Other Developmental Anomalies

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Clasped Thumb

Congenital clasped thumb may present with a number of findings including joint contracture [1–6], tendon [1, 6], and ligament insufficiency [4], intrinsic muscle abnormality [1, 2, 4], and thumb index web space contracture [1, 2, 4]. This deformity is usually associated with other generalized musculoskeletal malformations, including arthrogryposis, digitotalar dysmorphism, and Freeman–Sheldon syndrome. The initial classifications have not been useful in the clinical setting. Thus, a more practical approach has been proposed by McCarroll [4] and expanded by Mih [7] (Table 5.1).

Table 5.1 The classification of clasped thumb by McCarroll and Mih

Type	Features
I	Supple thumb Absent or hypoplastic extensor mechanism
II	– Joint contracture – Collateral ligament abnormality – First web space contracture – Thenar muscle abnormality
III	– Associated with arthrogryposis or its associated syndromes (mental retardation, aphasia, shuffling gait, and adducted thumbs) – Extensor mechanism may have minimal or no abnormality

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Clinical Features

Extension lag is present only at the metacarpophalangeal (MCP) joint, suggesting hypoplasia of the extensor pollicis brevis muscle tendon unit [5, 6]. Additional extensor pollicis longus and/or abductor pollicis longus anomalies have also been reported [7]. The diagnosis of clasped thumb is often delayed because an infant frequently holds the thumb within the palm for the first 3–4 months. The male-to-female ratio is reportedly 2.5:1 by Lin et al. [8] or 1:1 by Tsuyuguchi et al. [9]. This difference can be attributed to the wide variety of syndromes associated with clasped thumb and their different modes of inheritance, as well as the small number of reports in the literature that have documented the sex ratio of affected cases.

Treatment

The generally established treatment methods follow Weckesser et al. [10], Lipskeir and Weizenbluth [1], and McCarroll [4], who divided clasped thumb into two types (supple and complex) and treated the supple type with splinting and tendon transfer if splinting failed, while the complex type was treated by correcting the fixed contractures, reconstructing the lax ligaments, and tightening the skin as appropriate. Tsuyuguchi et al. [9] treated types I and II cases by splinting and operative treatment for type III

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and type II cases that did not respond to conservative management. The results of splinting are excellent for cases of type I clasped thumb [8, 9]. The small number of patients who were followed with a nonoperative treatment in this study was attributed to the delayed presentation of the patients, which was after the age of 1 year. Surgery is indicated for children who have failed splinting or present after 2 years of age. However, the degree of impairment must be considered during establishment of a treatment plan. Mild MCP joint extension lag does not hinder hand function and does not always require treatment (Fig. 5.1). Some deficiency of thumb extension that prevents grasp warrants treatment. Any associated thumb MCP joint or thumb/index finger web space contracture also requires treatment (Figs. 5.2 and 5.3). Initially, serial casting or splinting may be tried to release the taut skin and correct the contracture. Residual contracture correction must be managed surgically. In a type I clasped thumb, release of skin and subcutaneous tissue is usually sufficient to correct the contracture. Thumb/index finger space deficiency is treated with standard techniques of four-flap "Z"-plasty or dorsal rotation advancement flap [11]. A palmar skin deficiency requires a rotational flap in the palmar aspect of the thumb MCP joint. A modified dorsal rotation advancement flap [12] provides a long wide flap that

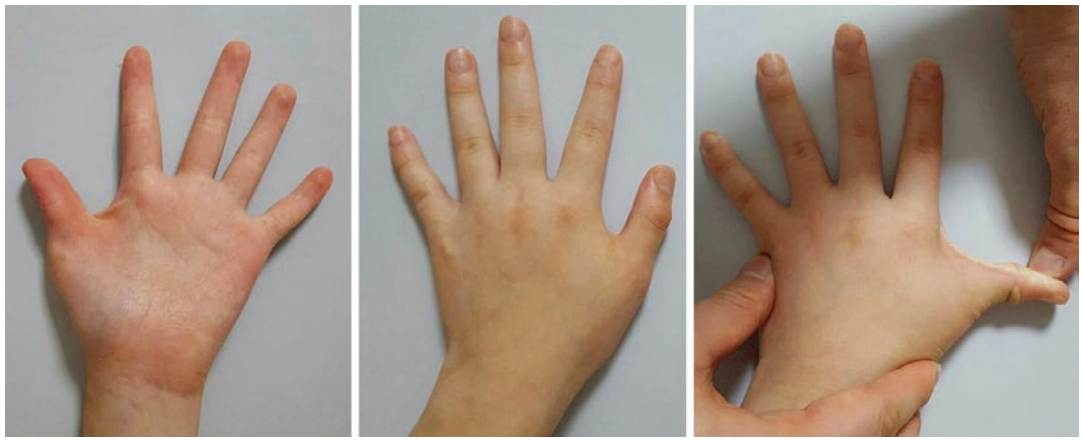

Fig. 5.1 A type I clasped thumb with mild web contractures

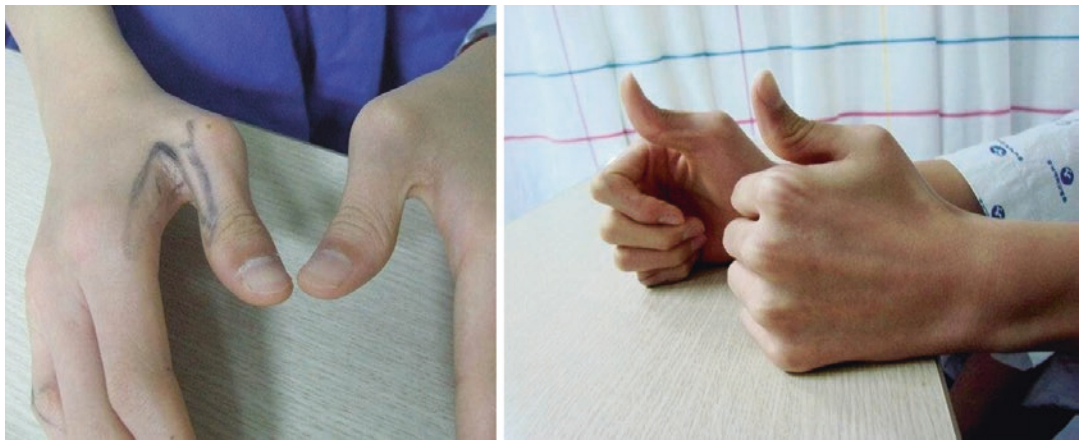

Fig. 5.2 A type II bilateral clasped thumb with web contractures

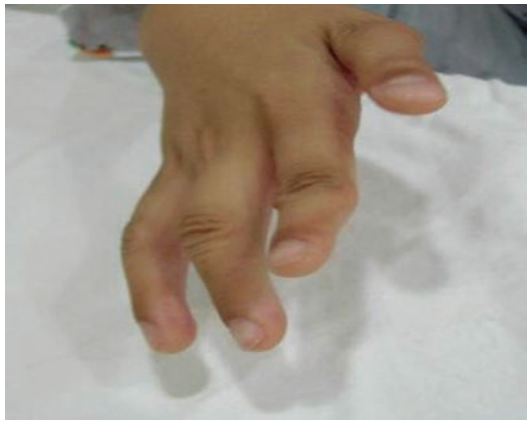

Fig. 5.3 Clasped thumb in arthrogryposis

releases the thumb index web space with suture lines far beyond the web. In addition, it releases the palmar skin, even when very tight in cases of severe narrowing of the web. A second incision is made over the dorsum of the thumb metacarpal and proximal phalanx. The extensor apparatus is explored. The extensor pollicis brevis tendon is usually present but is small and attenuated. A tendon transfer is performed to reinforce thumb extensor function. The extensor indicis proprius tendon is the first choice, but it may or may not be present [6, 7]. Absence of the extensor indicis proprius requires selecting an alternative donor tendon. Options are the flexor digitorum superficialis and the abductor digiti minimi muscle [7]. The tendon transfer is secured into the attenuated tendon and/or the base of the proximal phalanx (Fig. 5.4). Tendon transfer was done as an original method using the bone tunnel of Broadbent [5] or suturing the extensor indicis to the weak extensor tendon of extensor pollicis brevis, as introduced by Mih [7].

Treatment Considerations in Cases of Arthrogryposis

Hand function is highly variable, with intrinsic thumb flexion usually preserved even in the absence of other digital motion. There are several different patterns of hand deformities, some more commonly seen in specific arthrogryposis subtypes (Table 5.2). As with the general corrections for other deformities, fixed contractures need arthrodesis, whereas passively correctable contractures can be treated with a tendon transfer. In type 1, the CMC status remains an extension and the metacarpophalangeal (MP) shows a flexion contracture that is not flexible, and MP joint fusion (chondrodesis) is ideal. However, the more common thumb position is type 2 (flexed CMC, extended MP). In type 2, thumb opposition may vary from opposition to one or several fingers or to only the palm. If FPL motion is preserved despite the absence of other functions, reorientation of the thumb would be the more beneficial option for absence of proper prehension due to improper orientation of the thumb. The first web release is employed according to the degree of contracture. Traditional four-flap Z-plasty, dorsal rotational advance flap [12], and index rotational flap (Fig. 5.5) [13] are performed concurrently during surgery for chondrodesis/reorientation types 1 and 2. Modified new flaps have been introduced to cover a previous flap. Mahmoud et al. [14] designed a new flap to widen the web space further and correct the palmar contracture. They emphasized that the index rotational flap of Ezaki [13] provides insufficient skin at its apex, with the possibility of an incomplete correction and the frequent need for a thenar release incision. Abdel-Ghani et al. reported 4 years of follow-up outcomes in 39 patients with distal arthrogryposis [15]. They performed a modified rotational advance flap for the first web, an *a la carte* release of tight structures of the first web (intermetacarpal fascia, adductor pollicis muscle, first dorsal interosseous muscle, and capsule of the carpometacarpal joint of the thumb) to achieve full palmar abduction of the thumb, and finally stabilized the MP joint in extension by chondrodesis. They explained the importance of chondrodesis, as the presence of abnormal articular surfaces, poor inefficient muscles for transfer, and global instability of the MP joints making other reconstructions unpredictable. Chondrodesis also shortens the thumb ray, so it alleviates the need for release of deficient skin on the flexor aspect of the thumb

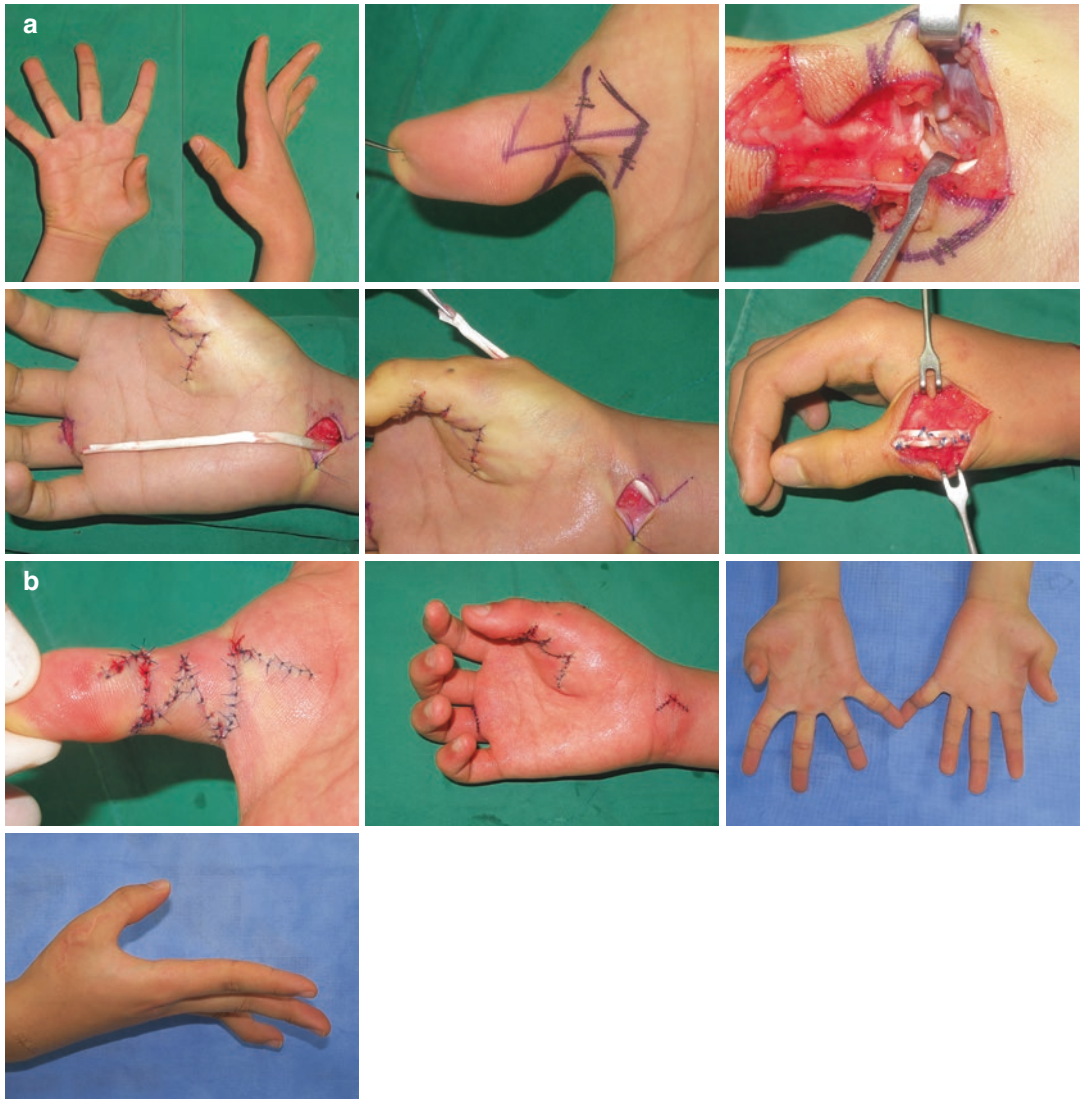

Fig. 5.4 A type II clasped thumb with web contractures. (**a**) First, volar contracture was released by multiple Z-plasty and advanced local flap. Capsulotomy and adductor pollicis/volar plate release were performed. Instead of extensor indicis proprius, fourth FDS was harvested and transferred to extensor pollicis brevis (modified Becker's method). (**b**) After 53 months after surgery, abduction and extension of the thumb were all satisfactory. Images courtesy of Dr. Woo SH, with original copyright holder's permission

Table 5.2 Classification of thumb contracture in children with arthrogryposis

	Joint status	
Classification	CMC	MP
Type 1	Extended	Flexed
Type 2	Flexed	Extended
Type 3	Flexed	Flexed

and may also alleviate the need for lengthening the flexor pollicis longus muscle. In addition, they recommend waiting until radiological appearance of the ossific nucleus of the proximal epiphysis of the proximal phalanx of the thumb. This allows exposure of subcortical bone without injuring the physis and ensures good bone-to-bone union.

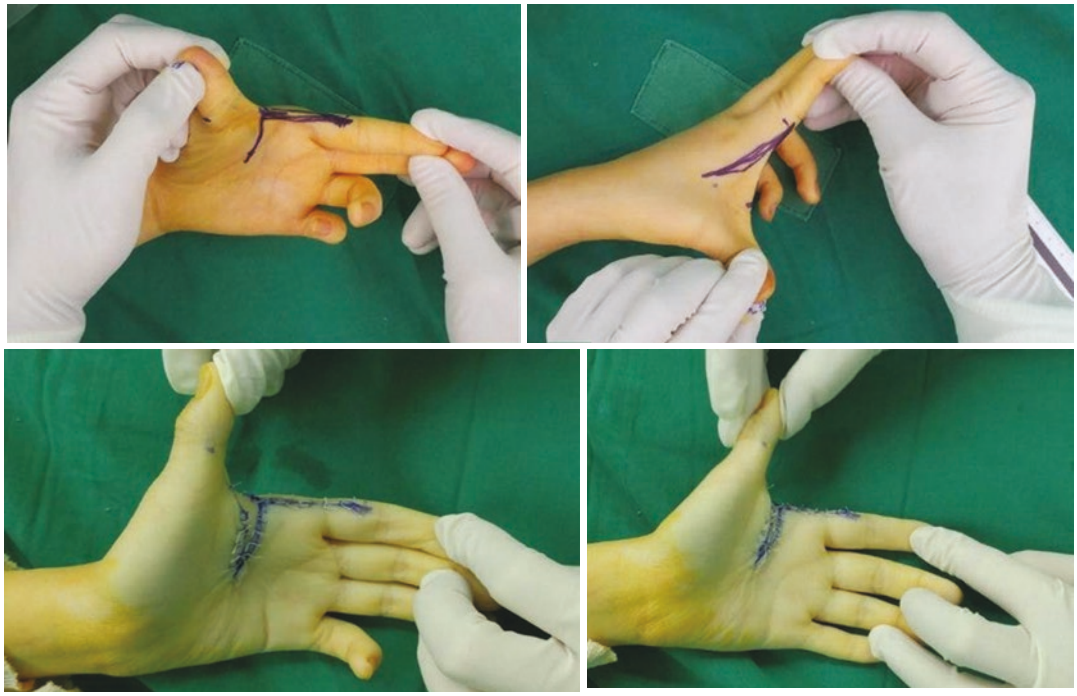

Fig. 5.5 Index rotational flap

Trigger Thumb

Incidence and Etiological Findings

It is controversial whether pediatric trigger thumb is a congenital or acquired condition. However, several neonatal studies support the idea that it is an acquired condition. Initially, Get et al. reported an incidence of one patient per 2000 live births, based on a somewhat small sample size. The term "congenital" is now being abandoned because widespread evaluations of newborns have failed to discover trigger thumb. Rodgers and Waters [16] examined 1046 newborns, and Slakey and Hennrikus [17] examined 4719 newborns to determine the congenital incidence of this condition; however, no infants with trigger digits were identified by either study. After those reports, Moon [18] examined 7700 neonates within the first few days of life, and Kikuchi and Ogino [19] examined 1116 babies within 14 days after birth and noted no trigger thumbs. Furthermore, this condition of postnatal growth would come to be regarded not as "trigger thumb" but as "fixed flexion deformities" of the interphalangeal joint. The pathogenesis of trigger thumb was explained by two interesting ultrasonographic studies. First, Verna et al. [20] prospectively analyzed the FPL tendon and A1 pulley using dynamic ultrasonography in patients with pediatric trigger thumb. They explained that triggering always occurred at the A1 pulley and that there was focal enlargement of the FPL without abnormalities in the A1 pulley. They suggested that the initial inciting event is developmental enlargement of the FPL tendon. A second study by Kim et al. [21] clarified the anteroposterior and radioulnar diameter of the FPL just under the A1 and normal A1 pulleys. They found that the radioulnar distance of the FPL tendon increased to a greater extent than did the anteroposterior distance of the FPL tendon in cases of trigger thumb. However, they

postulated that the developmental mismatch between the FPL tendon and the area under the A1 pulley is an important etiology of trigger thumb because the anteroposterior to radioulnar measurement ratio of the FPL tendon was similar between the trigger and contralateral thumbs; in this regard, the size of the FPL tendon in trigger thumbs seems to proportionately increase.

Natural History

It is controversial whether pediatric trigger thumb is a congenital or acquired condition. However, several neonatal studies support the idea that it is an acquired condition. Dinham et al. [22] reported that approximately 30% of trigger thumbs diagnosed prior to 1 year of age resolved, and about 10% diagnosed from 6 months to 1 year of age resolved spontaneously. Higher rates of resolution have been reported from the serial studies on Asian populations [23, 24]. In particular, Baek et al. [25, 26] reported resolution of 63% of thumbs at an average of 48 months follow-up and 75.9% of thumbs (an additional 12.9%) at an average of 87 months follow-up. They emphasized that pediatric trigger thumb has the potential to spontaneously resolve after a sufficient observation period of more than 4 years, without developing residual deformities.

Treatment

Nonsurgical management, including passive extension exercises as well as extension splinting, has been attempted but often results in persistently abnormal motion in 39–59% of patients [27]. Although steroid injections are commonly used to treat adult trigger fingers and are effective, they are not used in children and would be difficult to perform in an outpatient setting. Outcomes of splint therapy have been reported for a series of 40 trigger digits that were treated by applying a splint to hyperextend the interphalangeal (IP) joint during naptime and at night [28]. Twenty-three thumbs were relieved completely, and three patients required surgery. Slakey et al. [17] suggested that delaying surgery for up to 3 years would not be detrimental with regard to motion contracture. Kozin et al. [29] established their own guidelines, such as >1 year of age, with the need for stage IV surgery. The surgical procedure consisted of general anesthesia and loupe magnification. A transverse incision is made over the A1 pulley at the base of the thumb on the palm side. The radial digital nerve is identified and retracted. The A1 pulley is then identified and released. No further manipulation for a thickened nodule is needed. The wound is closed, and a soft dressing is applied with 1-week follow-up instructions. Percutaneous release [29, 30] of the A1 pulley has also been suggested, but this does not negate the need for general anesthesia (Figs. 5.6, 5.7, and 5.8).

Triphalangeal Thumb

Several classification systems have been used, but the simplest and most often used is the Wood classification, which is based on the shape of the extra phalanx [31] (Table 5.3).

The incidence of triphalangeal thumb is about 1 in 25,000, representing about 3% of upper extremity congenital anomalies. It can occur sporadically, be inherited as an autosomal dominant trait, or appear as part of a syndrome (Holt–Oram syndrome).

Treatment

There is no consensus on how this condition should be treated; thus, the definitive treatment depends on the type of triphalangeal thumb. The goal has been to reconstruct the thumb with adequate function prior to the development of oppositional pinch, which occurs between 12 and 18 months of age. Treatment of a minor length discrepancy or angulation can be delayed until later in life.

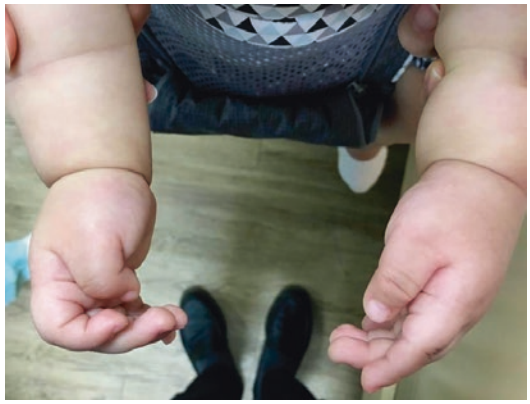

Fig. 5.6 Trigger thumbs diagnosed at postnatal 9 months were relieved by conservative treatment at 4 years

- Type I: In an earlier presented case, excision of the extra ossicle, with or without reconstruction of the collateral ligament, and with or without pinning was recommended to allow for joint remodeling of the new IP joint with time [32]. Usually, a dorsal approach is preferred, with a longitudinal incision through the extensor mechanism. The extra phalanx is isolated and its size delineated. Osteoperiosteal sleeves are elevated from each side including the collateral ligaments. The extra phalanx is removed, and the collaterals are reattached (Fig. 5.9). Wang and Hutchinson reported good long-lasting results with acceptable

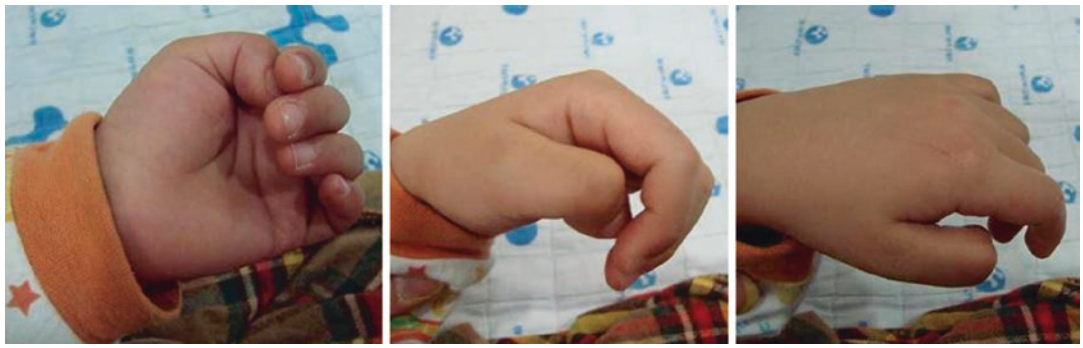

Fig. 5.7 Trigger thumb diagnosed at postnatal 6 months was not relieved by conservative treatment until 5 years old

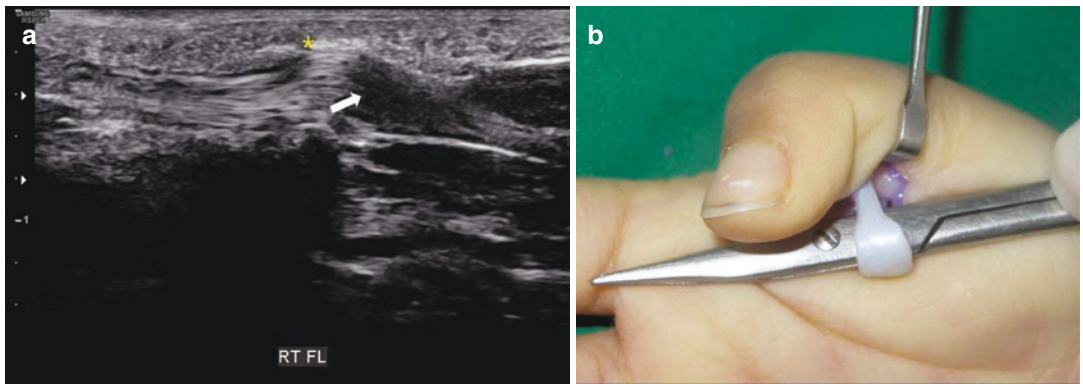

Fig. 5.8 (**a**) A sagittal sonographic view of a trigger thumb showing thickening of the tendon (white arrows) just proximal A1 pulley (yellow asterisk). The thumb was passively extended during sonographic inspections with no more passage of the thickened area under the A1 pulley. (**b**) After surgery, the thickened flexor pollicis longus was identified with free movement. Images courtesy of Dr. Woo SH, with original copyright holder's permission

thumb IP range of motion (ROM) and no pain after excision for 21 thumbs with average ages of 22 months [33]. For cases that are older, a closing wedge osteotomy through the delta and distal phalanges with angular correction and resection of the extra joint is suggested for bony growth and further visualization of the bone and joint cartilage [34].

- Types II and III: The principle is that the joint with the greatest motion is preserved and the joint with the least movement is fused. A large, wedge-shaped extra phalanx has sufficient length; thus, a simple excision would commonly evoke subsequent instability. Proper bone resections, from the extra-phalangeal and adjacent joint side, to shorten and realign, should be performed. If there is a definitive angulatory deformity, an asymmetric osteotomy could correct the deformity [35].
- Duplicated thumb: The dominant part of a duplicated thumb should be preserved. If the triphalangeal component is determined to be the dominant part, the procedure for the extra phalanx is based on the above explanation [29].

Table 5.3 The classification of triphalangeal thumb

Type	Clinical features
Type I	The extra phalanx is triangularly shaped, often also called a delta phalanx
Type II	Rectangular- or trapezoid-shaped extra phalanx, which is not as developed as a full phalanx
Type III	Full extra phalanx

Symphalangism

Symphalangism is a rare condition in which the IP joint presents as congenital ankyloses in the thumb. The radial component in cases of thumb duplication has been reported as symphlagism in a few studies [36, 37]. It was used to describe an autosomal dominant disorder affecting PIP joints of the fingers [38, 39]. These are related to NOG or noggin gene mutations [40, 41]. However, many other studies considered congenitally stiff DIP and MP joints to be symphalangism. The nonhereditary symphalangism often seen with symbrachydactyly has a sporadic pattern. Baek and Lee graded three degrees, according to the clinical manifestations and radiological findings [42] (Table 5.4).

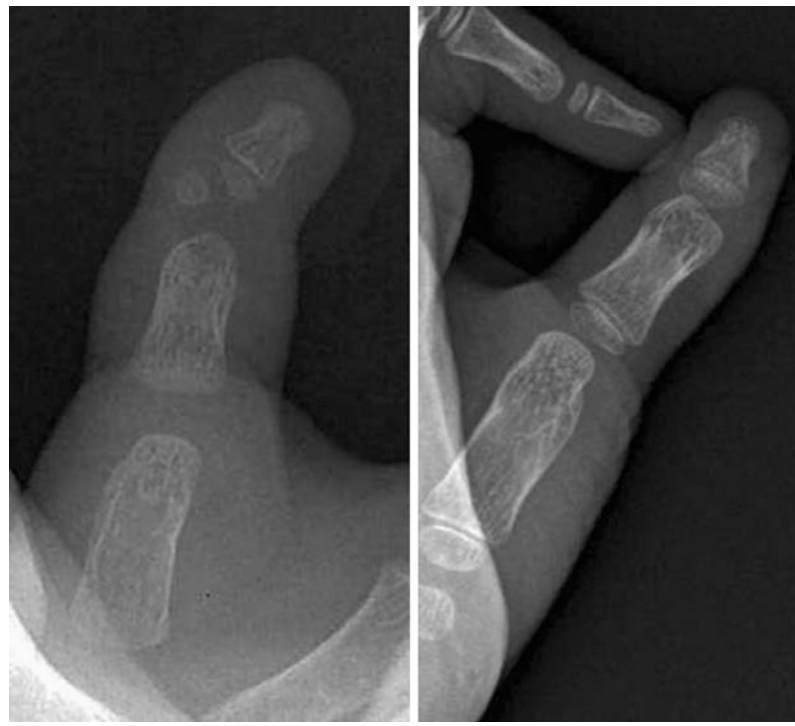

Fig. 5.9 Delta phalanx was excised at 18 months and remained satisfactory until 5 years

Table 5.4 Classification of symphalangism

	Grade I (fibrous symphalangism)	Grade II (cartilaginous symphalangism)	Grade III
Volar skin crease	Faint or absent	Absent	Absent
Active motion	Absent	Absent	Absent
Passive motion	10–20	Only jerk	Absent or jerk motion
Joint space in simple radiographs	Mild narrowing	Definite narrowing	Joint space absent
Phalangeal head in lateral view	Round	Flat	Fused to adjacent bone

Baek et al. [42] suggested grade I or early grade II thumb IP joint symphalangism before the age of 7 years could be an appropriate surgical indication. They explained that the thumb usually presents a slower progression than the fingers. Takagi et al. [43] resected all radial components of symphalangism and reported inevitable restricted motion at the IP joint type symphlagism (three cases) than at the MPJ joint type symphalangism (three cases). The dorsal capsule and dorsal halves of bilateral collateral ligaments divide after a Y-shaped dorsal incision. Gentle passive ROM exercises are initiated, with assistance, 2 days after surgery. Baek et al. explained that brain education may be needed to actively flex the joints, probably because the brain motor cortex for affected fingers has not yet formed.

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