Congenital Deformities of the Upper Extremity

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Abstract

Congenital deformities of the upper extremity are rare. They are often associated with other, more severe disorders of the cardiovascular, craniofacial, neurologic, and musculoskeletal systems. Most upper-extremity congenital anomalies are minor and cause no functional deficits, and surgical reconstruction is therefore unnecessary. If a severe cosmetic deformity is present or there is significant functional compromise, surgical treatment is indicated. The authors review the common congenital deformities of the upper extremity and offer treatment recommendations.

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A congenital anomaly of the upper extremity is present in 1 of every 626 live births.[1] In most cases, the deformity is minor and causes no functional deficit, but in 10% the patient requires treatment.[2] Such anomalies are often associated with cardiovascular, craniofacial, neurologic, and other musculoskeletal abnormalities. If treatment is needed, an integrated team approach, involving the physician, the therapist, and the parents, should be used.

Embryology

Development of the upper extremity precedes that of the lower extremity by 1 week and occurs in a proximal to distal sequence.[3] At 4 weeks, a bulge of the lateral body wall arises from the 8th through the 10th somites. This primitive limb bud consists of undifferentiated mesenchyme. The growth of the limb bud is under the control of the overlying ectodermal tissue, termed the apical ectodermal ridge. The hand paddle appears at 5 weeks' gestation. Programmed cell death then occurs between digits, and finger separation is complete by 8 weeks. The hands begin to move at 9 weeks' gestation, at which time skin creases begin to develop.

Etiology

The cause of 40% to 50% of congenital hand anomalies is unknown.[2] The remainder are due to genetic abnormalities or exposure to environmental teratogens. Genetic abnormalities include single-gene, multiple-gene, and chromosomal disorders.[1]

Classification

The classification system developed by Swanson et al[4] is currently accepted by the American Society for Surgery of the Hand and the International Federation of Societies for Surgery of the Hand. This system separates the congenital hand anomalies into seven major categories: failure of formation, failure of differentiation, duplication, overgrowth, undergrowth, constriction bands, and generalized skeletal abnormalities.

General Treatment Principles

Children have an exceptional ability to compensate for their deformities. Treatment should improve the child's function, not compromise it. If surgery is selected, the timing depends on the specific anomaly; usually, however, the procedure is performed between the ages of 6 and 18 months.[3] Conditions that threaten limb viability, such as constriction bands, should be treated earlier.[5]

Failure of Transverse Formation

Congenital Amputations

Congenital amputations are classified according to the level of involvement: complete arm, midarm, proximal forearm, wrist, midhand,

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and fingers.[1] The most common is a proximal forearm amputation.[6] Parts distal to the amputation level are either rudimentary or absent. The proximal limb segment is often abnormal as well. In the majority of cases, the abnormality is unilateral.

The reported incidence is 1 in 20,000 live births for forearm amputations and 1 in 270,000 for arm amputations.[5] The cause is usually unknown, although transmission as an autosomal-recessive trait has been reported.[6] Associated musculoskeletal anomalies may include clubfoot, meningocele, radial-head dislocation, and radioulnar synostosis.[5]

Treatment is usually conservative and starts in infancy.[1] A passive prosthetic device is fitted between 3 and 6 months. A change is made to an active terminal device as the child grows. The Krukenberg procedure, in which the radius and ulna are separated to function in a pincer fashion, is indicated for blind patients with bilateral upper-extremity amputations.[6] It may also be indicated for unilateral amputees in underdeveloped countries where prostheses are unavailable.[1]

Phocomelia

Phocomelia, or abnormal proximal-to-distal limb development, is classified into three types.[1] In the first, or complete, type, the hand is attached directly to the trunk. In the second, or proximal, type, the arm segment is absent, and there is direct forearm-trunk articulation. In the third, or distal, type, the forearm segment is absent, and there is direct hand-arm articulation.

The incidence of phocomelia is currently 1% of all congenital upper-extremity anomalies.[5] The deformity was more prevalent during the decades when thalidomide was prescribed for pregnant women.[1] Associated anomalies include radial-ray deficiency of the opposite limb, cleft lip, and cleft palate.[6]

Treatment is usually nonsurgical. Some patients benefit from use of a prosthesis and limb training. Skeletal lengthening and tendon transfer may improve function and stability.[5]

Failure of Longitudinal Formation

Radial Club Hand (Radial Deficiency)

Radial club hand is a preaxial deformity resulting from partial or complete absence of the radius. The forearm is short, and the wrist is radially deviated. Complete or partial absence of the thumb and radial carpus, combined with thenar muscle deficiency, is also common. The ulna is usually short and bowed. [1]

Bayne has classified this disorder into four types.[5] Type 1 is characterized by a short distal radius with delayed appearance of the distal epiphysis. In type 2 there is a hypoplastic radius with defective proximal and distal epiphyseal growth. Type 3 is characterized by partial absence of the middle and distal thirds of the radius. In type 4, the most common variety, there is complete absence of the radius (Fig. 1).

Radial club hand occurs in 1 of every 100,000 births and is bilateral in 50% of patients.[2] The cause is unknown, and most cases are sporadic. Environmental causes, such as thalidomide, have been implicated.[5] Radial club hand can be associated with cardiac abnormalities (Holt-Oram syndrome), aplastic anemia (Fanconi's anemia), thrombocytopenia, and the VATER complex (the syndrome that is characterized by vertebral anomalies, imperforate anus, tracheoesophageal aplasia, and renal anomalies).[6]

Treatment of type 1 and mild type 2 deformities is serial casting and stretching.[2] Occasionally, surgical release of tight fascial or tendinous structures is required to position the hand in a straight position over the forearm. Treatment for the more severe type 2 deformities, as well as for types 3 and 4, includes serial casting and stretching, followed by centralization or radialization of the carpus over the ulna.[2] There are a number of centralization techniques,[6] which usually are performed between 6

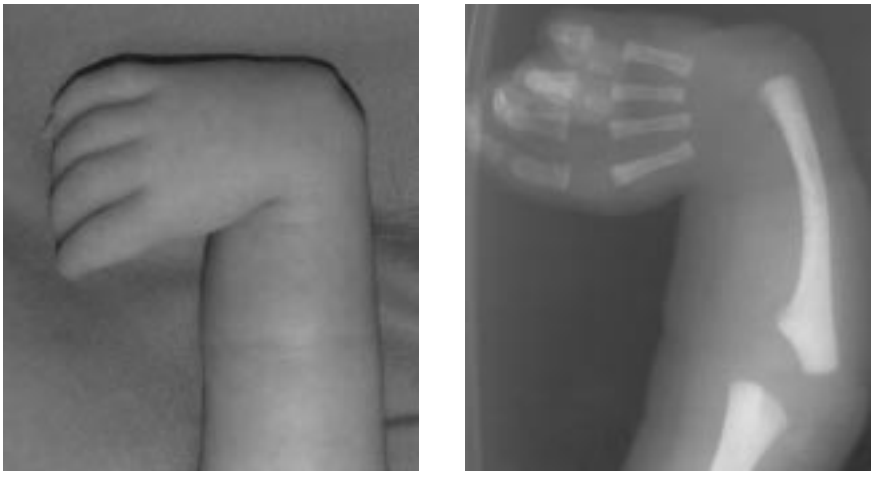

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Fig. 1 Type 4 radial club hand in a 2-month-old boy. **A**, Clinical appearance. **B,** Radiographic appearance.

months and 1 year of age.[5] Transfer of the extensor carpi radialis longus to the extensor carpi ulnaris is recommended to prevent recurrent deformity.[1] Lack of adequate elbow flexion for hand-to-mouth positioning is a contraindication to surgery.[5] Distraction lengthening (Fig. 2) is now being used experimentally by many surgeons for correction,[7] but is associated with a high complication rate.

The mild thumb hypoplasia seen in type 1 deformities usually requires no treatment. However, appropriate tendon transfers can occasionally improve function.[2] Pollicization of the index finger is recommended for more severe degrees of hypoplasia or complete absence of the thumb.[8] This is usually performed approximately 6 months after the centralization procedure.[2]

Ulnar Club Hand (Ulnar Deficiency)

Ulnar club hand is a postaxial deformity due to partial or complete ulnar absence. This deformity is classified into four types.[5] Type 1 is caused by hypoplasia of the ulna. Type 2 is characterized by partial absence of the ulna. In type 3 there is complete absence of the ulna. Type 4 has associated radiohumeral synostosis. Partial ulnar absence is the most common presentation.[5] The radial head is dislocated in 50% of cases.[1]

Congenital ulnar deficiency is very rare. Flatt[2] reports 1 case for every 3.6 cases of radial club hand. The majority of cases occur sporadically and are unilateral. Most associated anomalies are musculoskeletal, including proximal femoral focal deficiency, clubfoot, and spina bifida.[6]

Function in children with ulnar club hand is surprisingly good compared with the poor function in patients with radial club hand. Less than a third of patients have fixed ulnar deviation of the wrist, and most can place the wrist in the neutral position.[1]

Most patients are treated conservatively. Mild deformity in young children may be treated with stretching and serial splinting. A severe deformity with dislocation of the proximal radius may require excision of the proximal radius and fusion of the distal radius and proximal ulna (resulting in a one-bone forearm). This is recommended when elbow motion is restricted or elbow instability is present.[1] This procedure will stabilize the forearm but sacrifices forearm rotation, which is usually well compensated for by shoulder motion. For patients with radiohumeral synostosis and a severe internal rotation deformity, derotational osteotomy of the humerus to improve alignment may be considered.[5]

Central-Ray Deficiency

Central-ray deficiency is a failure of formation of the central rays of the hand and is described as a split, cleft, or lobster-claw hand. This condition is classified into two types: typical and atypical.[6] The typical pattern is a central V-shaped cleft due to absence of

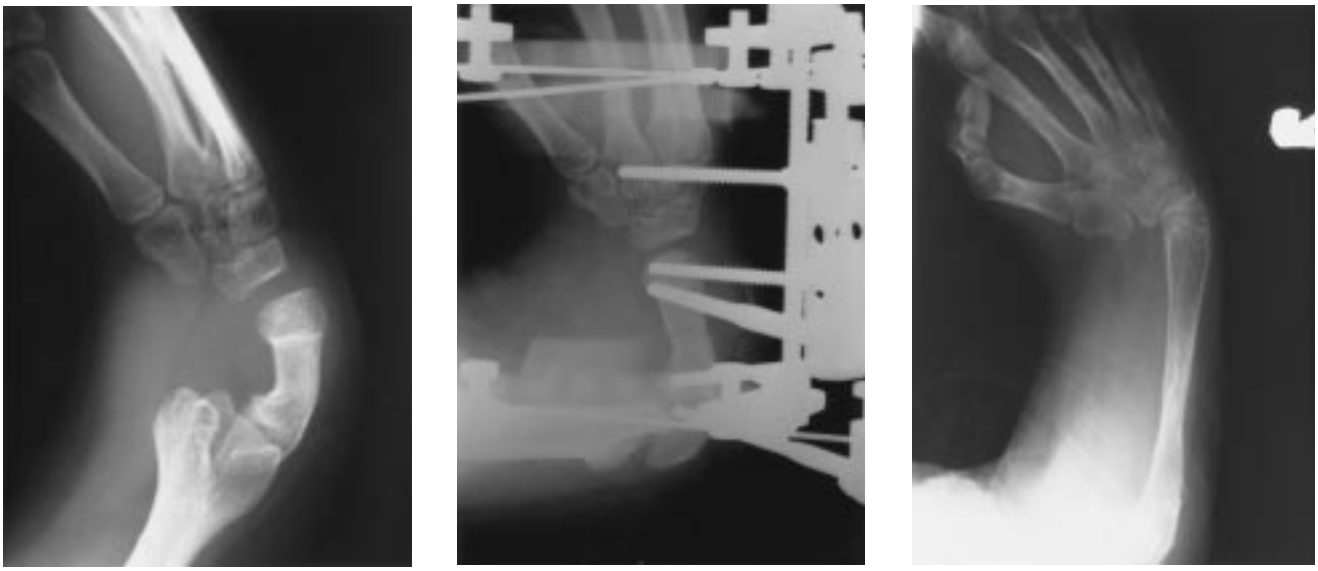

Fig. 2 Distraction lengthening for a severely shortened forearm in a patient with radial club hand. **A,** Preoperative radiographic appearance. **B,** Intraoperative radiograph obtained after osteotomy and fixator application. **C,** Fifteen months postoperatively, forearm length is improved, although residual radial wrist angulation is seen.

the long ray. The hand is divided into two radial and two ulnar digits. There may be associated syndactyly of these digits. The atypical pattern is seen when the three central rays are absent, leaving only a thumb and small finger. The radiographic findings are highly variable for both types. Split metacarpals and tranversely oriented bones often are present.

The incidence of cleft hand is 4% of all congenital upper-extremity anomalies.[2] Most cases occur sporadically and are unilateral. The typical variety can be transmitted as an autosomal-dominant trait and is usually bilateral,[6] whereas the atypical type is unilateral. Associated anomalies include cleft foot, cleft lip, cleft palate, congenital heart disease, imperforate anus, and deafness.[2]

Most patients do not require surgery, as function is satisfactory. For patients with a more severe deformity, closure is indicated. Many techniques have been reported, including Barsky's, which includes suture fixation of the index and ring metacarpals, reapproximation of the deep transverse metacarpal ligament, and occasionally metacarpal osteotomy.[9] Snow[1] and Miura and Komada[10] have described separate techniques for correcting the thumb and index syndactyly associated with cleft hand in cases in which pinch and hand function are compromised. In each technique, the index metacarpal is osteotomized at the base and transferred to the long metacarpal position. Flatt[2] recommends staged surgical procedures, with correction of syndactyly first, followed by cleft closure several months later.

Failure of Differentiation (Syndactyly)

Syndactyly (failure of digital separation) is one of the most common congenital hand deformities.[5] Syndactyly is classified as either incomplete or complete and as either simple or complex. The incomplete type is characterized by the joining of digits from the web space to a point proximal to the fingertip; the complete type, by the joining of digits from the web space to the fingertip. Simple syndactyly refers to only soft-tissue bridging between digits. Complex syndactyly refers to both soft-tissue and bone connections between affected digits.

Syndactyly occurs in approximately 1 of 2,000 births and is most common in white male children.[5] Involvement is bilateral in 50% of cases. The most common site is between the ring and long fingers; the least common, between the thumb and the index finger.[2] Eighty percent of cases occur sporadically; however, familial syndactyly does occur.[5] Associated anomalies include polydactyly, constriction bands, toe webbing, brachydactyly, spinal deformities, and heart disorders.[6] Syndactyly may occur as a component of a syndrome.[1] Chest-wall anomalies and syndactyly are seen in Poland's syndrome. Severe syndactyly combined with craniosynostosis, mental retardation, ankylosed interphalangeal joints, flattened facies, and hypertelorism is seen in Apert's syndrome.

Surgery is usually recommended and should be performed early to prevent progressive bone deformity and to complete the separation by school age. Separation of digits of unequal length is recommended early to prevent flexion and rotational deformities. Thumb–index finger syndactyly should be released at 6 months; ring finger–small finger syndactyly, at 1 year.[1] Separation of equal-length digits at 18 months is recommended by Flatt.[2] Children with severe complex syndactyly often require multiple procedures.

Several surgical principles are important in treatment.[2] The surgeon must be aware that anomalies of the neurovascular bundles can occur in complex deformities. One side of a digit should be corrected at each procedure to prevent vascular compromise. Separation of the other side of a digit should be performed 3 to 6 months after the original procedure. Zigzag incisions are used with a long volar or dorsal flap for web-space separation (Fig. 3). Full-thickness skin grafts are almost always necessary for wound coverage.

Intrinsic Digital Abnormalities

Camptodactyly

Camptodactyly is a flexion deformity of the proximal interphalangeal

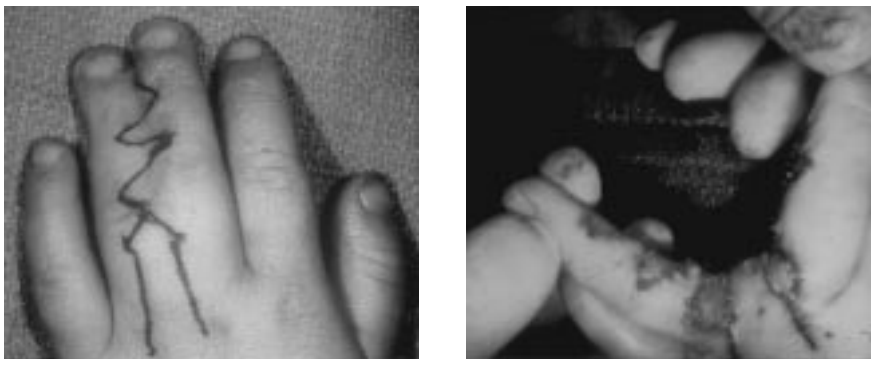

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Fig. 3 Correction of simple syndactyly. **A,** Planned incisions. **B,** Appearance after surgical correction and full-thickness skin grafting.

joint. The anomaly is most common in the small finger, but other digits may be affected.[1] The deformity may develop in infancy or adolescence; it usually increases during growth spurts and then stops in late adolescence. [6]

Camptodactyly occurs in less than 1% of the population. Most cases are sporadic,[5] although several causes have been reported,[1] including flexor tendon sheath contracture, flexor digitorum superficialis contracture, collateral ligament contracture, volar plate contracture, abnormal lumbrical insertion, phalangeal deformities, vascular abnormalities, and an abnormal extensor tendon mechanism.

Frequent stretching of the finger combined with splinting is recommended initially. For a progressive deformity in which flexion of the proximal interphalangeal joint is greater than 60 degrees, surgery is an option, but results are variable.[11] The procedure involves release of all tight volar soft-tissue structures, including the flexor digitorum superficialis and the abnormal lumbrical. Transfer of the flexor digitorum superficialis to the extensor mechanism[5] and osteotomy of the proximal phalanx[2] have also been reported. The latter procedure will change the arc of motion of the proximal interphalangeal joint, but will not improve it.

Clinodactyly

Clinodactyly is lateral deviation of a digit. It is most common in the small finger and appears as radial deviation of the middle phalanx or distal interphalangeal joint,[1] usually due to an abnormally shaped trapezoidal middle phalanx.

The incidence in otherwise-normal children ranges from 1% to 20%.[2] In persons with Down's syndrome, the incidence ranges from 35% to 79%.[2] Clinodactyly is more common in males and is usually bilateral.[6] It frequently presents as an autosomal-dominant trait, but may occur sporadically.[1] Associated anomalies include brachydactyly and macrodactyly.[1] This deformity should be differentiated from that caused by a delta phalanx.

Treatment is rarely indicated.[1] Stretching and splinting are not effective. Parents may request surgery for the cosmetic deformity. If this is thought to be warranted, an ulnar-based closing-wedge osteotomy is recommended, as it is the simplest technique.

Delta Phalanx

A delta phalanx is an abnormal triangular bone with a C-shaped epiphysis (Fig. 4). The abnormal shape of the growth plate is responsible for the progressive angular deformity. Delta phalanx appears most commonly as an extra phalanx of the thumb or as the middle phalanx of the small finger.[6] The affected thumb and small finger usually angulate toward the central axis of the hand.

The exact incidence of delta phalanx is unknown. It is more common in males and is usually bilateral.[2] Associated anomalies include syndactyly and polydactyly.[1]

Treatment with splinting and stretching is not effective. Surgical treatment is indicated for a severe cosmetic deformity and severe angulation causing functional impairment. A closing-wedge osteotomy of the delta phalanx combined with destruction of at least part of the physis is recommended. If possible, this procedure should be performed in the older child. [2]

Kirner's Deformity

Kirner's deformity is volar curvature of the distal phalanx. It is most common in the small finger and is not usually clinically apparent until 8 to 12 years of age.[1] This deformity occurs in approximately 1 of 410 live births, is more common in women,

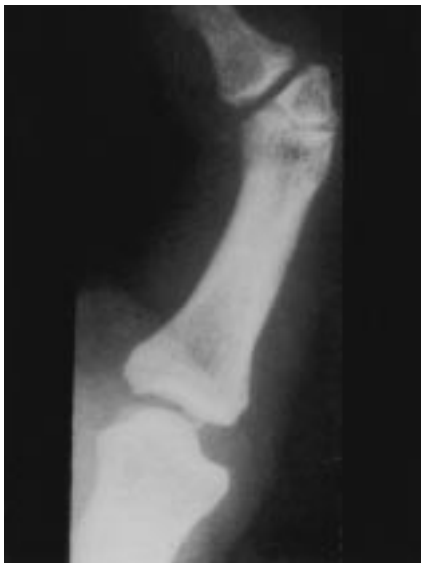


Fig. 4 Delta phalanx of the thumb.

and is usually bilateral.[6] Sporadic occurrence or transmission as an autosomal-dominant trait may occur.[5] Trauma, infection, and the residual effects of frostbite should be considered in the differential diagnosis. Associated anomalies include Cornelia de Lange, Silver's, and Turner's syndromes.[5] In the severely deformed finger, an osteotomy can be considered.[12]

Symphalangism

Symphalangism is the failure of finger interphalangeal joint development. Motion and skin creases are absent. Radiographs show a narrowed joint space or bone fusion.

Symphalangism represents 1% of all congenital upper-extremity anomalies.[5] An autosomal-dominant trait,[2] the deformity occurs most commonly in whites but rarely in blacks. Associated anomalies include syndactyly and foot deformities.[2]

Surgery is seldom warranted, as most patients adapt to the deformity. If function is severely compromised, arthrodesis in a more functional position may be consid-

ered.[1] However, Flatt's results indicate that surgery is rarely effective.[2]

Thumb Anomalies

Thumb Hypoplasia

Thumb hypoplasia ranges from minimal shortening to complete absence. The incidence is 4% of all congenital hand anomalies.[2] The deformity can occur sporadically, by genetic transmission,[5] or as part of a number of syndromes, most commonly those involving the cardiovascular system, the gastrointestinal tract, and the spinal column. The condition may also be seen in association with other congenital upper-extremity anomalies, such as radial club hand.[3] Blauth has classified thumb hypoplasia into five types.[2]

Short Thumb

The thumb is considered short when it does not reach the level of the proximal interphalangeal joint of the index finger. This is usually due to hypoplasia of osseous structures. Rarely is function compromised.

Short thumb is often associated with certain syndromes, including Fanconi's and Holt-Oram syndromes.[6] In these conditions, the thumb metacarpal is short and slender. When a short, broad metacarpal is seen, myositis ossificans progressiva, dystrophic dwarfism, hand-foot-uterus syndrome, and Cornelia de Lange syndrome should be considered.[6] The presence of a short, broad distal phalanx suggests brachydactyly, Rubinstein-Taybi syndrome, Apert's syndrome, or Carpenter's syndrome.[6]

Surgical treatment is rarely necessary. If the thumb is excessively short and compromises pinch, either web-space deepening or distraction lengthening should be considered.

Adducted Thumb

The adducted thumb is short, with a tight web space and deficient thenar musculature. There may also be deficiency of the collateral ligaments of the metacarpophalangeal (MCP) joint. Surgery, including release of the thumb web and tendon transfer to provide thumb opposition, is recommended. A two- or four-flap Z-plasty may improve a less severe tight thumb web. The more severe type requires fascial release with a dorsal flap or skin graft. Opponensplasty options include transfer of the abductor digiti minimi, the flexor digitorum superficialis, or the extensor digiti minimi.[6,13]

Abducted Thumb

This deformity arises because of the abnormally shaped insertion of the flexor pollicis longus into the distal phalanx.[2] One slip inserts into the volar distal phalanx, and the other passes dorsally and radially to join the extensor pollicis longus. This abnormal insertion is responsible for abduction with flexor pollicis longus contraction. Thumb metacarpal hypoplasia, thenar muscle absence, web-space deficiency, and severe MCP joint instability are present. The thumb metacarpal is adducted, but the thumb MCP joint is abducted due to MCP instability and the pull of the flexor pollicis longus.[2] Manske and McCarroll[14] have differentiated the abducted thumb into two types. In type A, the carpometacarpal joint is stable, and reconstruction is justified. In type B, the carpometacarpal joint is unstable, and pollicization is recommended.

Surgical treatment for type A deformity should correct the web-space deficiency, release the adduction contracture, restore stability to the MCP joint, release the abnormal flexor pollicis longus slip, and provide opposition.[5] Web-space correction usually requires a dorsal-flap procedure or skin graft. Adduction-contracture release is accomplished by palmar and dorsal release of the adductor pollicis fascia and possibly the first dorsal interosseous muscle. Both opponensplasty and correction of MCP joint instability can be accomplished by transfer of the flexor digitorum superficialis of the ring finger, as described by Riordan.[5]

Pouce Flottant

The pouce flottant is a short, unstable thumb that is positioned more distally and radially than normal on the hand. It is connected to the hand by a slender pedicle, which contains a neurovascular bundle. The proximal and distal phalanges are hypoplastic, and the metacarpal is either rudimentary or absent. Attempts to stabilize the thumb have not been as useful as amputation with index pollicization at 6 to 12 months of age.[6,8]

Absent Thumb

The absent thumb is treated with index pollicization.[8]

Thumb Duplication

In this condition, both the radial and the ulnar duplicates of the thumb display some degree of hypoplasia, although the radial duplicate is usually affected more.[2] The intrinsic muscles innervated by the ulnar nerve insert on the ulnar duplicate, and the intrinsic muscles innervated by the median nerve insert on the radial duplicate.[6] The digits may be angulated, and the joints may be stiff. Significant anatomic variations of the neurovascular pattern exist. Flexor and extensor tendons may be duplicated and eccentrically placed. The nail may be shared or completely duplicated. Wassel has classified this deformity into seven types[6] (Fig. 5). Type IV is the most common.

Thumb polydactyly occurs in 8 of every 100,000 births.[5] It is most commonly seen in the white and Asian populations, is more common in males, and is usually unilateral. Most cases occur sporadically; however, an autosomal-dominant pat-

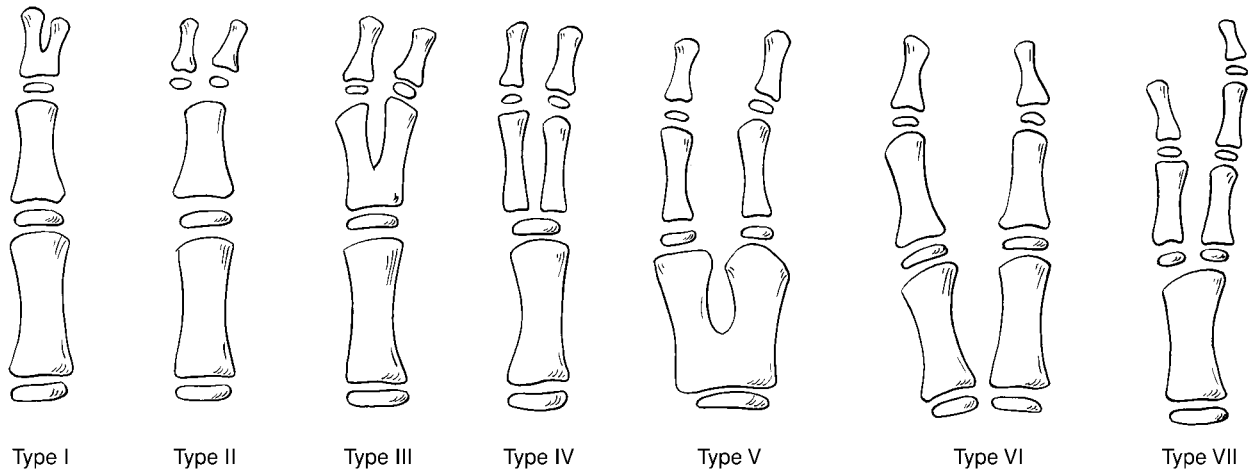


Fig. 5 Wassel classification of thumb polydactyly. Type I is characterized by a bifid distal phalanx; type II, duplicated distal phalanx; type III, bifid proximal phalanx; type IV, duplicated proximal phalanx (most common); type V, bifid metacarpal; type VI, duplicated metacarpal; type VII, thumb polydactyly with associated triphalangeal thumb.

tern is seen when associated with triphalangeal thumb. Associated anomalies are rare.[5]

Surgical treatment is indicated for this deformity. Flatt[2] recommends correction between 6 and 9 months. For types I and II with digits of equal size, Bilhaut recommends combining elements of both digits.[1] Care must be taken at surgery to ensure normal articular contour of the distal phalanx. For unequal digits, excision of the smaller digit with collateral ligament reconstruction is recommended.

Treatment must be individualized for more complex deformities. For type IV deformity, the least developed digit, usually the radial duplicate, is amputated. Appropriate intrinsic and extrinsic tendon transfers are performed, along with radial collateral ligament reconstruction.[6] Osteotomy or growth-plate arrest is occasionally required to improve bone alignment.[1]

Congenital Trigger Thumb

Trigger thumb is due to constriction of the A1 pulley, which affects gliding of the flexor pollicis longus. The thumb is held flexed at the interphalangeal joint, and there is often pain at the level of the volar MCP joint. A click may be noted with attempted extension. The anomaly represents 2% of all congenital hand deformities.[2] The involvement is usually bilateral, and 25% of cases are noted at birth. Occurrence is sporadic, and association with other anomalies is rare.[5]

According to Dinham and Meggitt,[15] approximately 30% of all congenital trigger thumbs detected at birth resolve by age 1. We therefore recommend observation for 1 year to allow for spontaneous correction. Splinting during this time is of little benefit. Surgical division of the A1 pulley is recommended if triggering persists after this period.

Congenital Clasped Thumb

Clasped thumb describes the flexed and adducted thumb position. All neonates hold the thumb in a flexed and adducted position until 3 months of age.[1] Normal children initiate movement at this time; however, affected children do not. The incidence of congenital clasped thumb is unknown. It is more common in males and is frequently bilateral.[6]

Weckesser et al[16] have classified congenital clasped thumb into four types. Type 1, the most common form, is characterized by absence of extensor function. Type 2 is an extension deficiency with associated flexion contracture. Type 3 has associated thumb hypoplasia. All cases that cannot be classified into the first three groups are included in type 4.

Initial treatment is serial casting in extension and abduction for 3 to 6 months. This will stretch contracted soft tissues and allow development of hypoplastic muscles.[2] Most patients respond to this regimen.[1] If conservative treatment fails, this indicates absence rather than hypoplasia of the extensor mechanism, and surgery is indicated. The extensor mechanism should be inspected at surgery; if it is found to be deficient, extensor indicis proprius transfer is recommended.[2] Associated thumb hypoplasia is treated as discussed previously.

Duplication (Polydactyly)

Polydactyly is the duplication of a digit. It is one of the most common congenital hand anomalies.[6] The incidence is 1 in 300 in the black population and 1 in 3,000 in the white population.[2] Polydactyly is separated into three types: preaxial, central, and postaxial. Preaxial is most common in white and Asian persons; postaxial, in black persons.[2] Postaxial is the most common overall.[6]

Stelling and Turek have classified polydactyly into three types.[5] Type 1 is characterized by an extra soft-tissue mass without skeletal elements. In type 2, a normal-appearing digit articulates with the phalanx or metacarpal. A type 3 digit has its own metacarpal. Preaxial polydactyly has already been discussed in the section on thumb duplication.

Central polydactyly involves the index, long, and ring fingers. The extra digit is usually a type 2 deformity, and the ring finger is most commonly involved.[5] This anomaly may be transmitted as an autosomal-dominant trait and is often associated with toe polydactyly and syndactyly.[6]

The treatment of central polydactyly is usually surgical. The anatomy of the neurovascular bundles varies considerably. Flexor and extensor tendons may be hypoplastic or absent. Excision of the least developed digit is recommended in the preschool years.[1] If the duplicated digit articulates with the metacarpal or phalanx, removal is indicated in the first year of life to prevent angular deformity.[1] If amputation is at the level of a joint, the collateral ligaments must be preserved or reconstructed to prevent instability. If the affected digit is stiff and nonfunctional, Flatt[2] recommends amputation and conversion to a three-fingered hand to improve function.

Type 1 postaxial polydactyly involves one or two genes with incomplete penetrance. Types 2 and 3 are inherited as dominant traits with marked penetrance.[5] The offspring of patients with type 1 deformity can have only this type. The offspring of patients with type 2 or type 3 can have any type.[5] White children with postaxial polydactyly are more likely to have an associated anomaly or syndrome.[6] Associated anomalies are uncommon in black children.

Treatment of type 1 postaxial polydactyly is simple ligation of the extra digit shortly after birth. Type 2 deformities are technically the most difficult to treat. Care must be taken to preserve important structures, such as the ulnar collateral ligament of the MCP joint of the small finger and the abductor digiti quinti insertion. For type 3 deformities, simple excision of the extra digit is recommended.

Overgrowth (Macrodactyly)

Macrodactyly, or gigantism, is the enlargement of all elements of an affected digit. Bone, nerve, blood vessels, and skin are also involved. This is in contrast to enlargement secondary to hemangioma, lymphangioma, arteriovenous fistulae, polyostotic fibrous dysplasia, and osteoid osteoma.[2] The involved digit is usually stiff, angulated, and unattractive. Hypertrophy of the median and ulnar nerves may occur due to the development of extra fat and fibrous tissue.[5]

Two forms of macrodactyly have been reported.[2] In the first, which is noted at birth, the affected digit subsequently grows at the same rate as other digits. In the second, and most common, form, the affected digit is large at birth and continues to grow disproportionately. This anomaly represents 1% of all congenital hand anomalies.[2] Ninety percent of cases are unilateral.[2] The index finger is most commonly involved.[6] Multiple-digit involvement occurs more often than single-digit involvement[6] (Fig. 6). The etiology is unknown; possible causes include an abnormal blood supply, nerve supply, or hormonal mechanism.[5] Associated anomalies are rare.

Surgical treatment is often necessary because the digit impairs function. A debulking procedure is done to remove excess skin and fat. Debulking procedures for each side of a digit should be separated by 3 months to avoid vascular compromise.[6] Osteotomy is required to correct angular deformities. Amputation may also be considered if the thumb and the other three fingers are normal.

The timing of the procedure depends on the size of the hypertrophic finger and the size of the parent's finger. An epiphysiodesis should be performed once the affected digit reaches the same size as the digit of the parent of the same sex. For digits longer than those of the parent of the same sex, bone-shortening procedures are indicated.[1] Tsuge[17] has described a shortening procedure in which portions of the distal and middle phalanges are combined. Barsky[18] recommends a less technically demanding procedure in which a portion of the middle phalanx is excised

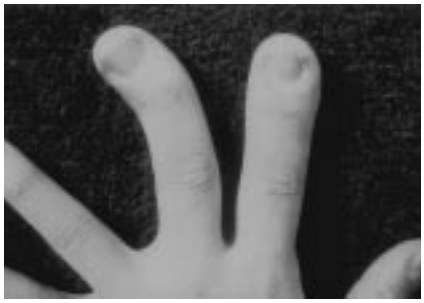


Fig. 6 Macrodactyly of the index and long fingers.

and the distal and middle phalanges are fused.

Carpal tunnel release with median nerve neurolysis may arrest progressive digital enlargement.[1] When a digital nerve is significantly enlarged, excision may be necessary for successful debulking.[17] The area supplied by the abnormal nerve usually has abnormal sensation, which is generally not altered by surgical resection.[1] Careful assessment is necessary before resection.

Constriction Bands

A constriction band is a deep skin crease encircling a digit, which causes varying degrees of vascular and lymphatic compromise. Patterson classified constriction bands into four types.[1] Type 1 is a mild transverse or oblique digital groove. Type 2 is a deeper groove with an abnormal distal part. Type 3 is characterized by incomplete or complete syndactyly of the distal part (acrosyndactyly). Type 4 is a complete amputation distal to the constriction.

Constriction bands occur in 1 of every 15,000 births.[5] Distal constriction bands are most common, and central digits are most frequently affected.[6] Occurrence is generally sporadic. The majority of cases are associated with other congenital hand anomalies, including syndactyly, hypoplastic digits, and brachydactyly.[2] Approximately 40% of patients have congenital anomalies that do not affect the upper extremity,[1] including clubfoot, cleft lip and palate, and craniofacial defects.[5]

The treatment of incomplete, type 1 constriction bands often is observation. Newborns with constriction severe enough to cause vascular impairment require surgical release with the use of multiple Z-plasty closure. The depth of the constriction band determines whether complete or partial band release is indicated. Clinical experience has shown that the entire band can be released if it is shallow. If the band is deep, only one side of a digit is released at each procedure to avoid vascular compromise. Procedures should be separated by approximately 3 months.[2]

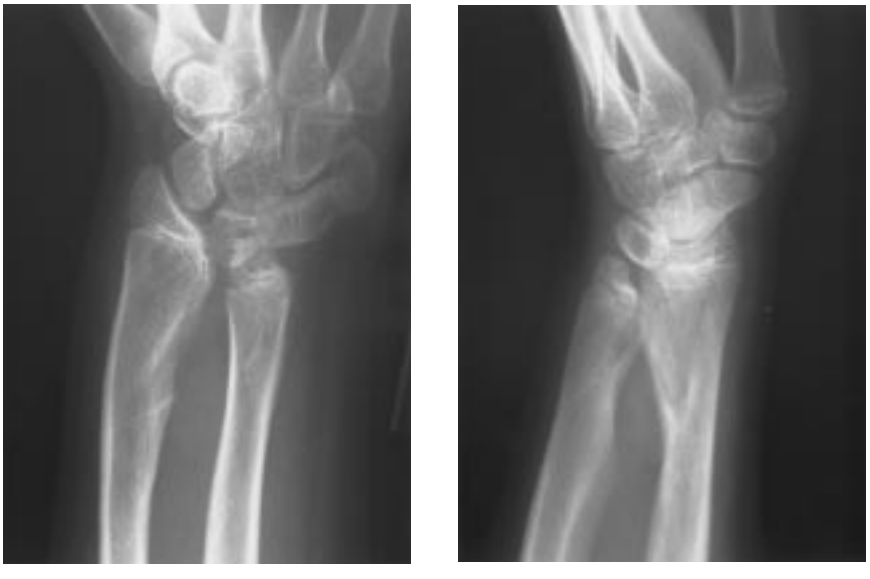
Acrosyndactyly separation should be initiated at 6 months of age, as multiple procedures are required. Release of bordering digits is accomplished first, followed by central-digit correction.[6]

Madelung's Deformity

Madelung's deformity is the abnormal growth of the volar and ulnar distal radial epiphysis. It is characterized by ulnar and volar tilt of the distal radial articular surface; premature fusion of the ulnar half of the radial physis; a short, bowed radius; a short ulna; and wedging of the carpus between the distal radius and ulna. The ulnar head is enlarged and dorsally subluxated (Fig. 7). The deformity usually becomes apparent in late childhood or early adolescence.[5] Variable degrees of limited motion and pain due to ulnocarpal impingement are seen.

The incidence is 2% of all congenital hand anomalies.[6] Girls are affected more often, and bilateral deformity is twice as common as unilateral deformity.[6] The etiology is unknown, although transmission as an autosomal-dominant trait has been shown.[5] An abnormality similar to Madelung's deformity may be seen in dyschondrosteosis, achondroplasia, multiple exostosis, multiple epiphyseal dysplasia, and Ollier's dyschondroplasia.[5]

Many patients become symptomatic during the adolescent growth spurt. Initial treatment should be splinting, as this alone can provide complete and permanent relief. Patients with persistent pain may require surgical treatment. Radial osteotomy combined with an ulnar-shortening osteotomy and radial osteotomy combined with distraction lengthening of the radius are two surgical options.



A B

Fig. 7 Anteroposterior **(A)** and lateral **(B)** radiographs of the wrist of a patient with Madelung's deformity.

Summary

Congenital deformities of the upper extremity are diverse. Most such anomalies are minor and cause no functional deficits, and surgery is therefore unnecessary. If a severe cosmetic deformity is present or there is significant functional compromise, however, surgical treatment is indicated. Complete understanding of the pathoanatomy of these anomalies is essential before embarking on surgical treatment.

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