Congenital and Developmental Abnormalities

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Abstract

Congenital hand differences (CHD) have been estimated to occur in 10% of children born with congenital abnormalities. CHD can be classified according to their pre-dominant abnormality using the Swanson classification. Failure of differentiation represents the most common group. Associated (nonlimb) abnormalities are common and it is important to identify those CHD requiring systemic evaluation. Radiology is important in the diagnosis and management of CHD with plain films providing the mainstay of imaging postnatally. The management of CHD should be within a multi-disciplinary environment.

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1 Introduction

Of the 1–2% of newborns born with congenital defects, 10% are born with upper limb malformations (McCarroll 2000). The incidence is higher in boys, with pre and post-term births, with multiple pregnancies and with increased maternal age (Giele et al. 2001). Congenital hand abnormalities are now commonly described as congenital hand differences (CHD). The majority (60%) of CHD has an unknown etiology but genetic factors may be attributable in 20% and environmental teratogens in 20% (Netscher and Baumholtz 2007). The embryologic limb bud can be seen at 4 weeks after fertilization, and the existing fetal limb structures continue to grow and develop for 8 weeks after fertilization. Most CHD are thought to arise during this 4-week interval (Kozin 2003).

CHD vary widely in their functional and aesthetic implications. Associated (nonlimb) abnormalities are also common, up to 26.7% in some series (De Smet 2002). It is important to identify those CHD which require further systemic evaluation and to differentiate them from isolated CHD. For example, radial deficiency occurs commonly in a number of syndromes such as the VACTERL association (vertebral abnormalities, ano-rectal and cardiac abnormalities, tracheo-oesophageal fistula, oesophageal atresia, renal defects, radial dysplasia and lower limb abnormalities).

Management must always be conducted in a multidisciplinary environment, involving surgeons, physiotherapists, orthotists, paediatricians, prosthetists, psychologists, anaesthetists, geneticists and radiologists. The purpose of this review is to provide an illustrated classification of congenital hand differences for radiologists, which will also be of interest to the other members of the multi-disciplinary team.

2 Imaging

CHD may be demonstrated by antenatal ultrasound particularly late in the first trimester and in the middle of the second trimester of pregnancy (Rypens et al. 2006) (Fig. 1). Three-dimensional ultrasound may be helpful in defining the abnormality more clearly (Ploeckinger-Ulm et al. 1996). Prenatal classification and characterization may help to identify fetuses that would benefit from more complete prenatal cardiac and karyotypic workup.

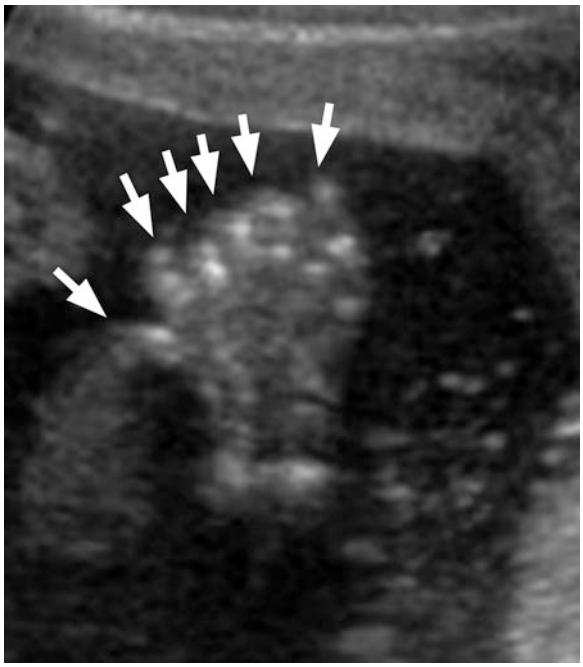


Fig. 1 2D Ultrasound image of a fetal hand of a fetus in the second trimester with trisomy 13 and post-axial polydactyly (Image courtesy of Dr Françoise Rypens)

Postnatally, plain films provide the mainstay of imaging, with ultrasound and magnetic resonance imaging (MRI) providing useful additional characterization in a minority of cases, principally in the clarification of the nature of masses and other causes of overgrowth.

3 Classification

The most widely accepted classification of CHD was proposed by Frantz and O'Rahilly and presented by Swanson (Frantz and O'Rahilly 1961; Swanson 1976). This was subsequently modified by the International Federation of Societies for Surgery of the Hand (IFSSH) in 1983 (Swanson et al. 1983), and further modified by the Japanese Society of Surgery of the Hand (JSSH) in 2000. The major categories are listed in Table 1 and are subsequently discussed.

Table 1 Abbreviated embryologic classification of CHD (adapted from Swanson 1976)

1	Failure of formation
2	Failure of differentiation
3	Duplication
4	Overgrowth
5	Undergrowth
6	Constriction ring syndrome
7	Generalised skeletal abnormalities

The seven categories differentiate the CHD according to the predominant abnormality, and further define the abnormality according to the nature of the embryonic failure during development. Group II (failure of differentiation) represents the most common group.

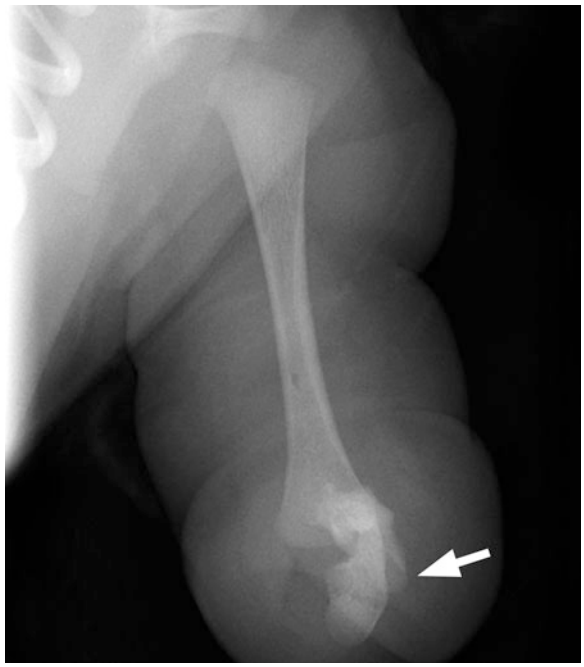


Fig. 2 Commonest level of transverse growth arrest at proximal radius and ulna (*arrow*)

4 Failure of Formation

Failure of formation of parts can be divided into transverse and longitudinal subgroups. Transverse deficiencies occur when growth stops abruptly, resulting in a "congenital amputation" and a shortened or truncated limb. Longitudinal deficiencies occur when structures are hypoplastic or absent along a longitudinal axis of the limb, resulting in abnormal formation and function biased down one 'side' of the limb (either the radial or ulnar sides, or 'centrally' between the two) (Kozin et al. 2004). In reality, most CHD within this group are mixed to some degree.

4.1 Transverse

This is defined according to the bone segment at which the growth arrest occurs (Van Heest 1996). The most common transverse deficiency occurs at the junction of the proximal and middle thirds of the forearm (Fig. 2).

These anomalies are usually unilateral, sporadic, and are rarely associated with other anomalies. Rudimentary "nubbins" or dimpling may be found at the distal end of the congenital amputation (Kozin 2003). This should not be confused with Constriction ring syndrome which may also manifest as an 'amputation' but requires the presence of a constriction ring either affecting the involved extremity or elsewhere (Wiedrich 1998).

4.2 Longitudinal

In this group the radial, central or ulnar side of the limb is abnormal. They may very rarely be characterized by absence of one or more 'segments' ('forearm' versus 'upper arm' versus 'both') of the upper limb (phocomelia), such as those defects attributed to maternal exposure to thalidomide. For example, when both segments are missing ('complete phocomelia'), the hand is attached directly to the shoulder.

4.2.1 Radial

Radial dysplasia is the most common longitudinal deficiency and occurs when structures on the radial side of the arm fail to develop properly. It is frequently bilateral (40–60%) with a male:female ratio of 3:2 (Van Heest 1996). It has been described as "an abnormal hand joined to a poor limb by a bad wrist" (Flatt 1994) (Fig. 3).

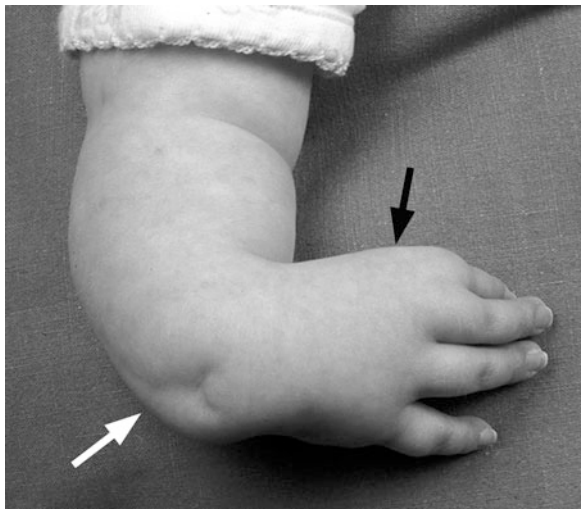


Fig. 3 Radial dysplasia displays an array of upper limb differences, the most striking of which are usually the radially deviated wrist (*white arrow*), absent thumb (*black arrow*), shortened forearm and stiff elbow

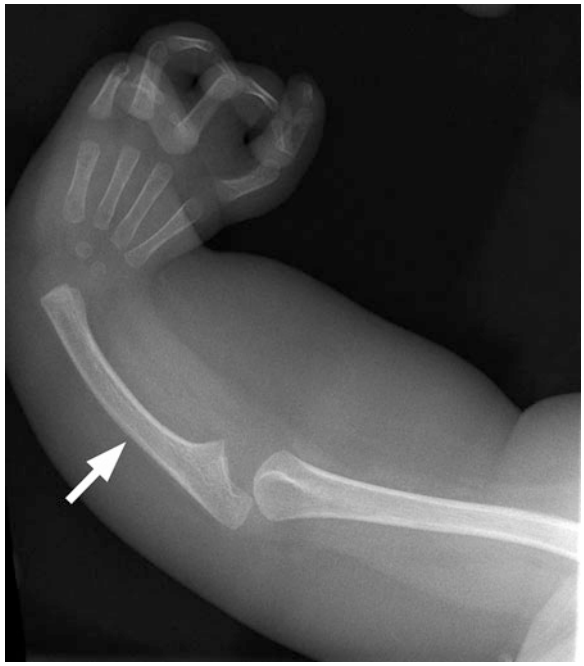


Fig. 4 Radial dysplasia has a spectrum of abnormalities of the radius, the commonest being complete absence of the radius. In such cases the ulna (*arrow*) is relatively hypoplastic also

Radial deficiency can range from mild hypoplasia to complete absence of the radius and thumb (the commonest type) (Fig. 4). The child presents with a hand that is radially deviated at the wrist. In the hand, the thumb is almost always affected and the fingers frequently have limited motion and function, the severity of which decreases from the radial to the ulnar side (James et al. 2004). The ulna is also usually shortened and bowed and may need later osteotomy (Fig. 5a, b).

A classification scheme has been described by Bayne and Klug for radial deficiency with proposed modifications by James et al. (Bayne and Klug 1987; James et al. 1999). For the purposes of imaging, it is useful to divide the deficiency into components and describe the characteristics of each component (James et al. 1999; Kozin 2003).

1. Thumb:
   - I. absent
   - II. hypoplastic
2. Carpal bones:
   - I. normal
   - II. absent
   - III. hypoplastic
   - IV. coalition

(Nb: Ossification centers of the carpal bones appear at a variety of ages ranging from 2 months for the capitate and hamate, to 4 years for the scaphoid, trapezium and trapezoid.)

3. Distal radius
   - I. normal
   - II. >2 mm shorter than ulna
   - III. absent physis
   - IV. absent
4. Proximal radius:
   - I. normal
   - II. radioulnar synostosis
   - III. radial head dislocation
   - IV. hypoplastic
   - V. absent

The elbows are commonly stiff, and the abnormal features frequently extend further proximally (Fig. 6). Recently Goldfarb and colleagues (Goldfarb et al. 2005) proposed a further subtype of proximal radial longitudinal deficiency, involving an abnormal glenoid, an absent proximal humerus (with the distal humerus articulating with the ulna) and radial sided hand abnormalities.

Treatment involves straightening (centralization) of the wrist, currently achieved by initial distraction (Fig. 7a) followed by centralization (Fig. 7b) (both of which require perioperative fluoroscopic

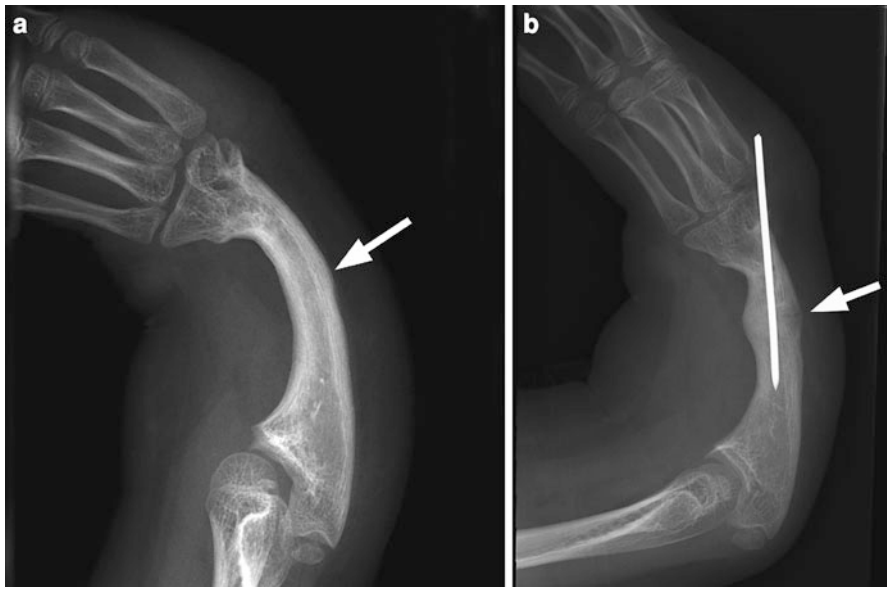


Fig. 5 **a** and **b** In radial dysplasia the ulna is frequently bowed, and may require correction as in this case which had been centralized 10 years previously. The osteotomy site (*arrow*) is secured with a large K-wire, removed at six weeks **a** pre-op **b** post op

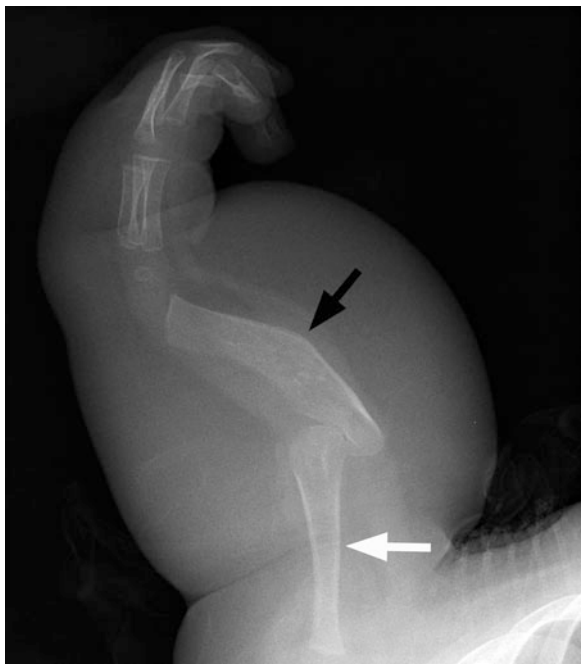


Fig. 6 Radial dysplasia with severe hypoplasia of the ulna (*black arrow*) and humerus (*white arrow*), both of which are shorter than the hand itself

screening), and later pollicisation (shortening and rotation of the index ray on its neurovascular pedicle to the position of the thumb, with tendon and intrinsic muscle rebalancing to create a 'thumb' from the index finger).

Importantly, radial longitudinal deficiencies often present as part of a syndrome. Associations include hematological conditions (e.g., thrombocytopenia-absent radius syndrome and Fanconi's anaemia), cardiac anomalies (e.g., Holt Oram syndrome), the VACTERL syndrome (see Sect. 1) and craniofacial syndromes (e.g., Nager and Duane syndromes) (Lourie and Lins 1998).

Children with radial longitudinal deficiencies should therefore have a thorough multi-disciplinary evaluation at the time of presentation to exclude associated abnormalities, understanding that some of these may not be immediately apparent (e.g., Fanconi's anaemia has a median age of onset of seven years of age) (Kozin 2003).

4.2.2 Central

This involves deficiencies of the central part of the hand and is also known descriptively as 'cleft hand', with traditional terms such as 'split hand' or 'lobster claw hand' having been rightly dropped because of their potentially emotive nature. Two types of cleft hand have been described, formerly known as 'typical' and 'atypical'. Although they may appear similar morphologically, they both have major clinical differences and probably different embryological derivations (Miura and Suzuki 1984; Ogino 1990a).

The typical cleft hand (now known as 'true' cleft hand) has a deep V-shaped defect (Fig. 8a, b). Most commonly the phalanges are missing. The metacarpals may be absent or malpositioned (Fig. 9), but are

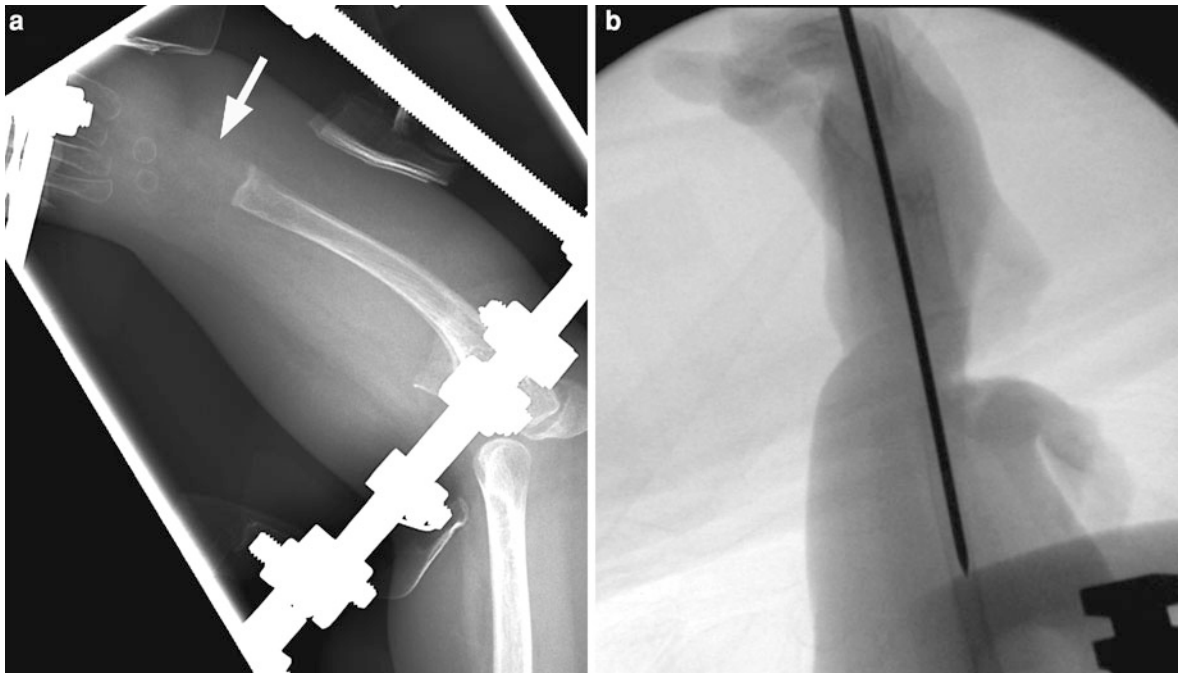


Fig. 7 Treatment of the radially deviated wrist is best achieved by (**a**) initial gradual lengthening of the soft tissues using an external distraction frame to bring the hand in line (*arrow*) over the end of the ulna, followed by (**b**) removal of the frame and formal centralization, with tendon transfers and temporary axial K-wire fixation, requiring intraoperative fluoroscopy

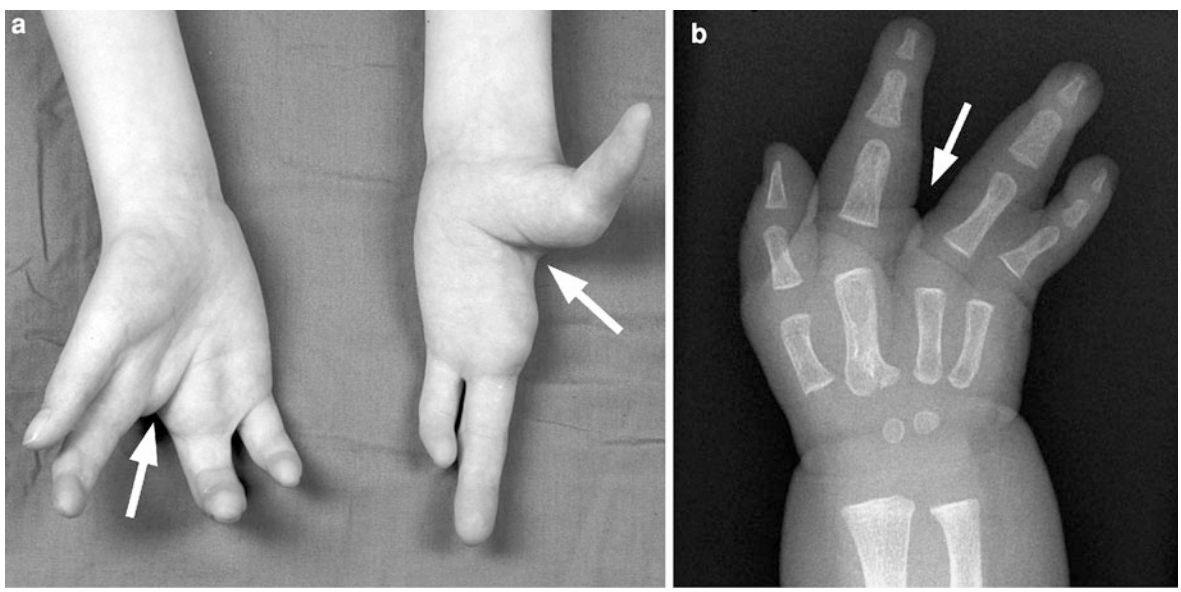


Fig. 8 **a** True cleft hand is usually bilateral, with V-shaped clefts (*arrows*), though the severity may differ between the two hands as in this case. **b** True clefts are usually centered on an absent third ray (*arrow*)

rarely hypoplastic. Joints at the site of the cleft can be very complex (Fig. 10), (Barsky 1964; Ogino 1990). This type of deficiency is often bilateral, and is usually inherited. There may be syndactyly of the ring-small or thumb-index web space, and associated foot involvement is common (Fig. 11). True central

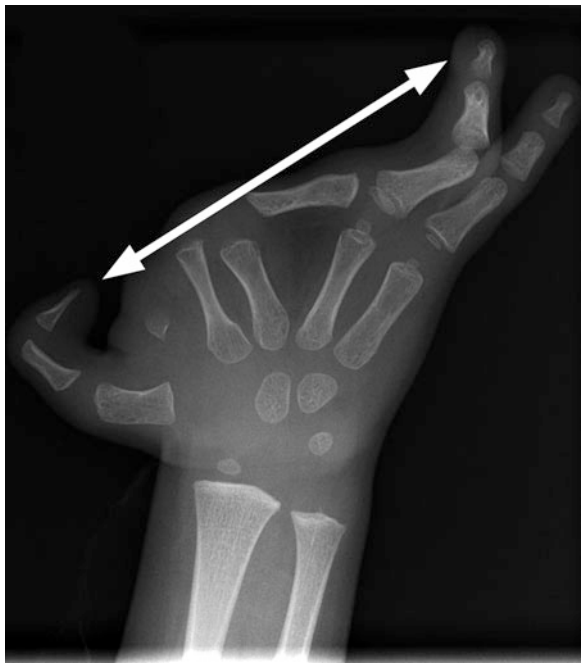


Fig. 9 In true cleft hand a wide spectrum of severity is seen, in which transverse metacarpals can cause progressive widening of the cleft (*arrow*)

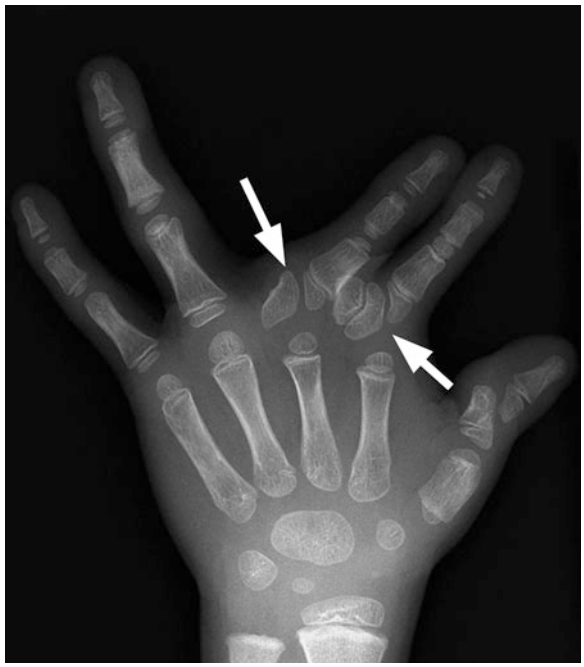


Fig. 10 This unusual variant demonstrates the complex joints (*arrows*) which can exist in cleft hands, requiring vigilance during surgical corrections

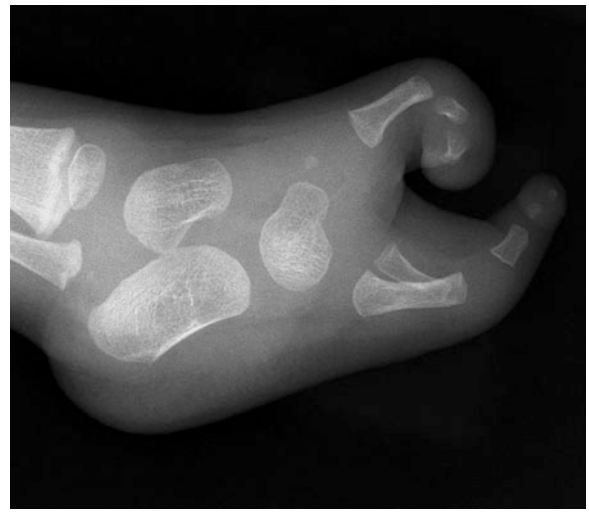


Fig. 11 Foot involvement is common in true cleft hand patients

dysplasia is also associated with cleft lip and palate but not Poland syndrome (Miura and Suzuki 1984; Ogino 1990a).

Atypical cleft hand has a shallower, U-shaped central defect that involves the central three digits (index, long and ring) (Fig. 12a), (Kozin 2003). It is a form of symbrachydactyly (shortened and webbed digits) that can vary in degree from the 'short finger type' (Fig. 12b), to absence of the central three digits (Fig. 12a), to a monodactylous type (thumb only, no fingers) (Fig. 12c, d), to complete absence of all digits, with only very rudimentary 'nubbins' (peromelic type) (Fig. 12e).

Atypical cleft hand is usually unilateral and sporadic. It is not associated with lower limb malformations or cleft lip and palate (Miura and Suzuki 1984; Ogino 1990a) but is associated with Poland syndrome (Fig. 13) (Ireland et al. 1976). The degree of hand deficiency in Poland syndrome does not correlate with that of the chest wall deformity, therefore clinical evaluation of the chest wall is required with all cases of symbrachydactyly (Kozin 2003).

4.2.3 Ulnar

This represents a spectrum of abnormalities along the ulnar surface of the upper limb. It is far less common than radial and central longitudinal deficiency (Miller et al. 1986; Schmidt and Neufield 1988) (Figs. 14a, b; 15; 16a, b).

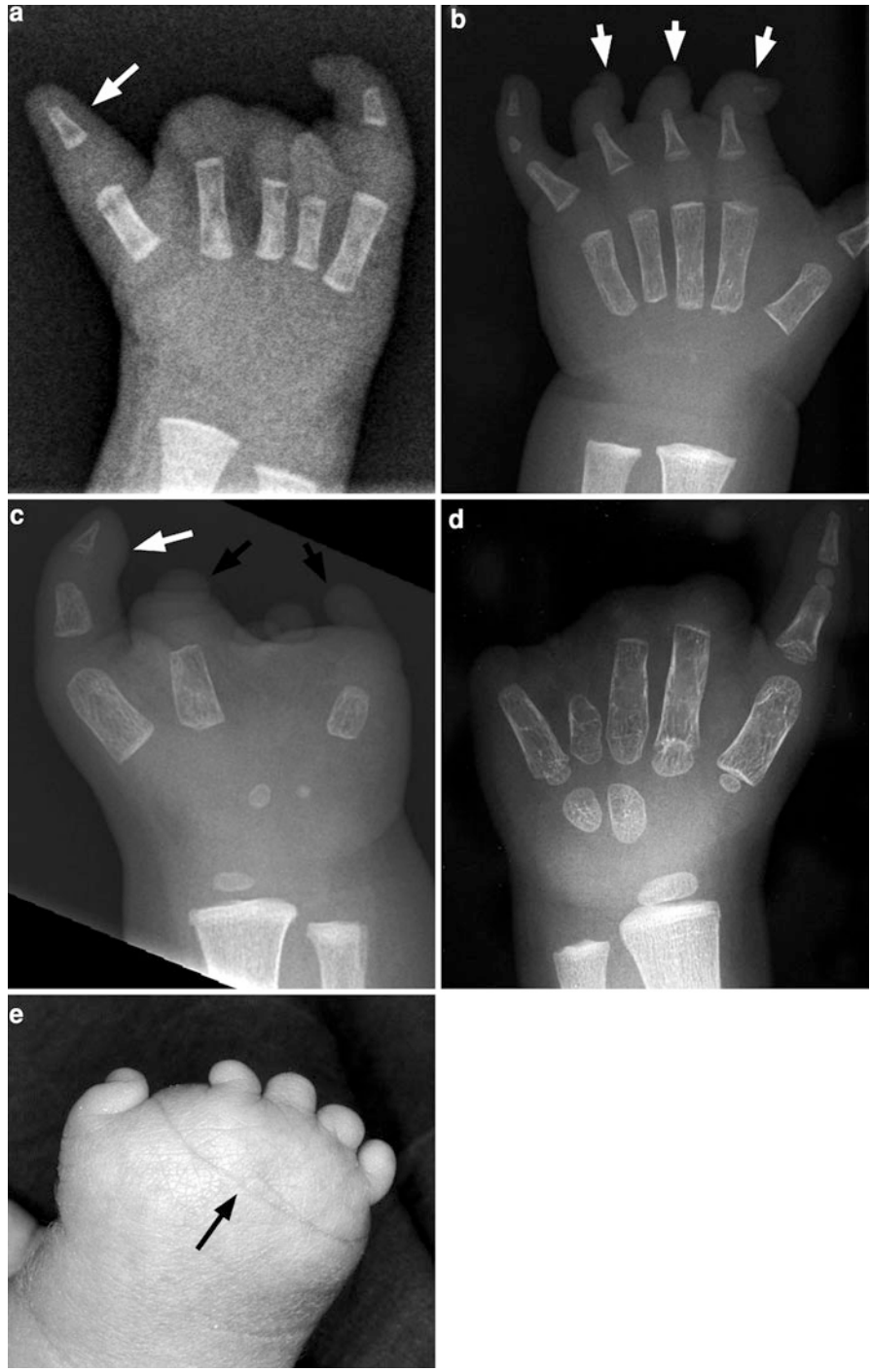


Fig. 12 Symbrachydactyly subtypes **a** Atypical cleft hand also known as'U-shaped cleft hand type' with absence of the central three digits, leaving only the thumb (*arrow*) and little finger, both of which are hypoplastic in this case. **b** Short finger type, with proximal phalanges and reasonable soft tissue envelopes distally (*arrows*) of the central three digits, which could be amenable to augmentation with bone grafts. **c** Monodactylous type, in which only the thumb remains (*white arrow*), but potentially useful soft tissue envelopes to the index and little fingers (*black arrows*). **d** Monodactylous type, with only the thumb, and no distal soft tissue. Function would be greatly improved by free toe transfers, or even custom made prostheses. **e** Peromelic type, with severe attenuation at metacarpal level (*arrow* denotes proximal palmar crease)

Radial sided hand anomalies may co-exist with ulnar longitudinal deficiency and hand abnormalities are present in 68–100% of cases (Miller et al. 1986). Ulnar longitudinal deficiency may be associated with other musculoskeletal anomalies (e.g., scoliosis), but is not associated with the visceral and haemopoetic disorders seen in radial deficiencies.

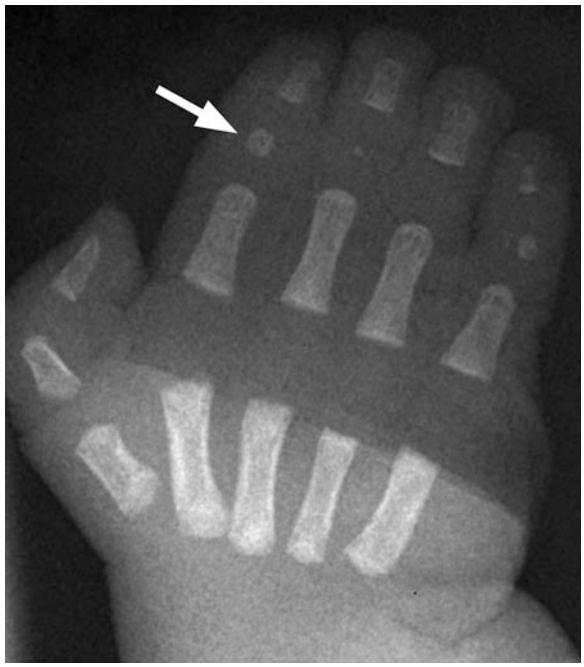


Fig. 13 Poland's syndrome with shortened digits due mostly to attenuation or absence of the middle phalanges (*arrow*), syndactylies and global hypoplasia of the hand, in association with other upper limb and chest deformities

5 Failure of Differentiation

5.1 Syndactyly

Syndactyly is one of the most common hand anomalies and results from incomplete separation of two or more adjacent digits. It has an incidence of 1:2,000 live births (Flatt 1994). The anomaly can be sporadic or familial (a family history is present in 10–40% of cases) and it may be associated with other syndromes including Poland syndrome and craniosynostoses such as Apert's syndrome (Fig. 17) (Kozin 2001).

It is classified as 'complete' if it reaches the distal interphalangeal joint (Fig. 16a), 'incomplete' if proximal to this joint, 'simple' if it involves only skin and soft tissue, and 'complex' if there is a bony fusion (usually distal) (Fig. 16b) (Van Heest 1996). Any combination of web spaces may be involved, but the commonest is an isolated third web-space involvement (between the ring and middle fingers). Syndactyly can also be classified as 'complicated' if it occurs in association with other abnormalities in the affected area, such as duplication (Fig. 18). Syndactyly should be differentiated from.'Acrosyndactyly' (distal fusion with a proximal fenestration) which occurs in association with Constriction ring syndrome (see Sect. 9).

Radiographs may indicate that separation of syndactyly is not always appropriate (Fig. 19a, b).

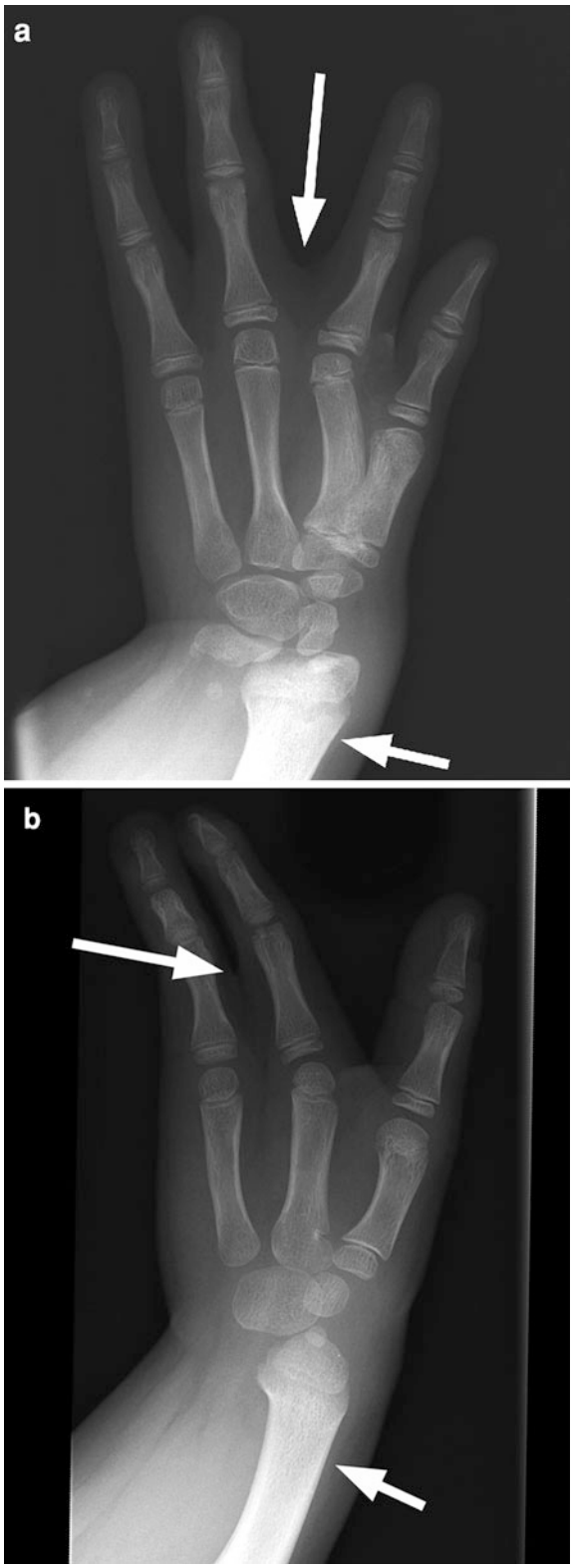
5.2 Arthrogryposis

This term has been used to describe conditions that present with congenital joint contractures (Mennen et al. 2005). This clinical entity has also been described by Sheldon as amyoplasia (Sheldon 1932) and most affected children (84%) present with involvement of all four limbs (Sells et al. 1996).

The contractures are thought to be the end result of decreased intra-uterine movement by the fetus which may occur because of neuropathies, myopathies, abnormal connective tissue or decreased intra-uterine space. It is a sporadic condition with an incidence of 1 in 10,000 live births (Hall 1997) (Fig. 20a, b).

5.3 Camptodactyly

This is a painless flexion contracture of the proximal interphalangeal joint (PIPJ) that usually affects the little finger and may be progressive. It is not related to trauma and is estimated to affect just under 1% of the population to some degree (Smith and Kaplan 1968) (Fig. 21). A classification system has been described by Benson with three main types (Benson and Kaplan 1994). Type I presents in infancy and is frequently confined to one or both little fingers, with approximately equal sex incidence. Type II presents in adolescence with females being affected more commonly than males. The deformity worsens with the adolescent growth spurt. Type III is more severe and usually affects multiple digits in both hands (Koman et al. 1990), and may be associated with chromosomal disorders and skeletal abnormalities. Most structures within the finger have been implicated, although abnormalities of the intrinsic lumbrical muscles of the superficial flexor tendon are not uncommon operative findings.



◀**Fig. 14** **a** and **b** Ulnar dysplasia has deficiencies of the ulnar side of the upper limb, with partial or complete absence of the ulna, commonly with the radius providing a one-bone forearm (*short arrows*), loss of ulnar digits, ulnar deviation at the wrist, and syndactylies (*long arrows*)

Plain radiograph assessment is vital, as the presence of classical bony changes around the joint predicts a very poor surgical outcome. These bony changes include flattening of the head of the proximal phalanx, widening of the base of the middle phalanx, and an indentation at the neck of the proximal phalanx (Ty and James 2009) (Fig. 22).

5.4 Clinodactyly

This refers to an abnormal deviation of a finger in the coronal or radioulnar plane which typically affects the middle phalanx of the little finger. The finger deviation is usually directed radially. Minor angulation of $<$10 degrees is so common that it may be regarded as a normal variant (Ezaki 1999).

Familial clinodactyly with an autosomal dominant pattern of inheritance usually presents bilaterally, and

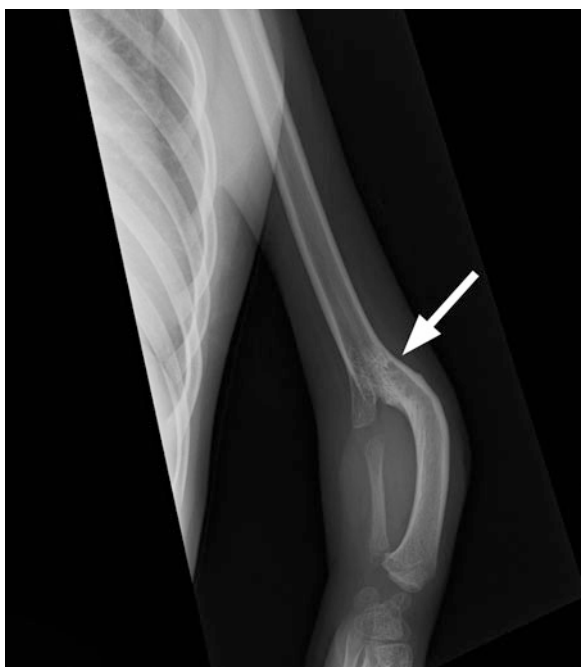


Fig. 15 Radiohumeral synostosis (*arrow*) can be seen in ulnar dysplasia

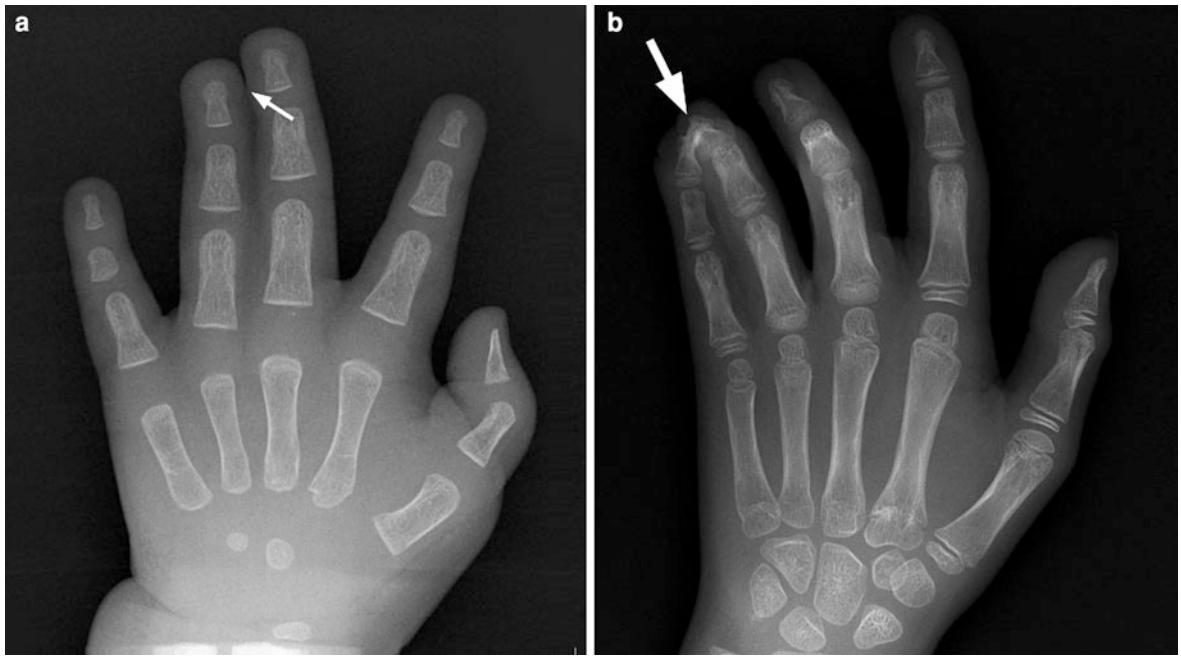


Fig. 16 Syndactyly **a** Simple, complete syndactyly with soft tissue union only, distal-to-distal interphalangeal joint (*arrow*). **b** Complex syndactyly with bony union which is usually at the distal phalangeal level (*arrow*), and requires earlier release than simple syndactyly as bony deformities can occur quickly with growth

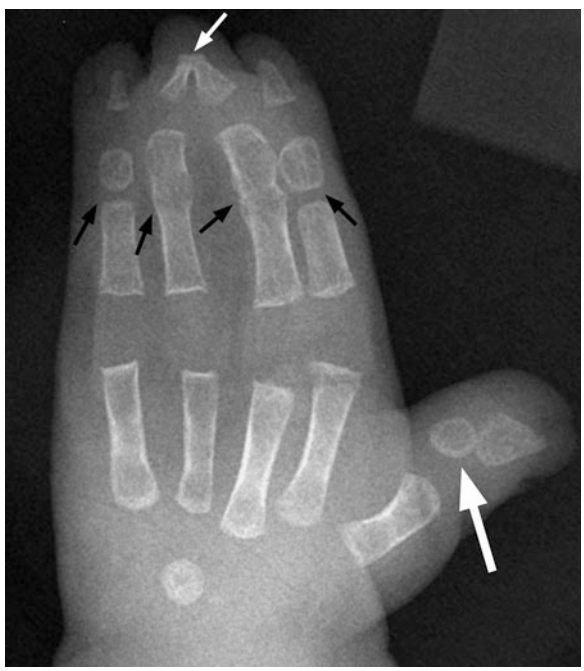


Fig. 17 Apert's hand with complex syndactyly (*short white arrow*), poorly developed or absent proximal interphalangeal joints ('symphalangism' (*black arrows*)), and a shortened deviated thumb caused by a delta phalanx (*large white arrow*)

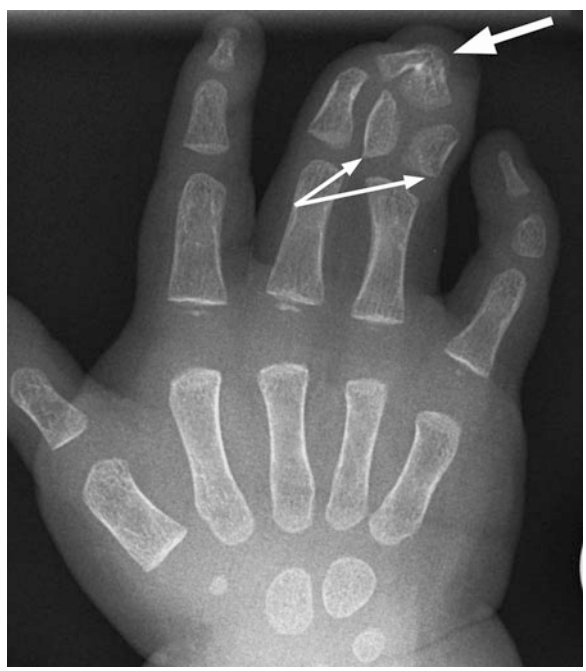


Fig. 18 Complicated syndactyly, with distal bony union (*large arrow*) and unusual duplication of the middle phalanx (*small arrows*)

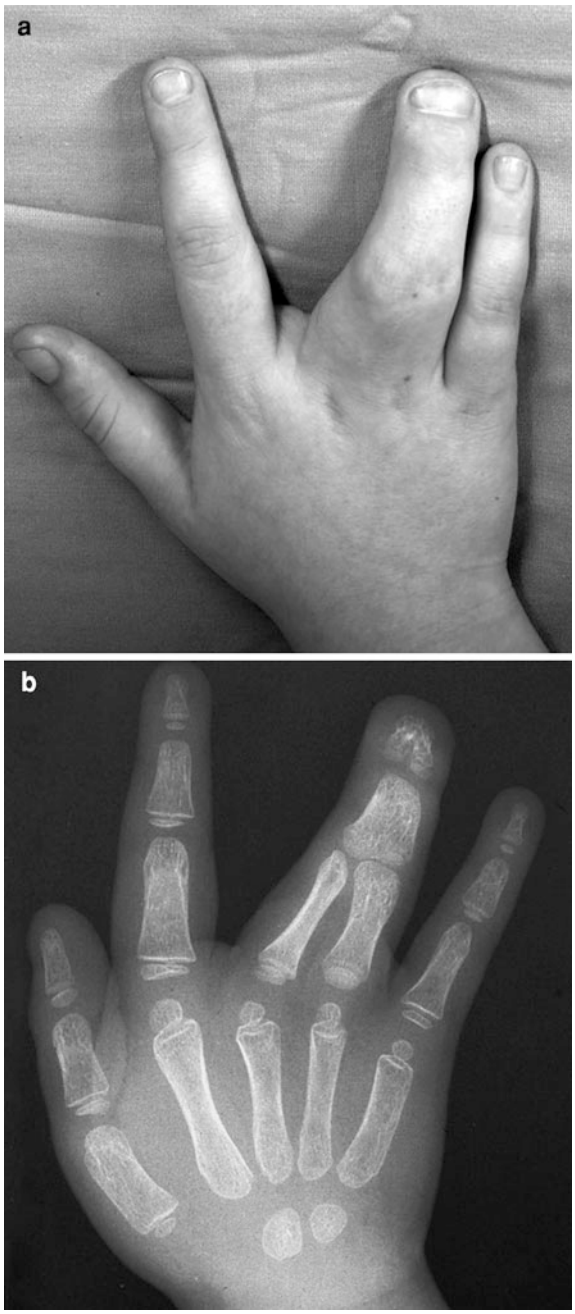


Fig. 19 Syndactyly release is sometimes contraindicated, as X-rays may reveal that the fingers work better together than if they were separated

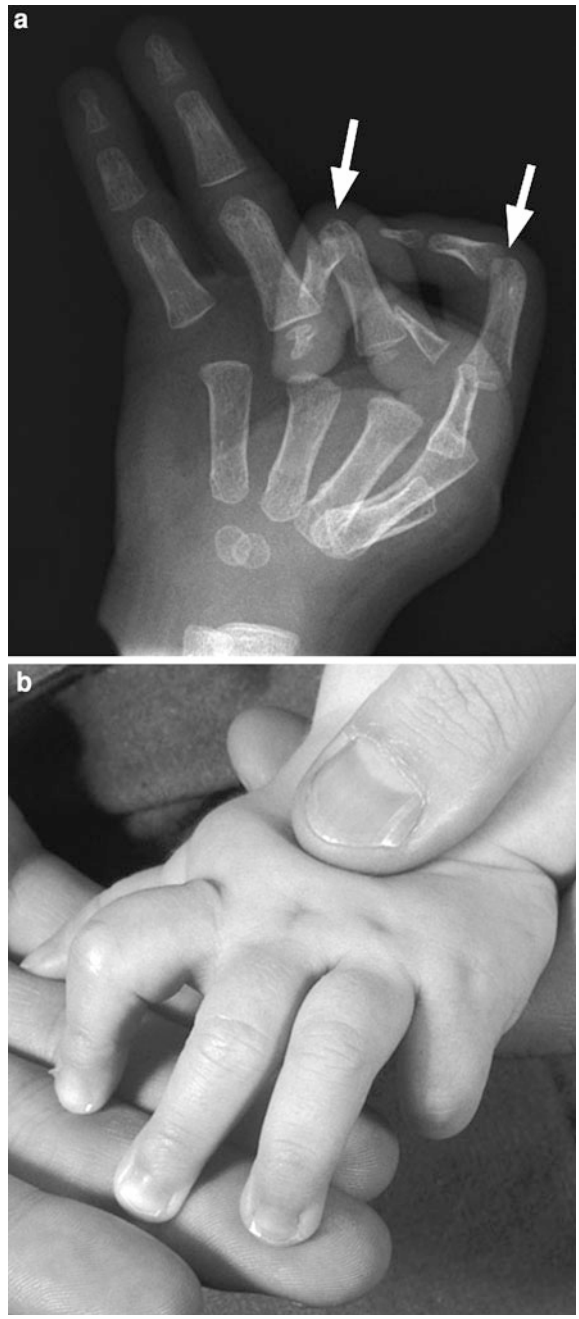


Fig. 20 Arthrogryposis is frequently associated (**a**) with multiple camptodactylies (*arrows*), sometimes affecting all digits (**b**)

is not associated with genetic syndromes. Clinodactyly may also be associated with syndromic and genetic conditions, most notably Down syndrome, in which the prevalence has been estimated at 35–79% (Flatt et al 1994).

The deformity is caused by an abnormally shaped middle phalanx, which may be triangular (a "delta" phalanx) or trapezoidal because of a C-shaped epiphysis (Jones 1964) (Fig. 23). Serial plain radiographs can be useful for objective monitoring of the angle of deviation. If progression occurs despite non-operative treatments

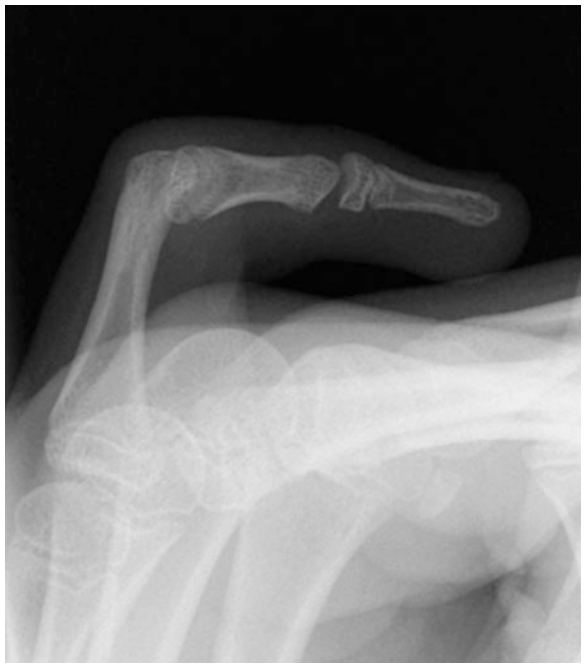


Fig. 21 Camptodactyly most commonly involves the little fingers, with stretching exercises and splintage forming the mainstay of treatment, with surgery often yielding disappointing results

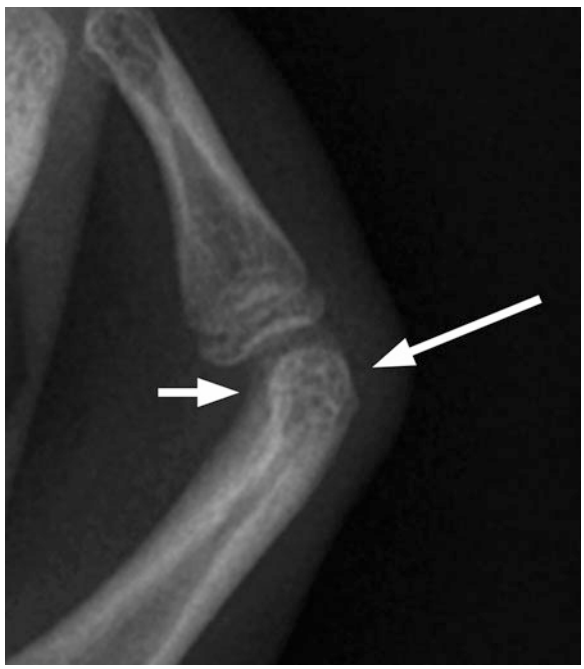


Fig. 22 Bony changes in camptodactyly chisel shaped head of proximal phalanx (*long arrow*), indentation of the neck of the proximal phalanx (*short arrow*), and widening of the base of the middle phalanx augur a poor surgical result

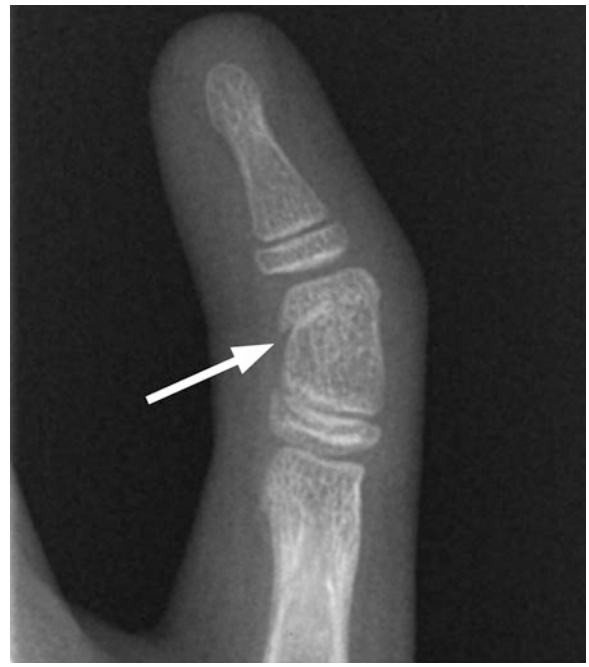


Fig. 23 Clinodactyly showing abnormal C-shaped epiphysis of middle phalanx (*arrow*)

(principally night splintage), surgery in the form of a wedge osteotomy may be required (Fig. 24a, b).

Delta phalanges can occur elsewhere in the hand and are frequently seen in the thumbs of Apert's patients, where they can cause marked deviation (Fig. 25) and often require surgery.

5.5 Congenital Thumb Flexion Deformities

Trigger and clasped thumbs are the two main causes of congenital flexion deformities of the thumb, with flexion, respectively at the interphalangeal and metacarpophalangeal joints. In trigger thumbs, a thickening in the flexor tendon may be felt at the A1 pulley level (clinically at the level of the metacarpophalangeal level).

In clasped thumbs there is a wide spectrum of differences, starting with attenuation then absence of the extensors, and progressing to flexion contractures with global thumb hypoplasias. Clinical evaluation usually suffices, though ultrasound may be useful in assessment of the tendons to aid in treatment planning. Hypoplastic extensors in a clasp thumb may respond to simple splintage whereas aplasias require tendon transfer.

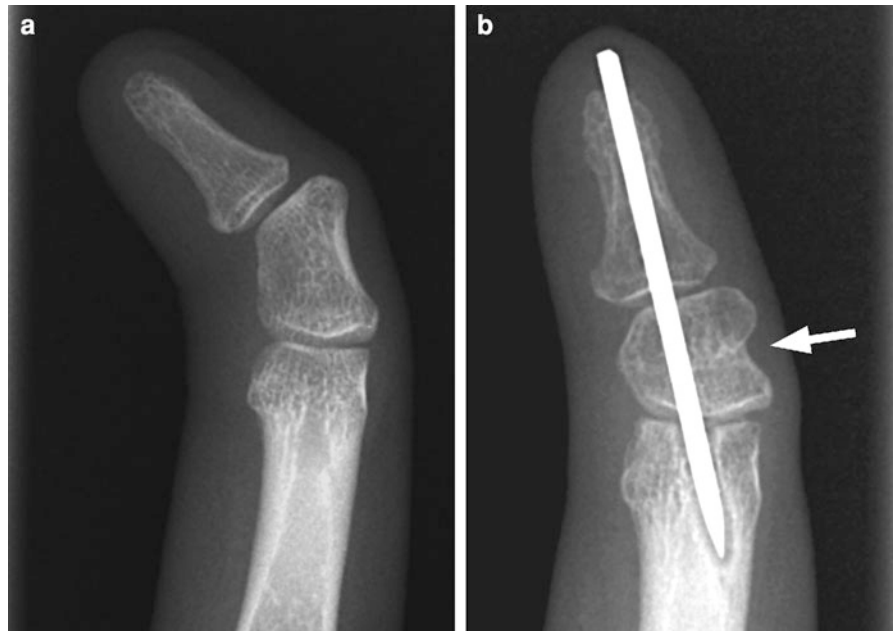


Fig. 24 Clinodactyly showing (**a**) a delta phalanx with fused epiphysis, treated by (**b**) a closing wedge osteotomy (*arrow*) and temporary K-wire

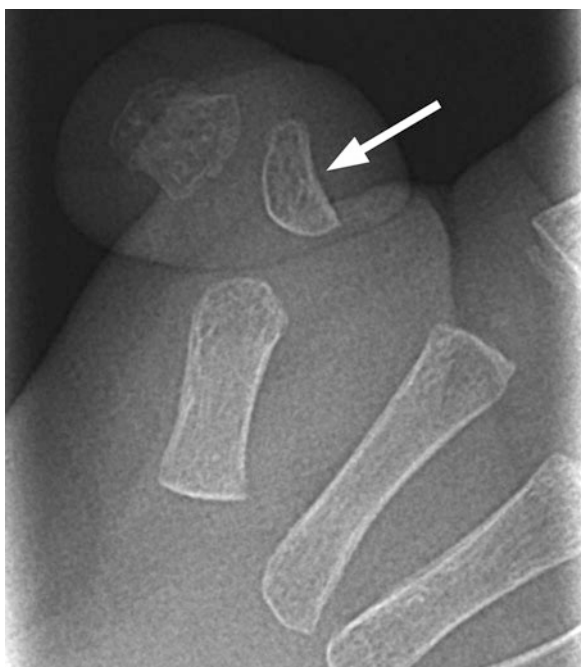


Fig. 25 Apert's thumb delta phalanx (*arrow*), which usually requires surgical correction

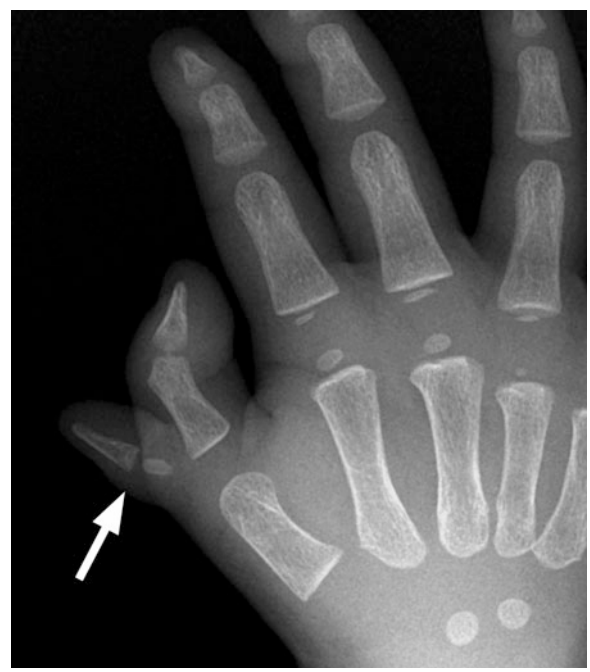


Fig. 26 Rudimentary radial duplicate (*arrow*)

6 Duplication

This probably results from splitting of the original embryonic part rather than a true duplication (Van Heest 1996). Polydactyly may be 'preaxial' (radial i.e., thumb), postaxial (ulnar i.e., little finger) or central.

6.1 Post-Axial Polydactyly

Duplications involving the ulnar aspect of the hand are more common in patients of African descent. The incidence is reported as 1 in 1,339 live births in Caucasians and 1 in 143 live births in African Americans (Watson and Hennrikus 1997).

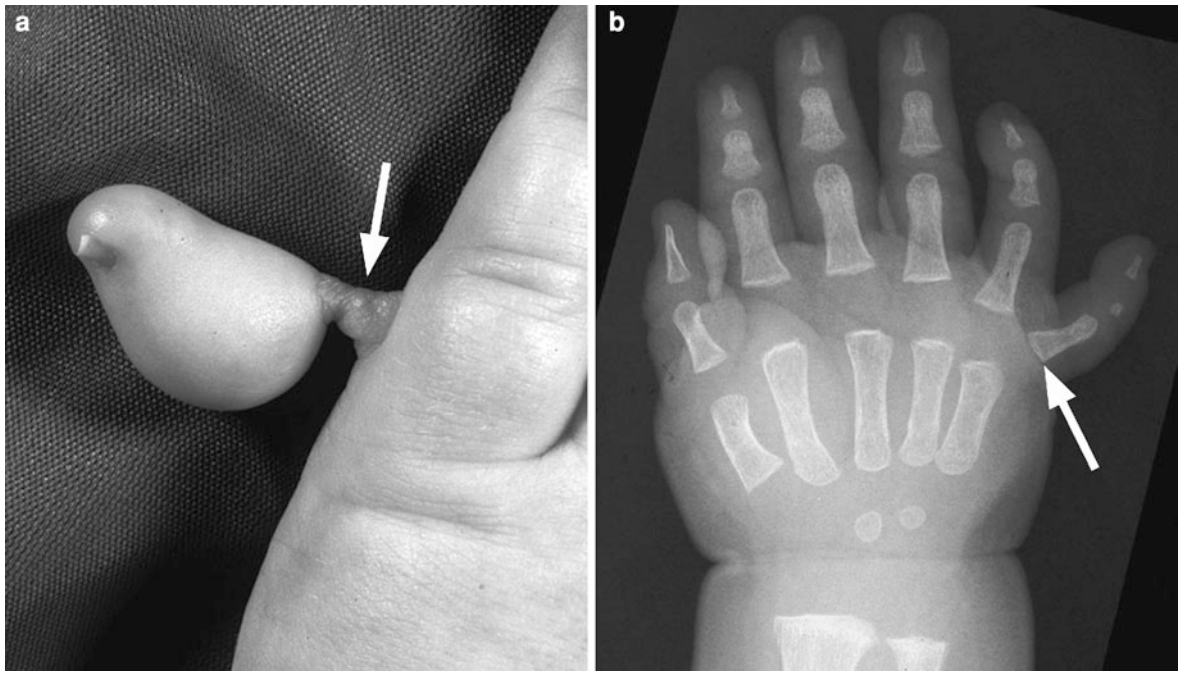


Fig. 27 Ulnar duplication is based on the Stelling classification. **a** *Type 1* skin bridge only (*arrow*) requiring only simple excision. **b** *Type 2* strong bony attachment, requiring preservation and reinsertion of the ulnar collateral ligament from accessory digit to fifth finger (position of insertion marked with *arrow*). *Type 3* is complete duplication of metacarpal

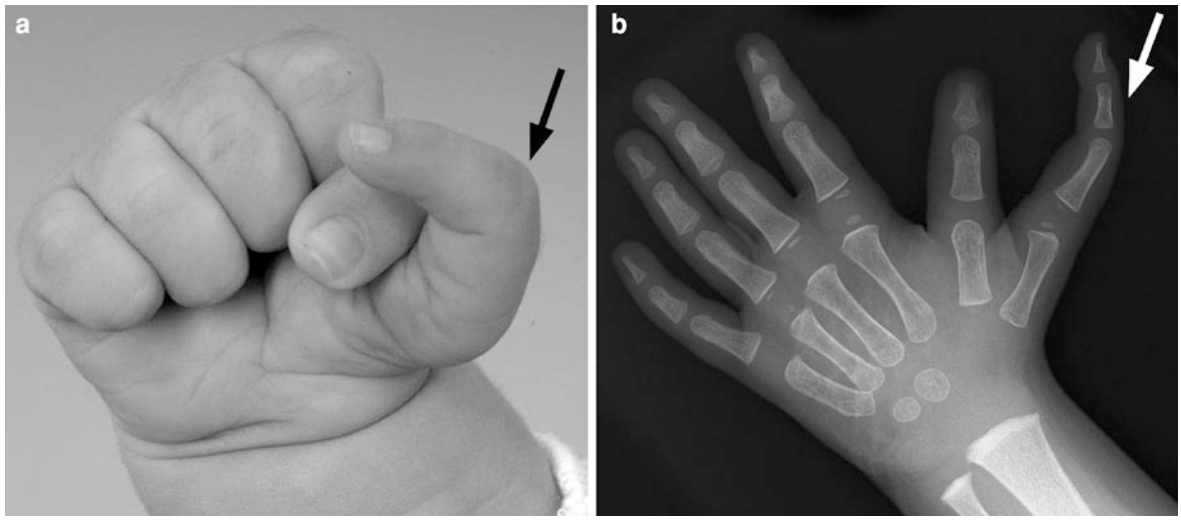


Fig. 28 Triphalangeal thumb (*arrow*)

Although it is commonly an isolated finding with autosomal dominant transmission and a favorable prognosis, it may also be associated with various syndromes. The presence of post-axial polydactyly in Caucasians is more suggestive of an underlying syndrome and merits systemic evaluation. Syndromes

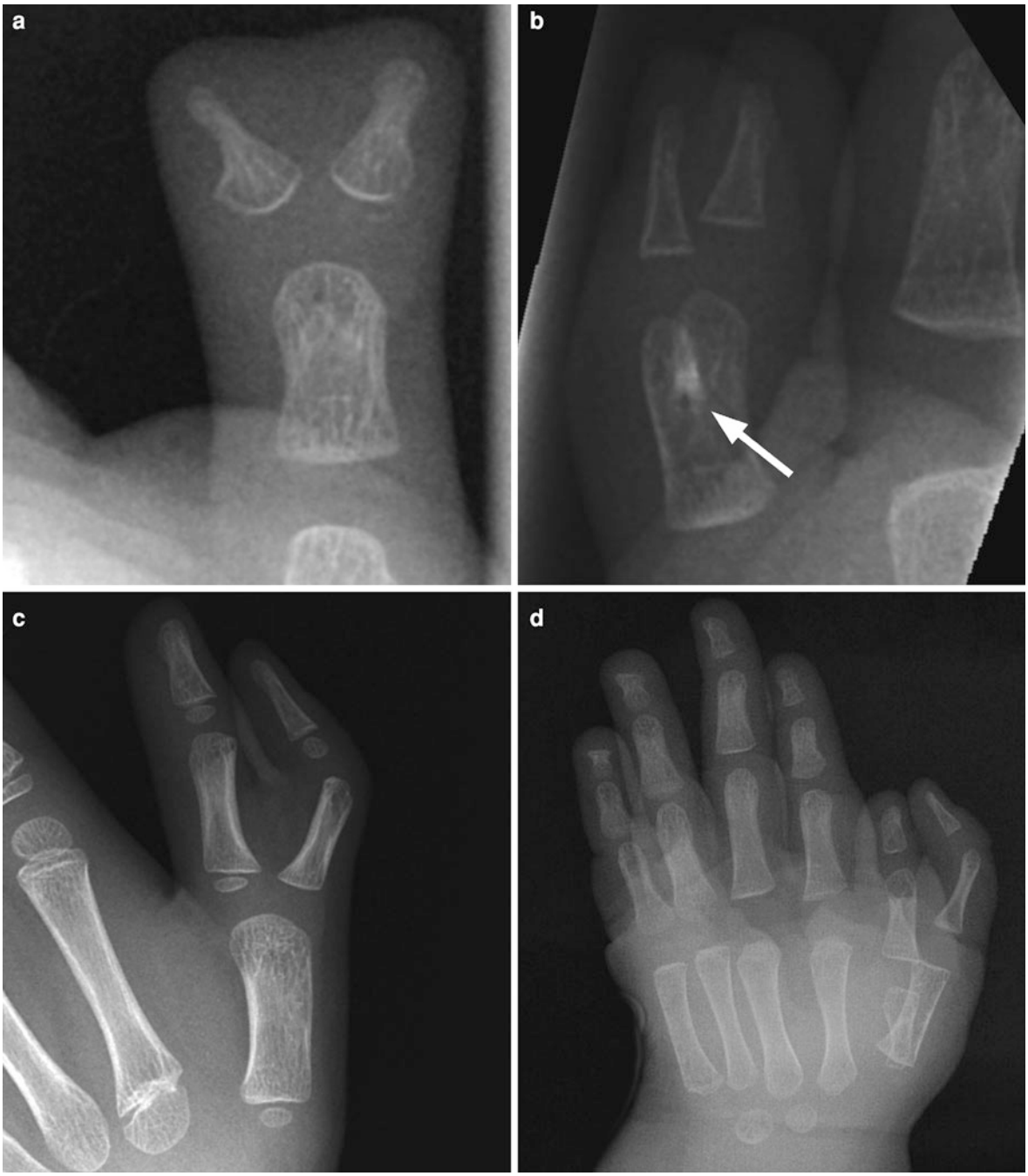


Fig. 29 The Wassel classification divides radial duplication into seven subtypes. **a** *Type 2* duplicated distal phalanx. **b** *Type 3* bifid proximal phalanx. **c** *Type 4* duplicated proximal phalanx. **d** *Type 6* duplicated metacarpal. *Type 1* is a bifid distal phalanx, *type 5* is a bifid metacarpal, and any duplication with a triphalangeal element is classified as a Wassel *type 7*

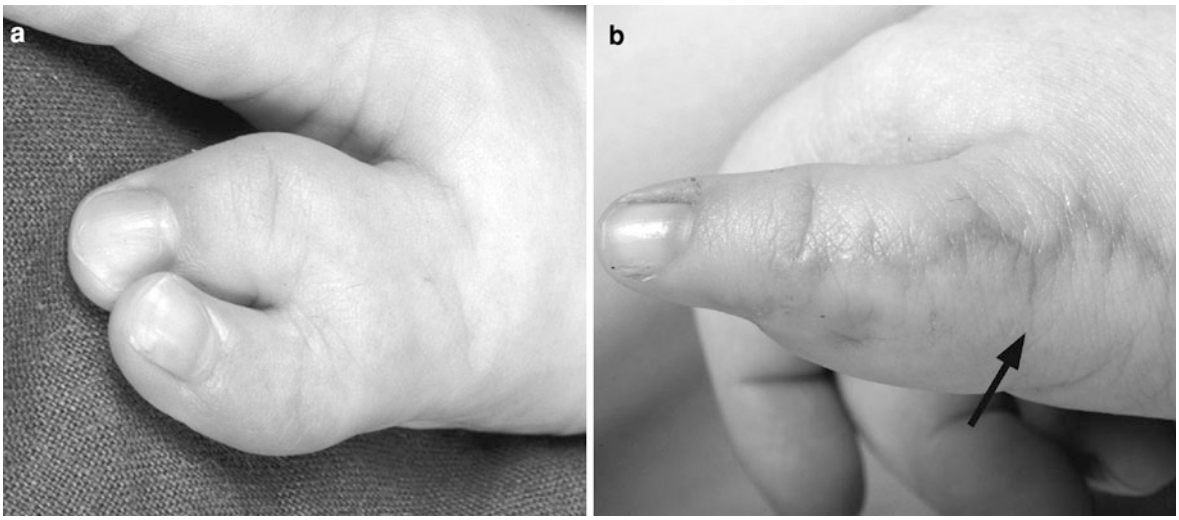


Fig. 30 Wassel 4 duplication at the metacarpophalangeal level is the commonest subtype (50%). **a** Well-matched duplicates, which would require skeletal amalgamation. **b** Post-amalgamation procedure of Wassel 4 duplication in which the radial duplicate was relatively hypoplastic, but which still requires preservation of soft tissues (*arrow*) from radial duplicate (radial collateral ligament, thenar muscle insertions and almost all of the skin), and reinsertion into the dominant ulnar duplicate. Only the hypoplastic skeleton, pulp and nail complex has been discarded

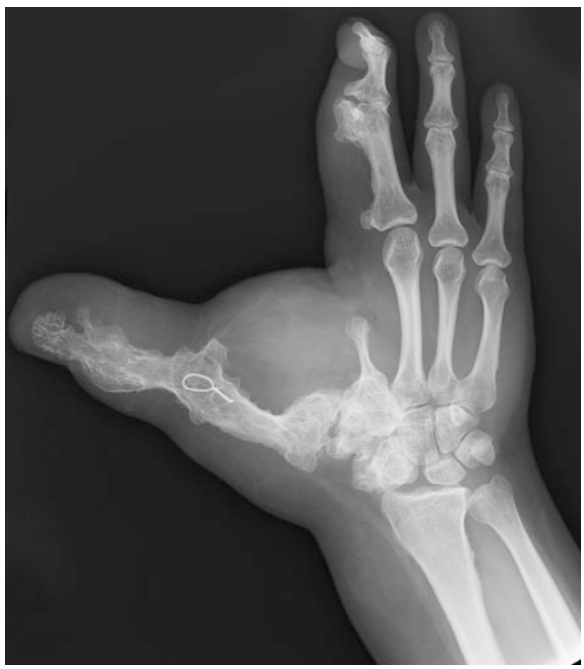


Fig. 31 Proteus syndrome

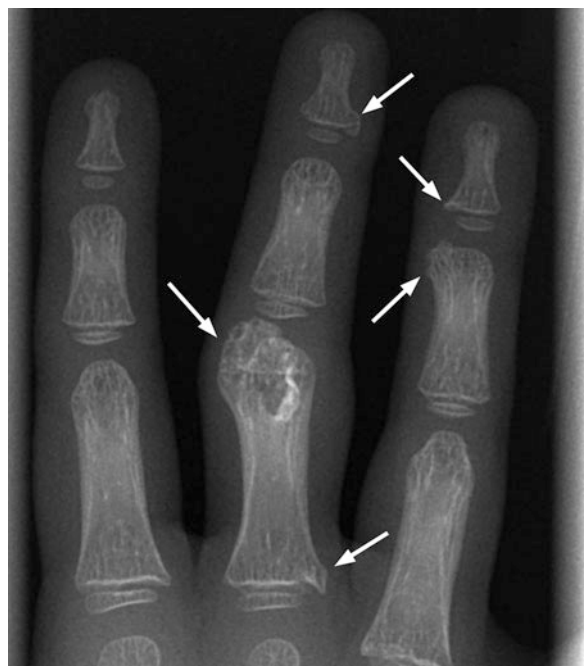


Fig. 32 Ollier's disease—multiple exostoses (*arrows*)

include trisomy 13, Meckel-Gruber syndrome, Bardet-Biedl syndrome, Smith–Lemli–Opitz syndrome, short rib-polydactyly syndromes and Ellis van Crevald syndrome (Lachman 2007).

Accessory digits may be well developed or rudimentary (Fig. 26) (Watson and Hennrikus 1997). Commonly the digit is rudimentary and held by a soft tissue bridge requiring only a simple excision

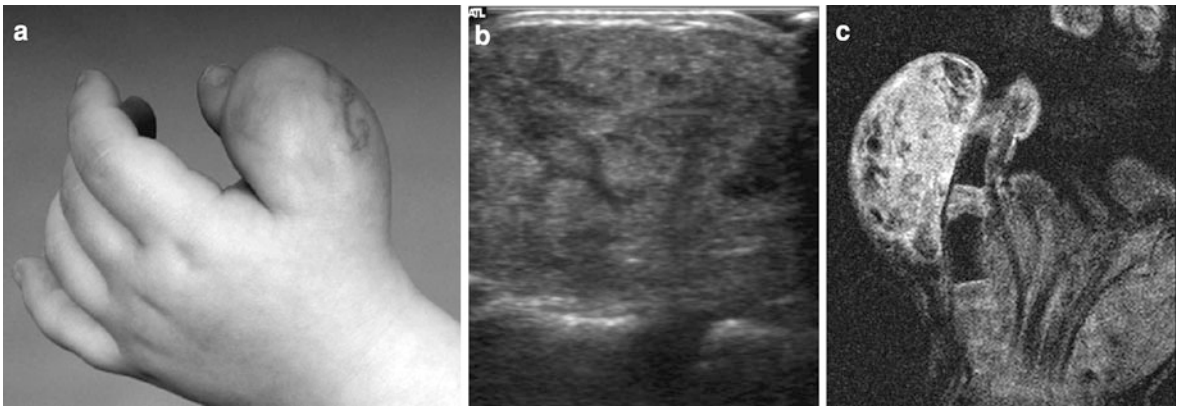


Fig. 33 **a** Large soft tissue mass on dorsum of thumb of 18 month old boy. Rapid growth raises possibility of malignant change, meriting initial urgent radiological investigation. Plain XR revealed no bony changes or calcification. This proved to be a benign lipoblastoma. **b** Ultrasound shows superficial heterogenous soft tissue mass. **c** Coronal T1 fat saturated sequence post contrast shows avid enhancement in the lipoblastoma adjacent to the thumb

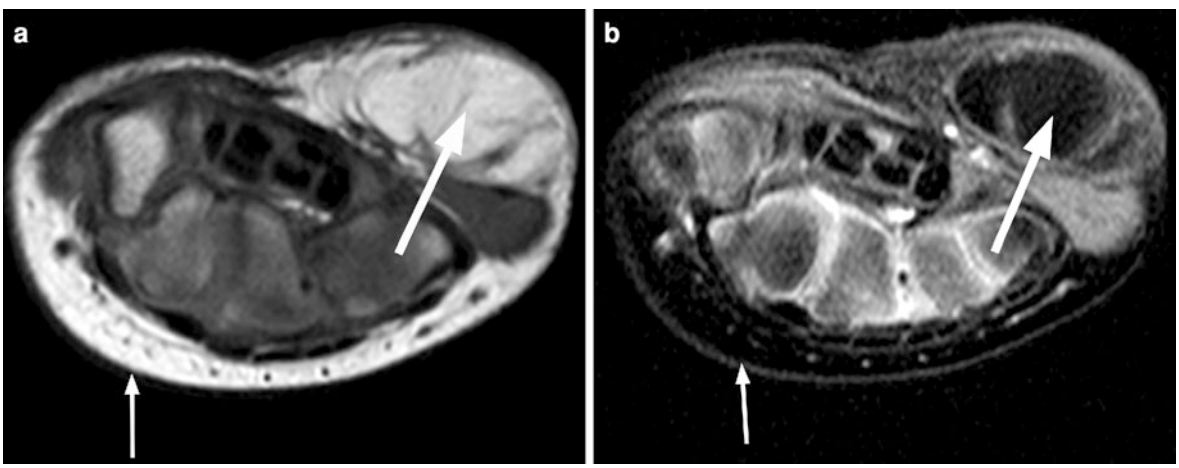


Fig. 34 MR of right hand shows. **a** on the T1 sequence the mass in the hypothenar eminence (*large arrow*) has the same signal as fat (*small arrow*). **b** on the T2 fat saturated sequence both fat and the mass saturate out, confirming the fatty nature of the mass

(Fig. 27a), though there can be bony attachments at proximal phalangeal or metacarpal level (Fig. 27b), requiring more involved surgery.

6.2 Pre-Axial Polydactyly

Although it is less common than post-axial polydactyly, it is more common in Caucasians and particularly in Orientals. The incidence is 1 in every 3,000 live births in the white population (Ress and Graham 1998). Duplication alone is usually unilateral and sporadic, however, if one of the duplicates is a triphalangeal thumb (Fig. 28a, b), a possible syndromic association should be considered (Rypens et al. 2006) and this can be inherited in an autosomal dominant pattern.

Thumb duplications are classified according to the level of duplication, and Wassel's classification is the most widely used and accepted (Fig. 29a, b, c, d). The most common type [Wassel type IV (Fig. 30a)] involves a duplicated proximal and distal phalanx sharing a common metacarpal articulation (Watt and Chung 2009).

Treatment usually involves creation of one good thumb out of the two duplicates, particularly if they

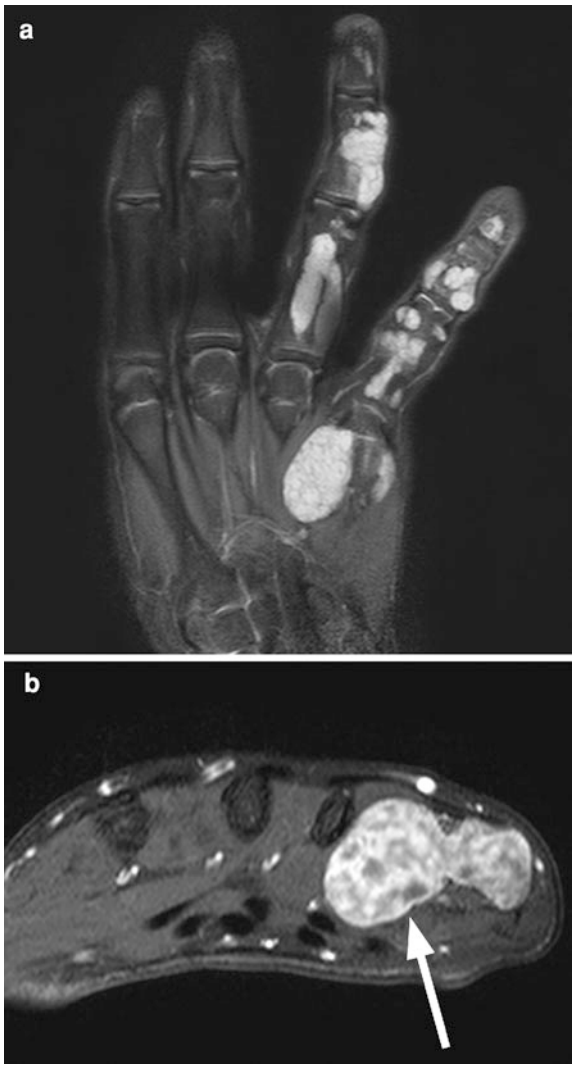


Fig. 35 Ollier's disease MRI. **a** T2 fat saturated coronal image shows the distribution of the high signal enchondromata **b** T1 fat saturated axial image post contrast shows a typical mottled chondroid type enhancement pattern in one lesion (*arrow*)

are of a fairly even size match, (i.e., amalgamation rather than excision) (Fig. 30b) and therefore plain radiographs are vital for surgical planning.

6.3 Central Polydactyly

This includes duplications that involve the index, middle or ring fingers and is often accompanied by syndactyly and cleft hand (Ress and Graham 1998; Tada et al. 1982).

6.4 Mirror-Hand and Mirror-Hand Spectrum

Duplication of the ulna, absence of the radius and thumb and duplication of the ring and small fingers about a common central finger characterize this exceedingly rare deformity. Only six cases of true mirror hand have been described in the literature (Watt and Chung 2009).

7 Overgrowth Conditions

These account for a small proportion of CHD and the region affected ranges from the entire limb, forearm, hand or digit. Overgrowth syndromes which may involve the hand include nerve territory orientated macrodactyly, lipomatous overgrowth, Proteus syndrome (Fig. 31), hemihypertrophy, Maffucci Syndrome, Ollier disease (Fig. 32), Parkes Weber Syndrome, Klippel-Trenaunay Syndrome and CLOVE syndrome (capillary malformations (C), lipomatous overgrowth of the trunk or extremities (LO), vascular malformations (V), epidermal naevi (E)) and generalized skeletal abnormalities) (Sapp et al. 2007).

In such cases, ultrasound and MRI can be very useful in clarifying the nature and soft tissue origin of the overgrowth (Figs. 33a, b, c; 34a, b; 35a, b). The typical MRI features of lipofibromatous hamartoma of the median nerve are illustrated (Fig. 36a, b, c).

7.1 Nerve Territory Oriented Macrodactyly and Lipomatous Macrodactyly

This is the most common overgrowth condition presenting to a hand surgeon. It is usually unilateral with a male predominance and no familial inheritance (Carty and Taghinia 2009). The lipomatous overgrowth may correspond with the ulnar or median nerve distribution within the hand (Fig. 37a, b). The affected nerves within the palm are characteristically grossly enlarged and compression neuropathies are common. The intrinsic muscles, joints, periarticular structures and skeletal parts are enlarged but blood vessels are of normal size. This latter fact is of great importance as the relative ischaemia in this condition can lead to difficulties in healing post surgery.

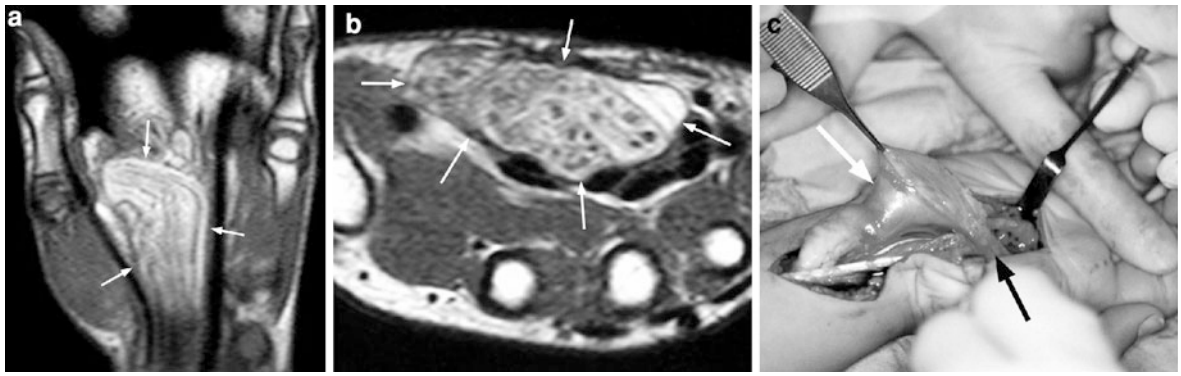


Fig. 36 Lipofibromatous hamartoma of the median nerve. **a** Coronal T1 shows high signal mass in the carpal tunnel (*arrows*) **b** Axial T1 sequence again shows the mass infiltrating the median nerve (*arrows*) **c** Operative photograph shows massive enlargement (*white arrow*) of the median nerve in the distal forearm and carpal tunnel caused by extensive and diffuse fatty infiltration, with relative sparing and more normal dimensions distally (*black arrow*)

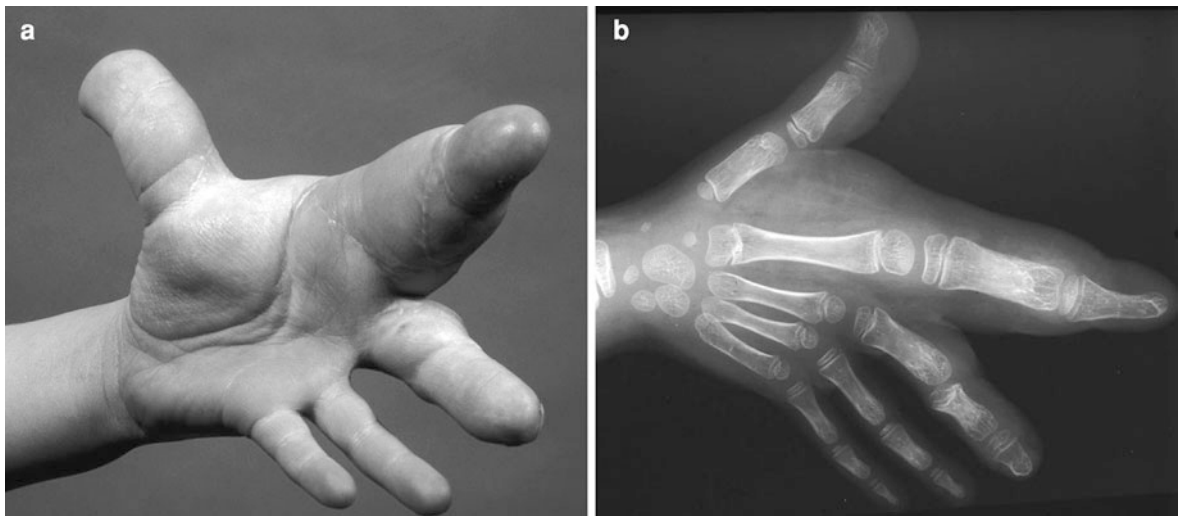


Fig. 37 **a** and **b** Nerve orientated macrodactyly following the median nerve distribution

8 Undergrowth Including Hypoplastic Thumb

This results from complete formation of the part during the embryonic period but incomplete growth and development during the fetal period (Van Heest et al 1996).

This group consists largely of the spectrum of hypoplasia of the thumb which can occur as an isolated entity, or more commonly in association with radial deficiencies. It is frequently bilateral and is often found in association with other hand differences including multiple other short digits, duplicated thumbs and a number of syndromes (Netscher and Baumholtz 2007).

The classification of thumb hypoplasia by (Blauth 1967) is useful in treatment planning, with less severe types requiring either no surgery or augmentation procedures (such as tendon transfers, ligament reconstruction and web-space deepening), whereas more severe forms (Fig. 38) require the creation of a new thumb by pollicisation of the adjacent index finger (Fig. 39a, b).

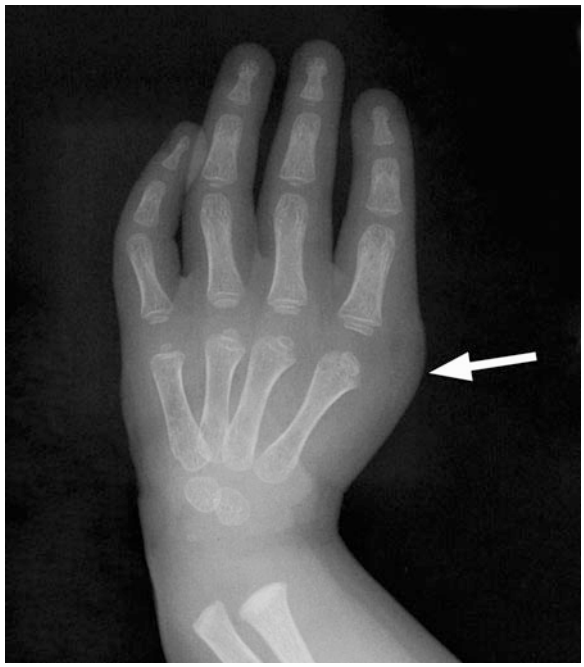


Fig. 38 Total aplasia of the thumb (Blauth grade 5)

Plain radiographs are vital in the assessment of the hypoplastic thumb as the absence of a well-formed thumb basal joint (Fig. 40), indicates that augmentation procedures would be unlikely to produce a useful thumb, and requires a pollicisation of the index finger, with removal of the poorly-developed original thumb.

Hypoplasia of the fingers most commonly affects the index and little fingers with the middle phalanx being the most commonly affected bone (Fig. 41). This CHD is frequently inherited in an autosomal dominant pattern, and is associated with numerous congenital syndromes including Treacher Collins, Apert, Poland, Cornelia de Lange and Bloom syndromes (Flatt et al 1994).

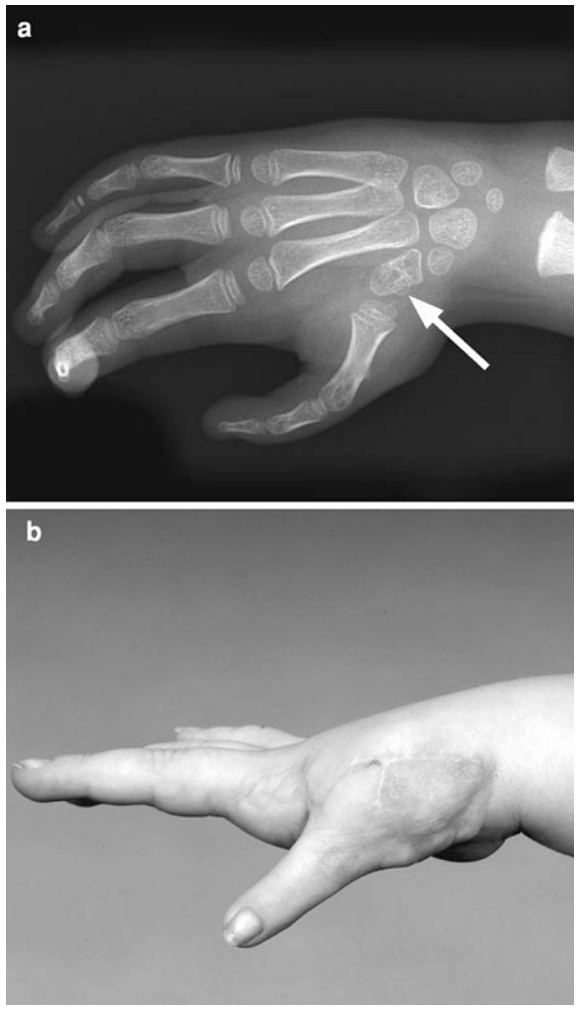


Fig. 39 Pollicisation of index finger for patient in Fig. 38 **a** Neo trapezium created by head of index metacarpal. **b** Pollicisation in such cases greatly upgrades the function of the hand

9 Constriction Ring Syndrome

This is a rare condition where the fetus may become entangled in the amniotic membrane although the exact etiology remains controversial. It had been postulated that early amniotic rupture leads to the development of fibrous bands, which entangle the digits and limbs leading to intrauterine amputations and malformations (Torpin 1965). It has also been described as amniotic band syndrome. Prenatal risk factors include prematurity, low birth weight, maternal drug exposure, maternal illness or trauma during pregnancy (Kawamura and Chung 2009). The reported incidence is 1 in 15,000 live births. There is no sex predilection and 60% have an abnormal antenatal history (Foulkes and Reinker 1994).

The abnormalities are characteristically asymmetric and the most common findings include distal ring constrictions (Fig. 42a, b, c), intrauterine amputations (Fig. 43a, b, c, d) and acrosyndactyly (distal fusion of digits with a proximal fenestration that communicates the dorsal side with the volar side) (Fig. 44). The fingers may be truncated, a small cleft remains at the

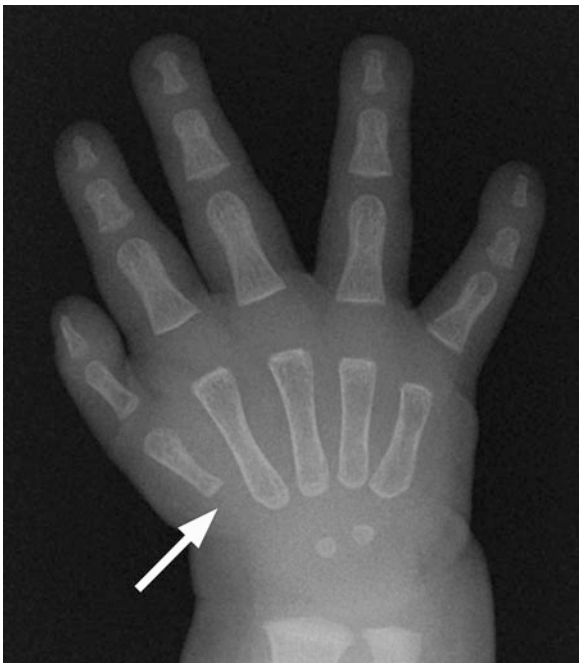


Fig. 40 A poorly developed basal joint in thumb hypoplasia (Blauth grade 3c) makes it very difficult to augment it into a useful thumb, though free metatarsophalangeal joint transfer from the foot has been used

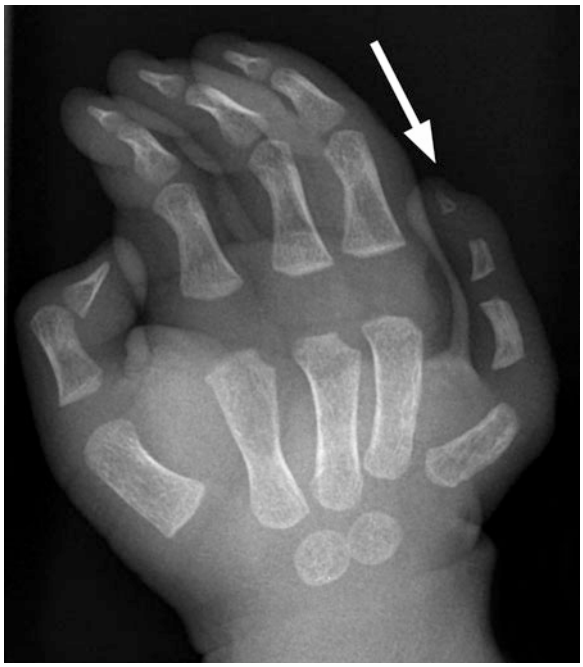


Fig. 41 Little finger hypoplasia. The marked attenuation of the fifth ray makes it difficult to augment, and was therefore treated in this case by ray amputation

site of the web space, and lower limb constriction bands and talipes deformities may be seen, (Kozin 2003; Wiedrich 1998; Foulkes and Reinker 1994). These features are not typically present with syndactyly resulting from inheritable and sporadic causes.

Interpretation of plain radiographs can be difficult in the more severe forms, in which the digits can be entwined and fused (Fig. 45a, b; 46a, b).

10 Generalized Skeletal Abnormalities

This includes conditions that do not fit into the other six categories described by Swanson, and includes skeletal hand deformities that are characteristic of a generalized bone and connective tissue disorder. The "tumour" like bone dysplasias including diaphyseal aclasia, and the fibrous disorders including neurofibromatosis and fibrous dysplasia are covered in separate chapters.

More than 250 skeletal dysplasias have been described with a complex classification system involving more than 30 groups organized by genetic and radiologic similarities. A comprehensive description of all the skeletal dysplasias involving the hand and wrist is therefore beyond the scope of this chapter, and reference should be made to a detailed text on this topic, (Lachman 2007; Spranger et al. 2002).

Antenatal ultrasound is able to diagnose many skeletal dysplasias (Dighe et al. 2008). In addition, low dose prenatal CT is starting to be used in large centers from the second trimester onward to provide further diagnostic information and aid antenatal counseling (Cassart 2010; Cassart et al. 2007). Post-delivery examination, plain film assessment, post mortem and genetic testing are, however, vital for determining a specific diagnosis for recurrence risk and counseling (Tretter et al. 1998).

A skeletal survey for the assessment of a skeletal dysplasia includes plain films of the skull, entire spine, chest, pelvis and extremities and there should be a systematic approach in the assessment of the bony skeleton.

It is important initially to determine the presence of any disproportionate shortening of the extremities and identify the presence of rhizomelia (proximal shortening of humerus or femur), mesomelia (middle segment shortening of tibia, fibula, radius and ulna) and/or acromelia (distal segment shortening of the hands and

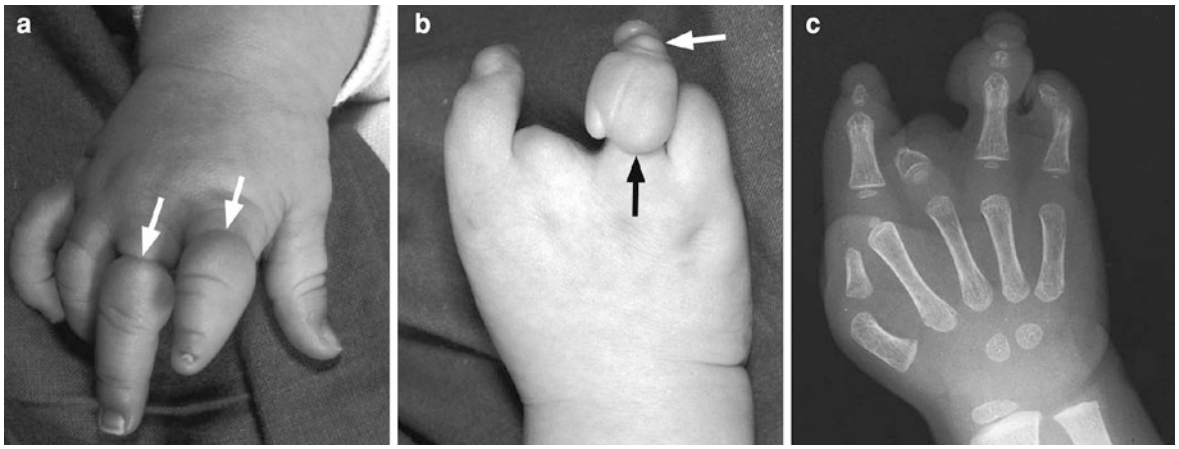


Fig. 42 Ring constriction syndrome. **a** Tight constrictions (*arrows*) with distal lymphoedema, needs relatively urgent release. **b** Marked distal lymphoedema, with rings at two levels on the same digit (*arrows*), plus autoamputations. **c** XR confirms involvement of all digits to varying degrees

feet). In reality, there is often a combination of shortening proximally and distally but the abnormality is categorized according to the predominant finding.

Disproportionate shortening may be present in isolation or as part of a more generalized skeletal dysplasia (Figs. 47, 48, 49, 50). The absence of hand and foot shortening in the presence of other abnormalities can suggest a diagnosis of a spondyloepiphyseal dysplasia congenita (Lachman 2008).

Secondly, assessment of the shape of the epiphyses, metaphyses and diaphyses and the appearance of epiphyseal ossification centers should be performed with particular attention to marginal irregularity, cortical thickness, flaring or flattening. Carpal bone ossification should be assessed for age and the presence of duplications, carpal fusions or accessory bones.

Findings should be organized and differential diagnosis and gamuts tables consulted in more specialist texts (Lachman 2007). Abnormalities should be differentiated from normal variants, always taking into consideration the clinical manifestations in conjunction with the radiologic findings.

This section will illustrate several more common examples with manifestations in the hand and wrist.

10.1 Achondroplasia Group

This includes three specific conditions, thanatophoric dysplasia, achondroplasia and hypochondroplasia. All have FGFR3 (fibroblast growth factor 3) mutations.

Thanatophoric dysplasia is also known as thanatophoric dwarfism and represents the second most common lethal bone dysplasia after osteogenesis type II. This condition is subdivided into types I and II, both of which are autosomal dominant mutations. In the extremities, there is generalized micromelia (shortening of all segments) and marked limb curvature (Fig. 51).

Achondroplasia is the most common, non-lethal skeletal dysplasia. It is an autosomal dominant condition with a spontaneous mutation rate of 80%. Most patients have a combination of rhizomelic, mesomelic and acromelic changes. In the hands, there is typically brachydactyly, metacarpal metaphyseal cupping and phalangeal metaphyseal widening (Lachman 2008) (Figs. 52, 53, 54).

Hypochondroplasia is also relatively very common, and shares many of same features of achondroplasia. The radiologic findings are milder and there is typically a later presentation, but there is always interpedicular widening in the lumbar spine.

10.2 Short Rib-Polydactyly Group

All conditions in this group are autosomal recessive and are similar radiologically.

Short rib-polydactyly dysplasias have the shortest ribs of all the dysplasias. In the hands, there is often severe brachydactyly with hypoplastic middle and distal phalanges. Polydactyly is commonly present.

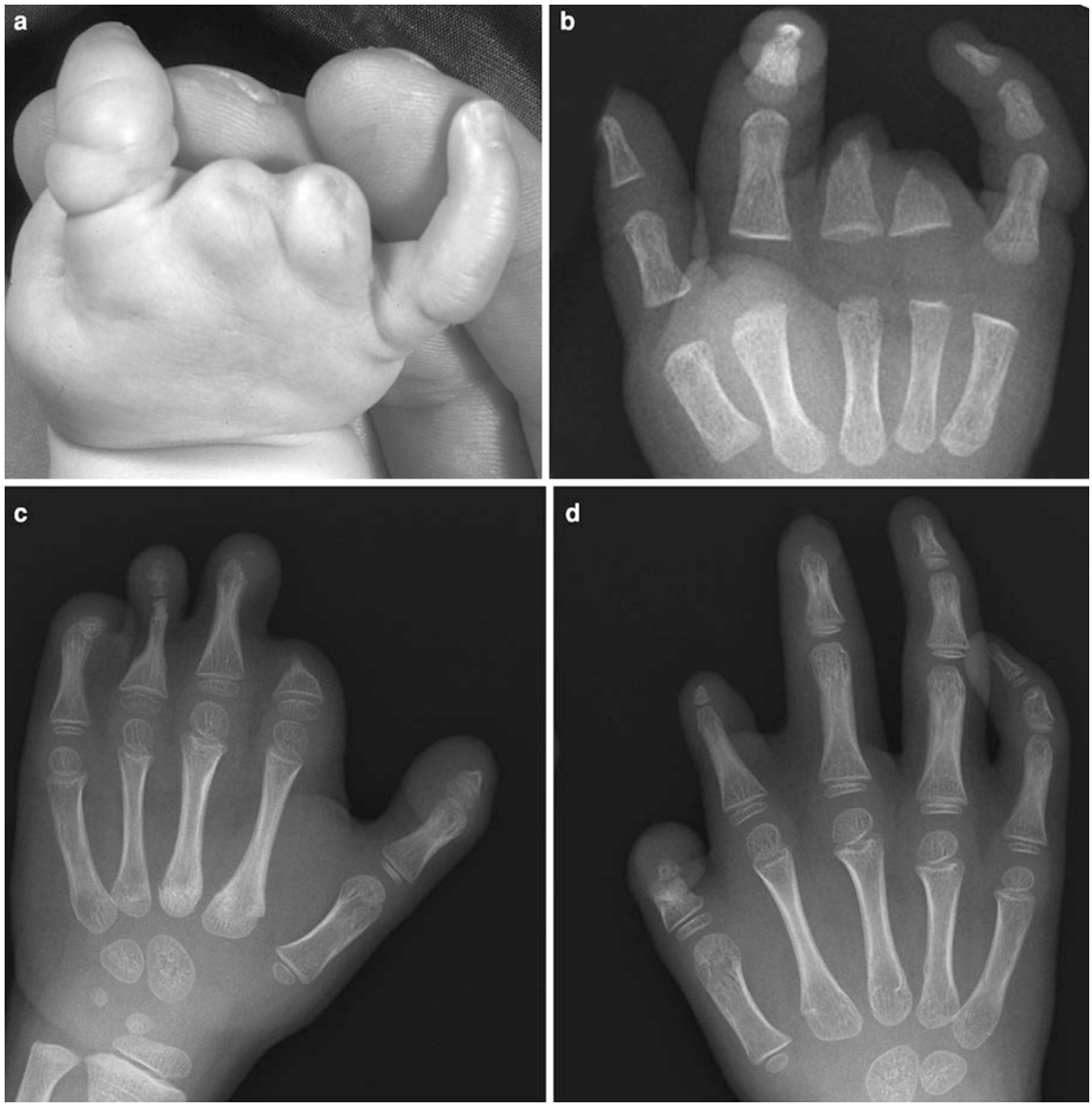


Fig. 43 **a** and **b** Autoamputations. **c** and **d** Bilateral asymmetrical involvement

Chondroectodermal dysplasia or Ellis van Crevald syndrome is a short-limbed dwarfism with dysplastic nails, hair and teeth accompanied by post-axial polydactyly and congenital heart disease. In the hands, as well as post-axial polydactyly, there are characteristic findings of carpal fusions, extra carpal bones and cone shaped epiphyses (Lachman 2008) (Fig. 55).

Asphyxiating Thoracic Dysplasia or Jeune Syndrome has similar radiologic appearances to Ellis-van Crevald syndrome with cone-shaped epiphyses in the hands. Uncommonly polydactyly is present.

10.3 Multiple Epiphyseal Dysplasia and Pseudoachondroplasia Group

In multiple epiphyseal dysplasia, the epiphyses of the hand are small, irregular and flattened and the carpal

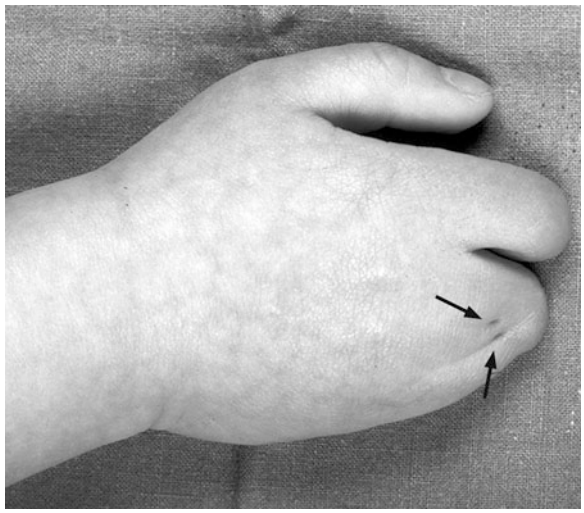


Fig. 44 Ring constriction syndrome with acrosyndactyly, and proximal fenestrations (*arrows*)

bones are small and irregular. There may be cone shaped epiphyses resulting in shortening of the short tubular bones of the hand.

Pseudoachondroplasia is a short limb, short trunk form of skeletal dysplasia. In the hands there is typically brachydactyly. The metacarpals are rounded proximally and there are mini-epiphyses in the hands with irregular carpal bones (Fig. 54).

10.4 Metaphyseal Disorders

Involvement of the hands occurs to varying degrees with the different metaphyseal dysplasias. Marked involvement of the hands is one of the main characteristics of metaphyseal chondrodysplasia McKusick Type. There is marked shortening of the hands, and flaring and cupping fragmentation of the metaphyses of the metacarpals and phalanges. The Jansen type metaphyseal chondrodysplasia demonstrates wide separation of epiphyses from metaphyses, with metaphyseal expansion and cupping. Schmid type metaphyseal chondrodysplasia, however, often has no hand involvement (Lachman 2008).

10.5 Dysostosis Multiplex Group

This group contains all the mucopolysaccharidoses, mucolipidoses and multiple storage diseases that produce a skeletal dysplasia.

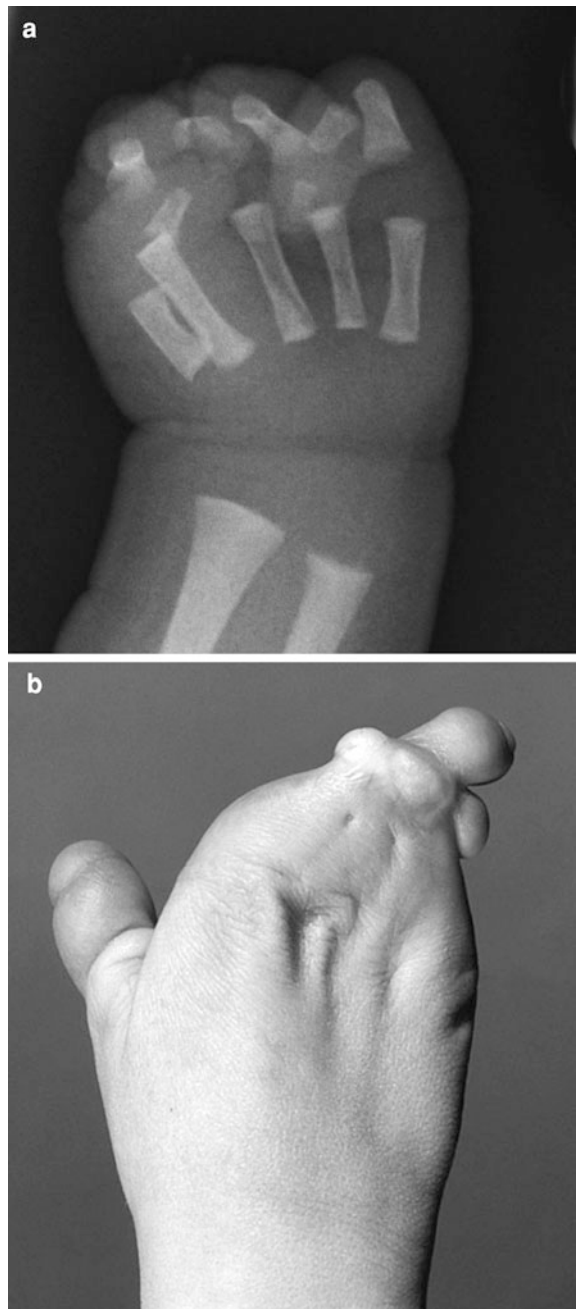


Fig. 45 **a** and **b** Ring constriction with complex arrangement of acrosyndactylies, necessitating very early release to prevent intractable bony deformities

Hurler syndrome (mucopolysaccharidosis type IH) has a recessive pattern of inheritance and most cases present in late infancy or early childhood. The hands characteristically exhibit brachydactyly, proximal

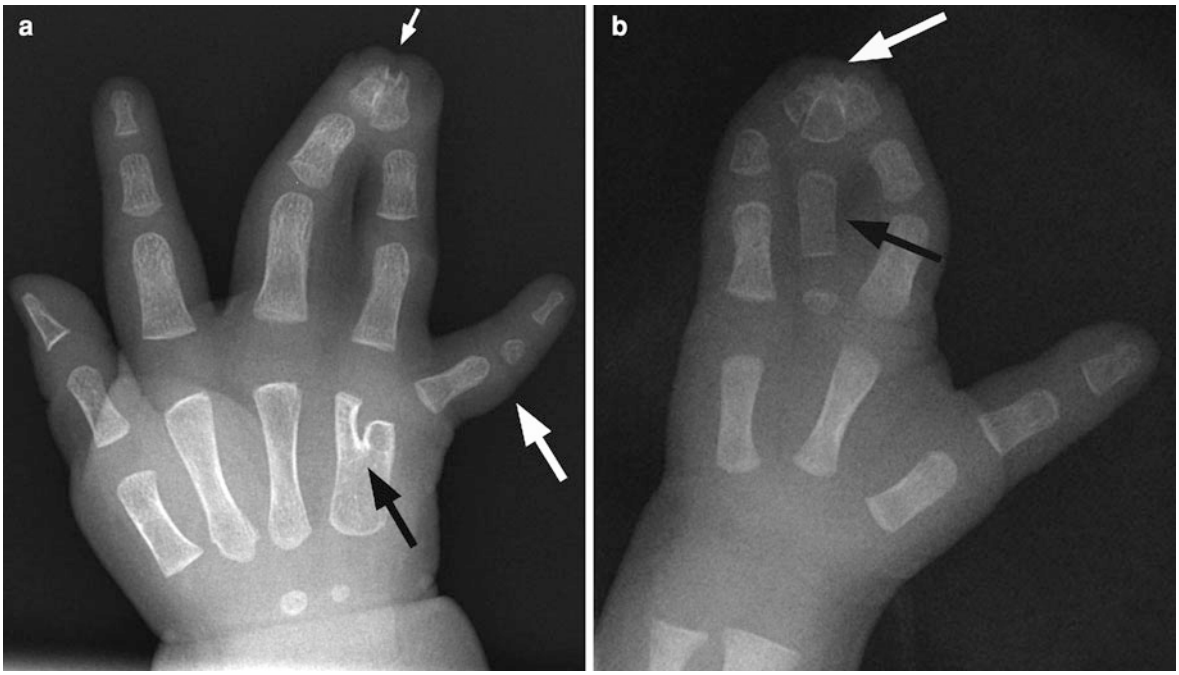


Fig. 46 Difficulties of classification. **a** Elements of syndactyly (*small white arrow*), hypoplasia (*large white arrow*) and bony synostosis (*black arrow*) in same hand. **b** Central duplication (*black arrow*), failure of formation (missing rays) and complex syndactylies (*white arrow*) in same hand

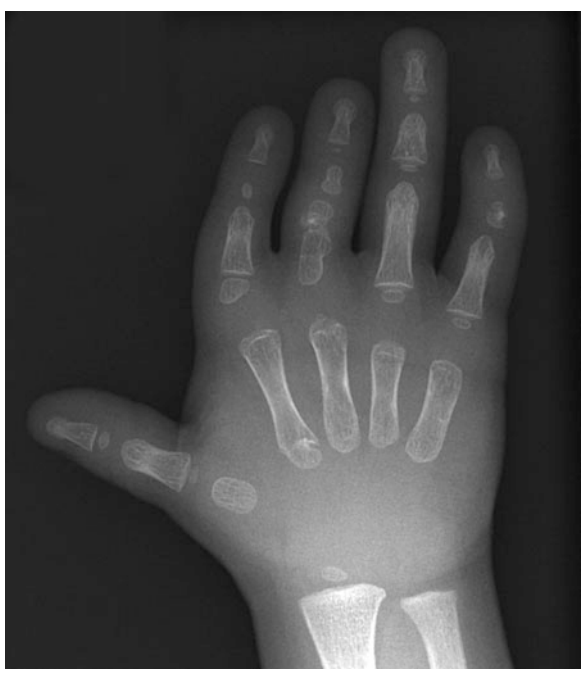


Fig. 47 Brachydactyly with absent carpal bone ossification, shortened metacarpals and shortened and dysplastic phalanges

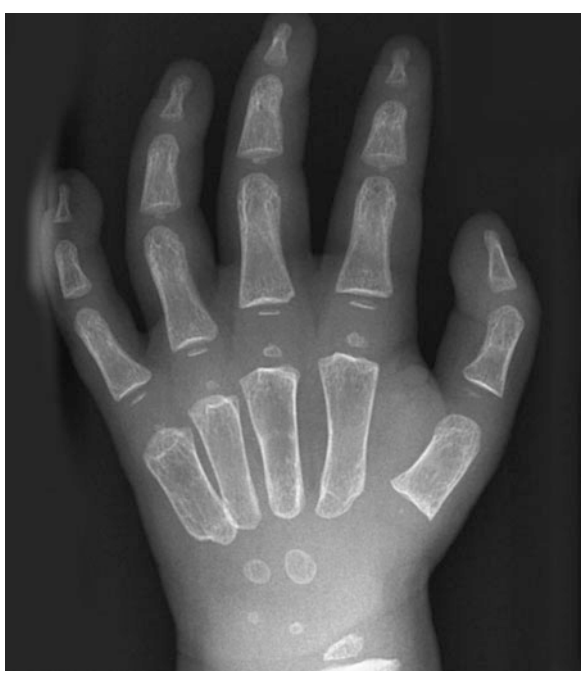


Fig. 48 Widened diaphyses of the metacarpals, proximal and middle phalanges with "pointing" of the proximal ends of the 2nd–5th metacarpals. The epiphyses are also dysplastic

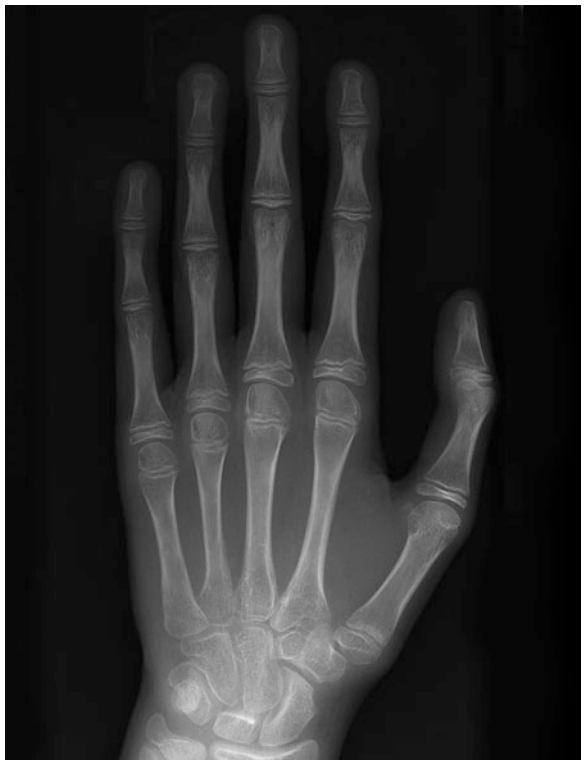


Fig. 49 Slender osteopenic bones with a metacarpal index of 10.4 in a patient with Marfans

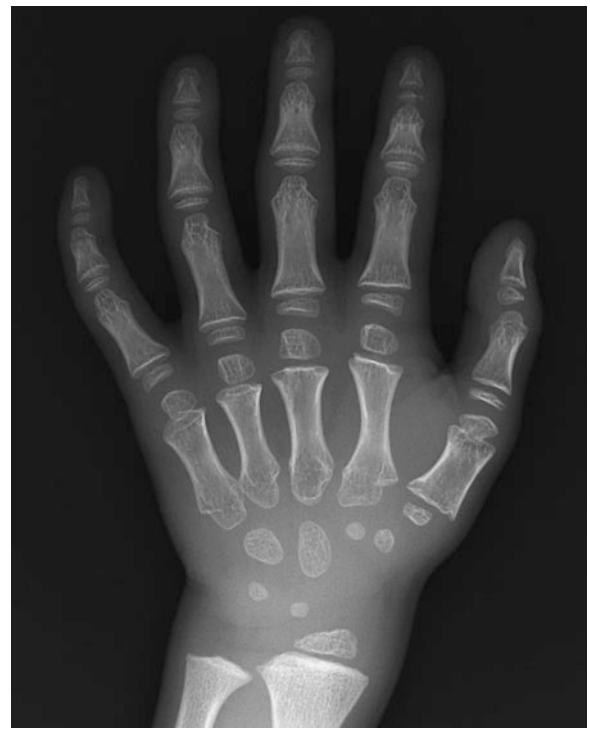


Fig. 50 Symmetrical shortening of the metacarpals with rounded, proximal metacarpals in a patient with Morquio syndrome. The carpal centers are small with delayed bone maturation

metacarpal "pointing", diaphyseal widening of metacarpals and proximal/middle phalanges and small irregular carpal bones with epiphyseal ossification delay (Lachman 2008) (Fig. 48).

Morquio syndrome (mucopolysaccharidosis type IVA and IVB) shows proximal metacarpal rounding, not "pointing" in the hands (Lachman 2008) (Fig. 50). The inheritance pattern is autosomal recessive (Hall 1997).

10.6 Dysplasias with Decreased Bone Density

These are represented primarily by osteogenesis imperfecta. Four main types have been described with osteogenesis imperfecta type II being the most common lethal skeletal dysplasia. In the extremities, in osteogenesis type II, there is generalized osteoporosis with or without fractures and shortened, widened long bones with thin cortices (Lachman 2008).

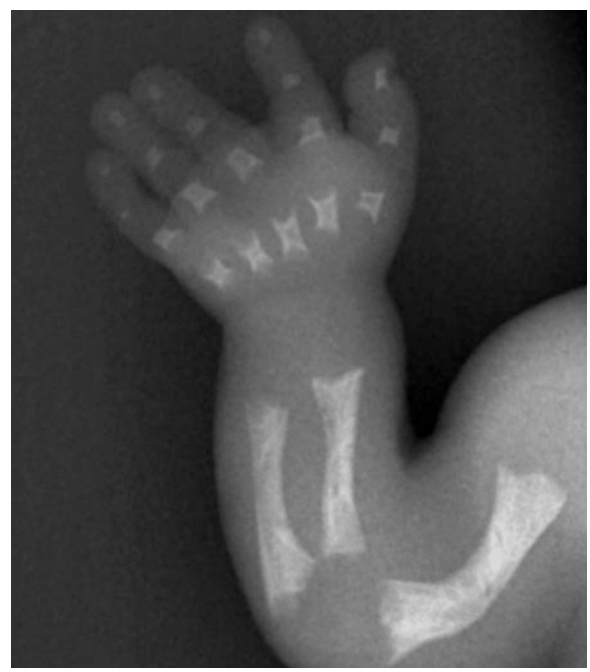


Fig. 51 Shortening and broadening of the tubular bones of the hand in a 20-week fetus with thanatophoric dysplasia type I

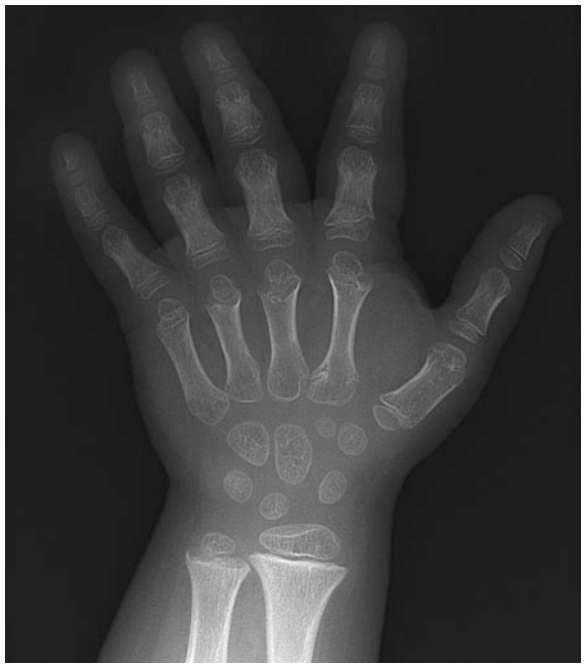


Fig. 52 Brachydactyly, metacarpal metaphyseal cupping and phalangeal metaphyseal widening in a patient with achondroplasia. In this patient, there is also flaring of the distal ulnar metaphysis

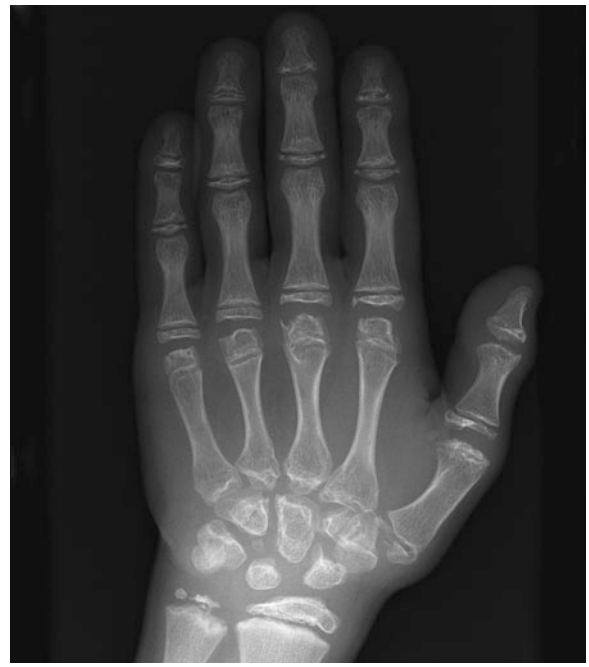


Fig. 54 Small carpals with irregular contours, shortened metacarpals and dysplastic, irregular epiphyses throughout the hand, some of which appear fragmented in a patient with pseudoachondroplasia

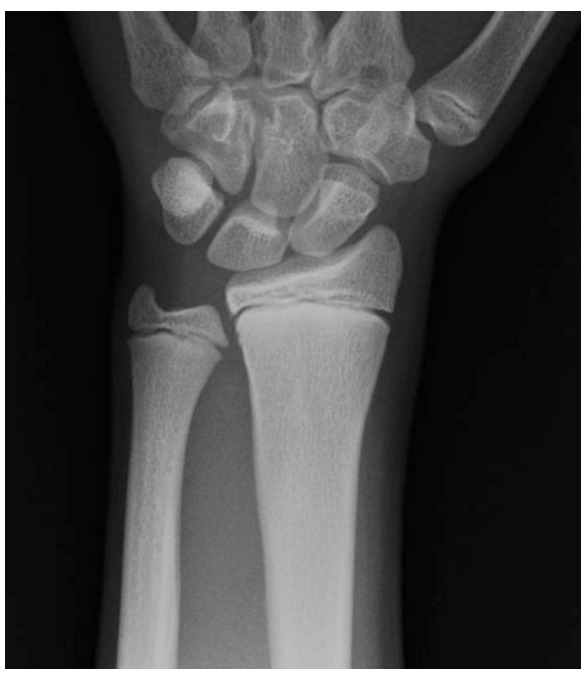


Fig. 53 Osteosclerosis of the bony skeleton and undertubulation of the distal radial diametaphysis in a patient with osteopetrosis

10.7 Sclerosing Bone Dysplasias

This can be divided into those conditions with increased bone density without alteration of bone shape (osteopetrosis and pyknodysostosis) and the craniotubular dysplasias (craniodiaphyseal dysplasia, craniometaphyseal dysplasia and Pyle dysplasia).

The long bones in osteopetrosis demonstrate metaphyseal expansion and osteosclerosis. There may be a "bone within a bone" appearance of the short tubular bones of the hand due to variation in disease activity (Lachman 2008) (Fig. 53).

Pyknodysostosis can be differentiated by the dysplastic or absent terminal phalanges, in combination with wide-open fontanelles and hypoplasia of the clavicles (Wynne-Davies et al. 1985).

10.8 Increased Limb Length

Marfans syndrome is a connective tissue disorder with autosomal dominant inheritance caused by mutations in the extracellular matrix protein fibrillin I. Typically, the

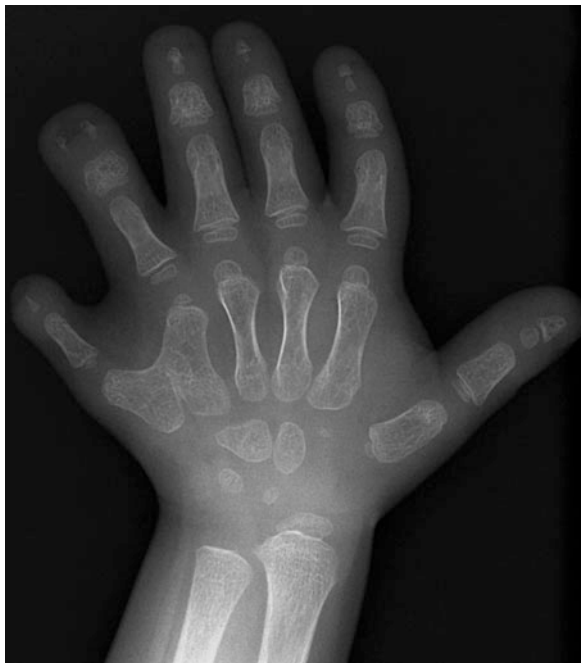


Fig. 55 Post-axial hexadactyly, middle and distal phalangeal hypoplasia, cone-shaped epiphyses of the middle phalanges, broadening of the hamate bone and early carpal coalition in a patient with Ellis van Crevald syndrome. The metacarpal of the duplicated digit is fused proximally to the metacarpal of the fifth finger. There is broadening of the fifth finger with duplicated distal phalanges articulating with a broadened middle phalanx

patient is tall with long, slender limbs and digits. Arachnodactyly can be determined by an abnormal metacarpal index above nine (sum of the lengths of the second to fifth metacarpals divided by the sum of the widths of the same bones) (McAlister et al. 2008) (Fig. 49).

11 Summary

While the classification of CHD presented is broadly and most commonly used in clinical practice it has many limitations, not least that some differences could comfortably be placed in several groups (e.g., Fig. 46a, b).

Radiology is vital for the assessment, treatment planning and follow up of all but the mildest cases of CHD, with plain films remaining the main imaging modality. Ultrasound and MRI are reserved for specific indications. Ultrasound can be extremely helpful in the assessment of tendons, and in the clarification of the nature of isolated swellings. MRI may provide additional information in selected cases, and is particularly important in the management of more complex overgrowth syndromes. Obstetric ultrasound now allows diagnosis of CHD earlier in fetal life, and advances in this modality will continue to raise the possibility of novel management opportunities and present new dilemmas.

Several CHD are associated with systemic syndromes and the reporting radiologist should be aware of these to highlight the need for further evaluation.

The management of CHD must be within a multidisciplinary environment with a team able to deal with all of the complex issues presented by this challenging group of patients.

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