Malformations of the Hand

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13.1 Introduction

Congenital anomaly of the hand is an area that not every hand surgeon engages in routinely. How do hand surgeons set priorities when dealing with congenital anomalies of the hand? Congenital anomalies are often complicated structural abnormalities, so the question of priority would really deserve close scrutiny. Before every surgical intervention, an analytical prioritization would help significantly to achieve a functional restoration. The relationship between structure and function of the hand is so intimate that in any structural correction of the hand, the priority should be on the functional restoration rather than a simple structural correction. In other words, with the exception of pure cosmetic corrections under very special circumstances, functional restoration or improvement overrules structural corrections. Functions of the hand should not be defined vaguely; these have been identified and labeled pretty well in the past. These functions include three different types of grip: power, diagonal, and hook; three different types of pinch: tip pinch, side pinch, and palp pinch, fulfilling different demands on physical activities, sensory feeling, and, lastly, expression. All surgical planning should pay full attention to the functional goal we want to achieve. Although the physical ability of the individual has a lot to do with the functional achievement based on intact hand structure, one still has to observe that structural integrity is not everything. By that, we are talking about the length and position of the digits, the mobility and stability of the joints, the strength of the components, and the sensation of the tactile parts. Without a proper length and favorable position, no digit could function. Without stability and mobility, no joint could function. Without reasonable strength, no hand could function properly, and, without sensation, a hand would not be able to protect itself, not to mention precision performance. These components need to be considered together. For example, good length without stability or mobility is meaningless.

Key Points

Congenital differences of the upper limb occur in approximately 0.16–0.18% of live births. Approximately 10% of these infants will have a partial or complete absence of the involved limb, leading to serious loss of function.

Because limb formation occurs concurrently with other organ development, it is

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important to be aware of associated abnormalities, including cardiac, hematopoietic, or tumorous conditions. Communication with the pediatrician is important in establishing a comprehensive diagnosis and for staging and planning any reconstructive procedures. A multidisciplinary approach will provide superior outcomes by addressing all aspects of the physical and emotional state of both the patient and the family.

Because congenital anomalies of the upper limb are a significant challenge, the hand surgeon or reconstructive surgeon, as the team leader and primary decision-maker, has a unique opportunity to positively and directly affect the child's growth and development.

A child achieves bimanual palmar grasp by the age of 9 months and learns three-digit pinch between 1 and 2 years. Patterns of hand–eye coordination have been established by age 3. Therefore, successful technical reconstruction may fail to alter already established fixed functional or psychological patterns if the reconstruction is not completed by approximately 4 years of age. Ideally, the reconstruction should be completed by school age to allow for easier social transitioning [1].

13.2 Principles Governing Pediatric Management

It would be wrong to consider children as small adults. Correction of deformities needs to take into consideration the age and the growth, not only the structural dimensions. Although growth plates and growing epiphysis are vulnerable to injury, it did not mean that they could not be touched. Perhaps as long as the epiphyseal plate is untouched, the epiphysis could be cut flat and stitched to its adjacent component in an attempt to achieve linear fusion, trying not to affect the optimal growth. Similarly, enlarged or abnormally faceted epiphyses could be shaved to give more congruent articulating surfaces without affecting the ultimate growth of the components handled. Tada and Yonenobi used this technique extensively in the Duplex thumb to bring more satisfactory alignment of the retained component after sacrificing the unwanted component. Young children do not tolerate pain and limitations of activities. Henceforth, dressing has to be double safe and better protected, pinning has to be secure and hidden, and casts need to go up one proximal joint (e.g., including the elbow) in order to avoid loosening. Taking care of the postoperative child should include paying attention to his psychological needs. The hand often reflects the mind. The hand is an important body image second only to the face. It is therefore easy to understand how a structurally abnormal hand could significantly affect a child. If possible, correction should be completed by school age [2]. It would be wrong to assume that severe psychologic disturbances exist only in children with severe anomalies and not in those who are not so seriously affected (duplication or syndactyly). In fact, the actual outcome of possible psychologic disturbances depends on environmental conditions, individual circumstances, and immediate family support. It should be mandatory for every child with a hand anomaly to receive a proper psychological assessment [3].

Radiologic images (perhaps inaccurate imaging of a growing child, unossified epiphyses) need to be interpreted with extra caution and with a sound knowledge of ossification centers; otherwise, fractures and displacements could be misdiagnosed. Likewise, correct interpretation about length issues cannot be accurately given. The varieties of imaging have expanded extensively in recent years so that congenital anomalies could acquire many benefits. Although conventional radiography and angiogram still served routinely for structural identification and delineation of vascular patterns, disappointments are not uncommon. Ultrasonic examinations could help in recognizing inflammation conditions such as tendonitis. The same device and technique could be useful to help identify thickened soft-tissue structures and to differentiate tissue planes

between the normal and abnormal tissues. Conventional tomography and computed tomography of the radioulnar joint and the wrist are used in patients with persisting complaints or doubtful findings on plain radiographs and difficult anatomical situations. Suspected ligamentous injuries of the wrist, including tears of the triangular fibrocartilage complex, are evaluated by wrist arthrography or magnetic resonance imaging, the latter requiring a highly skilled imaging and interpretation technique. Magnetic resonance imaging is the method of choice for the detection of osteonecrosis. The use of real-time sonography allows a reliable diagnosis of the cystic or solid nature of soft-tissue tumors and accurate estimation of their volume and their precise 3-dimensional localization. Sonography facilitates the location of foreign bodies and appears as a new promising technique for the evaluation of tendons [4].

Key Points

In children, the neurovascular bundle is much more stretchable than in adults. However, structural anomalies could well be associated with neurovascular anomalies. Awareness is much more important than special investigations, which cannot be taken as routine.

Tips and Tricks

To correct the alignment, very often the surgeon has to deal with joint surfaces and epiphyseal articulations. It is important to remember that as long as the growth plate is not touched, growth disturbance will not occur. If desired, therefore, the cartilage end could be shaved. Surgery these days tends to become gadget driven and gadget dependent. Surgical procedures should be followed closely, step by step. However, surgery performed for children with anomalies demands extra flexibilities on the operating table. Immediate decisions have to be made whether to stretch the neurovascular tissues or not; perhaps whether to end the surgery for a two-staged maneuver or to continue on to completion; whether to attempt perfection or to accept some defects; whether to use a fixation device or rely on casting alone; and so forth.

13.2.1 Reconstruction of Congenital Differences of the Hand

Expansion of the discipline of hand surgery and heightened interest in congenital problems have resulted in major advances in the treatment of congenital hand anomalies over the past 25 years. Increased experience with congenital anomalies of the hand has expanded the hand surgeon's knowledge of patterns and relationships between different anomalies, resulting in new methods of classification and more logical approaches to treatment. The principles of treatment of the more common anomalies, such as syndactyly, established by prior generations of hand surgeons have been refined in details of technique. New technologies, such as distraction lengthening and free vascularized transfers, have allowed the surgeon to treat new problems and old problems in new ways. In spite of our successes, much remains to challenge hand surgeons in this new millennium, especially in the construction of joints and the expanding field of fetal surgery.

13.2.1.1 Timing of Treatment

Treatment options and timing of treatment depend on the anomaly, although most advocate that surgical correction should be performed within the first 2–3 years of life to allow for maximal growth, development, and use. In addition, early treatment improves scarring and decreases the psychological impacts of congenital hand syndromes. Despite that, early operative procedures prove to be technically challenging. Noninvasive treatment options may be advisable in the first 1–2 years of life as the child grows.

These options include splinting, stretching, physical therapy, and prosthesis for increased function and cosmesis [1].

13.3 Epidemiology

Congenital hand conditions are common, occurring in 2.3 per 1000 of total births or 0.16% –0.18% of the population of the United States, with incidence varying according to region and ethnicity. Ekblom et al. reported a worldwide incidence of 21.5 per 10,000 live births. Lamb et al. and Giele et al. reported the overall prevalence of congenital hand anomalies to be 11.4–19.7 per 10,000 live births. Of the anomalies included, failure of differentiation is the most commonly reported, followed by failure of duplication and failure of formation anomalies. Polydactyly is the most common individual diagnosis. Although overall only slight variation exists in the prevalence of major anomalies among different regions or ethnic populations, there are exceptions. For example, ring constriction syndrome is 4 to 6 times more prevalent in Japan than in Scotland. Ulnar polydactyly is more common among those of African descent. In general, males are more likely to be affected by congenital hand anomalies than females with an overall ratio of 3:2. The prevalence of congenital hand anomalies increases with maternal age. Mothers older than 40 years are twice as likely to have a child with hand deformity than mothers younger than 30 years, 5–20% of upper limb anomalies occur as a part of a known syndrome, and 50% occur bilaterally. Up to 17% of patients will present with multiple upper limb anomalies, and up to 18% of patients will die before the age of 6 years because of other concurrent congenital disorders [5].

Because limb formation occurs concurrently with other organ development, it is important to be aware of associated abnormalities, including cardiac, hematopoietic, or tumorous conditions. Communication with the pediatrician is important in establishing a comprehensive diagnosis and for staging and planning of any reconstructive procedures. A multidisciplinary approach will provide superior outcomes by addressing all aspects of the physical and emotional state of both the patient and the family.

Therefore, successful technical reconstruction may fail to alter already established fixed functional or psychological patterns if the reconstruction is not completed by approximately 4 years of age. Ideally, reconstruction should be completed by school age to allow for easier social transitioning.

13.4 Embryology of Limb Development

During embryonic development, the upper extremity develops from the arm bud, a mass of mesoderm-derived mesenchyme covered by ectoderm. The Hox genes (HoxA, HoxB, HoxC, and HoxD) are responsible for regulating limb development in the human embryo. Sonic hedgehog, fibroblast growth factor, and Wnt-7a are some of the known signaling proteins that control Hox gene expression. Hox gene products act on competent mesenchymal cells within the limb bud, guiding these cells to form condensations at the appropriate time and location. These condensations form the precartilaginous skeletal foundation of the limb. The limb must form simultaneously across three anatomical axes: proximal to distal axis, dorsal to palmar axis, and anteroposterior (preaxial/ postaxial) axis. The apical ectodermal ridge forms as a thickening of ectoderm at the leading edge of the limb bud and, through its interactions with the underlying mesenchymal cells, is responsible for proximal to distal differentiation of the limb. The dorsal ectoderm helps to control the dorsal to palmar axis of differentiation, leading to distinct flexor and extensor surfaces of the hand and arm. The zone of polarizing activity is a condensation of mesenchymal cells on the preaxial surface of the limb bud. This zone signals the anteroposterior formation of the limb bud by setting up a gradient of signaling proteins along this axis. The arm bud begins as an outgrowth from the ventrolateral wall of the developing embryo and appears at approxi-

mately 30 days' gestation. Located opposite the fifth through seventh cervical somites, the arm bud precedes lower extremity development throughout embryogenesis. At 33 days' gestation, blood circulation develops within the bud, which has established a flipper-like appearance. By 38 days, blood vessels have become apparent growing from proximal to distal, and a constriction marks the separation of the forearm from the upper arm. Finger development is apparent by day 44, with five distinct mesenchymal separations. By day 52, the digits are completely separated because of apoptosis of the intervening mesenchymal tissue. This orderly resorption of tissue occurs through the release of lysosomal enzymes as cells migrate toward the digital condensations to participate in chondrogenesis. By approximately the seventh week of gestation, the limb bud rotates 90 degrees on its long axis with the elbow positioned dorsally. By the eighth week of gestation, embryogenesis is complete. After the eighth week, the small but completely formed upper limb continues to grow in size and primary ossification centers replace areas of cartilage to complete development [6–10].

13.5 Classification

Several classification schemes for congenital upper limb malformations have been conceived. The current classification scheme has been agreed on by the American Society for Surgery of the Hand and the International Federation of Societies for Surgery of the Hand and was first published by Swanson. This classification comprises seven groups based on abnormalities of embryogenesis: failure of formation of parts, failure of differentiation of parts, duplication, overgrowth, undergrowth, constriction ring syndrome, and general skeletal abnormalities. Many of these groups are further subdivided by the anatomical level of the malformation. In this chapter, the International Federation of Societies for Surgery of the Hand classification scheme is used to organize the discussion on congenital hand surgery [11–13].

13.5.1 Failure of Formation of Parts: Transverse Arrest

13.5.1.1 Transverse Deficiencies

Transverse deficiencies of the upper limb may occur at any level from the shoulder to the phalanges. Transverse arrest most commonly occurs at the level of the proximal third of the forearm and at the wrist. Digital appendages, or nubbins, are often found at the end of the limb. Transverse deficiencies are usually isolated, unilateral, and sporadic. These defects are thought to be the result of vascular disruption at some point during embryogenesis of the upper limb. Transverse deficiency differs from constriction ring syndrome; the latter tends to be hypoplastic at the same level in that proximal parts.

13.5.2 Proximal Transverse Deficiencies

With proximal transverse deficiencies, treatment is usually a prosthetic device. These devices may be static or dynamic and may be controlled by remaining skeletal structures or myoelectric impulses. For children with transverse deficiency at the wrist or metacarpal level, a volar paddle prosthesis may act as a post against which the remaining carpus or metacarpals may be flexed. In bilateral deficiencies, children often become adept at using their lower extremities to perform activities of daily living. Surgical options for proximal transverse deficiencies may include removal of functionless digital nubbins, stump revision to allow for prosthetic fitting, and excision of excess or functionless parts. In children with bilateral deficiencies and visual impairment, the Krukenberg procedure is advocated. This procedure separates the distal ulna from the radius, allowing for the opposition of the two bones during supination of the forearm.

Key Points

Prosthetic devices are the treatment of choice for children with proximal transverse deficiencies.

13.5.3 Transverse Arrest of the Digits Distal to the Metacarpal Level

Transverse arrest of the digits distal to the metacarpal level, sometimes referred to as symbrachydactyly, has been treated by conventional techniques of distraction lengthening and nonvascularized toe phalangeal bone grafting. Metacarpal or phalangeal lengthening uses the principles of distraction osteogenesis to form new bone. A distractor is placed spanning a metacarpal or phalangeal osteotomy or corticotomy, and the bone is distracted 0.5–1 mm per day for 3–6 weeks until the desired digit length is achieved. The bone gap may consolidate with regenerate bone or may require secondary autogenous or allograft bone grafting. Transverse deficiencies of the digits may also be treated with nonvascularized toe phalangeal bone grafting from the proximal phalanges of the second, third, or fourth toe. Up to 1.5 cm of length can be achieved with each proximal phalanx graft. Whether the epiphysis of a toe phalangeal bone graft continues to grow remains controversial. It has been recommended that toe phalangeal bone grafts should be performed before 15 months of age, that the bone should be harvested extraperiosteally, and that the collateral ligaments and tendons should be reattached to provide the optimal conditions for the physis to remain open and thus maintain growing potential. Free microvascular toe-to-hand transfer is becoming an increasingly accepted method for treatment of these patients. The first toe-to-hand transfer was performed by Nicoladoni in 1897 for a traumatic thumb amputation and required multiple stages to preserve the blood supply to the transferred toe. In 1955, Clarkson reported the first series of congenital toe-to-thumb transfers with 15 transfers in six patients. Because multiple stages required immobilization of the hand to the foot, the procedure fell out of favor. The first successful microvascular toe-to-hand transfer was reported by Cobbet in 1969 and led to the possibility of free toe transfers for congenital malformations. The first toe-to-hand transfer to reconstruct a congenital anomaly was performed by O'Brien et al. in 1978. In 1995, Vilkki reported a series of 18 successful congenital toe transfers, with an 11-year follow-up proving that toe transfer was beneficial in this population. Several congenital anomalies have been treated with toe transfer, including transverse deficiency, longitudinal deficiency, traumatic amputation, vascular malformations, and constriction ring syndrome. Studies have shown that growth potential is retained in the transferred toe. Epiphyseal plates remain open, and bone growth is comparable to that of the corresponding toe on the contralateral foot. A long-term study of toe-to-hand transfers in post-traumatic deformities has shown good hand function and acceptance of the transferred digit up to 20 years after the procedure. Transverse deficiency of the thumb is an ideal indication for free microvascular toe-to-hand transfer. Unlike longitudinal thumb deficiencies, the proximal thumb remnant tends to retain some normal anatomy, including a mobile carpometacarpal joint, thenar muscles, and proximal stumps of the flexor pollicis longus and extensor pollicis longus tendons. In such cases, a microsurgical second toe-to-thumb transfer is a better option than pollicization of the index finger. In children with a thumb but the absence of all four fingers or with the complete absence of all five digits, bilateral second toe transfers can be performed. The two toe transfers can provide three post pinch to a remaining thumb or one toe transfer can be used to reconstruct the thumb and the other toe transfer can be used to create a digit for pinch activity. The child's family should be carefully counseled regarding the limitations and potential complications before proceeding with this extremely difficult reconstruction [14, 15].

Tips and Tricks

- Toe phalangeal bone grafts should be performed before 15 months of age.
- The bone should be harvested extraperiosteally.
- The collateral ligaments and tendons should be reattached.

13.5.4 Failure of Formation of Parts: Longitudinal Arrest

13.5.4.1 Radial Longitudinal Deficiency

Radial longitudinal deficiency, or radial club hand, involves approximately 1 in 30,000 to 1 in 100,000 live births and is more common in boys than in girls and affects Caucasians more often than other races. The deformity is bilateral up to 50% of the time and, when unilateral, affects the right side more often than the left (Fig. 13.1a, b).

Radial longitudinal deficiency is often found in association with other malformations of the hematopoietic, cardiac, genitourinary, and skeletal systems, including Fanconi anemia, TAR syndrome (thrombocytopenia, absent radius), Holt–Oram syndrome (cardiac defects), and the VATER association (vertebral anomalies, anal atresia, tracheoesophageal fistula, renal defects). Radial longitudinal deficiency has been classified into four types based on the severity of the involvement of the radius. Thumb hypoplasia is often present, ranging from slight to total absence. The scaphoid and trapezium may also be absent. Radial sided digits often exhibit a flexion deformity or camptodactyly. The small finger is often unaffected, with relatively normal function. Short, fibrotic muscles run along the radial side of the forearm and insert into the ulna, causing severe bowing. The ulna may also be short, and the distal humerus is often hypoplastic, leading to stiffness of the elbow. Both the radial artery and radial nerve may be absent. The median nerve is always present and courses superficially below the skin in the radial concavity, making it prone to injury during operative exposure. Radial longitudinal deficiency results in very poor function in the affected hand because of the flexed position of the radially deviated hand, loss of wrist support, poor flexor/extensor tendon excursion, hypoplasia of the thumb, and stiffness of the elbow. These deformities increase with age, leading to deteriorating function. Psychologically, the appearance of the hand can be quite troubling for both the child and the parents. For these reasons, treatment should be done immediately after birth and consists of passive stretching exercises and serial casting to begin, centralizing wrist and hand on the remaining ulna. Contraindications to treatment include older patients who have adapted to their deformity and children with stiff elbows. In these circumstances, the radial angulation of the hand and carpus makes feeding and hygiene possible in the presence of a stiff elbow. Surgical treatment of radial longitudinal deficiency attempts to improve the appearance and function of the hand by stabilizing the carpus on

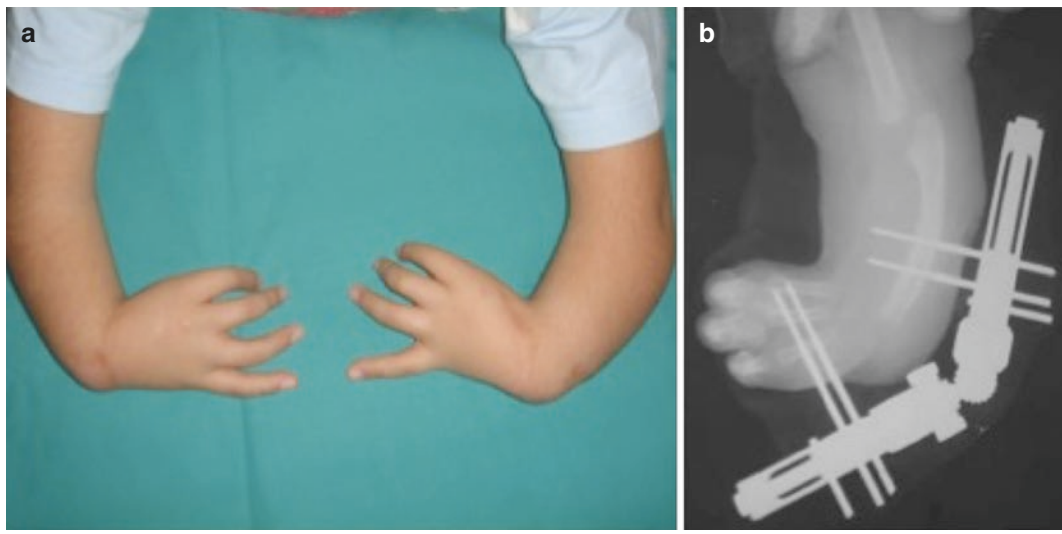

Fig. 13.1 Radial longitudinal deficiency; (**a**) clinical picture, (**b**) post-operative x-ray

the end of the ulna. Historically, centralization of the carpus over the ulna and bone grafting of the absent radius have been attempted, but centralization of the carpus only on the ulna remains the definitive treatment. Centralization is usually performed in the first year through a Z-plasty incision over the radial aspect of the wrist to release the tight skin envelope. After identifying the median nerve, the carpus is freed from the radial fibrotic muscle mass and then centralized over the ulna after transecting the brachioradialis, flexor carpi radialis, and extensor carpi radialis longus tendons. The lunate may need to be excised to fit the carpus over the end of the ulna. A longitudinal Steinmann pin is used to hold the middle finger metacarpal bone and carpus over the ulna for several months. Radial deforming tendons such as the flexor carpi radialis may then be transferred to the extensor carpi ulnaris to help rebalance the carpus [2].

Tips and Tricks

Buck-Gramcko has advocated radialization of the carpus in which the deformity is overcorrected to the ulnar side by placing the ulna along the axis of the index finger metacarpal bone. Preoperative distraction may be performed initially to allow the carpus to be radialized or centralized without the need for resection of carpal bones. If significant ulnar bowing is present, a corrective osteotomy or multiple osteotomies may also be performed and fixed with the same Steinmann pin to help straighten the long axis of the forearm.

13.5.4.2 Ulnar Longitudinal Deficiency

Ulnar longitudinal deficiency occurs in 1 in every 100,000 live births. The deformity is often sporadic and does not have the syndromic associations of radial longitudinal deficiency. However, approximately 50% of the patients will have some type of musculoskeletal abnormality, including the contralateral upper limb or the lower limbs. There is a clinical spectrum from hypoplasia of the ulna with an intact epiphysis to total absence of the ulna with radiohumeral synostosis. In all cases, a fibrous anlage tether replaces the missing ulna and inserts into the ulnar aspect of the carpus or the distal radius epiphysis. The flexor carpi ulnaris is absent, the ulnar and median nerves are present, but the ulnar artery is often absent. Unlike radial longitudinal deficiency, the wrist is stable, allowing for relatively normal digital function. The radial head may be dislocated, leading to pain or loss of function at the elbow. With the most severe deficiencies, the humerus is internally rotated and the forearm pronated, compromising the positioning of the hand. Treatment of ulnar longitudinal deficiency consists of serial casting to improve the wrist and elbow positions. Excision of the anlage is indicated for greater than 30 degrees of angulation or when the deformity is progressive. The anlage is approached through a lazy-S incision and resected off from the carpus or distal radius. Kirschner wires may be used to hold the wrist in a neutral position. Tendon transfers are not required; however, in severe bowing, a radial osteotomy may be required to help straighten the long axis of the forearm. When there is a loss of function at the elbow, the proximal radial head is resected, and a one-bone forearm is created by osteosynthesis of the distal radius to the proximal ulna. In the case of radiohumeral synostosis, a derotational osteotomy of the humerus may be required to place the hand into a more functional position. Arthroplasty of the elbow is not advised because of the low likelihood of success [16].

13.5.4.3 Central Ray Deficiency

Central ray deficiency, or cleft hand, was originally classified as either typical or atypical. Typical (true) cleft hand is caused by failure of development of the central digit of the hand, the middle finger, including the metacarpal bone, which leads to a deep V-shaped cleft. The border digits are occasionally involved in syndactyly with a tight first web space. Transverse bones separating the index and ring fingers are often present. Atypical cleft hand is now considered to be a variant of symbrachydactyly. The central digits of the hand are shortened or absent, with

vestigial nubbins remaining. The "cleft" is broad and flat, unlike the V-shaped cleft of typical central ray deficiency, usually leaving a thumb and ulnar border digits. Cleft hand is usually inherited as an autosomal dominant trait, with reduced penetrance and variable expressivity among family members. There may be associated abnormalities, including cardiac, visceral, ocular, auditory, and musculoskeletal, including cleft feet. Manske and Halikis have classified cleft hands based on the involvement of the first web space, which is the most predictive factor of hand function and therefore helps guide surgical treatment. Flatt described cleft hand as a "*functional triumph but a social disaster*." Treatment is directed at closing the cleft to improve the appearance of the hand and to treat any syndactyly that may exist. Transverse bones are removed from within the cleft, as these will continue to grow and push the cleft farther apart with age. If the ulnar border digits are syndactylized, they can be released at the time of cleft closure [17–20].

Tips and Tricks

The Snow–Littler procedure may be used to release the first web space syndactyly by releasing the thumb from the index finger and then transposing the index finger ray onto the middle finger metacarpal remnant, thereby achieving web release and cleft closure simultaneously. Alternatively, closure of the cleft by transposition of the index finger into the middle finger position as described by Miura and Komada could be used and may be technically simpler.

13.5.5 Undergrowth

13.5.5.1 Hypoplastic Thumb

Hypoplastic thumb may also be characterized as a variant of radial longitudinal deficiency and is often associated with radial club hand. Children with a hypoplastic thumb may begin to develop a widening of the second web space to achieve rudimentary pinch between the index and middle fingers. Children with a complete absence of the thumb may develop pronation of the index finger for the same reason. Therefore, treatment is often recommended by the second year to establish more normal prehensile patterns. The Blauth classification is used to categorize thumb hypoplasia. Type I is a slightly shorter normal functioning thumb and does not require any treatment. Types II and IIIA, in which the carpometacarpal joint is stable, can be treated with deepening of the first web space and a tendon transfer to improve opposition. The web space is deepened by a traditional four-flap Z-plasty procedure [21].

The Huber transfer uses the abductor digiti minimi to recreate the thenar eminence and replace the hypoplastic intrinsic muscles. The flexor digitorum superficialis from the ring finger may also be transferred through a window in the transverse carpal ligament to restore thumb opposition. Types IIIB, IV, and V require complete reconstruction because of an unstable or absent carpometacarpal joint. Index finger pollicization is the treatment of choice for these children (Fig. 13.2a, b) [22].

Pollicization was originally described by Littler and modified by Buck-Gramcko. In this technique, skin flaps are designed to widen the web space between the new thumb and middle finger. The index finger is elevated as an island flap on its radial and ulnar neurovascular pedicles, dorsal veins, and tendons. An osteotomy of the second metacarpal bone at the level of the distal epiphyseal plate is performed, and the metaphyseal flare at its base and the intervening shaft are removed. The finger is pronated between 140 and 160 degrees, and the metacarpal bone is placed in 45 degrees' abduction palmar to the base of the index finger metacarpal bone. The metacarpal head then becomes the new carpometacarpal joint. The first dorsal interosseus muscle is reattached to become the abductor pollicis brevis, the first palmar interosseous muscle becomes the adductor pollicis, the index extensor digitorum communis functions as the abductor pollicis longus, and the extensor indicis proprius becomes the extensor pollicis longus [23].

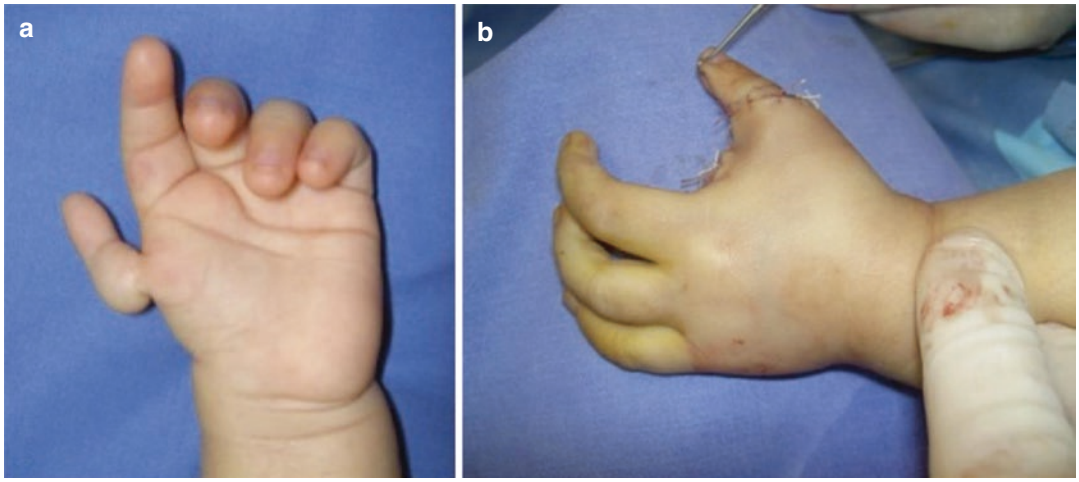

Fig. 13.2 Hypoplastic thumb; (**a**) pre-operative picture, (**b**) Pollicization: immediate post-operative result

13.5.6 Failure of Separation of Parts

13.5.6.1 Syndactyly

Syndactyly results from failure of digital separation and is one of the most common congenital hand malformations. It occurs in approximately 1 in 2000 births and is common in white male children. It occurs bilaterally in 50% of cases, and 10–40% of these cases show a family history of inheritance as an autosomal dominant trait. Inherited forms are associated with genetic defects involving particular candidate regions on the second chromosome. In isolated syndactyly, the third web space is most commonly affected, whereas the first web space is the least commonly affected. Syndactyly is classified as either complete or incomplete and as either simple or complex, depending on the degree of skin and/or bone involvement. In complete syndactyly, the web extends out to the nails, whereas incomplete syndactyly stops short of the fingertips. Simple syndactyly involves only the soft tissues, whereas complex syndactyly involves the phalanges, most commonly involving the fusion of the distal phalanges. Syndactyly associated with other anomalies such as polydactyly, constriction rings, toe webbing, brachydactyly, spinal deformities, and heart disorders is termed complicated syndactyly (i.e., Apert syndrome). Surgical release of syndactyly is recommended early to allow normal growth of the digits and normal grasp and pinch. Timing of syndactyly release is based largely on surgeon preference, although most begin the separation by 12 months of age with the goal of finishing all releases by the time the child is of school age. Early release of syndactyly involving the first web space, complex syndactyly involving the distal phalanges, and syndactyly producing a flexion contracture of the longer digit may require release by 3–6 months of age. Syndactyly involving more than one web space, such as in Apert or Poland syndrome, requires a decision on the sequence of staged releases because usually only one side of a digit should be released at one time to avoid vascular compromise of the digit that would occur if both sides of the digit were released simultaneously. The border digits, thumb, and small finger are usually released first, followed by the central three digits several months later. In complete syndactyly, the web space is reconstructed with a proximally based dorsal rectangular flap. The design of interdigitating skin flaps must be planned carefully, and triangular, zigzag, and rectangular incisions have all been used. Separation will not usually provide sufficient skin to resurface the circumference of each digit, and thus skin grafts are required. Full-thickness skin grafts are preferred to split-thickness grafts, as they are less prone to contracture. In incomplete syndactyly, various other techniques, including simple Z-plasty, four-flap Z-plasty, or double-opposing Z-plasty

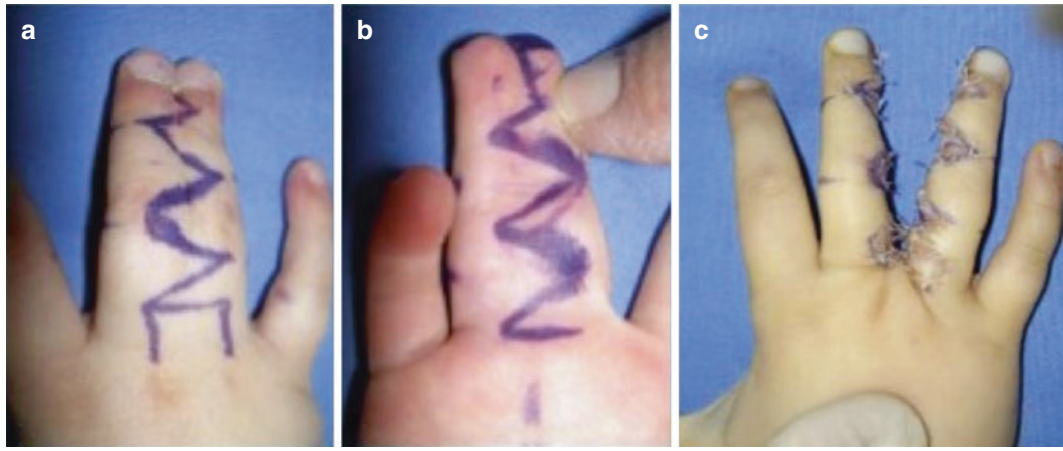

Fig. 13.3 (**a**, **b**) preoperative drawing, (**c**) immediate postoperative result Syndactyly

(Fig. 13.3a, b, c), may allow separation of the digits and deepening of the web space without requiring full-thickness skin grafts [24–28].

Key Points
Surgical release of syndactyly should be performed as early as possible to allow normal growth of the digits and normal grasp and pinch.

13.5.6.2 Radioulnar Synostosis

Congenital proximal radioulnar synostosis results from the failure of developing cartilaginous precursors of the forearm to separate late in the first trimester. It is bilateral in 60% of all patients. The incidence is unknown, and in most cases, it occurs sporadically, but it can be inherited as an autosomal dominant trait with variable penetrance. Children usually present between 2 and 6 years of age, with the absence of forearm rotation and a slight elbow flexion contracture. Children are often at school age before a diagnosis is made; this is due to the fact that the wrist has the ability to compensate for the lack of pronation/supination of the forearm. Clinical suspicion warrants radiographs of the forearm, which reveal the proximal radioulnar synostosis. The radial head is often subluxed or dislocated. Surgical management depends on the severity of the synostosis and the resulting functional impairment. Extreme pronation or supination that interferes with function is an indication for surgery. In addition, a forearm fixed in greater than 60 degrees of pronation generally requires surgery. Derotational osteotomy either at the site of synostosis or in the diaphysis of the radius and ulna to fix the forearm in neutral or slight pronation has been advocated. However, resection of the synostosis and interposition of autologous tissue or allograft between the radius and ulna is favored. However, separation is tenuous, as the synostosis tends to recur. Many interposition materials, to place, at the time of separation have been studied, including synthetic materials, autologous tissues, and allograft tissue. Synthetic materials, such as silicone and polyethylene sheeting, and autologous tissues (i.e., nonvascularized or vascularized tissue), such as free fat grafts, radial forearm fascial flap, and free lateral arm adipofascial flap, have been used. In addition, some surgeons have recommended perioperative irradiation, although this is usually used for post-traumatic radioulnar synostosis [29, 30].

Key Points
Extreme pronation or supination that interferes with function is an indication for surgery.

Surgery is recommended when the forearm is fixed in pronation >60°.

13.5.6.3 Symphalangism

Symphalangism is the term used to describe the failure of interphalangeal joint development and fusion of the proximal phalanges to the middle phalanges and was first described by Cushing in 1916. This condition represents 1% of all congenital upper extremity anomalies and is frequently transmitted as autosomal dominant. Flatt and Wood classified symphalangism as true symphalangism without additional skeletal abnormalities, symphalangism associated with symbrachydactyly, or symphalangism with syndactyly. Clinically, there is the absence of motion, and there are not skin creases in the affected digits. The proximal interphalangeal joint does not develop with growth. The affected fingers do have some flexion, as the metacarpophalangeal and distal interphalangeal joints are present and have a normal range of motion. Attempts have been made to reconstruct or replace the proximal interphalangeal joints, but results have not been favorable. If a child has a poor grasp secondary to symphalangism, a wedge of bone can be removed from the level of the proximal interphalangeal joint and the phalanges fused in 45 degrees of flexion.

13.5.6.4 Duplication (Polydactyly)

Polydactyly can occur on the preaxial (radial) or postaxial (ulnar) side of the limb or centrally, with postaxial polydactyly being the most common type. Preaxial polydactyly is more common in white population, and postaxial polydactyly is more common in African Americans. The supernumerary digit in postaxial polydactyly is either well developed (type A) or rudimentary and pedunculated (type B). Those that are rudimentary and represent a small nubbin of tissue can be managed by ligating the base of the pedicle in the nursery. This will lead to necrosis of the nubbin, which will eventually fall off. The more developed type A digits require formal surgical ablation and may require reattachment of the ulnar collateral ligament at the metacarpophalangeal joint or the abductor digiti quinti tendon. Preaxial polydactyly or thumb duplication occurs in 8 in 100,000 births (Fig. 13.4).

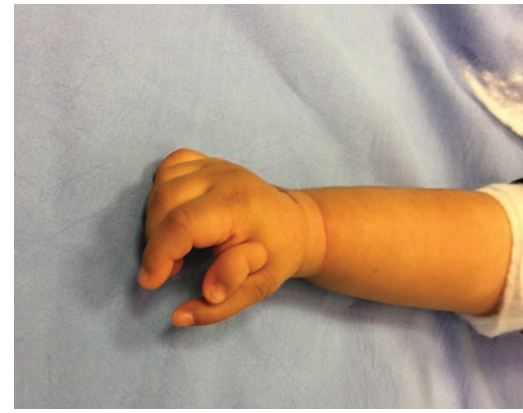

Fig. 13.4 Preaxial polydactyly or thumb duplication

Both the radial and ulnar duplicated thumbs show some degree of hypoplasia, although the radial duplicate is usually more affected. Wassel has categorized thumb duplication into seven types. Type I is characterized by a bifid distal phalanx, whereas type II is a duplication at the level of the interphalangeal joint. Type III is a bifid proximal phalanx, and type IV, the most common, is a duplication at the level of the metacarpophalangeal joint. Type V is characterized by a bifid metacarpal, and type VI is a duplication at the level of the carpometacarpal joint. Type VII describes thumb polydactyly with an associated triphalangeal thumb. Treatment of thumb polydactyly is based on the type of duplication. Types I and II can be treated with either resection of the radial duplication or central resection (Bilhaut operation) from each of the duplicated thumbs while preserving their outer portions. Unbalanced thumbs are generally managed with resection of the radial duplication, and balanced thumbs are managed with central resection. Treatment of duplication types III and IV must be individualized. In general, the best phalangeal portions of both thumbs are incorporated to create the best thumb. The radial duplication is usually amputated, as it is less developed, followed by radial collateral ligament reconstruction of the metacarpophalangeal joint and reattachment of the thenar muscle insertion to the radial base of the proximal phalanx of the remaining thumb. Treatment of types V and VI involves amputation of the radial duplication along with intrinsic muscle

reattachment and collateral ligament reconstruction if necessary [31–34].

Osteotomies may occasionally be required to realign the metacarpal with the proximal phalanx. Central polydactyly is a duplication involving the index, long, or ring finger. This is the least common type of polydactyly and may occur in isolation or as part of a syndrome. Duplication of the ring finger is the most common, followed by the long and index fingers. Central polydactyly may be hidden within concomitant syndactyly, which is termed synpolydactyly (Fig. 13.5a, b).

Treatment depends on the extent of involvement. A fully formed and functional central polydactyly does not necessarily require excision. Central polydactyly that is partially formed and/or has limited motion may require a ray resection. Given the potential spectrum of abnormality, the neurovascular bundles must be meticulously dissected so as not to compromise the remaining central digits [35].

13.5.7 Overgrowth

13.5.7.1 Macrodactyly

Macrodactyly, or gigantism, describes enlargement of all components of an affected digit and represents 1% of all congenital hand anomalies. Most cases are sporadic, without evidence of inheritance, 90% of cases are unilateral, and the index finger is most commonly involved. Several mechanisms have been proposed, including abnormal innervation leading to unimpeded growth, increased blood supply to the digit, and an abnormal humeral mechanism stimulating growth. There seem to be two forms of macrodactyly: the static type, which is noted at birth and in which the affected digit grows at the same rate as the other digits; and the more common progressive type, in which the digit is large at birth and grows disproportionately. Macrodactyly may be associated with hypertrophy of the median, ulnar, or digital nerves, which may result in symptoms of compression neuropathy that may require

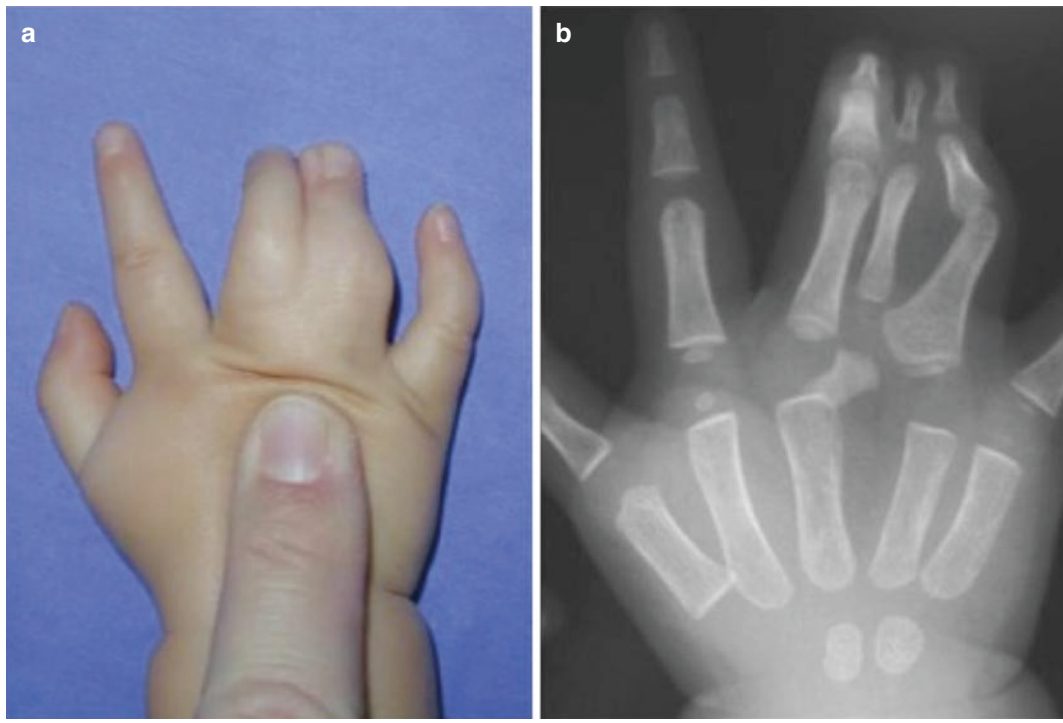

Fig. 13.5 Synpolydactyly; (**a**) clinical picture, (**b**) x-ray

decompression. Macrodactyly is extremely difficult to treat, but surgical intervention is often necessary, as the digit(s) lack function and may interfere with other normal digits. In addition, children with macrodactyly are subject to teasing and social embarrassment [36–38].

Key Points

Surgical options include debulking of the digit and/or disrupting further growth by obliterating the epiphyseal plates. Given the difficulty of treating this anomaly and the mediocre functional results, amputation should be strongly considered when only one or two digits are involved. If amputation is not warranted or if the parents refuse, staged debulking may be considered.

13.5.8 Congenital Constriction Ring Syndrome

13.5.8.1 Constriction Rings

Constriction rings may encircle a single digit or multiple digits or the entire limb of a newborn, causing varying degrees of vascular and lymphatic compromise. Constriction rings occur in 1 in 15,000 births. The constrictions may be either circumferential or incomplete and may occur anywhere on the body, although the limbs are most commonly affected. The cause of this condition is not fully understood. According to the intrinsic mechanism, it is caused by a vascular disruption in the embryo. According to the extrinsic mechanism, amniotic disruption causes the release of amniotic bands that encircle and strangulate the limb or parts of a limb in utero. Patterson classified constriction rings into four types. 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 parts, which is termed acrosyndactyly. Type 4 is a complete amputation distal to the constriction. Treatment of a digit or limb threatened at birth by distal ischemia caused by a proximal constriction ring requires urgent release of the ring.

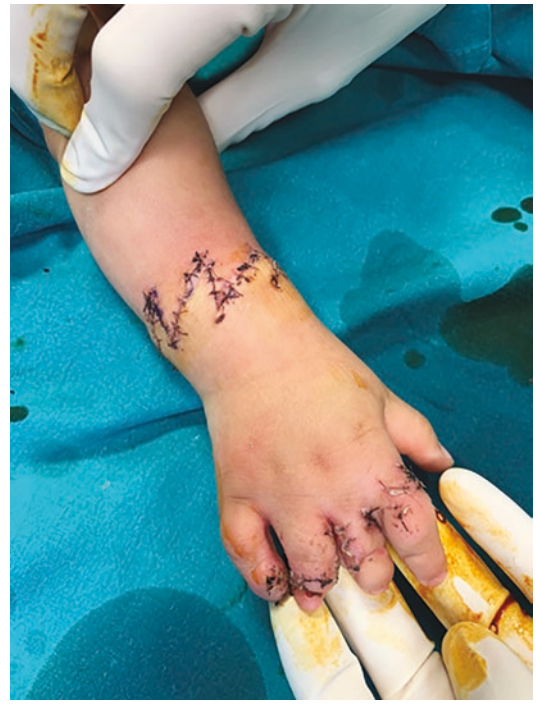

Fig. 13.6 Constriction ring, immediate post-operative result

Constriction rings may also affect the underlying nerves, necessitating decompression. In addition to releasing the constriction ring, the skin and subcutaneous tissues are rearranged with multiple Z- or W-plasties (Fig. 13.6). Some surgeons advocate the release of constriction rings in two stages. Half of the circumference of the ring is excised at the first stage, and the skin is lengthened with multiple Z-plasties. The remaining 50% of the constriction ring is released in a similar manner at a second stage [39–41].

Tips and Tricks

Constriction rings may be successfully released circumferentially around a limb or digit in a single stage.

If the constriction rings have transected extensor or flexor tendons, reconstruction with tendon grafts and/or tendon transfers may be necessary.

Amputation may occasionally be required.

13.5.9 Flexion Deformities

13.5.9.1 Camptodactyly

Camptodactyly is a flexion deformity of the proximal interphalangeal joint that occurs most commonly in the small finger, although other fingers may be affected. The metacarpophalangeal and distal interphalangeal joints are not affected. This flexion deformity occurs in less than 1% of the population, and most patients are asymptomatic and do not seek treatment. Two-thirds of the cases are bilateral, although the degree of flexion may not be symmetrical. The pathogenesis of this deformity remains unknown, although every structure surrounding the proximal interphalangeal joint has been implicated, including the skin and subcutaneous tissues, the ligaments (collateral, transverse, and oblique retinacular ligaments), the volar plate, the flexor tendons, the lumbricals, the interossei, and the extensor apparatus. Camptodactyly has been divided into three types. Type I deformities are the most common and are limited to the small finger. These become apparent during infancy and affect boys and girls equally. Type II deformities do not become apparent until preadolescence (ages 7–11) and affect girls more than they affect boys. Type II camptodactyly does not generally improve and may progress to a severe flexion deformity. Type III deformities are more severe, involving multiple digits of both extremities, and are generally associated with a variety of syndromes. Treatment of camptodactyly depends on the severity of the deformity. Initially, physical therapy and splinting (static and dynamic) may be used to extend the finger. If the contracture progresses to greater than 60 degrees of flexion, surgery may be indicated. This includes exploration and release of any abnormal structure found limiting proximal interphalangeal joint extension, including skin, fascia, ligaments, and/or tendons. Transfer of the flexor digitorum superficialis to the extensor apparatus has been described to decrease proximal interphalangeal joint flexion and increase proximal interphalangeal joint extension [42–45].

13.5.9.2 Congenital Clasped Thumb

Congenital clasped thumb (also known as isolated congenital thumb-palm deformity) represents a spectrum of thumb anomalies. It is more often bilateral and is seen in boys twice as often as in girls. The mild form is caused by the absence or hypoplasia of the extensor mechanism. Moderate to severe forms are related to joint contractures, collateral ligament abnormalities, first web space contracture, and thenar muscle hypoplasia. A clasped thumb is commonly found in arthrogryposis or its associated syndromes. In the classification system proposed by McCarroll and expanded by Mih, type I clasped thumb is flexible and has absence or hypoplasia of the extensor mechanism; type II clasped thumb is more complex, with additional findings of joint contracture, collateral ligament abnormality, first web space contracture, and thenar muscle abnormality; and type III clasped thumb is associated with arthrogryposis or its associated syndromes. The initial treatment of clasped thumb involves serial casting in extension and abduction for 3–6 months. The goal of surgical management is to bring the thumb out of the palm and restore grasp by addressing any or all of the abnormalities of the thumb web space, intrinsic muscle contracture or deficiency, extensor tendon deficiencies, and joint stability [46–49].

13.5.9.3 Arthrogryposis

Arthrogryposis (also known as arthrogryposis multiplex congenital) is a syndrome of nonprogressive joint contractures that is present at birth. Multiple variants of arthrogryposis vary in presentation and severity, and the cause is unknown. This may affect all joints in all limbs. Commonly, the wrist and fingers are flexed and the thumb adducted and flexed into the palm. Treatment should be individualized to achieve independent function. Manipulation of the deformities by a hand therapist shortly after birth may improve the range of motion and overall outcome. If progress is not achieved by 6 months of age, surgical management should be considered. Delaying surgery until after 1 year of age makes improvement more difficult, as the contractures become more

severe. Most surgeons advocate one-stage procedures that address the bone, joints, and soft tissue, as this gives the best results [50–52].

13.5.10 Pediatric Trigger Finger

Key Points

Trigger thumb is one of the most common pediatric hand conditions and responds universally to simple surgical release.

Trigger fingers are more complex, often owing to systemic conditions or anatomical abnormalities, and consequently require a wide and ample treatment.

13.6 Trigger Thumb

The etiology of trigger thumb in children remains uncertain. The main accredited hypothesis is that there is an anatomical mismatch between the diameter of the tendon sheath and the diameter of the flexor pollicis longus (FPL) tendon.

The condition normally presents with fixed flexed thumb interphalangeal joint (IPJ), with the presence of a small nodule on the volar face of the metacarpophalangeal joint (MPJ), Notta nodule. Sometimes it is possible to find some cases in which the thumb is fixed in an extended position, with no IPJ active flexion.

The relatives find the condition accidentally because the triggering is painless.

Sometimes it is supposed to be the result of a trauma or of a subluxation, but normal radiograms and ultrasound exclude it. Moreover, the presence of Notta nodule is diriment.

Regarding treatment, there is no unique direction.

Normally, instructions vary from a first period of 3–6 months of splinting to open surgical release in cases of splinting failure.

Exploring the literature, some authors suggested that all the therapeutic attitudes could be considered correct and could in some way lead to problem resolution. They analyze results of simple observation, versus stretching and exercises, versus night splinting with or without daily exercises versus open surgery and find out that all the series lead in some way to different percentages of clinical resolution, evidently in different periods of time treatment.

Anyway investigating correctly the data, it is evident that splinting or observations or stretching leads to a complete resolution preferentially in mild cases, and normally resolution is obtained with longer treatments.

On the contrary, open surgery leads in the majority of cases to recovery in a shorter time with really low rate or no complications.

Normally relatives willingly accept the period of orthesis to try to have a simple way toward resolution. Anyway usually, due to little compliance of the babies and to a long treatment, relatives usually switch happily to surgical solutions.

Surgery is quite simple; it could be performed under slight sedation and local anesthesia. It is performed through a small transversal incision at the MPJ volar surface of the thumb, the identification of the tight A1 pulley, and its surgical release. Often, it is possible to identify the Notta nodule, but once the pulley is open, no procedures are required on the nodule. Immediately after the complete pulley release, the finger shows a complete extension of IPJ [53, 54].

13.7 Trigger Finger

Pediatric, or congenital, trigger finger presents as a digit, other than the thumb, that locks in flexion.

As pediatric trigger thumb, although described as congenital by some authors, there are no clear records of this condition being present at birth. It has been reported as presenting between the ages of 3 weeks and 11 years. Many papers suggest that the pathological cause is due to anatomical anomalies, but this does make it hard to explain why the condition is not present at birth.

The management of this condition has varied from conservative splinting to operative exploration and correction of the offending structures.

In the literature, there is confusion on outcomes of splinting in trigger finger due to the fact that papers often compare the conservative treatment of trigger thumb and trigger finger together.

What is clear from the literature is that etiology of congenital trigger finger is different from congenital trigger thumb and adult conditions. It is reported that anatomic mechanical condition, such as mutual relationships among flexor digitorum profundus (FDP)) and flexor digitorum superficialis (FDS), or anatomical anomalies of pulleys, causes and sustains the triggering. The application of the operative principles applied in pediatric trigger thumb and adult trigger finger consisting in releasing of the A1 pulley only could lead to insufficient results.

Children who present with trigger fingers could have an underlying condition responsible for the triggering. Triggering has been associated with mucopolysaccharidosis, juvenile rheumatoid arthritis, Ehlers–Danlos syndrome, Down syndrome, and central nervous system disorders such as delayed motor development [55].

Surgery is quite often indicated, and a stepwise approach through Bruner's incision is therefore necessary.

Surgery could be performed under soft sedation and local anesthesia. The surgical approach allows the possibility to have a complete view of the flexor apparatus; both tendon structures and the pulley system must be carefully analyzed, and triggering must be evoked during surgery in order to be sure that the procedure undertaken has eliminated each possible cause of tendon friction [56].

13.7.1 Physiotherapy

Aristotle defined the hand "the tool of tools." The hand is for an individual a work, communication, and cognitive tool, right from the prenatal life: through touch, we learn, discover, get excited, and communicate.

The rehabilitation of the child involves not only the little patient but also his family and all of the medical staff who will take care of his health as a team.

The protagonist of the rehabilitation project is not just the hand but the whole little patient in the harmonious development of the evolutionary stages happening in the family context.

The hand therapist builds an ad hoc path using his knowledge, cultural background, and experience, but above all his own creativity and imagination because it is important not to forget that the patient in this case is a child.

In the specific area of pediatric hand affected by congenital malformation, rehabilitation must be offered in the form of a game, with suggestive activities that try to involve and stimulate the little patient. Playing thus becomes a fundamental tool that the hand therapist has at his disposal; it must therefore meet criteria not only of functionality but also of attractiveness and stimulus requirements for the attention and involvement of the child.

This is where the therapist has to use all his creativity. There is no pathology-dependent or predetermined activity for everyone, but, depending on the goal you want to achieve, the most suitable game is identified. Many times observation and play turn out to be the key to rehabilitation, always implementing new strategies and proposals that attract the attention of the young patient and that respect the developmental and cognitive stages reached.

Pediatric hand rehabilitation is not just "playing" or "gymnastics," as it is often defined, but a methodology that develops and supports the child's skills within specific and individualized paths.

Treatment of upper limb malformations, therefore, is part of a broad and rich context that aims at the well-being of the child in his whole being.

13.7.2 Psychological Aspects

Hands have a central role in social and emotional relationships because they vehicle emotionality; they are the protagonist in nonverbal communi-

cation, but they are also constantly visible to others as well as to their owner. When a person faces trauma or malformation problems, this last aspect is really important from a motivational point of view: the subject cannot avoid confronting his/her own difficulties and reactions raised in the social context.

Whether originated, the pathology of the upper limb affects and modifies many areas of individual life and requires an important psychological effort of acceptance and adaptation.

The relationship between patient and health specialists grows and shapes in this difficult context, especially when patients are children.

Several observations proposed by psychologists, surgeons, and physiotherapists working on this topic highlight the need for a synergistic approach involving different professionals.

Emotional manifestations are common reactions to stressful events, and in most situations, they can find support and reach acceptance within the proposed therapeutic protocol. However, if such emotions become stable and do not spontaneously evolve and resolve, it is essential to evaluate the duration and intensity of these emotions and their impact on quality of life and therapeutic protocol. It is essential that the psychologist attends during the first consultation and then, subsequently, is available to the patient or her/his family throughout the therapeutic protocol.

The interview with the psychologist allows the patient to face emotional aspects, helping the child's parents or the patient to expose and understand doubts and perplexities. Usually, uncertainties concern aspects strictly related to the therapeutic options and protocol, but patients and parents are also worried about life situations involving social skills, general child development—if the patient is a child, and educational aspects.

The whole family is included in the therapeutic pathway starting from the psychological interview following the first medical examination with the surgeon and all the following access to the medical care before and after surgery and at follow-ups.

As expected, even the young patient must be involved; this can be done in many different ways according to his/her age, since, based on it, the relevant main topics will be different.

During the interview between psychologist and parents, it is really important to discuss this topic in order to help parents in accepting the child's questions and supporting him/her during the inclusion in new contexts, for example, by talking and explaining to teachers and educationists.

The attention of the psychologist must therefore always be directed to the whole family system and its subsystems: the individual, the couple, interpersonal, and the sibling systems.

A multidisciplinary approach is essential considering the complexity of congenital hand conditions and hand diseases. The team has to deal carefully and respectfully with the patient and the family, keeping in mind their needs.

This requires that many professionals integrate together in order to provide a complete and adequate response to a patient's needs and not only to his/her malformed or traumatized hand.

13.8 Conclusions

Congenital differences of the upper limb represent a significant and unique challenge for the hand surgeon. In all cases, the ultimate goal is to provide a functional limb that can be integrated into the child's overall development. This goal may be met surgically or through specialized therapy and rehabilitation. Every case is unique, and each patient (and parent) will have a different capacity to adapt. These differences should be taken into account before embarking on a long, often difficult reconstructive course. It should be made clear from the outset that the child will never have a "normal" hand. Once realistic expectations have been set, reconstruction and/or rehabilitation can commence.

Pearls and Pitfalls

Proximal Transverse Deficiencies

Treatment is often a prosthetic device.

Surgical options are only to improve stump as removal of functionless digital nubbins.

Transverse Arrest of the Digits Distal to the Metacarpal Level

The objective of treatment is to create a pinch and grip. Distractions lengthening of metacarpal or phalangeal and nonvascularized toe phalangeal bone grafting are performed mostly in the case of oligodactyly or hypoplastic digits. In the case of monodactyl, adactyl, or transverse deficiency of the thumb, free microvascular toe-to-hand transfer could be the best option.

Nonvascularized toe phalangeal bone grafting should be performed before 15 months of age. The bone should be harvested extraperiosteally, and the collateral ligaments and tendons should be reattached.

Free microvascular toe-to-hand transfer, providing pinch to the adactylous hand by microsurgical toe transfer, is accomplished in two stages: first with a digit in the thumb position and then with a digit positioned for pinch using the second toe transfer most commonly.

Failure of Formation of Parts: Longitudinal Arrest

Radial Longitudinal Deficiency

Radial longitudinal deficiency is often found in association with other malformations. Thumb hypoplasia is often present, ranging from slight to total absence.

Treatment takes place step by step. First of all, the rigidity is corrected, then the wrist deviation, and lastly the thumb hypoplasia.

Treatment starts immediately after birth and consists of passive stretching exercises and splinting.

In the case of persistence of significant stiffness, soft tissue distraction is made with an external fixation device. Around 1 year of age, centralization or radialization is performed.

Centralization consists of the centralized carpus over the ulna after transecting radial fibrotic muscle and bands. The new position of the wrist is fixed by a pin through the middle finger metacarpal bone, carpus, and the ulna. Removal of the lunate and tendon transfer as flexor carpi radialis to extensor carpi ulnaris could be performed according to the degree of wrist stiffness and deviation. Lastly, pollicization of the ring finger is made to give a pinch.

Ulnar Longitudinal Deficiency

Treatment consists of serial casting to improve the wrist and elbow positions. For greater than 30 degrees of angulation, excision of the anlage (fibrotic bands) is indicated and K-wires may be used to hold the wrist in a neutral position. In the case of elbow stiffness, the proximal radial head is resected and a one-bone forearm is created by osteosynthesis of the distal radius to the proximal ulna. In the case of radiohumeral synostosis, a derotational osteotomy of the humerus may be required to place the hand into a more functional position.

Central Ray Deficiency

Treatment is directed to closing the cleft, creating a wide first web space and treating any syndactyly that may exist to improve the appearance of the hand. To achieve this, different surgical techniques are described.

The Snow–Littler procedure may be used to release the first web space syndactyly by releasing the thumb from the index finger and then transposing the index finger ray onto the middle finger metacarpal remnant, thereby achieving web release and cleft closure simultaneously [57].

Miura and Komada propose a simpler closure of the cleft by transposition of the index finger into the middle finger position [58].

Hypoplastic Thumb

Treatment of hypoplastic thumb depends on the grade of hypoplasia. Type I does not require any treatment. Types II and IIIA are treated with deepening of the first web space and a tendon or muscle transfer to improve opposition. Types IIIB, IV, and V require an index finger pollicization because there is an unstable or absent carpometacarpal joint.

Failure of Separation of Parts

Syndactyly

Surgical release of syndactyly should perform as early as possible, mostly in the case of syndactyly between 4° and 5° digits and 1° and 2° digits, to allow normal growth of digits and normal grasp and pinch. The goal of all surgical techniques is the reconstruction of the web space with a dorsal flap and separation of the digits using an interdigital zig-zag flap.

Duplication (Polydactyly)

Polydactyly can occur on the preaxial (radial) or postaxial (ulnar) side of the limb or centrally.

In each group, the treatment depends on the type of duplication. Except for rudimentary and pedunculated digits, treatment consists in ablation of supernumerary digit and rebalance of the dominant digit.

Overgrowth

Macrodactyly

Surgical options include debulking of the digit and/or disrupting further growth by obliterating the epiphyseal plates. Given the difficulty of treating this anomaly and the mediocre functional results, amputation should be strongly considered when only one or two digits are involved. If amputation is not warranted or if the parents refuse, staged debulking may be considered.

Congenital Constriction Ring Syndrome

The constrictions may be either circumferential or incomplete and may occur anywhere on the body, although the limbs are most commonly affected.

To release the constriction ring, multiple Z- or W-plasties are set up. The procedure could be done in one step by the release circumferentially or into two steps. When the constriction ring affects, an arm is important to release the fascia.

Trigger Thumb

Trigger thumb is one of the most common pediatric hand conditions and responds universally to simple surgical release.

Trigger fingers are more complex, often owing to systemic conditions or anatomical abnormalities, and consequently require a wide and ample treatment.

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