arthroplasty designs show improvements over older models, they do not totally duplicate these interactions. This results in a variety of compromises in motion, balance and durability, compared to the natural wrist joint. Thus, regardless of the need or desire for arthroplasty, the patient must accept and commit to a lifetime of restricted activities imposed by an artificial wrist. The patient must also recognise the risk of implant failure and the possible need for revision surgery.

Brief History

In 1967, Alfred Swanson developed a silicone implant, which became the first widely used wrist arthroplasty [9]. Wrist motion resulted from a combination of implant flexibility and pistoning within the medullary canals of the radius and third metacarpal. These implants often provided initial pain relief and some motion, but restoration of wrist height and hand balance was unpredictable and follow-up revealed subsidence within the bone and a high incidence of implant breakage, reaching 52 % at 72 months [10–12]. Silicone synovitis also became an important issue later, although its incidence was lower than with individual carpal bone implants [13].

The next generation of implants were articulated and utilised cobalt chrome, or titanium and high-density polyethylene. Early articulated designs incorporated bearings with small surface areas to maximise joint motion, however imbalance and instability were common. A variety of stem designs were developed for cement fixation in the radius and carpus, with carpal components typically fixed in the metacarpal canals. Unfortunately, a high incidence of carpal component loosening occurred, marked by metacarpal erosion and implant penetration. Periprosthetic bone resorption of the distal radius was also common. An early example was the very popular ball and socket implant, introduced by Meuli in 1972 [14]. During the same period, Volz developed an articulated non-hinged prosthesis with the convex surface on the carpus and the concave surface on the radius. These implants also produced satisfactory early clinical results, but longer term follow-up revealed continued problems with imbalance, subsidence and loosening [15, 16]. The Biaxial wrist prosthesis (DePuy, Warsaw, IN) had similar fixation, but introduced an ellipsoid-shaped articulation that demonstrated improved wrist balance and the potential for uncemented fixation of the radial component. This implant became very popular and early results were good in most patients, although loosening remained a substantial problem. Eight of 11 failures in one series of 58 Biaxial implants (DePuy, Warsaw, USA) were secondary to distal component loosening and subsidence, which often resulted in penetration through the dorsum of the third metacarpal and substantial bone destruction [17]. The implant has now been withdrawn from sale by the manufacturer.

Another attempt to preserve anatomy was the Anatomic Physiologic (APH) implant (Implant-Service Vertreibs-GmbH, Hamburg, Germany), designed with a titanium articulation [18]. Midterm follow-up unfortunately revealed a very high failure rate, with 39 of 40 undergoing revision at an average 52-month follow-up. Isolated loosening of the carpal component was the most common mode of failure. Titanium wear debris was found in the soft tissues of all revisions and was believed to be a major contributing factor to early periprosthetic bone resorption [19].

The Universal wrist (Kineticos Medical, Incorporated, Carlsbad, CA), designed by Menon, introduced a number of new concepts in total wrist implant design to the USA and is considered the predecessor to current resurfacing-type designs [20]. Although refinements have

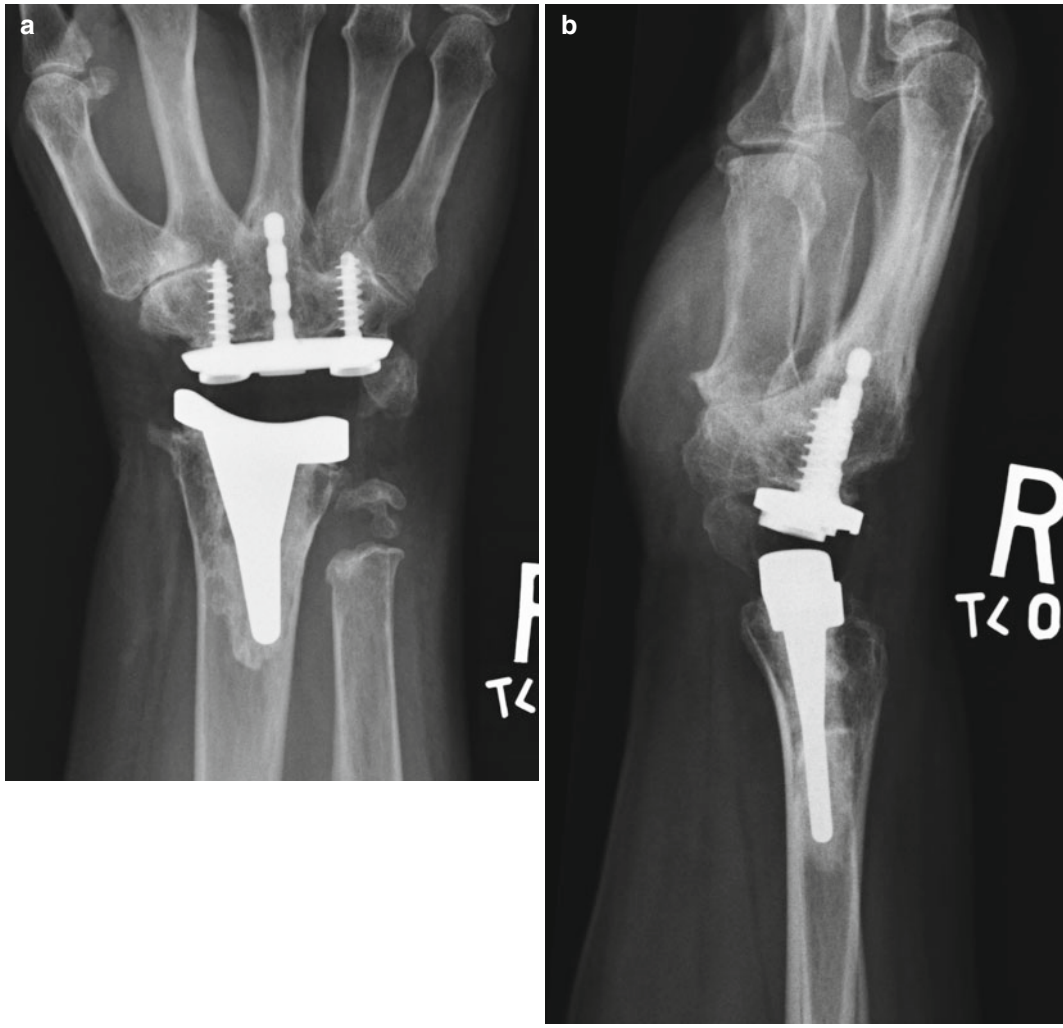

Fig. 16.1 (**a**, **b**) KMI total wrist in 62 year old female with inflammatory arthritis at 10 year follow up. No evidence of loosening or osteolysis

been made to his design, many of the basic concepts remain, particularly relating to the distal component (Fig. 16.1).

Current Designs

The overall experience with different designs over the last three and a half decades provides evidence that specific criteria must be met to optimise the clinical results. The present generation of prostheses combines many of the basic concepts of the Universal and Biaxial implants to improve both wrist balance and minimise distal component loosening. Although the natural wrist kinematics are much simplified, the designs attempt to replicate the natural centre of motion, resulting in a functional range of motion and better balance. The articulation is broad, generally ellipsoidal in shape and semiconstrained to provide a functional range of motion and yet resist imbalance and instability, so as to allow rapid recovery with minimal formal rehabilitation or splint protection [21].

Distal component fixation is primarily within the carpus and not the metacarpal canals and screws are used to augment fixation. A key to durability of distal component fixation is an

intercarpal fusion, to create a two-bone wrist joint,, thus eliminating the multiple articulations of the natural wrist. The radial component is shaped to minimise bone resection and preserve the joint capsule, to enhance prosthetic stability and wrist balance.

The distal implant component, which must be relatively thin and appropriately shaped, is made of titanium for better strength and osteointegration. The proximal component is made of cobalt chrome, with a polished articular surface, to provide a low friction interface for reduced wear. The articular bearing is made of high density polyethylene for improved wear characteristics. The articulation is unconstrained, but the concave surface of the radial component provides a stable joint.

Fixation by osteointegration rather than cement is an option for both components, in an attempt improve durability and reduce bone destruction if revision becomes necessary. All systems have the option to preserve the ulnar head and distal radioulnar joint (DRUJ).

There are currently three implant systems available in the USA that have used these criteria, but each has unique features. The Remotion total wrist (SBI, New York, NY) offers a mobile bearing attached to the carpal component that theoretically improves motion and load transfer, thus reducing stress contributing to loosening [22]. The Universal 2 system (Integra Life Sciences, Plainsboro, NJ) is a versatile system that replicates the natural wrist centre of motion and can be used for a wide range of indications whilst providing excellent joint stability. The Maestro (Biomet, Warsaw, IN) allows complete resection of the proximal carpal row and has a polyethylene surface proximally. It is also approved for hemiarthroplasty, using the distal component alone.

Patient Selection Criteria

As with implant arthroplasty of other joints, such as the hip and knee, patient selection is the single most important controllable factor affecting the outcome of total wrist arthroplasty. A variety of patient factors must be included in the decision process, although the most important are age, activity demands, bone and soft tissue quality, joint deformity, disease activity and reliance on the limb for overall body mobility, such as the use of walking aids. In addition, the relative benefits, compared to other procedures, should be considered.

The ideal candidate for a total wrist arthroplasty is a patient with a low-demand lifestyle who requests relief of pain and is seeking modest wrist motion to maintain function for non-stressful activities. Elderly patients with generalised rheumatoid arthritis have been considered the ideal patients for total wrist arthroplasty, particularly those with bilateral wrist arthritis, who would otherwise require bilateral wrist fusions. Also, rheumatoid patients with involvement of the hand, elbow and shoulder often request maintenance of motion in the wrist to better preserve limb function and to avoid a feeling of total limb stiffness. However, rheumatoid patients with highly active synovitis, producing bony erosions or joint hyperlaxity, have a substantially higher risk of implant instability and loosening and, thus, are better treated by arthrodesis. Regular use of the upper extremities for support during ambulation or transfers is a further contraindication; intermittent use of crutches or a cane, however, is acceptable if the patient uses a wrist splint.

Patients with post-traumatic or degenerative osteoarthritis may also be candidates for wrist arthroplasty. Because these patients typically have good bone quality, muscle strength and wrist alignment, the early and midterm results can be excellent. However, these patients should only undergo arthroplasty to maintain dexterity for activities of daily living and specific low-demand activities, rather than to increase activity levels and perform stressful tasks with less pain. Osteoarthritis patients who have highly demanding lifestyles are not good candidates. Many patients with post-traumatic arthritis are young and very active and thus are not candidates for arthroplasty, because of the high stresses they will likely impose on the wrist.

Absolute contraindications for total wrist arthroplasty include a minimally functional hand,

recent infection and lack of wrist extension due to ruptures of the extensor carpi radialis brevis and longus tendons or radial nerve palsy.

Clinical Pearl

Patient selection is the key to a successful, durable outcome for total wrist arthroplasty. A thorough discussion regarding the risks and benefits and alternative procedures is required so that the patient can make a proper informed decision.

Patients should undergo a routine preoperative evaluation. In patients with rheumatoid arthritis, the cervical spine is evaluated for instability with flexion and extension views to ensure their safety under anesthesia. Physical examination of the wrist confirms active wrist extension and functional digits. Failure to recognise preoperative soft-tissue contractures may lead to persistent wrist deformity, reduced wrist motion and imbalance of the extrinsic finger tendons. Posterior-anterior (PA) and lateral views of the wrist are reviewed to assess bone stock, joint deformity and probable implant size. There must be adequate bone stock and quality to support the implant, especially the distal component. Implantation in patients with severe osteopenia, bone erosion, or joint deformity is more challenging and the implant fixation is likely to be less durable. Previous surgical fusion or proximal row carpectomy are relative contraindications; these patients must have adequate carpus remaining and intact wrist extensors.

Lower limb surgery, such as total hip or knee arthroplasty, should be done prior to wrist replacement to avoid weight bearing on the wrist replacement during rehabilitation of the lower limb. Procedures on the digits should be completed after wrist arthroplasty to optimise joint alignment and tendon balance in the hand.

If bone loss or active erosive disease of the carpus is suspected, but not confirmed by preoperative imaging, arthrodesis should be discussed with the patient pre-operatively, as this may be the only alternative from the intraoperative assessment. Partial or complete resection of the distal ulna is undertaken when there is symptomatic arthritis of the distal radioulnar joint. Otherwise the head of the ulna can be left in place. Implant size is estimated using radiographic templates, but final size is often determined during surgery.

Surgical Technique

Step 1: Setup and Incision

Although the technique for Universal 2 (IntegraLifeSciences, Plainsboro, NJ) is described, the basic principles apply to all other wrist arthroplasty systems. Prophylactic antibiotics are administered. The operation is performed under general or axillary regional anesthesia, using a nonsterile arm tourniquet. A strip of transparent adhesive film is applied to the dorsum of the hand and wrist to protect the skin from damage during instrumentation.

A dorsal longitudinal incision is made over the wrist, in line with the third metacarpal. The skin and subcutaneous tissue are elevated from the extensor retinaculum and held with retraction sutures. Care is taken to protect branches of the superficial radial nerve and dorsal cutaneous branches of the ulnar nerve. If the distal ulna is to be resected, the extensor carpi ulnaris compartment is opened along its volar margin. Alternatively, if the distal radioulnar joint is to be preserved, the extensor digitorum quinti is opened dorsally. The extensor retinaculum is elevated radially to the septum between the 1st and 2nd extensor compartments. An extensor tenosynovectomy is performed if needed and the tendons are then retracted.

Step 2: Joint Exposure

A distally based rectangular flap of joint capsule is raised over the dorsal wrist. The sides of the flap are made along the far medial and lateral aspects of the wrist joint. If the ulnar head is to be resected, the proximal edge of the capsule is raised in continuity with the dorsal capsule of the distal radioulnar joint (DRUJ) and the periosteum

over the distal 1 cm of the distal radius, to create a long and broad flap for closure over the prosthesis. If the distal ulna is to be preserved, then the interval between the capsule and the dorsal distal radioulnar ligament is carefully divided and the capsule is raised distally to preserve the horizontal components of the triangular fibrocartilage complex. The brachioradialis and the tendons of the first dorsal compartment are elevated subperiosteally from the distal part of the radial styloid. The wrist is then fully flexed to expose the joint. Synovectomies of the radiocarpal and distal radioulnar joints are performed when needed.

Clinical Pearl

Full exposure of the joint is required for proper implantation of the components. Newer generation systems provide instrumentation to help provide more accurate and reproducible surgical technique.

Step 3: Preparation of the Radius

Using a bone awl or drill, a hole is made through the articular surface of the radius, about 5 mm below its dorsal rim and under Lister's tubercle. The hole is enlarged and the radial alignment guide advanced into the medullary canal. Fluoroscopy is used to confirm that the guide rod is centred within the canal. The cutting guides are applied and aligned to resect a minimum amount of distal radius, so as to create a flat distal surface. The radius cut is completed, but the sigmoid notch is preserved by stopping the cut a few millimeters short of the distal radioulnar joint. The radial component is designed with an ulnar sided flare to avoid the distal radioulnar joint. The alignment rod is reinserted and the appropriate size broach head is used. Its orientation is important to reproduce the natural, slightly supinated position of the carpus relative to the radius. A trial radial component is inserted. The carpus can be reduced on the radial component to assess soft tissue tension. If it is excessive, then further resection of the radius may be necessary, although this is usually delayed until the carpal preparation is completed.

Step 4: Preparation of the Carpus

If the scaphoid and triquetrum are mobile, carpus preparation is facilitated by first temporarily pinning these bones to the capitate and hamate. The lunate is excised by sharp dissection. Using the drill guide, a guide wire is inserted in the capitate along the long axis of the 3rd metacarpal. Fluoroscopy is used to confirm the correct position of the wire. A cannulated drill is used to create a hole. The cutting guides are applied and aligned to make the cut through the proximal 1 mm of the hamate, a small amount of the capitate head and about half of the scaphoid and triquetrum. The trial component is inserted. Screw holes for the carpal component can be made at this time, or at the time of final implant insertion, depending on surgeon preference. The holes are typically not perpendicular to the carpal component, with the radial screw extending into the base of the 2nd metarcarpal and the ulnar screw passing into the body of the hamate, but not into the 4th metacarpal.

Step 5: Trial reduction

A trial polyethylene is applied to the carpal plate and range of motion and stability are checked. The prosthesis should demonstrate approximately 35° of flexion and extension, with modest tightness at full extension. If the volar capsule is limiting extension, then the radius may be shortened slightly, but no more than 2 mm at a time. When a pre-operative flexion contracture is present, a step-cut tendon lengthening of the wrist flexors may be necessary. Conversely, when tension is insufficient, the palmar joint capsule is inspected and repaired, if detached. If the capsule is intact but instability persists, then a thicker polyethylene component may be required.

Step 6: Implantation

Prior to implantation, three horizontal mattress sutures of 2-0 polyester are placed through small bone holes along the dorsal rim of the distal radius, for later capsule closure. If the ulnar head was resected, then sutures are also placed

through the dorsal part of the ulna neck. The dorsal portions of the articular surfaces are removed from the triquetrum, capitate, hamate, scaphoid and trapezoid and previously resected bone is packed into the spaces to achieve an intercarpal arthrodesis. The final implants are impacted into place and the final screws are inserted. The appropriate polyethylene component is applied to the carpal plate and a final assessment of motion and stability is made.

Step 7: Closure

If the distal ulna was resected, the palmar capsule on the ulnar aspect of the wrist is brought dorsally and sutured to the end of the ulna, through drill holes. The dorsal wrist capsule is reattached to the distal margin of the radius and ulna, using the previously placed sutures. If the capsule is deficient, the extensor retinaculum is divided in line with its fibers and one half is placed under the tendons to cover the prosthesis. Meticulous closure of the capsule is mandatory to ensure prosthetic stability and to protect the tendons from irritation. The remaining retinaculum is repaired, leaving the extensor carpi radialis longus and brevis and the extensor pollicis longus superficial to the retinaculum. The skin is closed over a self-suction drain and the wrist is immobilised in a bulky gauze dressing and a plaster splint. Because the procedure may be associated with substantial postoperative swelling, strict elevation of the hand and immediate active finger motion are very important.

Post-operative Management and Rehabilitation

The dressing and plaster splint are removed between 2 and 10 days post-operatively and a removable well-moulded wrist splint is applied. A supervised exercise program is initiated, consisting of full digital motion and gentle active wrist and forearm motion, that emphasises wrist extension, but avoids forceful wrist flexion until 4 weeks post-operative to protect the capsule repair. The patient is gradually weaned from the splint after 4 weeks and strengthening is added. Full activities are permitted after 8 weeks, depending on discomfort and swelling. The patient is advised to avoid impact loading of the wrist (e.g. use of a hammer, playing tennis) and repetitive forceful use of the hand. Permanent restrictions including lifting more than 10 lb, except on an occasional basis and 2 lb on a regular basis.

Outcomes

There have been relatively few published reports on the outcomes of presently available prostheses, although several studies have been published on the outcomes of the Universal wrist designed by Menon. In Menon's first report of 37 Universal prostheses, with a mean follow-up of 6.7 years (range 4–10 years), none of the cases demonstrated radiographic evidence of distal component loosening [20]. A further follow-up study, that included 57 implants, again demonstrated no evidence of carpal component loosening [23], Subsidence of the radial component was observed, but was not progressive or symptomatic. Consistently good pain relief (90 %) and a functional range of motion was achieved, with an average postoperative motion of 36° extension, 41° flexion, 7° radial deviation and 13° ulnar deviation. Dislocation was the most common complication, with 5 occurring in the first 37 cases and a total of 6 among the 57 cases in the later follow-up. The initial higher incidence of dislocation was partly attributed to the lack of availability of different implant sizes and thicknesses of polyethylene inserts at that time.

A prospective study of 22 Universal prosthesis implanted by two surgeons, with a 1–2 year follow up, demonstrated results similar to Menon's. Patients achieved an average of 41° flexion and 35° extension [24]. Disabilities of the Arm, Shoulder and Hand (DASH) outcome survey scores improved 24 points at 2 years. Three prostheses (14 %) were unstable and required further treatment; all three were in patients with active rheumatoid disease with severe wrist laxity [22].

Ward et al. reported a prospective consecutive series of 25 wrist arthroplasties in 21 patients with rheumatoid arthritis [25]. Nineteen wrists in

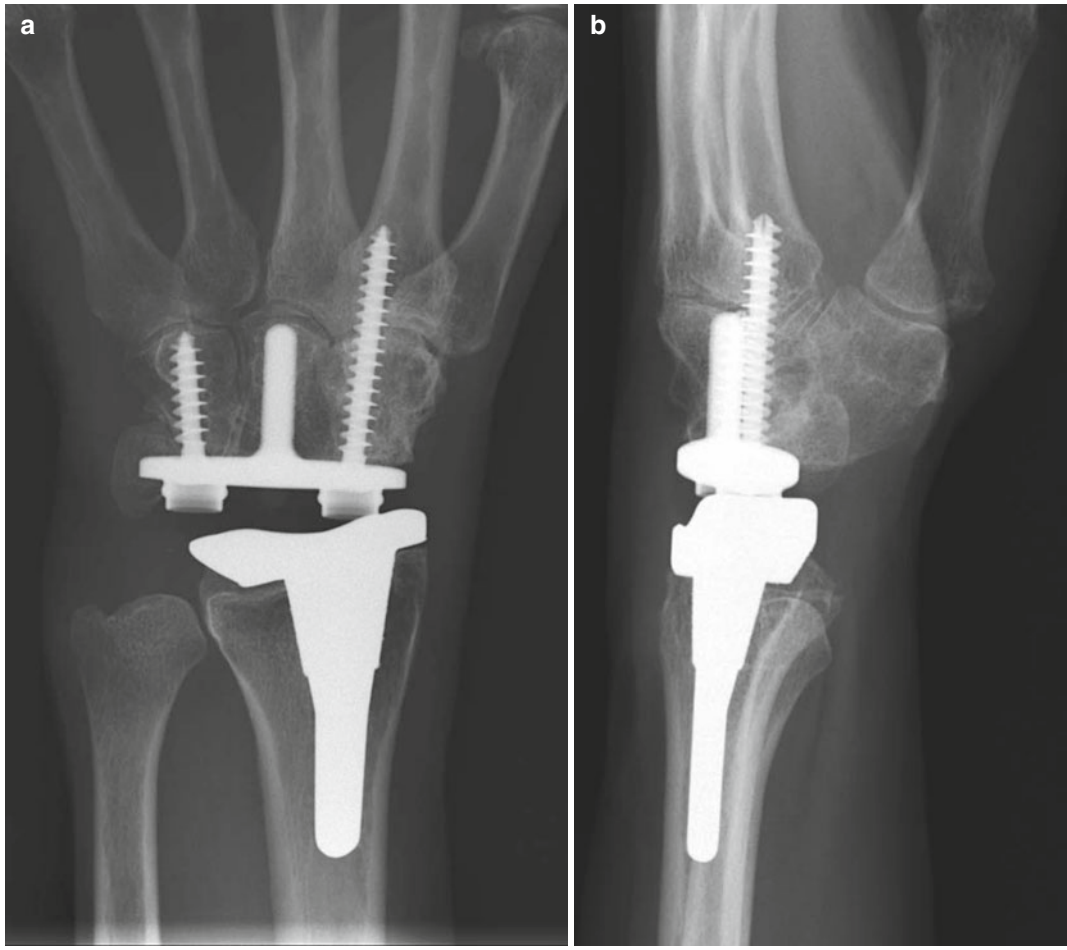

Fig. 16.2 (**a**, **b**) Universal 2 total wrist in a 59 year old female with rheumatoid arthritis at 2 year follow up. No evidence of loosening or osteolysis

15 patients returned for clinical and radiographic examination at a mean of follow-up of 7.6 years (range 5.0–10.8 years). Mean wrist flexion and extension at final follow-up were 42 and 20° respectively, for a mean improvement in the total flexion-extension arc of 14°. Average Disabilities of the Arm, Shoulder and Hand (DASH) score improved from 62 points pre-operatively to 40 points at 5 years after surgery. A total of 9 wrists in 8 patients had undergone revision surgery for a loose carpal component. Two additional wrists in two patients had radiographic evidence of carpal component subsidence at final follow-up. The authors concluded that the Universal wrist prosthesis provided a functional range of motion and good patient-reported outcome measures in patients with a functioning implant. Carpal component loosening led to a high incidence of failure, resulting in revision surgery in 47 % of the patients by 10 years. However, most failures occurred in patients with severe and persistently active rheumatoid disease, who would by today's criteria no longer be considered candidates for wrist arthroplasty, while those with less severe and better controlled disease had durable outcomes.

Our early results were reviewed for the Universal 2 prosthesis (Fig. 16.2) in 25 wrists: 20 patients had rheumatoid arthritis, 2 had post-traumatic arthritis and 3 had osteoarthritis [26]. Twenty were women and 5 were men. All prostheses were implanted uncemented. Results

showed functional motion, with an average of 37° flexion, 33° extension, 22° ulnar deviation and 9° radial deviation. Motion often did not maximise for 6 months. Pain relief was rated good by all patients, but mild ulnar sided wrist discomfort persisted in five. Average DASH score improved 20 % and Patient Rated Wrist Evaluation (PRWE) score improved 35 %. No cases showed radiographic implant loosening, but three osteopenic patients had 2–5 mm of subsidence, which plateaued after 1 year. The carpal component stem fractured in three patients who had the first version of this implant. All were revised and obtained a functional result. The implant has subsequently been redesigned with a greater diameter (stronger) stem and full porous coating over the entire distal surface (stem and plate), to better share and transfer the load to the carpus. No fractures have been found with the new version of the carpal component.

Ferreres et al. published a review of 21 consecutive Universal 2 implants, with an average 5.5 year follow-up [27]. Most of the patients had inflammatory arthritis, but 4 patients had chondrocalcinosis, grade IV Kienböck's disease, or degenerative arthritis. Only one patient reported not being satisfied with the procedure and 11 reported being very satisfied. Average flexion and extension range of motion was reported as 42° and 26° respectively. No patient had a dislocation or revision surgery in the follow-up period, but 2 had radiographic osteolysis and 1 had subsidence of the distal component.

Herzberg reported the early results, using the Remotion prostheses [22]. The series included 20 wrists, of which 13 were in patients with rheumatoid arthritis. The patients were prospectively followed for a minimum of 12 months, with an average of 32 months. There was one case of carpal and one radial component loosening both in rheumatoid patients. Rheumatoid patients had an average 41 % improvement in clinical scores, with 7 excellent, 5 good and 1 poor result. The non-rheumatoid patients had an average 27 % improvement in clinical scores, with 2 excellent, 2 good, 2 fair and 1 poor result. Overall there was significant improvement in pain and function, but only modest improvement in strength. The authors concluded that the Remotion wrist showed better outcomes than older designs and similar to the Universal 2.

Clinical Pearl

Reports of newer generation implants show promising early and midterm outcomes in both rheumatoid and non-rheumatoid patients for function and pain. However, patients should recognise that avoidance of stressful activities is likely to reduce the risk of implant loosening.

Partial Wrist Arthroplasty (Hemiarthroplasty)

Although total wrist arthroplasty has been used in selected patients with advanced arthritis, high-demand patients are rarely considered primarily due to a high risk of implant loosening, particularly of the distal component. A distal radius hemiarthroplasty may obviate the need for the strict restrictions required of patients treated by total wrist arthroplasty. It may provide another motion-preserving surgical option for active patients with severe wrist arthritis, especially those with distal radius articular degeneration as a result of osteoarthritis, or those with post-traumatic arthritis.

Using this concept, the author has performed a combined proximal row carpectomy and hemiarthroplasty, using the radial component of the Universal 2 wrist [28]. The first patient was a 42 year old female, with rheumatoid arthritis and severe bilateral wrist arthritis with distal radius erosion and volar carpal subluxation, with satisfactory condition of the capitate articular surface (Fig. 16.3). She obtained good pain relief and an 80° flexion-extension arc at 2 year follow-up. The second patient was a male with a SLAC wrist, who at 1 year follow-up denied pain and had a 69° flexion-extension arc. The procedure has subsequently been done in 26 wrists in 24 patients, with up to a 3 year follow-up. Initial selection criteria for the first 5 patients included a capitate head, having only minimal

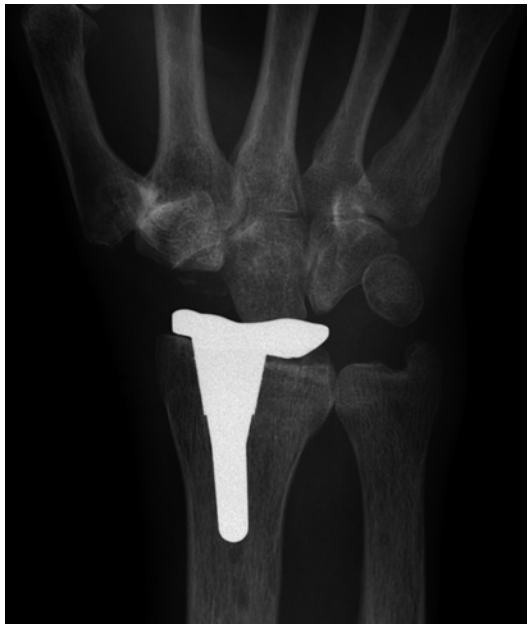

Fig. 16.3 Hemiarthroplasty using the radius component only of a Universal total wrist combined with a proximal row carpectomy in a 65 year old male at 3 year follow up

articular cartilage changes. Subsequently this was changed to a capitate without erosion or cystic changes. The diagnoses were rheumatoid arthritis in 2 patients (3 wrists), SLAC wrist in 18 (19 wrists) and post-traumatic in 4 patients (4 wrists). The procedure generated a minimum of 30° each of flexion and extension in 22 of the 26 wrists. The patients that failed to achieve this range of motion were 2 patients with rheumatoid arthritis, 1 with a SLAC wrist and 1 with post-traumatic arthritis. The patient with rheumatoid arthritis ultimately underwent a revision to a complete arthrodesis, due to continued synovitis and erosions throughout the carpus. The patient with the SLAC wrist had preoperative cystic changes in the capitate and is showing erosion of the capitate at 1 year follow-up, but remains satisfied with the procedure at this time. The other two patients remain satisfied with their outcomes, including good pain relief, despite reduced motion. In retrospect, the two wrists showing post-operative changes of the capitate also had poor structural bone quality pre-operatively and should not have been indicated for the procedure. However, both patients had a satisfactory result on the first wrist and requested the same procedure on the opposite side. The remaining patients are showing no measureable capitate erosion, or loss of motion.

Complications and Management of Failed Wrist Arthroplasty

A comprehensive review of strategies for the prevention and management of intra-operative and post-operative complications in total wrist arthroplasty is beyond the scope of this article, but can be found in other reports [29, 30]. Potential intra-operative complications include fractures and tendon injury. Early post-operative complications include wound-healing problems (hematoma, wound edge necrosis, dehiscence), extensor tendon adhesions, wrist stiffness, wrist imbalance, distal radioulnar joint problems (impingement, instability, arthrosis), prosthetic instability and infection. The most common long-term serious complication is implant loosening, particularly of the distal component in the newer designs. Osteolysis can occur due to polyethylene wear, with concurrent metallosis, if the polyethylene wear results in exposed metal surfaces (Fig. 16.4). Metallosis can also occur from screw carpal plate interface motion, due to carpal component loosening. The true incidence of these complications remains unknown in the absence of adequate long term reports.

Revision arthroplasty, arthrodesis and resection arthroplasty are options for salvaging a failed total wrist arthroplasty due to imbalance, loosening, or instability [29, 30]. Revision arthroplasty is an option for aseptic loosening if there is adequate bone stock, or if bone grafting is feasible. The thickened capsule must be widely released to allow wrist flexion and extraction of the components. If there has been substantial subsidence, then lengthening of the wrist flexors and extensor tendons may be required. Iliac crest bone graft may be needed to fill defects and re-establish the basic architecture of the carpus. When using the Universal 2 prosthesis for revision, the graft can be transfixed to the remaining carpus, using the carpal component

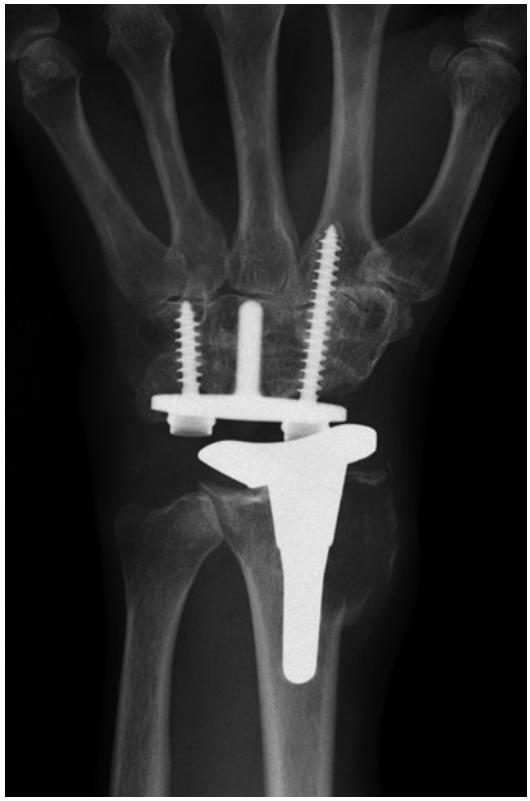

Fig. 16.4 Universal 2 total wrist in a 64 year old female with rheumatoid arthritis at 7 year follow up showing severe osteolysis (same patient in Fig. 16.2). Carpal component remained well fixed. Joint was debrided and defects filled with allograft. Polyethylene bearing component was revised

fixation screws. Because the decision to perform a revision depends primarily on the integrity of the bone and soft tissues, it may not be possible to make a final decision until direct inspection, at the time of surgery. Thus, the surgeon must be prepared for arthroplasty and for arthrodesis. Patients who have poor bone stock, severe capsule defects, or particulate synovitis are rarely good candidates for revision arthroplasty. Removal of the implant components, intercalary bone graft, using the cancellous portion only of a femoral head allograft shaped to fill the defect and dorsal arthrodesis plate is an effective technique, though the time to fusion can be several months (Fig. 16.5). An established infection should be treated by implant removal and primary or delayed conversion to an arthrodesis.

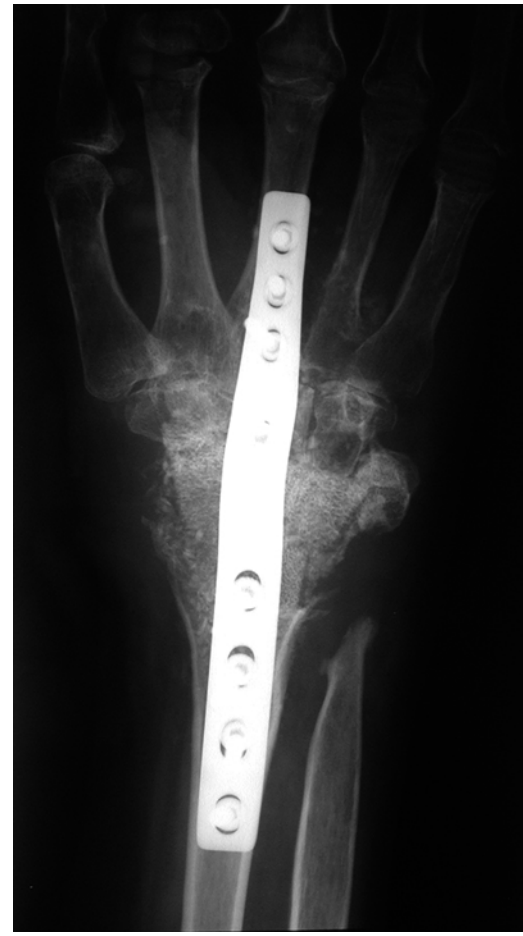

Fig. 16.5 Wrist arthrodesis for a failed total wrist using the cancellous portion only of a femoral head allograft and a dorsal wrist arthrodesis plate

Clinical Pearl

Conversion of a failed arthroplasty to a complete wrist fusion is a reliable procedure using modern surgical methods, which restores joint position and hand function.

Summary

Total wrist arthroplasty preserves motion and improves hand function for daily tasks and lower-demand vocational and other activities. It is often preferable to fusion, when both wrists are arthritic.

Early implant designs were troubled by complications, including wrist imbalance and loosening. However newer prosthetic designs provide a functional range of motion, better wrist balance, reduced risk of loosening and better implant stability. The procedure is more likely to be successful in a lower demand patient. Complications are less common in patients with adequate bone stock who do not have severe wrist deformity or highly active disease. In properly selected patients, short term complications are uncommon. The success of total wrist arthroplasty depends on appropriate patient selection, careful pre-operative planning and sound surgical technique.

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DRUJ Replacement Arthroplasty

17

Vivien C. Lees

Keywords
Arthroplasty • Distal radioulnar joint (DRUJ) • Proximal radioulnar joint (PRUJ) • TFCC • Biomechanics • Kinetics • Kinematics • Prosthesis

Introduction

It is a general observation that as new prosthetic options are introduced and confidence in the efficacy and longevity of the prostheses is gained that the indications for the use of such prostheses are extended and the prostheses are used earlier in the disease process with clinical benefit. This pattern of development is what we are currently experiencing with respect to DRUJ replacement arthroplasty. Recent years have seen an increasing awareness of the anatomical and biomechanical significance of the distal radioulnar joint (DRUJ). With this has come a more critical approach to surgical management of DRUJ disorders and a realization that all forms of "excisional arthroplasty" can only restore forearm rotation at the expense of forearm stability. This, in turn, has led to renewed interest in prosthetic replacement of the ulnar head, a procedure that had previously fallen into disrepute because of material failures with early implants.

Partial or complete loss of the ulna head produces significant disruption of forearm biomechanics through impingement of the ulna remnant on the radius [1]. The clinical problem is manifest as ulna side wrist pain, loss of ability to lift and load as in power grip maneuvres and variable loss of forearm rotation. Not every patient having these ablative surgeries does poorly and those that fare better may have either low demand lifestyle / occupation and/or may be able to recruit brachioradialis muscle action on lifting and power grip (brachioradialis being the only muscle that can lift the radius off the ulna stump). Instability is associated with symptomatic clunking and will eventually lead to arthrosis secondary to the abnormal movement. The term convergent instability has come into common use but it should be appreciated that all instability is convergent; this fact being inherent on the underlying biomechanics. Prior to the development of arthrosis anatomical ligament reconstruction is the treatment of choice [2, 3] Once arthrosis supervenes the options lie between conservative treatment and, in early osteoarthrosis, realignment by ulna shortening

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[4] or replacement of the ulna head. In the presence of instability and significant arthrosis full replacement arthroplasty of both ulnar and radial components should be considered. Where instability is not part of the primary problem or can be adequately addressed through simultaneous soft tissue reconstruction there are a range of ulna head prostheses now available.

Even to this day, the literature contains statements that salvage treatment for the degenerate DRUJ is best achieved through resection arthrodesis [5]. The Sauvé-Kapandji operation is still being recommended for salvage of distal radioulnar joint pathology [6]. It is recently advised that primary distal radioulnar joint pathology be treated by the various partial ulna head excisions (Darrach's procedure and its variants – Bower's hemiresection interposition arthroplasty, wafer procedure, matched ulna resection) with prosthetic replacement reserved as salvage for patients in whom such primary surgery fails [7]. Excisional arthroplasties cannot and do not restore normal biomechanics in terms of loading and lifting strengths.

Applied Anatomy and Biomechanics Relevant to Prosthetic Design

What follows is a brief synopsis of those pertinent aspects of the anatomy and biomechanics that need to be factored in to DRUJ prosthetic design. It is beyond the scope of this article to provide an entirely comprehensive description or to acknowledge all those who have made an undoubted contribution to our understanding of the same.

Forearm rotation occurs between the head of the radius and the radial notch of the ulna proximally and head of the ulna on the sigmoid notch of the radius distally. These joints known as the proximal radioulnar joint (PRUJ) and distal radioulnar joint (DRUJ), respectively, act functionally as hemi-joints facilitating forearm rotation. The DRUJ is a uniaxial pivot or trochoid joint with articulating surfaces between the disproportionately sized convex head of the ulna and concave sigmoid notch of radius. Differential curvatures of these structures confer mobility at the price of stability [8]. The DRUJ is geometrically a non-constrained articulation subject to dorsal and palmar translational movement. The differentially-curved surfaces permit the sigmoid notch to translate palmarly or dorsally on the fixed seat of the ulna throughout the range of forearm rotation. This is an important point for prosthetic design as a constrained joint, with otherwise similar features, would not permit the same freedom on rotation. Furthermore, the sigmoid notch has been shown to have wide variability in its conformation both in transverse and coronal planes [9]. This variability could conceivably impact the degree of stability of some of the ulna head prosthetic designs.

Stability of the DRUJ depends largely on the soft tissues and the triangular fibrocartilage complex (TFCC) is key in this regard. Most of the partial and whole ulna head prostheses depend on the integrity of the soft tissue complexes around the DRUJ for their success and as these soft tissues are often already compromised at the time of presentation the choices lie between total replacement arthroplasty as the primary procedure or partial replacement alongside one of the soft tissue reconstructions elsewhere reviewed [7]. The TFCC is a 3-dimensional structure (comprising the triangular fibrocartilage, extensor carpi ulnaris subsheath, ulnolunate ligament, ulnotriquetral ligament and ulnar collateral ligaments) [10, 11] that essentially attaches the hand to the ulna via the ulna styloid and is key in both the transmission of load from hand and carpus to the ulna, and in binding the radius to the ulna during loading manoeuvres. Ligamentous condensations of the TFC itself are also referred to as the distal radioulnar ligaments (DRUL) and are important for joint stability. It is important to appreciate that the ulna is the fixed structure of the forearm about which the radius revolves. It is a misconception to talk about 'instability of the ulna head' as it is the radius that is unstable on the ulna. The ulna is key to the load-bearing function in the hand and forearm [12]. The radius, meanwhile, carries the hand into different positions in 3-dimensional space, depending on its position of rotation relative to the ulna. The principle function of the

hand and forearm is to grasp, lift and manipulate objects necessarily placing varying loads through the limb. During power grip and lifting maneuvres load is transmitted across the DRUJ and distributes between the two forearm bones. The vectors of force can be described in terms of axial and transversely oriented components. It follows that assessment of outcomes following partial or total DRUJ reconstruction or replacement arthroplasty should report on power grip and the ability to lift load.

Clinical observation had suggested that it was important to preserve the integrity of the ulna head and that removal of the ulna head would lead to the impingement of the ulna stump against the radius on lifting a weight (Fig. 17.1a, b) [13]. This so-called impingement phenomenon is caused by the brachialis muscle contracting to flex the elbow in the neutral position of forearm rotation, and because of its attachment to the proximal ulna, levers the ulnar head or ulnar stump towards the sigmoid notch or shaft of radius, respectively. The brachioradialis muscle has the opposite action and with elbow flexed lifts the radius off the ulna head /stump. Clinical outcome may be influenced in part by factors relating to balance of muscles determining forces across the DRUJ. Instability/impingement of the ulna stump is a prime cause of therapeutic failure for surgical intervention on DRUJ-related pathology.

Our own experimental series sought to document what we saw as the important role played by the DRUJ in transmitting both axial and transversely applied loads of the hand and forearm (power grip and the ability to lift weight). Testing our hypothesis we have undertaken biomechanical studies on a cadaver model of the human forearm leaving intact the ligamentous stabilising structures of the TFCC. The kinematics and load-bearing characteristics of the DRUJ were examined [14]. Figure 17.2 summarises the results from the series of 12 cadaveric arms that were measured. Figure 17.2a shows the family of force transmission curves generated from the Tekscan®™ data with increase in applied load up to 10 kg (limit of this experimental system). Force transmitted across the DRUJ clearly

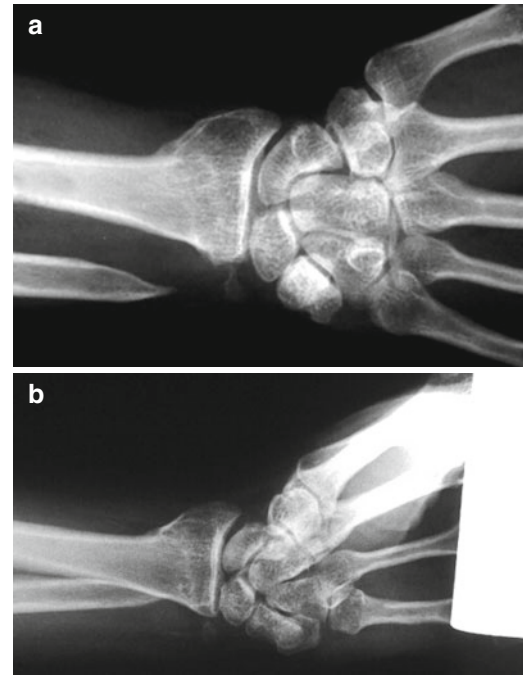

Fig. 17.1 Ulnar impingement in a 46-year-old man who had Sauvé-Kapandji procedure 10 years previously. The patient had presented with continuing distal forearm pain. (**a**) Anteroposterior view of the wrist as routinely requested on clinic review (**b**) Stress loading view demonstrating the ulnar impingement (Reproduced with permission of Sage from Lees and Scheker [13])

increases as the forearm rotates peaking at 60° supination. Similar families of curves were seen in respect of contact areas within the joint (data not illustrated). Figure 17.2b shows the strain gauge data with the distribution of axial forces between the two forearm bones showing what appears to be a reciprocating pattern of load distribution between the radius and ulna. When the total loads are averaged for each of the forearm bones it is observed that the ulna takes 32–24 % of the load. This is in contrast to some publications in the literature and in accordance with others [14]. Excision of the ulna head mimicking a Darrach's procedure defunctioned the forearm such that the ulna transmitted no further load (Fig. 17.2c). It is fairly widely appreciated nowadays that optimum kinetics of the forearm requires an intact ulna head. Where the ulna head is lost through disease it is appropriate to consider prosthetic replacement.

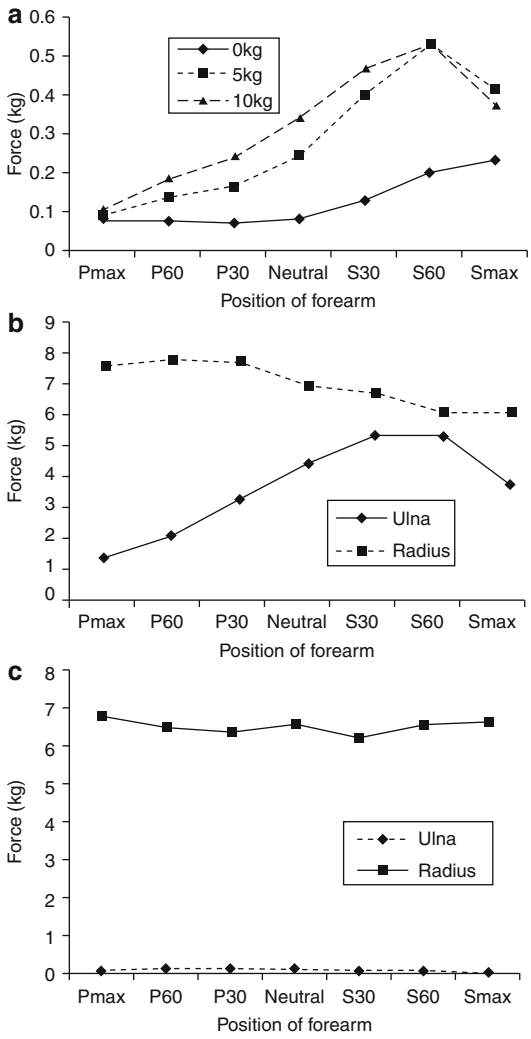

Fig. 17.2 (**a**) Tekscan data showing greater force transmitted across the DRUJ in supination. The family of related curves shows increased force transmitted with increasing axial load throughout the range of forearm rotation. (**b**) Strain gauge data showing approximately reciprocal curves of force transmission along the radius and ulna (10 kg of axially applied load). (**c**) Strain gauge data showing effect of excision of the ulna head on normal forearm mechanics. Under same conditions as 8b there is virtual complete loss of force transmission through the ulna (Reproduced with permission of Sage from Shaaban et al. [14])

Implant Mechanics

Austerman et al (2007) [15] undertook laboratory studies on cadavers to look at the effect of ulnar implant stem materials and lengths on bone strains using cemented stems and then measuring strains to applied loads using strain gauges. Under these test conditions titanium stems were closer to native bone strains than stainless steel and interestingly the shorter 3 cm stems showed load transfer characteristics that were closer to those of native bone than the longer (5 or 7 cm stems). The equivalent measurements were not undertaken for non-cemented press-fit stems that are currently in use such that the significance of this work to these non-cemented prostheses currently in use is not known.

A prototype ulna head prosthesis, with integral suture attachment site to allow temporary stabilisation of the head, was tested in an *in vitro* system on a cadaver model to evaluate the kinematics of partial versus total ulna head replacement [16]. Both types of prosthesis were indistinguishable from the intact joint controls but simple excision of the ulna head (Darrach's) produced DRUJ instability and radioulnar convergence. Similar results were obtained using two other prototype ulna head prostheses trialled in cadaver models where it was observed that these prosthetic replacements produced more normal biomechanics than distal ulna resection, albeit that there was some reduction in pronation and subluxation of the prosthetic head on the sigmoid notch in supination [17].

An instrumented ulnar head prosthesis was designed by Gordon et al (2006) [18] for measurement of force transmission across the DRUJ with the cadaveric forearm specimen fitted into a custom joint simulator following surgical placement of the implant. Computer-controlled pneumatic actuators were used to simulate the action of forearm musculature. The results suggested that forearm position and degree of contact of the ulna head with the radius articulation were the most important factors determining load transmission.

Treatment Options

Historically simple silicone caps were tried for replacing the ulna head [19]. A dome-shaped ulnar head prosthesis had a stem to be inserted in

the medullary canal and a cuff that fitted over the edge of the ulnar stump. Over time this implant proved insufficiently durable and failed in too many cases. Furthermore, Fatti and Palmer [20] reported that within 5.8 years of placement of Swanson silicone rubber interpositional wrist arthroplasties (not specifically those of the ulna head) silicone synovitis was present in the majority of cases reviewed. Essentially, simple silicone prosthetic wrist prostheses such as the Swanson silicone ulnar head replacement ceased to be a treatment option because of this complication.

Classical treatment of disorders of the DRUJ involved the use of a variety of arthroplasty techniques. These include full excisional arthroplasty, hemi-resection or partial resectional arthroplasty, ulnar shortening, or prosthetic replacement arthroplasty [21]. For the reasons outlined earlier the excisional arthroplasty techniques are no longer recommended except in the lowest demand hands.

Prosthetic Replacement Arthroplasty – Prostheses. Indications, Techniques and Outcomes

It would be fair to say that the last 20 years has seen a great deal of activity in terms of prosthetic design with quite a number of new prostheses entering the market. These are described here as the partial (ulna head replacement) and total (radial and ulnar components) prosthetic replacement arthroplasties. Prostheses can be unconstrained, semi-constrained, and constrained. A fully constrained prosthesis does not permit forearm rotation. An unconstrained prosthesis depends on the integrity of the soft tissues.

The general indications and contraindications for DRUJ prosthetic replacement arthroplasty are summarised in Table 17.1. More specific detail on indications as recommended by the designers is given with the description of each individual prosthesis along with preliminary results, and 5 year results where available. Where materials science and biomechanical studies have been published these are also included. The surgical techniques of prosthetic implantation have been illustrated with reference to the Herbert-Martin™, Eclypse™ and APTIS™ covering the principal types of design.

Table 17.1 Indications and contcraindications for prosthetic replacement arthroplasty of DRUJ

Indications
Primary
Primary osteoarthrosis – pain not adequately relieved by conservative management
Osteoarthrosis secondary to distal radius fracture with malalignment or chronic instability
Inflammatory arthritis (reasonable bone stock)
Crystal deposition disease
Recurrent instability with impingement following anatomical ligament reconstruction
Secondary – for salvage in case of failure following:
Darrach's and variants
Bower's hemiresection interposition arthroplasty
Matched ulna resection
Sauvé-Kapandji procedure
Malalignment of DRUJ not corrected by osteotomy (radius fracture, Madelung's deformity)
Revision for previous ulna head or total replacement arthroplasty
Contraindications
Previous history of infection in the joint or infection complicating previous replacement arthroplasty
Partial resurfacing type prosthesis rely on presence of adequate ligaments for effective function and if ligaments not present consider total replacement arthroplasty
Patient has job or recreational activities that will lead to heavy loading of the joint with risk of damage to prosthesis over time
The various prostheses are not interchangeable with respect to their indications and contraindications.

Partial: Ulna Head Replacements

The Herbert-Martin Ulna Head Prosthesis

The Herbert-Martin™ ulnar head prosthesis (Martin Medizin-Technik, Tuttingen, Germany) was first marketed in 1995 and 10 year results are now reported in the literature [22, 23]. The prosthesis consists of a ceramic head and non-cemented titanium stem that is fitted to the shaft of the ulna. It is reported that this prosthesis can improve forearm function following previous ulnar head excision and that there have been no signs of failure of the prosthesis in the medium to

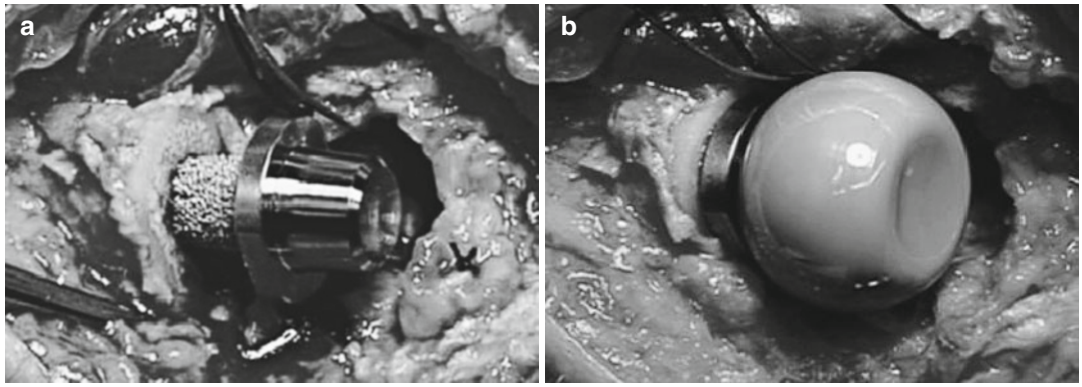

Fig. 17.3 (**a**) Herbert head stem sited in the ulna shaft prior to impaction. (**b**) Prosthesis inserted prior to reduction onto the sigmoid notch (Reproduced with permission from Mühldorfer-Fodor [25])

long term. This prosthesis has been widely used and there is considerable experience accrued in its utility.

The technique of insertion begins with dorsal exposure of the distal radioulnar joint through the floor of the 5th dorsal extensor compartment. An ulnar-based capsuloretinacular flap is raised by sharp dissection off the ulnar neck proximally and off the dorsal part of the triangular fibrocartilage complex (TFCC) distally. More detailed description of surgical approaches to the DRUJ has been elsewhere detailed by Garcia-Elias and Hagert [24]. Osteotomy of the distal ulna corresponding to the preoperatively planned size of the prosthesis, is performed and the ulna head, while preserving the attachment of the TFCC within the capsuloretinacular flap. The ulnar medullary canal is reamed prior to insertion of a trial prosthesis. The trial prosthesis is accurately fitted into the shaft such that it lies 1–2 mm ulna minus at wrist joint level. The position is checked fluoroscopically. After implanting the definitive stem and ulnar head [Fig. 17.3] the capsuloretinacular flap is tightly reattached to the dorsal rim of the sigmoid notch through drill holes [25]. Patients with pre-existing impingement of a distal ulnar stump may have insufficient tissue to raise an adequate capsuloretinacular flap. This situation may be aggravated where there have been one or more previous surgeries with loss of the ulna head as an anatomic landmark. Postoperatively an above elbow cast is applied with 70° elbow flexion, 40° forearm supination, and 20° wrist extension for 2 weeks. Subsequently, an ulna gutter splint is applied with forearm rotation limited at 40°. Six weeks postoperatively unlimited active range of motion is allowed and normal activities are gradually commenced. Return to all activities is allowed 12 weeks postoperatively. Stability of the distal radius can be restored where there is sufficient soft tissue present and where appropriate tightening of the soft tissue is performed at the time of definitive implant placement.

Clinical Pearl

Design and modify access incisions for DRUJ arthroplasty respecting and, using where possible, previous scars to avoid skin necrosis due to parallel or perpendicular incisions. Avoid tacking sutures in the corners of skin flaps.

Incipient skin necrosis overlying a prosthesis can be pre-empted by excision and local flap transposition

Ensure tight press fit of ulna components in ulna diaphysis selecting the largest size that will fit

Rehabilitation would best include exercises for strengthening brachioradialis as the only muscle that lifts the radius off the ulna head

Ulna head prosthetic replacement can be used as a salvage procedure for failed Sauvé-Kapandji with painful radioulnar convergence [26].

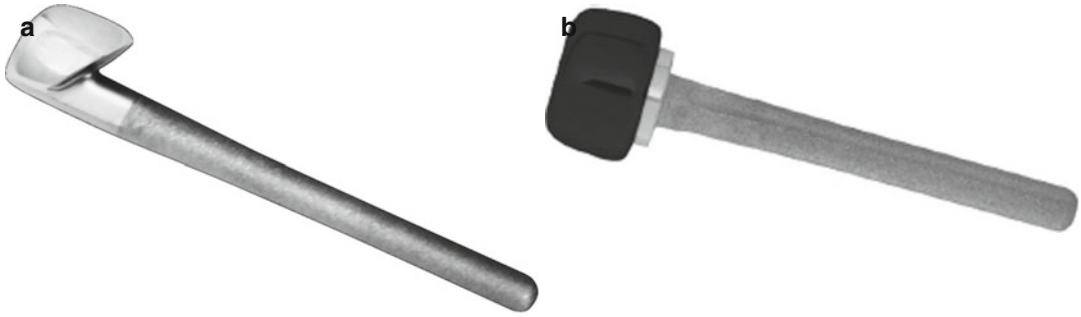

Fig. 17.4 First choice DRUJ system by Ascension. (**a**) Partial ulna head resurfacing implant. (**b**) Modular version designed for use in revision procedures where the ulna head has already been removed either partially or in entirety (Reproduced with permission from Kopylov and Tägil [29])

Following the placement of the Herbert-Martin™ ulnar head prosthesis in ten patients, all had relief of pain and instability with improvement in grip strength from 27 % preoperatively to 55 % that of the normal hand postoperatively. Two patients had fracture of the radioulnar fusion mass that had been left in place at the time of prosthetic placement.

Long-term outcomes of the Herbert-Martin™ ulnar head prosthesis are now available with two studies including one from the inventor. Kakar et al (2012) reported on 47 cases followed for a mean of just under 5 years demonstrating implant survival rate of 83 % [27]. Evidence of sigmoid notch changes in response to rubbing of the implant was seen in numbers of cases as was stress shielding of the implant within the ulna diaphysis leading to bone resorption. There was modest loss of range of rotation from preoperative values for both pronation and supination. Grip strength improved from 17 to 21 kg and was associated with reduction in pain scores. Of those implants that had previously failed there seemed to be a trend for higher failure rates in patients with flat-faced sigmoid notch as compared with C-shaped or ski-slope type notch conformation (presumed less inherent stability). In the other study [28] 16 cases were available for review at a mean of 11 years from the authors' earlier series of patients treated by prosthesis for failed resection arthroplasty of the ulna head. Importantly, the early results reported for this cohort do appear to be stable in the long term with no additional surgeries, similar pain scores, range of movement and grip strengths demonstrated in these long term reviews. Those implants that are stable and successful in the short term remain stable in the long term with no evidence of loosening.

First Choice DRUJ System (Ascension)

Kopylov and Tägil (2007) described a partial ulna head resurfacing implant for use in primary cases and a modular ulnar component for use in previously operated cases [29]. The First Choice DRUJ™ system has two forms; one for primary procedures [Fig. 17.4a] and a modular prosthesis for revision procedures [Fig. 17.4b]. In common with other ulnar head prostheses the designers stress the importance of having either intact soft tissues to confer stability in the final result or undertaking appropriate soft tissue reconstruction at the time of implant surgery to restore that stability. Following the initial description there does not appear to have been any substantive report on the use of this implant in a clinical series. Cadaveric biomechanical studies demonstrated that the prosthesis closely replicated the anatomy of the resected part of the ulna head [30]. It was anticipated that placement of a near anatomical partial ulna head implant without extensive dissection of soft tissues would optimise clinical results. The prosthesis was designed to allow retention of the ulnar neck, ulnar styloid, extensor carpi ulnaris groove, ulnocarpal ligament attachments, extensor carpi ulnaris sheath, and the triangular fibrocartilage complex attachments to the ulnar styloid. Thus, while all articular surfaces of the ulnar head are replaced, the

ligaments and other bony anatomy responsible for DRUJ stability are maintained. Because the DRUJ anatomy is preserved, the joint mechanics are not altered. Furthermore, since the procedure is performed through a minimal exposure and immediate implant fixation can be achieved, rapid rehabilitation is possible.

Indications for the partial ulnar head from the Ascension First Choice DRUJ™ system are those for a partial resurfacing arthroplasty including skeletally mature patients with primary or secondary osteoarthritis and rheumatoid patients with well-controlled disease process and good bone stock. Patients with negative or positive ulna variance can have this corrected to a more neutral ulna variance during placement of the prosthesis and it is clear that an ulna positive situation should be avoided to reduce the risk of ulna impaction syndrome. Excessive *acquired* ulnar positive variance maybe a contra-indication for the partial ulnar head replacement implant; these patients are better suited for total head replacement using the modular version of the implant.

Published results from 10 patients included 7 treated for primary osteoarthritis and 3 for post-traumatic arthritis. In a retrospective review of patient records at an average 6 months follow-up, there were no intraoperative or postoperative complications. Pain relief was good in all patients; however, none were completely pain free. Motion was also improved in all, with patients achieving at least 75° of pronation and 65° of supination. Wrist flexion and extension were unaffected. There were no cases of distal radioulnar joint instability (BD Adams 2008, manufacturer's data – Ascension).

E-centrix™ Ulna Head Prosthesis

Sauder and King (2007) described an ulnar head prosthesis with eccentric design to best anatomically recreate the original ulna head [31]. The geometric centre of the ulna head has been shown to lie offset from the ulnar shaft by a mean 2.5 ± 1.4 mm. Furthermore, computer tomography studies have shown ulna shaft canal diameter does not correlate with head diameter making modularity of these two components a desirable feature. These authors have developed the E-centrix™

Fig. 17.5 E-centrix prosthesis shown here is the modular version with the extended ulna collar for use in revision procedures. The neck of the prosthesis contains holes for suture attachment. (Reproduced with permission from Sauder and King [31])

(Wright Medical Technology, Arlington, Tenn) comprising a head made of cobalt chrome and a titanium stem to allow uncemented press fit with bony ingrowth. The stem of this prosthesis has suture holes to allow an initial suture fixation of the prosthesis to soft tissue DRUJ stabilisers (Fig. 17.5) Titanium has a low modulus of elasticity thus reducing stress shielding of the ulna. This design seeks to reduce the bulk of the prosthesis and in theory should allow for tighter more secure soft tissue tensioning. The most common complications reported by the authors were stiffness and continuing instability. Attention to sizing of the ulna head would appear to be critical to success of the technique. Report in the literature of a definitive clinical series of this implant use is awaited.

Kinematic studies of forearm movement in a cadaver model addressing the issue of whether the prototype eccentric ulna head prosthesis could restore normal kinematics to the distal forearm were undertaken as part of the developmental work done on this prosthesis [16]. It was shown that the prosthesis used restored normal kinematics as compared to the surgically unaltered specimens. By contrast, full excision of the ulna head produced distal radioulnar joint instability in the form of radioulnar convergence and increased anteroposterior translations.

Avanta uHead™ Prosthesis

The uHead prosthesis (Small Bones Innovations Inc., Morrisville, PA, USA (formerly Avanta)) consists of a cemented modular prosthesis with metal head having holes for soft tissue attachment. Implant arthroplasty of the distal ulna combined with an adequate soft-tissue repair was recommended to improve pain, function, and strength of the wrist and forearm [Fig. 17.6a, b] [32]. The prosthesis is

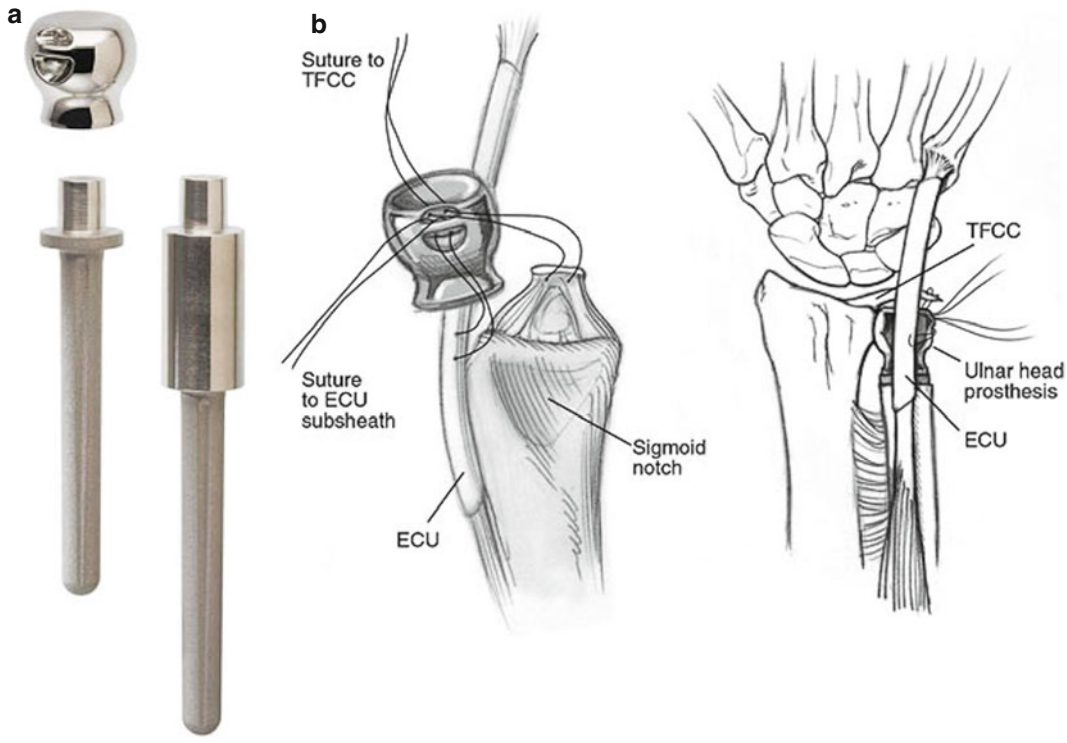

Fig. 17.6 (**a**) The uHead prosthesis is shown in both the regular and the modular version. (**b**) This diagram illustrates in detail how the ulna head component is secured with suture attachment to the soft tissues prior to securing it onto the previously placed ulna stem. (Reproduced with permission from Sauerbier et al. [32])

modular with a cobalt chrome head that fits onto a plasma sprayed stem that press fits into the reamed ulna shaft. The head of the prosthesis has suture holes to allow attachment to soft tissues and two different stems sizes to accommodate primary and revision cases. The surgical technique has been described along with emphasis on the repair of the soft tissues stabilising the joint, in common with the other prostheses of this type. Prosthetic replacement of the distal ulna restored stability to the DRUJ in patients with partial or complete excision of the ulnar head or DRUJ arthrosis and relieved symptoms from radioulnar impingement. The incidence of complications and revision surgery to date in these authors' early series were low.

Two year results following uHead™ implants performed on 19 wrists in 17 patients suggested pain scores decreased by 50 %, grip strength improved to 4 kg and all wrists remaining stable [33]. There were 6 complications in the 19 cases with 1 case requiring a tendon stabilisation procedure, 1 case of progressive degeneration of the sigmoid notch and 2 cases of stem loosening that resolved with cementing the loose stem in place. By 4.5 years the same group reported their intermediate experience with recurrent instability in 4 cases, 2 of which needed further intervention and were successfully restabilised whilst one implant was replaced for breakage after a fall [22].

Salvage of a previously performed Darrach's procedure using the uHead prosthesis and soft tissue stabilising procedures (pronator muscle interposition (Johnson procedure) and tendon tether procedure (Breen-Jupiter tenodesis)) as a means of preventing radioulnar convergence were investigated and compared in a cadaver model using the Mayo dynamic PC-controlled forearm simulator [32]. The uHead prosthesis restored stability while the soft tissue stabilising procedures did not differ significantly from the Darrach's procedure under the testing conditions of the construct. The implication of the work is that implanting an

ulna head endoprosthesis for patients suffering painful forearm instability after ulnar head resection is likely to be a more worthwhile approach than trying to correct instability with soft tissue procedures. Comparison of the uHead™ prosthesis and Herbert ulna head prosthesis in a cadaver model using a Fastrack sensor system to capture the amount of displacement of the radius with applied load demonstrated a degree of subluxation and loss of part of the range of pronation following either of the ulna head replacements [17]. However, biomechanics were much closer to normal than was observed when simple ulna head excision was performed.

More recently Small Bones Innovations Inc. have developed a sigmoid notch replacement to be used with the Avanta uHead™ where complete DRUJ replacement is indicated (manufacturers data). This is an unconstrained system and reports on use are awaited.

The Eclypse™ Partial Ulna Head Prosthesis

The Eclypse (Bioprofile, Grenoble, France) partial ulna head prosthesis has been developed and recommended for isolated DRUJ arthrosis. It comprises a pyrocarbon spacer slotted over a titanium peg that fits into the distal ulna. The spacer is free to rotate and translate relative to the stem thus accommodating the 'play' between the distal ulna and the concavity of the sigmoid notch [Fig. 17.7a]. Pyrocarbon has very low coefficient of friction that makes it a highly durable material for prosthetic use. The construct can be sited without disrupting the foveal insertion of the TFCC ligament complex [34]. The purpose of the implant is to prevent convergence of the radius and ulna through the full range of forearm rotation. The 'peg in a hole' design allows sufficient play in the system to accommodate physiological proximodistal translation of radius on the ulna and slight axial rotation of the implant. The initial report contained the details of three clinical cases whose outcomes at 11 months follow up included relief of presenting pain and ability to lift weights up to 4 kg. Unlike total ulnar head prostheses partial implants permit replacement of only the damaged portion of the ulnar head without loss of the ligamentous stabilizers of the joint that are necessary for physiological function. It follows that the indications for use of this prosthesis include idiopathic osteoarthritis of the DRUJ, inflammatory disorders that have compromised the integrity of the ulna articular surfaces and posttraumatic loss of congruency of ulna head (sigmoid notch intact). It is not recommended that this prosthesis be used for salvage of the failed Darrach's procedure and its variations as by definition the ulna styloid and its ligamentous attachments including the TFCC have been lost.

The technique of Eclypse implantation [Fig. 17.7] is outlined. With the patient under regional anaesthesia and upper arm tourniquet control a dorsoulnar skin incision is made centred on the protruding ulnar head, the extensor retinaculum is exposed as far as the fourth dorsal extensor compartment and the fifth dorsal compartment incised longitudinally. An ulnar based flap of extensor retinaculum is elevated and the EDQM retracted radialwards [Fig. 17.7b]. The diagram shows how the retinacular flap should be elevated without breaching the DRUJ capsule and leaving the sheath around the ECU intact. An ulnar-based capsular flap is then raised to expose the joint surfaces extending from the dorsal ridge of triquetrum through the midline of the radiolunotriquetral ligament along the line of the intersection of the septum between the fourth and fifth dorsal compartments and the radiocarpal joint. The ulna head can now be exposed without detaching the intact TFCC by fully pronating the forearm and applying a dorsal translation force from the palmar aspect.

Two osteotomies are then performed and the correct orientation of the osteotomy cuts is key to the success of the procedure [Fig. 17.7c]. A transverse cut is made perpendicular to the axis of the ulna. The second longitudinal cut at the level of the fovea needs to be perpendicular to the plane of flexion-extension of the elbow. Following removal of the damaged portion of the ulna the ulna head is exposed as before and an awl is introduced at the base of styloid/fovea near the centre of the metaphysis and parallel to the shaft of ulna. A 4 cm drill hole is made along the distal ulna shaft and a suitable sized stem press fitted in place [See Fig. 17.7a for detail of

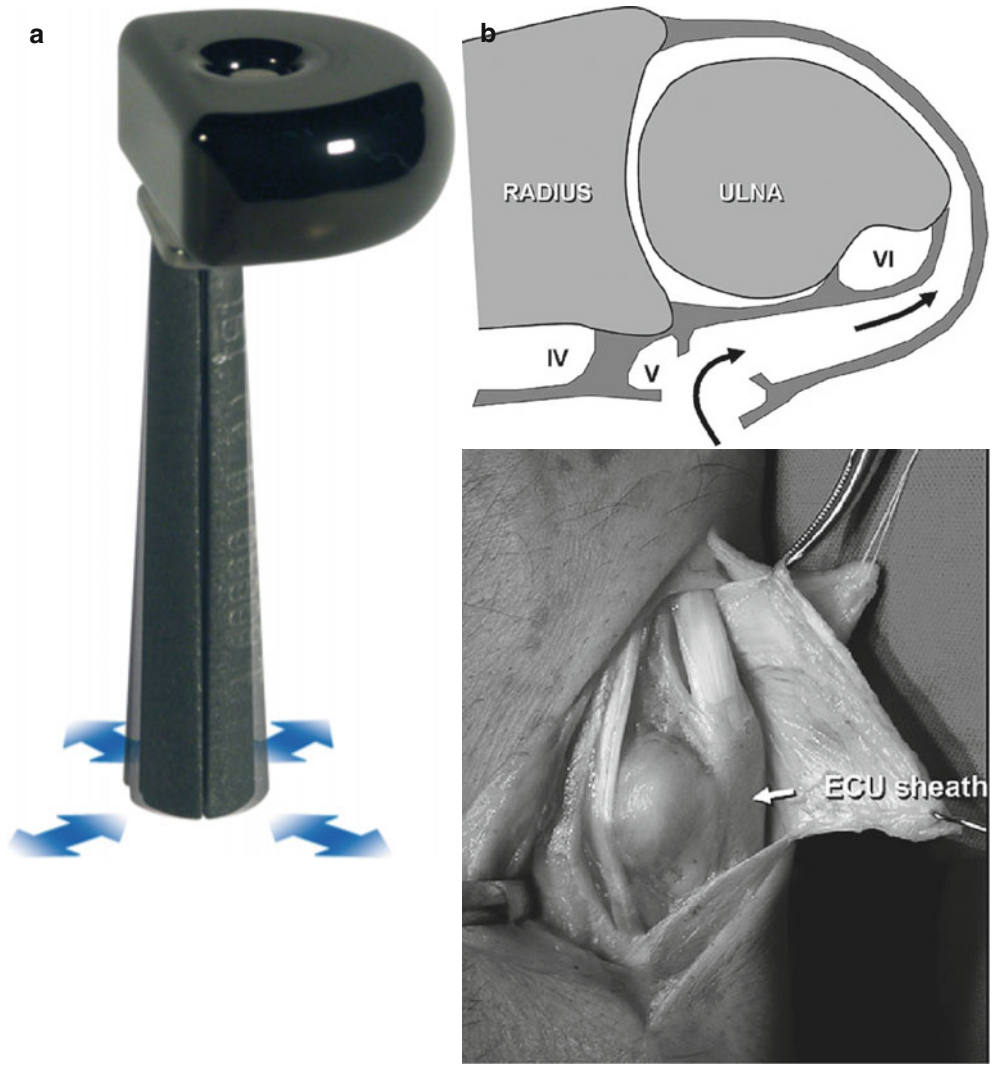

Fig. 17.7 The Eclypse prosthesis: (**a**) Eclypse prosthesis showing the legs of the ulna stem that splay out proximally following insertion to stabilise the stem in the ulna shaft. The design accommodates new bone formation between the four rectangular elements of this stem design. [Eclypse manual]. (**b**) The surgical approach to the joint is shown here diagrammatically and in surgical dissection. An ulnar-based retinacular flap is elevated leaving the DRUJ capsule and ECU sheath intact. (**c**) The sawbones model illustrates the cuts that need to be made in the head and ulna with emphasis on the orientation for correct placement of the prosthesis. (**d**) Demonstration of lifting capacity through the full range of rotation shown here in a patient 10 months following surgery using a custom-made device and 4 kg load (Reproduced with permission from Garcia-Elias et al. [34])

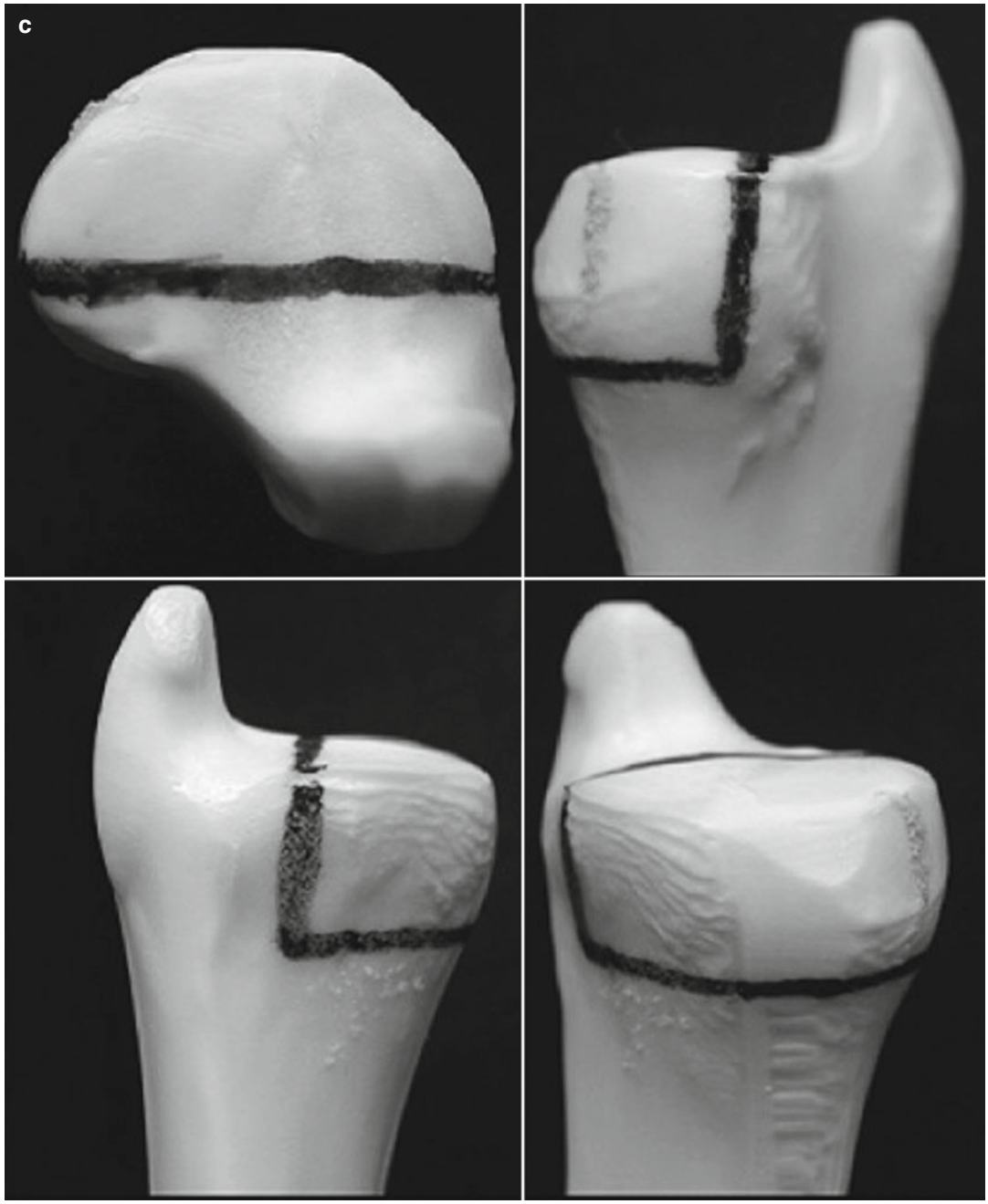

Fig. 17.7 (continued)

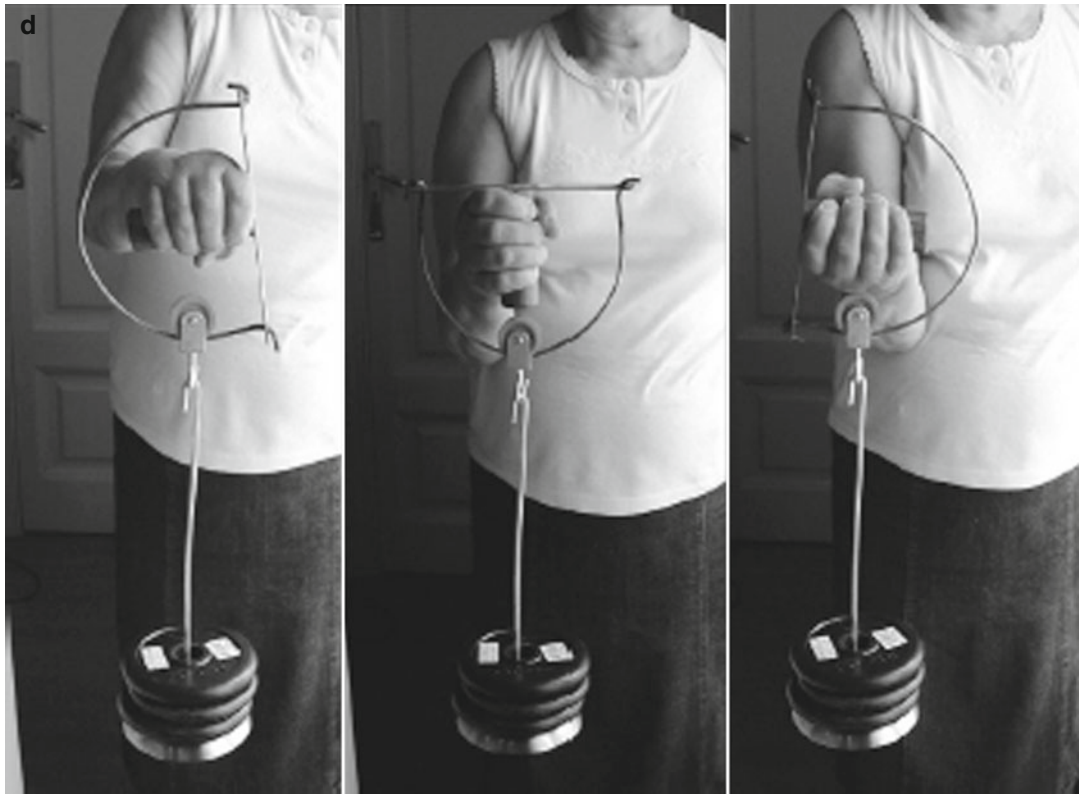

Fig. 17.7 (continued)

implant design]. The stem is not meant to be a stabiliser of the pyrocarbon head but rather an anti-dislocation device. The peg in the hole of the spacer is not likely to be subject to large forces as these are dissipated by the intact soft tissue structures. Following placement of the stem one of three possible size heads are used with the size determined at the time using trial sizers. The correct size is likely to match the volume of bone resected from the head but will be sufficient to ensure stability and full mobility of the joint. Following placement if supination is limited then release of the palmar DRUJ capsule and ulna attachment of pronator quadratus may be needed. On closure the capsular flap and the retinacular flaps to the 4th/5th dorsal compartment septum are carefully repaired.

The implant stem design depends on bone ingrowth around the 4 legs of the stem and the implant needs to be protected for the first 6 weeks following implantation for this to happen. Above elbow splintage is used for 3 weeks with active protected mobilisation and below elbow splintage for a further 3 weeks combined with hand therapy inputs.

Results were favourable at a minimum 10 month review of the 3 index cases reported in the article, with patients demonstrating ability to lift 4 kg throughout range of forearm rotation [Fig. 17.7d] [34]. A more extensive multicentre trial is underway at the present time but has yet to report at the time of writing.

Total: Ulnar and Radial Components

The ATPIS™ total DRUJ replacement arthroplasty (APTIS Medical, Louisville, KY) is presented as a bipolar self-stabilizing endoprosthesis that totally replaces the DRUJ and its stabilising ligaments [35] [Fig. 17.8a]. This prosthesis differs from the ulnar head prostheses by replacing all three components of the DRUJ, namely the ulnar head, sigmoid notch and triangular fibro-

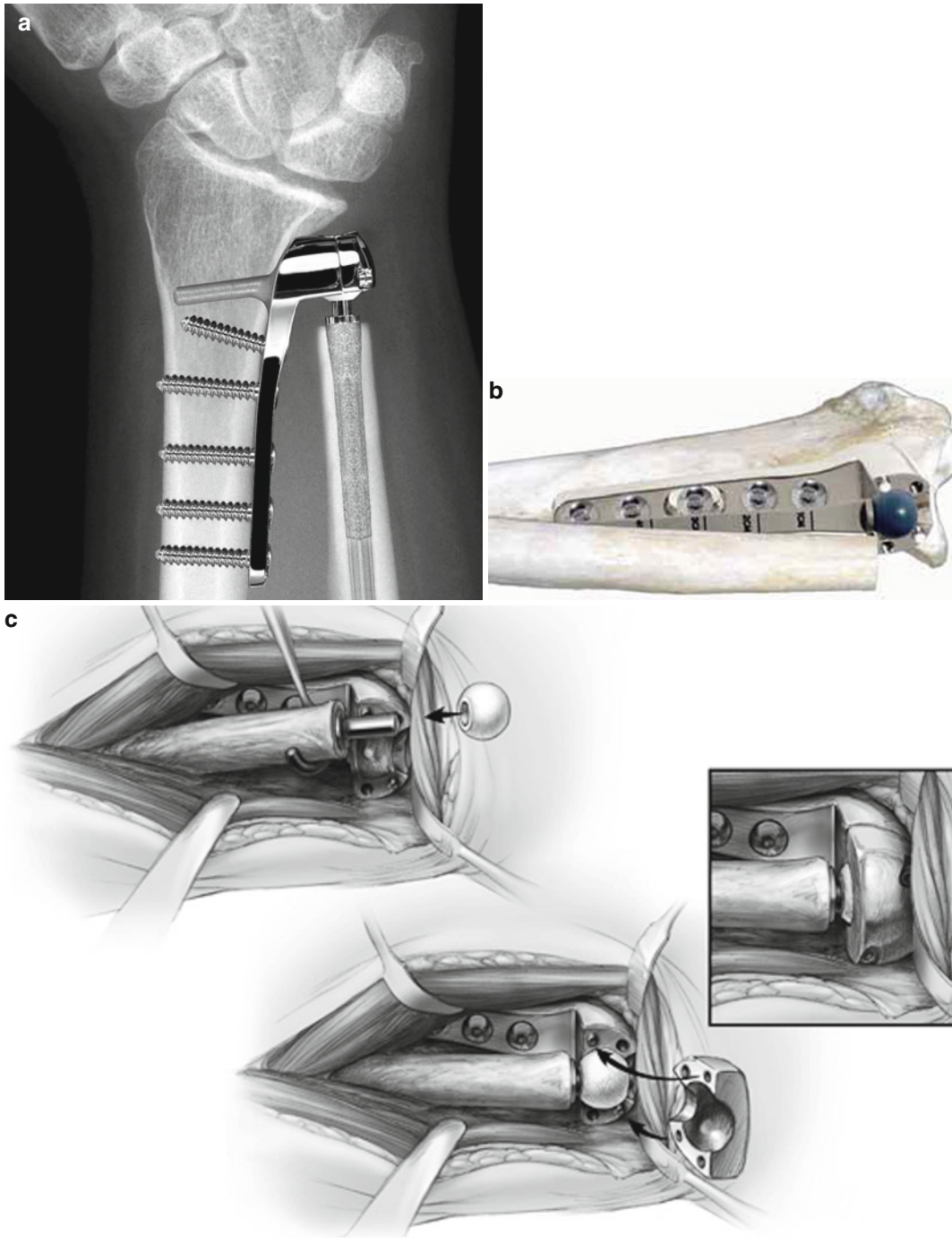

Fig. 17.8 (**a**) The APTIS prosthesis. (**b**) Diagram of sectioning measurement of ulna shaft using the trial ulna component. (**c**) Detail on implantation of the prosthesis showing the stage of placement of the polythene ball on the previously sited ulna stem component. The radial component has already been secured. The metal cage is screwed into place following placement of the ball (Reproduced with permission from the APTIS manual)

cartilage. The design allows for the physiological longitudinal migration of the radius through its range of pronosupination and provides stability of the entire forearm as well as the ability to lift loads in excess of that achieved with other prostheses [36].

The prosthesis design accommodates a full range of pronosupination, the foreshortening of the radius with pronation by means of travel of the polystyrene ball on the peg of the ulna component with the physiological lateral translation of the ulna occurring at the humero-ulna joint (trochlea). There is 30° of freedom in the movement of the polystyrene ball within the radius cage component such that physiological movement is accommodated. Early critics of the prosthetic design suggested the radial component might loosen on the radius [7]. For this to happen there would need to be torque between the plate and the radius. This does not appear to have happened in practice as the radial component permits free rotation of the ball inside the cage with the stem of the ulna free to piston proximally and distally through the range of pronosupination. Out of 900 implants that have been used worldwide the inventor has not been made aware of any instances of loosening separate from four infections that have occurred (LR Scheker, personnal communication 2013).

The technique of insertion of the APTIS joint replacement is summarized as follows: under regional anaesthesia with tourniquet control the forearm is prepped and wrapped in iodine plastic wrap to minimize contact of implant and skin. An 8-cm hockey stick incision is made along the dorsoulnar aspect of the forearm overlying the 5th and 6th dorsal extensor compartments. Skin and subcutaneous tissue are elevated as far as the radial wrist extensors in subfascial plane. An ulna-based fascial flap is raised that is transposed later on wound closure to separate the prosthesis from the ECU tendon. The approach is continued to access through the interval between the ECU and extensor digiti quinti minimi with the latter elevated off the ulna and interosseous membrane for at least 8 cm proximally. The ECU sheath needs to be fully opened distally to its insertion on the fifth metacarpal base to avoid tendon impingement on the distal implant. A preliminary excision of the ulna head is undertaken to access the sigmoid notch but it is important at this point not to take off too much of the ulna – the correct level of section is just proximal to the articulating joint surface of the ulna head. If the TFCC is found intact then it is worth leaving this *in situ*. The interosseous membrane is sharp dissected off the radius for about 8 cm and the ulna may then be displaced palmarwards to allow access to the sigmoid notch of the radius. A radial trial plate is then fitted with a minimum of 3 mm of sigmoid notch lying distal to the end of the trial. The trial should lie cleanly in the sagittal plane and if needed the palmar lip of the sigmoid notch may need burring flat to achieve this. The trial plate is secured with K-wires and the position checked on image intensifier, ensuring the trial plate is centralized on the image to avoid misleading parallax from the image intensifier. The radial plate has an oval screw hole and the first screw is now placed followed by the drilling for the distal peg. The definitive radial plate can now be applied and the remaining 3.5 mm screws sited. Further images are obtained at this stage as it is not possible to adjust screw length on the radius plate once the ulna component is sited.

Attention is now turned to the ulna and, in the forearm pronated position, a measuring device is fitted to the hemi-socket of the seated radial component and the ulna juxtaposed to the stem of the measuring device. The correct level of section of the ulna stump is then just proximal to the distal rim on the stem of the measuring device [Fig. 17.8b]. Following section a 1.6 mm K-wire is inserted into the medullary canal to act as a guide for the reaming of the ulna. The medullary canal is reamed and broached with care taken to avoid heating the bone. The ulna component is introduced and seated by tapping on a plastic impactor. An ultra high molecular weight polythene ball is placed over the distal peg on the prosthetic stem and finally the cover that constitutes the other half of the hemi-socket of the radial component is secured in position with two small screws [Fig. 17.8b]. The complete construct is once more checked under image intensifier and the wound closed by standard technique.

A soft bulky dressing is applied and the patient mobilizes from surgery.

Pearls

For the APTIS prosthesis release the entire length of the ECU from the sixth dorsal compartment to avoid distal ECU attrition on the underlying prosthesis

Use PA, lateral and 30° oblique views to ensure proper alignment of plate and screws on radius

Amongst the more typical indications for replacement of the ulna head the ATPIS prosthesis has been used in Madelung's deformity of the wrist to correct persisting painful stiffness of the DRUJ following alignment procedures for the radiocarpal joint [37].

Good medium term results have been reported for the APTIS prosthesis in a series of 31 patients at mean follow up of 5.9 (minimum 4) years. Twenty-six of 31 patients returned to their previous occupation/activities and a further four patients returned to their previous job with a weight restriction in place. Thirteen patients were able to lift up to 9 kg in the forearm neutral position and 7 kg in the fully pronated position. All but two of the patients could lift a minimum of 2 kg load and the two exceptions had pre-existing CRPS Type 1 limiting the outcome in these cases. Grip strength improved from mean of 10–24 kg. Visual analogue pain scales improved from 4.2/5 preoperatively to 1/5 post-operation. Mean range of motion in pronation was 79° and mean supination 72° [36]. Results from one of the author's own cases is illustrated [Fig. 17.9a–d].

2-Part DRUJ Prosthesis

More recently another total DRUJ prosthesis has been designed [38] comprising 2 parts with a chromium cobalt ulna stem (ceramic coating distally) that pistons through a radial component that comprises a ring with a peg that goes into the distal radius and is also made of chromium cobalt. The ring of the radial component lies in the coronal plane and receives the distal end of the ulna stem. Bony fixation is obtained by means of the hydoxyapatite coating on the prosthesis via osseointegration.

Out of 19 clinical cases that were mainly performed for salvage of previous Darrach's and Sauvé-Kapandji procedures 5 cases of the earlier prototype failed, typically at 1 year post-implant. There were subsequent failures in 12 of the remaining cases. In those prostheses that were retained there was some improvement in grip strength and some reduction in pain but patients were not pain free in line with other authors' experience after prosthetic replacement arthroplasty. Where the prosthesis was retained it does appear stability was achieved. Longer terms results from the authors of their most recent version of the prosthesis will be of interest.

Pitfalls

The dorsal sensory branch of the ulna nerve passes distally within the access incisions likely to be used for DRUJ arthroplasty. Great care must be taken to identify and preserve the nerve

Ensure radiographs centre the DRUJ on the image intensifier to avoid parallax and misjudgement on the level of resection of ulna head

Soft tissue stabilisation procedures that fail to stabilise in all positions of forearm rotation are likely to fail over time i.e. all non-anatomical reconstructions and tethers

Avoid use of a single component (ulna head only) prosthesis where the soft tissue stabilisers of the DRUJ are already compromised and unreconstructable

Use of a stem that is narrower than the medullary canal of the ulna may lead to long term remodelling of the ulna which then widens in response to the movement of the ulna stem (resolve by placement of a new wider stemmed prosthesis with bone graft or bone cement to achieve integration/stabilisation of the prosthesis

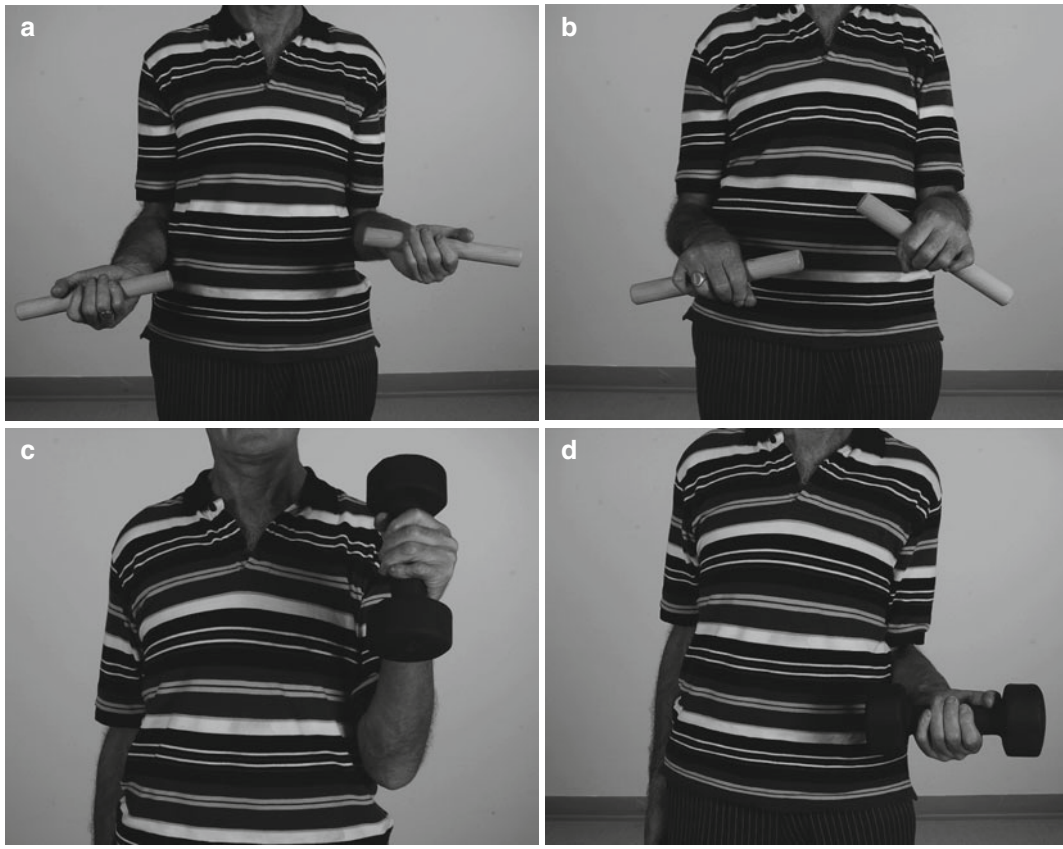

Fig. 17.9 Patient having APTIS joint on left side 2 years previously for osteoarthritis of the DRUJ. (**a**) and (**b**) Demonstrate the range of motion achieved. (**c**) The patient readily lifts a 10 kg weight with the forearm in midrotation. (**d**) The 10 kg weight can also be supported in the supinated forearm position. (Author's case: patient permission to publish)

Results

The results for individual prostheses are detailed in each relevant section but there are some studies that have looked at specific implant-related phenomena. One of these is the incidence of resorption and erosion around the head of metallic ulna head implants [39] In a series of 17 ulna head implants performed for a variety of indications that were followed prospectively for an average follow up of 36 months all patients had bone resorption around the collar of the implant that increased through the first postoperative year and then remained stable thereafter. Overall, patients improved in terms of pain, function, arc of forearm rotation and demonstrated modest improvement in grip strength. The longer-term consequences of this pattern of bone erosion have yet to be determined.

While some ulna head prostheses can restore ability to lift modest weight e.g. Eclypse™ 4 kg, this does depend on the stability achieved in the particular patient. Some designs do not restore the ability to lift even minor weights, albeit that forearm rotation has been restored and pain reduced. Grip and lifting strength are not always mentioned but are good indicators of the efficacy of the prosthesis. It is important that such measurements are included in future reports.

Most interventions for replacement of part of the DRUJ do not restore normal function but can nonetheless produce useful clinical improvement. Thought should be given when entertaining prosthetic replacement as to what will have to be done next as and when the particular prosthesis used fails. Typical prosthetic complications

are reported including loosening, continuing pain and weakness of grip. The incidence of infection is in line with what would normally be expected from implant arthroplasty. Patients with pre-existing CRPS appear not to do particularly well after prosthetic replacement arthroplasty [35].

Salvage of the Failed Prosthetic Replacement Arthroplasty

Any prosthesis can only buy time against the moment of implant failure and what we have to understand is whether we can use a second version of the same implant at that time or indeed go on to use another type of implant. Three revision cases using Herbert-Martin and Avanta prostheses where revision implant arthroplasty had been undertaken still having good/excellent outcomes (modified Mayo wrist score) were reported [22]. One study has looked at the disassembly force required to remove an uncemented ulna head implant as measured by the Morse taper decoupling strength on an Instron machine (the cobalt chrome head-cobalt chrome stem (closest correlation), cobalt chrome head-titanium stem couple and ceramic head-titanium couple (representing the three most commonly used ulna head prostheses) and found that the decoupling force required correlated closely with the amount of force that was needed to put the implant in the first place [40].

The decision to replace one ulna head arthroplasty by another will depend on the perceived reasons for the original device failure. Where it is clear that the soft tissues are inadequate and cannot be restored with consequent persisting instability then a further ulna head replacement is not indicated. Partial resurfacing of the ulna head may be treatable in theory by conversion to a complete ulna head although reports on this sequence are sparse. Ulna head-only arthroplasty may be salvageable by conversion to total replacement arthroplasty. With respect to the APTIS™ prosthesis a new ulna stem has been replaced successfully following high-energy trauma with breakage of the original stem. To date there are no reports of the radius plate component loosening. Infection complicating the insertion of an APTIS™ prosthesis has been successfully treated in one report [35] following primary implant removal with antibiotic beads placement for a 3 month interval prior to successfully inserting a second prosthesis.

Notwithstanding the decision to proceed to partial or total prosthetic replacement, there will come a time when there is a prosthetic failure without the possibility of further prosthetic replacement. Such circumstances will include infection, loss of adequate bone stock, traumatic injury and device failure over time. In these circumstances it will become necessary to fall back on the traditional salvage options of wide excision of the ulna or creation of a one-bone forearm accepting the limited clinical outcomes that will result from the same.

Conclusions

Our knowledge is at a point where the treating surgeon faces a dilemma. On reviewing the evidence base not all prosthetic replacements are successful and we know that excisional techniques on the ulna head also have significant failure rates. There appears to be little justification for putting patients through Darrach's procedure and its modifications as a prelude to prosthetic replacement. Where we do need to change thinking is to advocate the primary use of implants rather than resectional procedures for treatment of DRUJ disorders. DRUJ replacement arthroplasty is coming of age and represents a genuine treatment option for disorders of this joint that cannot otherwise be treated by operations that respect anatomical principles and restore anatomical structures. There are a number of designs for replacement of the ulna head with some favourable and other less favourable outcomes reported. Larger series are needed on some of these prostheses to assess the utility of the particular prosthetic design. No doubt there will be further designs described in future.

It is tempting to offer an algorithm for DRUJ replacement arthroplasty in which several prosthesis feature with indications as suggested by their designers. This would probably prove a mistake and it will be better to gain familiarity with those prostheses that are clearly designed

with reference to the underlying biomechanics of this complex joint. It is clear that the limitation on use of the various ingenious ulna head prostheses is the state of the soft tissues. It is important to understand that the different prostheses are not interchangeable with respect to the indications for their use. With pre-existing instability it is necessary to secure or anchor the prosthesis and to reconstruct the soft tissues at the same time. Such anchoring/soft tissue plication may not hold in the medium to long term as several authors have attested and recurrent instability as the sutures give way appears to be a clinical problem. Arthrosis secondary to instability may best be treated by total replacement arthroplasty in the first instance.

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