Management of Thumb Hypoplasia

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EPIDEMIOLOGY

Conway and Bowe\textsuperscript{20, 75} have reported the incidence of congenital deformity involving the hand-forearm unit as 1 in 626 live births. Naujoks\textsuperscript{22} reported similar findings of 34 in 17,800 live births, and Hegdekätt\textsuperscript{24} has reported it to be 1 in 200 live births. Major defects of development involving the thumb were reported by Entin\textsuperscript{23} as 16\% of his series of upper-limb congenital anomalies.

Congenital failure of thumb formation is a rather commonly observed birth defect and may in certain cases be part of a more complicated picture. Thumb abnormalities, for example, are often seen in association with pre-axial longitudinal failure of formation of the radius (Fig. 1: radial agenesis) and have been reported with variable penetrance and expressivity in Lamb's\textsuperscript{48} series of 117 consecutive cases. Syndrome complexes that involve cardiovascular, genitourinary, and musculoskeletal manifestations may also have associated thumb abnormalities, including Holt-Oram syndrome, Fanconi syndrome, diastrophic dwarfism, and Cornelia de Lange syndrome.

Each of the three tubular bones of the thumb osteoarticular column may be affected differently, depending on causative genetic or environmental factors. Certain characteristic deformities reflect specific syndromes, e.g., the short, flat, broad distal phalanx of Apert's acrocephalosyndactyly (Fig. 2A and B) and Carpenter's syndrome, or the slender terminal phalangeal hypoplasia characteristic of both Holt-Oram and Fanconi syndromes.

Longitudinal growth deficiency of the thumb proximal phalanx is usually not an isolated phenomenon and can occur in combination with some type of general phalangeal growth disturbance involving the medial fingers, e.g., brachydactyly (most commonly attributable to growth arrest of the middle phalanx) and synbrachydactyly (Fig. 3A–D), or more complex musculoskeletal syndromes, such as brachydactyly in association with absence of the sternocostal portion of the pectoralis major muscle (Fig. 4: Poland's syndrome).

Congenital absence, shortness, instability, or deformity of the thumb can result potentially in profound compromise to grasp and pinch. The rationale for surgical reconstruction should be based on consideration not only of the functional, but of the emotional impact of the child's congenital deformity. The demanding technical considerations involved in surgically creating an anatomic potential for grasp or pinch are only part of a decision-making process; a process that might, unfortunately, include complete lack of patient cooperation. Unpredictability of growth potential in those tissues to be used for reconstruction, and a general predisposition for recalcitrant anatomic abnormalities encountered at the time of surgery also confuse the decision-making process. Add to this the absence of normal cerebrocortical thumb representation, and the surgeon can readily appreciate problems inherent in functional reconstruction of

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partial or complete congenital thumb deficiency.

DEVELOPMENT OF A CONTEMPORARY SURGICAL APPROACH TO CONGENITAL THUMB RECONSTRUCTION

Through the last century of thumb reconstructive surgery, an appreciation has existed for the importance of an opposable unit to the prehensile human hand. Since 1874, when Huguier reported deepening a remaining cleft between a partial traumatic thumb and index finger amputation in an adult, exhaustive surgical efforts have been directed towards salvage and reconstruction of an opposable thumb unit. The issue of thumb reconstruction has been controversial throughout the last century. Established doctrines are challenged frequently, and thoughtful new principles and techniques regularly introduced into the literature. Few of these innovations have endured to become contemporary hallmarks of reconstruction of the congenitally deficient thumb.

Differences between congenital failure of thumb formation and traumatic thumb loss cannot be overstated, and must be appreciated by the reconstructive hand surgeon during his or her assessment of the functionally compromised patient. The child with congenital thumb deficiency has a markedly reduced, or absent cerebrocortical representation for thumb function, which is normally a rather large portion of the sensory cortex (Fig. 5A and B). To compensate for inadequacies of prehensile thumb function, the child afflicted by complete or partial thumb
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...immb is a frequent ...some degree to some degree agenesis. Es-
...d evolved frequently, ...nd techniques over the ...ew of the evolu-
...tional failure of ... thumb loss can-
...aplasia may develop insidiously a progressive 
...uction for the congenitally compromised hand are based on surgical experience following 
...n the latter part of the 19th and early 20th centuries, two distinct surgical philosophies evolved: (1) local thumb 
...essentially a deepening of the first web space as first described by Huguier; and (2) distant pedicle flap attachment with delayed secondary detachment, described in 1897 by Nicoladoni.64 
...ld the thumb following traumatic amputation by staged, attached great toe-to-thumb transfer (Fig. 7), a reconstructive effort of substantial technical magnitude for its time, requiring an enormous degree of patient motivation and cooperation. In 1966, Tubiana and coauthors78 reported that on long-term follow-up of Nicoladoni's three patients in whom this technique was used, each refused subsequent bone grafting procedures and remained with unstable,
Figure 5. A and B. The importance of the thumb to normal human hand function is obvious on the sensory homunculus. Children with congenitally absent thumbs have no cerebrocortical thumb representation.

floppy thumbs after detachment. Clarkson as recently as 1955, however, reported the largest known series using Nicoladoni's principles of two-stage distant pedicle toe transfer in 15 digits transferred in six patients, one of whom underwent a 5-toe transfer for the treatment of complete congenital transverse failure of formation at the midpalmar level. Results were reported as completely successful, requiring only a later web-space deepening between the great and second toes.

The idea of using distant pedicle flaps in

Figure 6. Congenital thumb absence is associated with progressive widening of the index-middle finger web space, mild pronation of the index finger, and broadening of the index pulp; these adaptive changes facilitate elementary grasp and pinch between index and middle fingers.
rebuilding amputated thumbs was also proposed by Luksch in 1903, first describing the use of the contralateral index finger as a staged pedicle transfer. His work was based on an earlier extensive reconstructive experience by Guermonprez, which unfortunately went unnoticed and unappreciated for more than a half-century. Joyce, in 1918, reported reconstruction of the amputated thumb with the ring finger of the opposite hand in a manner similar to Guermonprez's work, relying on the principles of a distant pedicle flap with delayed detachment.

The "osteoplastic" reconstruction, was used extensively prior to World War II, using either one or two stages to attach the injured thumb stump to a random tubed pedicle of vascularized skin and subcutaneous tissue from the abdomen or groin, with iliac crest cortico cancellous bone grafting for skeletal reconstruction.

The historic principles that evolved during the early decades of this century continued in 1931 when Bunnell reported the first use of an index finger remnant (distal metacarpal and proximal phalanx) to reconstruct the lost basilar joint of a thumb. But not until 1949 did Gosset, in Paris, report the first transfer of an index finger on its neurovascular pedicle alone to a remote position on the hand. In introducing this technique, Gosset not only revolutionized contemporary traumatic thumb reconstruction, but pioneered seminal principles of congenital reconstruction for the absent thumb.

An international exchange of medical ideas and information after World War II found Gosset's techniques being used worldwide in centers for hand reconstruction. His initial techniques have been modified over the past four decades from those that involved simple lateral displacement of the index ray without bony shortening, to metacarpal recession and rotation, intrinsic muscle transfers, and skin flaps especially designed for a wide first web space, free of scar contracture.

Before these principles gained wide acceptance, Barsky and others sought to improve prehensile grasp in the congenitally deficient hand by surgical enhancement of the natural progressive pronatory deformity of the index finger in these children. Barsky advocated simple surgical deepening of the index-middle finger web space, with rotational osteotomy of the index metacarpal; the pulps of the "repositioned" index finger could be opposed for pinch and primitive grasp (Fig. 8A-C). Although at the time these principles were quite sound and were widely practiced, following World War II the technique was quickly replaced by neurovascular pedicle transfer with index recession and repositioning, adapted from techniques being practiced by Bunnell, Littler, and others for traumatic thumb loss.

For historic accuracy, Gersuny in Vienna in 1887, was the first to describe the principle of the neurovascular island pedicle flap. However, his reports identified no named artery. Not until 1893, when Dunham of New York described a permanently vascularized island pedicle flap with identifiable blood vessels, were principles established. The technique was widely applied throughout the first half of this century in the field of plastic surgery and craniofacial reconstruction, and became the basis for Gosset's later historic work in France.

A plethora of respectable contributions to the field of digital neurovascular pedicle transfer followed Gosset's original description. Success with index ray transfer led to the use of the middle, ring, and little fingers for reconstruction of thumb position, length, and sensibility. As recently as 1955, even the traditional osteoplastic staged pedicle-graft reconstruction received a boost by Moberg's introduction of the "island pedicle" transfer of vascularized, sensible, cornified skin from an uninjured portion of the hand, used to restore stereognosis to the tactile portion of the osteoplastic thumb. Thus, this type of staged reconstruction used combined principles of preliminary tubed abdominal pedicle, iliac crest cortico cancellous bone graft, and
neurovascular island pedicle transfer from another digit (usually the ulnar border of the middle finger), with subsequent cosmetic debulking. This technique was widely practiced.17

The thalidomide tragedy in Europe resulted in a devastating number of drug-induced congenital deformities of the upper and lower extremities between 1959 and 1962. Over a 10-year period, Buck-Gramcko11 performed an unprecedented series of 100 index finger pollicizations for congenital failure of thumb formation, mostly drug-induced. Through careful study and revision of both principles and technique, Buck-Gramcko presented the results of his work at the 1971 meeting of the American Society for Surgery of the Hand in a landmark paper on congenital thumb reconstruction. He emphasized in his report that digital transposition should be performed during infancy, as Riordan had advocated.11, 35, 69, 71 Using contributions of Gosset,30 Hilgenfeldt,36 Littler,47-50 Harrison,34, 35 and Riordan,55, 56, 71 Buck-Gramcko designed a surgical approach for the thumbless child, using the index finger, that resulted in stability, mobility, and optimum position and strength for prehensile grasp and pinch. His theoretical and practical contributions, including intrinsic muscle transfers, carpometacarpal

Figure 8. A–C, Barsky's technique for surgical improvement of grasp and pinch in the hand afflicted by congenital absence of the thumb involved index-middle finger web space deepening and abduction/rotation osteotomy to "reposition" the index finger. Principles were sound but, historically, the results were disappointing.

Figure 9. A and B, Complete failure of formation of the thumb has neither soft tissue nor bony thumb remnants. The entire osteoarticular column, including trapezium and scaphoid, are absent (Buck-Gramcko grade V thumb hypoplasia).
reconstruction, and the geometry of skin flaps to avoid first web space contracture, have become hallmarks of contemporary congenital thumb reconstruction.

CONGENITAL FAILURE OF THUMB FORMATION: THE APLASTIC THUMB AND THE "POUCE FLOTTANT"

Phenotypic expressivity of congenital deformity of the thumb has wide variations, including failure of formation of the entire osteoarticular column (including trapezium and scaphoid) (Fig. 9A and B) and the so-called floating thumb ("pouce flottant") (Fig. 10A and B).

A useful classification of the spectrum of congenital thumb involvement was suggested by Müller in 1937 as a "teratologic sequence," and later by Blauth in 1967. Buck-Gramcko (D. Buck-Gramcko, MD, personal communication, 1989) has refined and detailed these earlier contributions, and classified thumb hypoplasia into five reasonably distinct categories (Fig. 11A–E). Using this classification, severe thumb involvement with absence of the entire osteoarticular column and all soft tissue would be Grade V hypoplasia, or aplasia (the final most severe stage of thumb reduction in Müller's "teratologic sequence").

Grade IV hypoplasia describes the "pouce flottant" or "floating thumb" as a rudimentary appendage connected to the hand by a small skin tag, and usually containing two rudimentary phalanges and a single well-developed neurovascular bundle. The remainder of the thumb osteoarticular column is absent; the origin of this severely deficient appendage is along the radial midaxial line of the hand, and quite distal (Fig. 12A and B). The appendages are variable in size.

Assuming fundamental understanding and cooperation of both patient and family, complete absence of the thumb (grade V hypoplasia) should be treated early in life by pollicization of the index finger using the principles of Gosset, Littler, Riordan, and Buck-Gramcko. Progressive moderate pronation of the index ray, widening of the index-middle interdigital space, and broadening of the index pulp, all predispose the untreated patient to functional scissoring between index and middle fingers (Fig. 13).

Index ray development in patients afflicted by either complete thumb absence or pouce flottant (grade V or IV thumb hypoplasia) demands pollicization. Attempts to mobilize poorly developed and contracted joints to re-establish a proper orientation to the base of a grade IV floating thumb are unjustified, even without considering the problems inherent in multiple tendon transfers.

The constriction at the base of the pouce flottant permits digital survival during the postnatal period. The thumb unit is severely hypoplastic, usually devoid of metacarpal, trapezium, and scaphoid, and has neither intrinsic nor extrinsic tendons; the proximal and distal phalanges may be only cartilage remnants (see Fig. 11D). A thumb nail is usually present and developed to an extent similar to the general development of the entire distal thumb unit. It is important to emphasize that within the narrowed base of the grade IV hypoplastic thumb is a distinct neurovascular pedicle (Fig. 14A and B).

The pouce flottant should be amputated, either as an independent procedure or in conjunction with pollicization. In the newborn nursery, the base of the thumb can be anesthetized locally and a sterile silk tie applied tightly
A, Grade I thumb hypoplasia has an essentially complete complement of bones in the osteoarticular column; thenar and first web-space musculature is normal, joint range of motion is normal, and only the gross size of the thumb is diminished. B, Grade II thumb hypoplasia is manifest by an even smaller thumb stature than grade I hypoplasia and either reduction of volume or complete absence of the thenar muscles. Because of imbalance between thumb abductors and adductors, first web-space contracture is common. The bones of the osteoarticular column are narrow, and instability of the ulnar collateral ligament of the metacarpophalangeal joint is common. Basilar thumb instability may also be found, although the radial carpus (scaphoid, trapezium, and trapezoid) is usually present. C, Grade III thumb hypoplasia is associated with severe first web-space contracture secondary to a usual complete absence of thenar muscles. The thumb is short, and global instability of the metacarpophalangeal joint is common. Osseous and articular relationships at the thumb base are variable in their formation. D, Grade IV thumb hypoplasia is the classic *pouce flottant* (floating thumb). A well-formed neurovascular pedicle can be found coursing through the pedicle; the origin of the pedicle is quite distal along the radial mid-axial line. All thenar and adductor thumb musculature is absent, except for the second metacarpal origin of the first dorsal interosseous (abductor indicis). The trapezium and scaphoid are usually absent; the trapezoid is often normal. E, Grade V thumb hypoplasia is complete congenital absence of the thumb (aplasia); even the abductor indicis are absent or deficient in more than 50% of cases. The index finger usually undergoes pulp widening, digital pronation, and index-middle finger web-space widening, with progressive curvature of the index ray. These cases are ideal candidates for index finger pollicization.
Figure 12. A and B, Distal and dorsal origin of the pouce flottant makes functional reconstitution difficult. To perform prehensile tasks, the origin of the thumb unit must be surgically moved more proximal and volar, and bone grafts used to create a stable base.

Figure 13. Patients with severe thumb hypoplasia prefer the scissoring action of index-middle abduction/adduction for pinch and minor grasp. Left untreated, even the "rich" hypoplastic thumb will frequently be bypassed because of its subnormal stability and mobility and its less than normal cerebrocortical representation.

Figure 14. A, In spite of severe underdevelopment, gross instability, and a narrow constricting pedicle at its base, the pouce flottant has a normal complement of digital nerves and a single large digital artery with its venae comitantes (arrow). B, Arteriogram of a patient with grade IV hypoplasia clearly demonstrates a single large unconstricted digital artery passing through its narrow base.
at the base to induce ischemic necrosis. A small amount of long-acting anesthetic prevents the pain associated with crushing a well-developed digital nerve with the tie.

Match reported filleting open the floating unit at the time of pollicization, and incorporating the skin of the deficient thumb into the new first web space (Fig. 15). Whether the part is ablated as an independent procedure or removed at the time of pollicization, efforts should not be made to reconstruct a grade IV hypoplastic thumb with a functional unit as a surgical goal. The results of such efforts are poor both functionally and cosmetically. The child—often after a plethora of surgical procedures—will bypass the reconstructed unit, preferring a more natural index-middle finger scissoring action. Dexterity of the medial four fingers outweighs any effort to create a functional thumb unit from the pouce flottant.

There are, however, exceptions to any rule. Particularly in cultures in which a five-fingered hand has extreme importance, select cases may justify efforts to reconstruct a Grade IV hypoplastic thumb. In these rare instances, the site of origin of the thumb base must be shifted to a more palmar and proximal locus, ensuring stability by either autogenous corticocancellous iliac crest graft or longitudinal fibular strut, inserted between the base of the index metacarpal and the hypoplastic thumb rudimentary proximal phalanx (Fig. 16 A and B). Concomitant reconstruction of a wide first web space is obviously necessary. Tajima has advocated a large dorsal skin flap designed over the index metacarpal and proximal phalanx, which can be rotated with the floating thumb to create an acceptable first web; skin graft is applied to the flap donor site. Osseous stability in these grade IV hypoplastic thumbs can also be achieved using a free nonvascerealized transfer of the metatarsophalangeal joint of the fourth toe, potentially providing a degree of thumb mobility (Fig. 17 A–E). Because of the importance of the five-fingered hand in Japanese culture, techniques oriented towards salvage and reconstruction of the congenital floating thumb have emanated principally from that country. Reconstruction is obviously a major surgical effort, which involves stabilizing the digit by bone graft or whole joint transfer, complex skin flaps and grafts, and three subsequent individual tendon transfers as potential balanced motors. Thumb extension can be achieved by transfer of the extensor indicis proprius; abduction and/or opposition by the Huber transfer of the abductor digiti quinti (modified by Littler); and flexion/adduction by direct transfer of the ring finger flexor digitorum superficialis (Fig. 18 A–E).

The surgeon must realize that results of these reconstructions are disappointing at best. Poor joint development should be an absolute contraindication for tendon transfer: by definition, tendon excursion is severely limited by poor joint mechanics, and the natural pattern of functional index-middle finger scissoring continues. The potential for index pollicization may also be lost by irreparable surgical damage to skin incurred during primary efforts to make the grade IV hypoplastic thumb functionally useful. None of White’s priorities for thumb reconstruction can be met: despite excellent sensibility, opposition cannot be effectively generated from a rigidly fixed thumb base. Pollicization of the index finger becomes a very attractive alternative.

POLlicization FOR CONGENITAL THUMB DEFICIENCY

Far more challenging than the theoretically similar reconstruction for traumatic thumb loss,
In those rare instances where reconstruction of a floating thumb is indicated, stability can be attained by a corticocancellous pelvic strut graft placed between the distal osteoarticular remnant and the base of the second metacarpal. Improper reconstruction of the base of the floating thumb affords absolutely no increased potential for prehensile grasp or pinch.

Figure 16. A, In those rare instances where reconstruction of a floating thumb is indicated, stability can be attained by a corticocancellous pelvic strut graft placed between the distal osteoarticular remnant and the base of the second metacarpal. B, Improper reconstruction of the base of the floating thumb affords absolutely no increased potential for prehensile grasp or pinch.

Figure 17. A–E, The Tajima technique for reconstruction of a pouce flottant achieves basilar joint stability by nonvascularized metatarsophalangeal joint transfer from the fourth toe (A) and establishes a wide first web space by large dorsal rotation flap design (B and C). After achieving temporary bony fixation with Kirschner wires (D), full-thickness skin graft is applied to the flap donor site (E).
pollicization for *congenital* thumb deficiency may require use of a digit that itself may be anatomically deficient, particularly in the case of radial club hand. Pollicization was first successfully adapted to congenital hand reconstruction in 1960 by Zancolli, but the thalidomide tragedy of 1959–1962 led Buck-Gramcko to his unparalleled experience and confirmed the usefulness of index finger pollicization as the procedure of choice for treatment of grade IV and V congenital thumb formation failure.

The functional goals of pollicization are reached only with painstaking attention paid to technical details. The incisions must be designed to allow shortening, rotation, and repositioning of the digit to be pollicized. Skin flaps must form a well-contoured first web space, free of restraining scar. Stabilization of the base of the new thumb can be achieved by a variety of techniques; but the propensity for severe basilar joint hyperextension must be addressed if the index metacarpophalangeal (MCP) joint...
is used as the new thumb carpometacarpal (CMC) joint. Buck-Gramcko recognized this problem, rectifying it by hyperextending the condyloid surfaces of the index MCP joint at the time of surgical stabilization, thereby redirecting the new carpometacarpal arc of motion to the flexion rather than hyperextension range (Fig. 19A). Osteotomy is performed through the distal index metacarpal physis and the entire proximal index metacarpal is removed. The transposed index ray should be secured in 120° pronation, 40° palmar abduction, and 15° extension. I prefer disarticulation of the index carpometacarpal joint, leaving a sleeve of capsule to which the hyperextended MCP joint can be securely anchored (see Fig. 19A and B).

**MOTORING THE POLLICIZED DIGIT**

Although the time-tested principles of digital transposition on neurovascular pedicles, first recommended in this country by Littler, in 1953, are still used as the basis for pollicization, new techniques for motoring the transposed digit are regularly being introduced in the literature. The currently popular intrinsic transpositions of first dorsal and first volar interossei for thumb abduction and adduction stem from the accumulated experiences of Riordan and Buck-Gramcko.

Figure 20. Principles of meta-carpal shortening and tendon transfers for index pollicization were developed by Buck-Gramcko following the thalidomide tragedy in Europe. The technique relies upon extensor indicis proprius (EIP) shortening by a length equal to the portion of index metacarpal resected; the extensor digitorum communis (EDC) II is rerouted to abduct the new thumb metacarpal base, with abduction and adduction restored by the 1st dorsal interosseous (DI) and 1st volar interosseous (VI), respectively.

Buck-Gramcko has reported it unnecessary to shorten the extrinsic index (new thumb) flexor system or to remove the lumbrical from the index profundus tendon, despite resection of the entire proximal index metacarpal. He has, however, advised shortening the extrinsic extensor tendons (extensor indicis proprius and index extensor digitorum communis) by length equal to the segment of metacarpal removed. Manske and McCarroll disagree with this recommendation and report that in their experience the extensor indicis proprius muscle-tendon unit also spontaneously absorbs the slack of skeletal shortening. These authors believe that flexors and extensors should be left surgically unshortened.

The currently popular Buck-Gramcko pollicization technique suggests distal detachment of the insertions of the abductor indicis (first dorsal interosseous) and first volar interosseous from the index proximal phalanx (Fig. 20). These motors can then be advanced distally into the radial and ulnar lateral bands of the dorsal apparatus and function in an abduction-adduction plane (1st DI for abduction; 1st VI for adduction). The design of skin flaps serves simply to accommodate the increased muscle volume at the level of the new thumb metacarpal, and to minimize scar contracture in the new first web space.

In personal experience with 45 pollicizations for congenital thumb deficiency, I have found...
complete or partial absence of the abductor indicis (first dorsal interosseous) in more than 50% of cases. In these patients, active palmar abduction of the new thumb can still be achieved by transfer of the extensor digitorum communis more distally on the volar border of the proximal phalangeal diaphysis (Fig. 21). This technique substitutes extensor digitorum communis (EDC) for the absent abductor indicis for thumb abduction. If necessary, supplementary use of either the ring flexor digitorum superficialis or the abductor digitii quinti as an opponensplasty can be made, either at the time of pollicization or secondarily.37,38

An alternative technique of pollicization, which I use on occasion, simplifies skin flaps and minimizes dissection into the index proximal phalanx and its dorsal apparatus. The muscle belly of the first volar interosseous is rotated and distally advanced along the periosteum of the adjacent third (middle) metacarpal, leaving its insertion unchanged. This technique still relies upon the 1st VI for functional thumb adduction, but without detachment and distal advancement of the tendon to the ulnar lateral band as Buck-Gramcko described (Fig. 22A-C; see Fig. 21).

The surgeon must be aware of the potential

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**Figure 21.** The Buck-Gramcko technique for index pollicization requires the presence of a healthy first DI and distal dissection into the radial and ulnar lateral bands of the digit. In the absence of a first DI, an alternative for thumb abduction is to advance the EDC II more distal on the proximal phalanx of the index finger. Adduction can be alternatively achieved by proximal advancement of the muscle belly of the first VI (palmar I) on the shaft of the third metacarpal, sparing dissection into the digit itself (see text).

**Figure 22.** Incisions used for the pollicization technique described in Figure 21.
for anatomic variations in any type of congenital hand reconstruction, particularly with respect to availability of useful motors for the pollicized digit. If present (and if substantial), the first dorsal interosseous should be used preferentially for palmar abduction, as detailed by Buck-Gramcko; however, distal and volar advancement of the index EDC should always be considered an option when a poorly developed 1st DI exists. Tendon transfers described earlier in this chapter for motoring the pouce flottant are readily adaptable to a deficient pollicized index finger (e.g., abductor digiti quinti or ring flexor superficialis for opposition, or extensor digiti quinti proprius for thumb extension in the absence of developed extrinsic index extensors). The surgeon's own ingenuity and skill in selecting the most appropriate transfer will yield the best functional thumb unit.

HYPOPLASIA OF THE THUMB, EXCLUDING POUCE FLOTTANT AND COMPLETE ABSENCE

While grade I hypoplasia is essentially a normal functioning thumb, except for being slightly smaller than normal size, the grade II hypoplastic thumb is considerably smaller and less stable. Thenar musculature is usually deficient and palmar abduction limited; the first web space is narrow, and thumb pronation is restricted (see Fig. 12A). The ligaments of the metacarpophalangeal (MP) joint (particularly the ulnar collateral ligament) are poorly developed, which further compromises pinch.

Cases of grade II hypoplasia manifest a plethora of abnormalities. Deficiency in extrinsic flexor and/or extensor systems may also be included, particularly an often-observed tendinous connection between the flexor pollicis longus and extensor pollicis longus tendons (G. Lister, MD; S. Stahl, MD, personal communication, 1989). Any combination of abnormal tendon vectors or insertions, absence or unusual presentation of pulleys, or complete absence of tendon systems may be found in the grade II hypoplastic thumb category.

Grade III thumb hypoplasia exhibits a more profound reduction in thumb size, stability, and length (see Fig. 12C). Instead of simple MP collateral ligament attenuation observed in Grade II hypoplasia, the MP joint of the grade III thumb is flail, with usually only a rudimentary CMC joint. Extrinsic tendon systems are absent, and there is essentially no participatory function of the thumb unit in routine hand function (Fig. 23).

Figure 23. Profound intrinsic and extrinsic muscle deficiency usually accompanies Buck-Gramcko grade III thumb hypoplasia. Because of poor motor development, and position-irregularity of the base, this thumb will be permanently bypassed in favor of index-middle finger scissoring.

RECONSTRUCTION

The philosophy of surgical reconstruction of the grade III hypoplastic thumb (see Fig. 12C) is based on the potential for pinch-and-grasp enhancement, using the patient's own thumb. If the thumb lever arm is short only because of compromised longitudinal skeletal development (brachydactyly, as opposed to transverse failure of formation), it can be lengthened, as long as carpometacarpal stability and mobility are reasonable (grade II). Additional length can be provided in a variety of ways; one is by extraperiosteal transfer of the fourth toe proximal phalanx into the collapsed soft-tissue digital tube of the hypoplastic thumb (Fig. 24, A and B). A chevron incision is used, and the dorsal apparatus of the toe is split longitudinally; the proximal phalanx can then be disarticulated and removed en bloc. Care must be taken to avoid injury to the extrinsic toe flexors, which conform intimately to the plantar contour of the proximal phalanx. Once removed, the longitudinal extensor tenotomy is repaired and the skin closed. The resulting shortened toe is functionally stable, and not cosmetically disfiguring.

Carroll and Green reported no active growth in their series of free phalangeal transfers removed by subperiosteal dissection. Goldberg and Watson, however, suggested up to 90% preservation of normal growth from the transplanted epiphyseal plate (age-dependent) using extraperiosteal dissection. Subperiosteal harvesting and transfer can result in partial resorption of the transplanted phalanx; extraperiosteal transfers undergo no resorptive change, and a variable rate of growth.

Once filled with transplanted bone, further thumb length can be attained by osteotomy and
distraction using an external fixation device (Matev, Kessler) designed to gradually create an intercalary defect, secondarily bone grafted when an appropriate length has been achieved (Fig. 25A–D). This technique, useful in both congenital and traumatic thumb reconstruction, is usually combined with first web space deepening by simple $60^\circ$ Z-plasty, 4-flap Z-plasty (Broadbent and Woolf), or a dorsal index-finger rotation flap with full-thickness skin graft to the donor site (Fig. 26). Donor bone graft for the intercalary skeletal defect

Figure 24. A and B, Thumb lengthening by nonvascularized toe phalangeal transfer can be performed by extraperiosteal harvesting of the fourth or fifth toe proximal phalanx through a chevron incision; a temporary percutaneous Kirschner wire can be used to maintain length until healed (3 to 4 weeks). C, Extraperiosteal toe proximal phalangeal transfer to lengthen and stabilize a grade III hypoplastic thumb (arrow).

Figure 25. A–D, Extraperiosteal toe phalangeal transfer to the thumb, index, ring, and little finger metacarpal bases in a child with transverse failure of formation: Once bone transfer to the digital tube of the hypoplastic thumb has been performed, additional length can be secondarily achieved by osteotomy and progressive lengthening in an external distraction device (B and C); the intercalary defect is filled by corticocancellous iliac crest bone graft (D).
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Figure 26. After thumb lengthening, first web-space release is achieved by first DI recession and partial adductor myotomy. Skin coverage of the new first web can be by one of a variety of methods, including dorsal rotation flap from the index finger with adjunctive full-thickness skin graft.

Figure 27. An 8-year-old child after 2 of three planned tendon transfers for grade III hypoplasia. Results were so disappointing that plans for the final stage were abandoned.

Figure 28. A and B, Mild hypoplasia of the thumb justifies efforts to reconstruct it, particularly if only minor tendon transfers are necessary. It is difficult to recommend ablation of a digit with near-normal skeletal development.
Figure 29. The principles of opponensplasty rely on a change in motor vector from the pisiform to the radial border of the thumb metacarpophalangeal joint. In this example, the FDS IV is harvested through an incision in the distal palmar crease (A and B), rerouted deep to the FCU at the wrist (C), and passed subcutaneously from the pisiform to the thumb (C and D) under appropriate tension.

exceedingly difficult task for the hand surgeon; but predictable functional failure in lengthening, repositioning, and tendon transfers, will make ablation and index pollicization an attractive alternative occasionally. Reluctance to ablate and pollicize by both the surgeon and the parents is understandable; but the realistic potential for incorporation of a reconstructed hypoplastic thumb into coordinated prehensile activities of the hand must be given paramount consideration in any treatment plan proposed.

CONGENITAL ABSENCE OF THE THENAR MUSCLES

A commonly observed manifestation of congenital failure of formation of soft tissues of the hand is absence of the thenar muscles, often part of grade II and invariably part of grade III hypoplasia (see Fig. 12B and C). These patients are unable to lift the thumb from the plane of the palm for grasp, and present with a narrow first web space.

For simple thenar muscle group deficiency with full passive thumb range of motion (ROM), opponensplasty using the ring or middle finger flexor superficialis is recommended. These motors are usually available, certainly expendable, and have the appropriate cross-sectional area and muscle amplitude for strong opposition. Using the distal flexor carpi ulnaris as a pulley, the "angle of attack" of the transfer on the thumb provides a palmar abduction/flexion pull from the vicinity of the pisiform (Fig. 29A–D).

Figure 30. Sequential technique for release of the con- tracted first web space: (1) recession of the first metacarpal origin of the 1st DI muscle, and (2) myotomy of the transverse head of the adductor pollicis.
transfers can be considered. First web space release may require recession of the first dorsal interosseous from the first metacarpal, or myotomy or recession of the transverse head of the adductor pollicis, as described by Matev35, 86 (Fig. 30). Division of the oblique CMC and/or transverse intermetacarpal ligaments supporting the base of the thumb is usually unnecessary. If the initial thumb position is severely adducted, adjunctive full-thickness skin grafts may be necessary to cover exposed raw areas, regardless of the flap design used to reconstruct the new first web space (Fig. 31). Skin rearrangement, whether by Z-plasty flaps, rotation flaps,26 or full-thickness skin grafts, provide both breadth and increased depth.

ABSENCE OF THE EXTRINSIC TENDONS OF THE THUMB

An unusual anomaly of the thumb associated with hypoplasia involves failure of formation of either the long extensor (Fig. 32A and B) or long flexor (Fig. 33A and B) tendon. Inheritance is autosomal dominant, with a high penetrance and expressivity (Fig. 34).

Patients with absence of the extensor pollicis longus present with an interphalangeal (IP) flexion attitude (see Fig. 32A), similar to that observed in congenital trigger thumb. Conversely, failure of formation of the flexor pollicis longus manifests itself as inability to actively flex the IP joint, with absence of distal volar digital skin creases (see Figs. 33A and 34).

If the IP joint is laterally stable and passively flexible, the ring flexor digitorum superficialis transfers can be considered. First web space release may require recession of the first dorsal interosseous from the first metacarpal, or myotomy or recession of the transverse head of the adductor pollicis, as described by Matev35, 86 (Fig. 30). Division of the oblique CMC and/or transverse intermetacarpal ligaments supporting the base of the thumb is usually unnecessary. If the initial thumb position is severely adducted, adjunctive full-thickness skin grafts may be necessary to cover exposed raw areas, regardless of the flap design used to reconstruct the new first web space (Fig. 31). Skin rearrangement, whether by Z-plasty flaps, rotation flaps,26 or full-thickness skin grafts, provide both breadth and increased depth.

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Figure 33. Absence of distal digital thumb flexion creases in the presence of full passive range of motion is the hallmark of congenital failure of formation of the flexor pollicis longus (A and B); the flexor pollicis longus is vestigial and can often be excised from the system, allowing the ring flexor digitorum superficialis to be transferred directly through the oblique pulley to the terminal phalanx (C and D).

can be transferred to create active thumb flexion (see Fig. 33C). Although the terminal flexor tendon is vestigial, a reasonably well-developed pulley system is usually present; the existing nonfunctional tendon system can be excised, and the ring superficialis transferred through the oblique pulley to the distal phalanx of the thumb (see Fig. 33D).

Tendon reconstruction dorsally is more difficult because of the wide variations in deficiency of the extensor pollicis longus and dorsal apparatus. The extensor indicis proprius with or without free tendon graft (see Fig. 18A) can often be used to restore active extension to the passively extensible thumb IP joint. However, if the joint remains stiff even after conscientious therapy, arthrodesis is indicated. Fusion is performed between the distal metaphysis of the proximal phalanx and the epiphysis (secondary ossification nucleus) of the distal phalanx, preserving the physis and growth potential of the more distal bone (Fig. 35 is an example of the same technique used at the thumb MP joint).

RECONSTRUCTION OF THE CONGENITALLY DEFICIENT OR ABSENT THUMB BY TOE-TO-THUMB VASCULARIZED TISSUE TRANSPLANTATION

Additional length can be achieved in a congenitally deficient thumb by various methods, including (1) the insertion of a free nonvascularized toe proximal phalanx into an existing collapsed soft-tissue digital tube (see Fig. 25A), (2) toe phalangeal transfer as intercalary graft (see Fig. 24C), or (3) lengthening of existing bone by osteotomy and progressive distraction by means of an external fixation device, with secondary intercalary bone graft (see Fig. 25B–D).

Over the past two decades, another technique has been added to the surgeon's armamentarium: free-tissue transplantation using microvascular surgical disciplines. Introduced by Cobbett, in London in 1969, for thumb reconstruction, the early technique was based on pioneering microsurgical research by Buncke in primates 3 years before. Cobbett's
Management of Thumb Hypoplasia

The longitudinal growth potential of phalanges arthrodesed at the metacarpophalangeal or interphalangeal joint can be preserved by fusing the distal metaphysis of the proximal bone to the secondary ossification nucleus of the distal bone, without violating the physis. In this example, the arrow indicates the healed fusion between a distal metacarpal and the epiphysis of a thumb proximal phalanx. Note the persistent open physis of the proximal phalanx.

Single case involved vascularized transplantation of a great toe after traumatic loss of a thumb. The surgery and postoperative course of this landmark case was quite stormy.

Following publication of Cobbett’s experience, techniques were refined, with many authors reporting successful series of microvascular toe-to-thumb transplants to restore function following traumatic amputation. But not until 1978 did O’Brien, and coauthors, report the first case of a free-vascularized great toe transplantation for congenital absence of the thumb. Yoshimura, in 1980, reported similar success in two congenitally deficient patients. May and coworkers in 1981 reported a successful experience using techniques of microvascular free-tissue transfer to build a thumb in a hand afflicted by multiple digital aplasia. Arteriography may be helpful in assessing which congenital patients are suitable candidates for microvascular free-tissue transfer. If proper patient selection is employed, marked functional and cosmetic improvement can be attained (Fig. 38A and B). The skilled surgeon’s armamentarium using microvascular transfer and free-tissue transplantation can be represented by either nonfunctional tissue distal to congenital constriction bands (A) or by complete digital absence distal to a certain level (B). In either type, the anatomy of the hand proximal to the defect is normal and should be a suitable recipient bed for free vascularized toe-to-hand transplantation.
Figure 37. Presence of nail elements in a congenitally deformed hypoplastic digit defines a longitudinal, rather than transverse, failure of formation. Proximal vessel and nerve anatomy is anomalous and unpredictable, making these children poor candidates for microvascular free-tissue reconstruction.

The congenitally deficient hand remains one of the more controversial facets of hand surgery. Riordan\(^\text{59, 71}\) and Flatt\(^\text{25}\) have suggested that pollicization can be safely performed at 6 to 12 months of age. Buck-Gramcko has reported good results at an even earlier age (D. Buck-Gramcko, MD, personal communication, 1985). However, because the cardiopulmonary system is sufficiently developed by 1 year postpartum to make general anesthetic risks minimal, and because bimanual grasp develops normally by 12 months of age, perhaps pollicization should be reserved until after the age of 1 year.

Another consideration is that although size itself is not a major contraindication for earlier pollicization, normal involution of embryonic endosteal circulation during the first year of life makes tourniquet control of intraoperative hemorrhage more effective if the surgeon delays surgery until 1 year of age.

Figure 38. A and B. Appropriately selected patients can undergo free vascularized toe-to-thumb transplantation for congenital failure of thumb formation; in this case, the second toe was used as donor tissue. With suitable indications, tenorrhaphy, neuorrhaphy, and bony reconstruction can afford sensibility, stability, and mobility to the reconstructed unit. Phases of all bones of the transplant remain open, and growth potential parallels that of the normal bones of the hand.
Manegement of Thumb Hypoplasia

Figure 39. A and B, The rewards of reconstruction of congenital aplasia or hypoplasia of the thumb are functional improvement in prehensile grasp and pinch. In these examples, three hands in two affected patients show typical results that can be achieved by pollicization. Any of the techniques described in this article—when used appropriately—can enhance the functional capability of the hand.

SUMMARY

This chapter emphasizes the dilemma of salvage and reconstruction of the congenitally aplastic or hypoplastic thumb without normal cerebrocortical representation for prehensile grasp and pinch. The philosophy of reconstruction of the congenitally anomalous thumb is clearly divergent from thumb reconstruction following trauma. It is difficult to advise parents who seek surgical correction of the severely deficient thumb ray that amputation is the procedure of choice, to be followed by transfer of an otherwise normal digit to become a functional thumb unit. The psychological impact of these recommendations may be devastating to parents. Informed understanding of the likely progressive development of index-middle finger scissoring, pronation of the index ray with spontaneous broadening of the pulp, and the identification in the abundant subcutaneous tissue of the normal infant hand. By 2 years of age, an infant’s friable arteries and veins have developed into vessels of substantial thickness, suitable for microvascular anastomosis. Gilbert also feels that although it is not clearly understood how the new thumb is cerebrocortically integrated for grasp and pinch, these routine activities seem easier for the patient when surgery is performed at a younger age.

What is the appropriate age for reconstruction? The surgeon should take into account these four points.

Development of the cardiopulmonary system

Involution of the embryonic endosteal circulation

Size of structures and vessel friability, particularly in microvascular cases

Normal development of bimanual grasp as a functional landmark

The rewards of reconstruction of congenital aplasia or hypoplasia of the thumb are functional improvement in prehensile grasp and pinch. In these examples, three hands in two affected patients show typical results that can be achieved by pollicization. Any of the techniques described in this article—when used appropriately—can enhance the functional capability of the hand.

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deteriorating use of an existing hypoplastic thumb may make the decision for ablation easier for parents. It is critical that these decisions be made through careful education and understanding, considering always the overall grasp-and-pinching capability of the hand as a whole.

Reconstruction of the aplastic or hypoplastic thumb is an exciting and challenging area of hand surgery. The rewards are improvement in grasp and pinch either by functional integration of the reconstructed part, or by complete replacement of the deficient thumb by tissue from adjacent or distant donor sites. Success is measured not only in terms of cosmetic appearance, but as enhanced capacity of the child’s hand in all activities of daily living (Fig. 39A and B).

REFERENCES

Management of Thumb Hypoplasia

51. Lutsko I: Über eine neue Methode zum Ersatz des verlorenen Daumens. Verhandlungen der Deutsche Gesellschaft Chirurgie 32:221, 1903
64. Nicoladoni C: Daumenplastik. Wein Klin Wochenschr 10:663, 1897
68. Pierce GW: Reconstruction of the thumb after total loss. Surg Gynecol Obstet 45:625, 1927

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