

Reorientation Osteotomy for the Atypical Clasp Thumb in Children With Arthrogryposis

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Abstract: Congenital thumb contractures are a well described but complex deformity to manage. Thumb contractures are common in patients with arthrogryposis. Many patients with arthrogryposis develop a supination, adduction, and flexion contracture at the thumb carpometacarpal joint. Despite some thumb function, their thumb position and contracted first webspace precludes effective pinch or grasp, with the thumb opposing only to the palmar space. An extension, reorientation metacarpal osteotomy that places thumb in an efficient position has the potential to improve function in of these patients. This report provides a new classification system for congenital thumb deformities that can dictate management based upon thumb joint positions. We describe a previously unreported, arthrogryptic thumb contracture that can be managed with an extension/pronation metacarpal osteotomy with simultaneous widening of the first webspace.

Key Words: arthrogryposis, congenital thumb contracture, metacarpal osteotomy

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HISTORICAL PERSPECTIVE

Arthrogryposis is a descriptive term for congenital joint contractures in at least 2 joints. The incidence is 1 per every 3000 live births and it is associated with hundreds of congenital disorders.^{1,2} Subcategories of arthrogryposis

include sporadic amyoplasia and various autosomal dominant disorders referred to as distal arthrogryposis.^{3,4} Although the contractures are generally nonprogressive, arthrogryposis can be a disabling disorder affecting independent function, feeding, and personal hygiene. Treatment plans must be individualized, as contracture severity and type varies between patients. Multidisciplinary approaches involving surgical teams, occupational and physical therapy begin as early as possible to maximize therapeutic gains. Surgical procedures that can increase function are critical to patients with arthrogryposis, as normal intellect and sensation make them excellent rehabilitation candidates.⁵

The hand of a child with arthrogryposis typically exhibits a thumb in palm deformity and is one of the most varied and complex thumbs to manage.^{6,7} This thumb contracture has also been referred to as clasped thumb, adducted thumb, or pollex varus and is associated with an abnormal first webspace and intrinsic musculature, attenuated extrinsic tendons, and joint stiffness.⁷ The most commonly described thumb in palm posture involves flexion of the interphalangeal (IP) and metacarpophalangeal (MP) joints with carpometacarpal (CMC) joint extension (Fig. 1A).⁸ This deformity limits grasp and pinch function. Depending on the severity and underlying anatomic characteristics of the contracture, treatment options vary from stretching and splinting to surgical management. Many patients with arthrogryposis, particularly amyoplasia,



FIGURE 1. Clinical photograph of the right thumb of a patient with a classic clasp thumb (A), type 1 by the suggested classification. By contrast, a type 2 thumb exhibits a flexion, adduction, and supination thumb deformity at rest (B) with nonfunctional active thumb opposition to space (C). Courtesy of the Shriners Hospital for Children, Philadelphia.

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TABLE 1. Classification System for Congenital Thumb Contractures Based on Position of CMC and MP Joint Position With a Modifier of “A” and “B” Indicating Whether or not the MP Joint Contracture is Passively Correctable

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

CMC indicates carpometacarpal; MP, metacarpophalangeal.
 Courtesy of Dan A. Zlotolow, MD.

demonstrate a different set of thumb contractures than the classic clasped thumb. To our knowledge, this has not previously been discussed in the literature. Flexion of the CMC joint combined with extension of the IP and MP joint results in a thumb that is not truly in the palm, however, the coincident adduction and supination render the thumb incapable of opposition, even when good flexor pollicis longus function exists. This deformity results in opposition to space when attempting grasp or pinch (Figs. 1B, C). The first webspace is contracted, with the thumb resting adjacent to and in the plane of the index finger. In these patients, the often recommended thumb reconstruction algorithm does not apply. Performing a chondrodesis of the MP joint in patients with this deformity does not make clinical sense as the MP joint rests in extension. Fusing the CMC joint is also not an option as this is sometimes the only mobile segment.

We have developed a thumb reorientation procedure that incorporates elements of the more common thumb in palm reconstruction, such as expanding the first webspace, in combination with an abduction, pronation, and extension osteotomy at the thumb metacarpal base. In cases of arthrogryposis where the thumb MP joint is in extension, a metacarpal base osteotomy preserves motion at the CMC joint and reorients the thumb in an

improved functional position. Rotational osteotomies of the thumb metacarpal have improved grasp and pinch function in patients with “thumb in the plane of the hand” congenital deformity. Langer et al⁹ used an abduction/pronation metacarpal base osteotomy to restore opposition and pinch in these affected hands. Adopting these same concepts and applying them to patients with an adducted and supinated thumb can improve pinch function in patients with arthrogryposis. The Langer technique was not standardized and provided no detailed method of the procedure. Variable flaps were used to widen the first webspace based on individual clinical presentations.⁹ Conceptually, the Langer technique and the below described technique are similar and involve opening the first webspace and an osteotomy at the base of the first metacarpal. This paper details a technique with a dorsal closing wedge osteotomy with an index rotation flap and describes a new treatment algorithm for thumb deformities in children with arthrogryposis.

In addition to thumb position, restoration of the normal first webspace depth is important for grasp, pinch, and prehension.^{10,11} Although mild contractures have excellent results with z-plasty procedures, skin deficiencies in >1 plane require more extensive coverage. Local, regional, and distant flaps have all been described for treatment of first webspace contractures.^{11,12} The index rotational flap allows for correction in 2 planes and has gained popularity since first described by Ezaki and Oishi.¹³ The low morbidity and primary closure of the donor site has increased its use for first webspace contractures associated with congenital thumb differences.

Indications

This procedure is indicated for patients with a supinated, adducted, and flexed thumb CMC joint. In these patients, the thumb opposes to space instead of to the fingers, preventing functional pinch or grasp. Management and classification of congenital thumb contractures are predicated upon the position of the CMC and MP joints. A new, simplified classification consisting of the 3 major types of thumb deformities in children with arthrogryposis is presented in Table 1. Each type

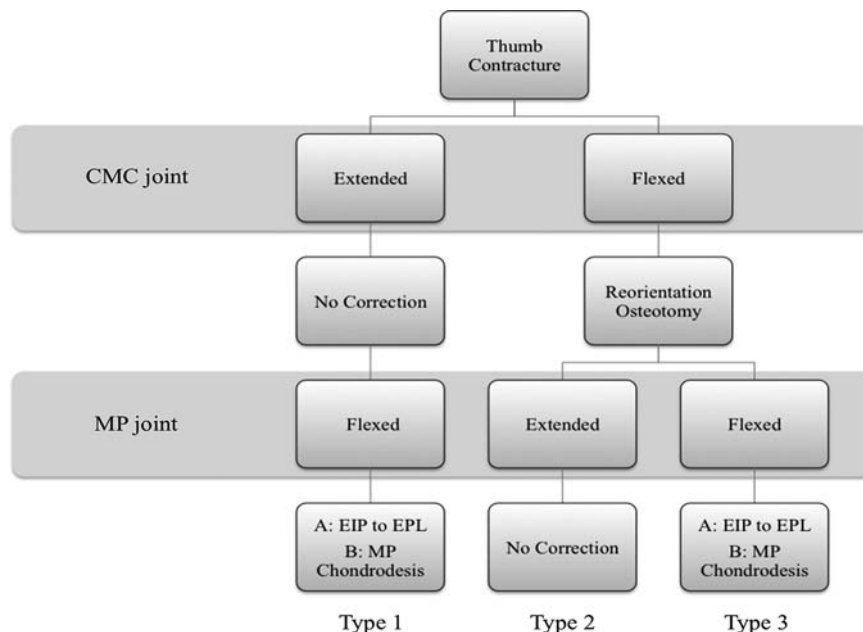


FIGURE 2. Flowchart detailing the management for thumb contractures based upon carpometacarpal (CMC) and metacarpophalangeal (MP) joint position. EPL indicates extensor pollicis longus; EIP, extensor indicis proprius. Courtesy of Dan A. Zlotolow, MD.



FIGURE 3. Intraoperative photograph outlining the index rotational flap and metacarpal osteotomy incision sites. Courtesy of the Shriners Hospital for Children, Philadelphia.

is classified based on CMC and MP joint position. A modifier of “A” and “B” indicates whether or not the MP joint contracture is passively correctable (Table 1). Type 1A thumbs are best treated with a tendon transfer if a donor is available. If no donor is available for a 1A, or the patient has a 1B thumb, an MP joint chondrodesis and index rotation flap are most appropriate. Type 2 thumbs are not amenable to the same treatment as type 1 thumbs, as the primary flexion posture is at the CMC joint (Fig. 2). The position of the CMC can only be corrected with a CMC fusion or an extra-articular reorientation osteotomy. The advantage of a fusion is that there is no risk of deformity recurrence. However, in children with limited mobile segments, the advantage of preserving joint motion is even more significant. An extension osteotomy at the base of the metacarpal is intended to correct the flexion posture of the CMC joint while maintaining motion. The osteotomy described below also addresses the adduction and supination of the thumb, whereas the index rotation flap widens the contracture of the first webspace.

Contraindications

Each patient with arthrogryposis has variable contracture severity, rendering a standardized surgical approach impossible. We advocate caution when proceeding to surgery when patients who exhibit an adducted and supinated thumb are still functional. If the patient is able to function independently, we opt for continued observation. If the patient has no flexor pollicis brevis function, thumb reorientation is contraindicated as active pinch and/or grasp are not achievable. In patients with minimal hand function, the adductor pollicis may be the only working muscle and abduction of the thumb can compromise the patient’s hand use.

The family should be aware that any change to a child with arthrogryposis, particularly in older children, could have unforeseen consequences in the child’s global function. The patient’s shoulder, elbow, and wrist and hand function should be considered in concert with the primary goals of self-feeding and perineal care. Although we do not recommend a specific age for this procedure, we have found that delaying surgery



FIGURE 4. Intraoperative photograph of the exposure of first dorsal interosseous and adductor pollicis muscles with release of the overlying abnormal fibrous bands. Courtesy of the Shriners Hospital for Children, Philadelphia.



FIGURE 5. Intraoperative photograph of the closing wedge osteotomy site between the extensor pollicis longus and extensor pollicis brevis tendons and distal to the physis, with the retractors protecting the extensor tendons and the periosteum. Courtesy of the Shriners Hospital for Children, Philadelphia.



FIGURE 6. Intraoperative radiograph demonstrating antegrade K-wire placement via the osteotomy site (A), followed by retrograde advancement of the 2 K-wires to secure the thumb in an extended, pronated, and abducted position (B). Courtesy of the Shriners Hospital for Children, Philadelphia.

until the patient is at least 18 months of age allows the hand to grow to a size that makes for more predictable results.

TECHNIQUE

Creation of the index rotational flap is addressed first during the thumb reconstruction. Under tourniquet, this flap is developed as previously described with sharp and blunt dissection to create a deepened thenar webspace.¹³ We have

found that having the ulnar apex of the incision more distal than the radial apex allows for a better contour along the thumb (Fig. 3). Careful dissection and identification of the dorsal sensory branch of the index finger, the radial index neurovascular bundle proper, and the princeps pollicis artery decrease the risk of iatrogenic injury. Although commonly hypoplastic, the first dorsal interosseous muscle and the adductor pollicis are visualized and can often be sufficiently lengthened by releasing the overlying fascia and fibrous bands

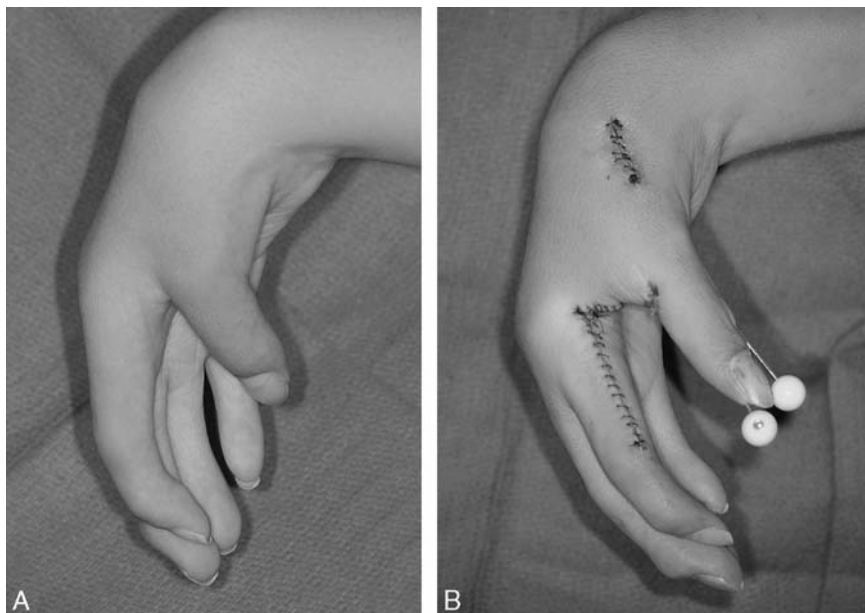


FIGURE 7. Preoperative (A) and postoperative (B) photographs demonstrating the change in thumb position, with a widened first webspace, and fixation of thumb into a more functional position for grasp and pinch. Courtesy of the Shriners Hospital for Children, Philadelphia.

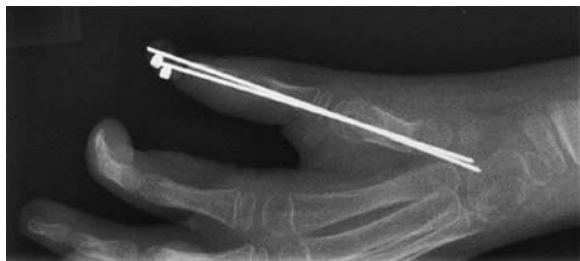


FIGURE 8. Six-week postoperative radiograph demonstrating a healed osteotomy site. Courtesy of the Shriners Hospital for Children, Philadelphia.

(Fig. 4). Occasionally, there is no muscle present, and the fibrous tissue must be completely divided to allow abduction.

A separate dorsal incision is made directly overlying the base of the thumb metacarpal (Fig. 3). Dissection is carried down to the thumb extensor tendons, where a longitudinal periosteal incision is made between the extensor pollicis longus and extensor pollicis brevis tendons just distal to the physis (Fig. 5). A subperiosteal dissection is used to expose the bone and Holman retractors are used to protect the soft tissues. A metacarpal base extension and pronation rotational osteotomy is performed with a sagittal saw (Fig. 5). A first cut is made perpendicular to the long axis of the metacarpal, but is left incomplete. The second cut is made in the distal fragment to remove a wedge from the dorsal aspect of the metacarpal. The first cut is then completed and the surfaces of the osteotomy are reduced to evaluate the amount of correction achieved. The thumb is usually rotated into the maximal allowable pronation, as pronation of the thumb of between 90 and 120 degrees is difficult to achieve in these patients due to soft-tissue constraints. If adequate pronation cannot be obtained, then the osteotomy can be shortened further to loosen the soft-tissue envelope. The osteotomy should be modified before definitive fixation to place the thumb in the best position for both grasp and tip pinch. Often, this position puts the hand in a “hand puppet” posture. Key pinch may be possible but is difficult to achieve alongside grasp in patients with minimal function. Be aware of finger motion and optimize the thumb position to oppose to the position of the fingers. Every child is different and no standard position exists.

The osteotomy is fixed with 2 Kirschner wires (K-wires) introduced in an antegrade and then retrograde manner (Fig. 6). The K-wires are left out of the skin for later removal in the office. With an open first webspace and the thumb now positioned optimally for pinch, the index rotation flap is closed primarily with a gut suture (Fig. 7). A sterile dressing and long arm cast are placed over the surgical area with the wrist in maximal extension and radial deviation to provide passive

stretch of the contraction. A short arm cast can be used in older children who are unlikely to remove their own cast.

Postoperative care includes cast removal at 4 to 6 weeks with x-ray evaluation. If adequate healing at the osteotomy site is observed (Fig. 8), the K-wires are removed and the patient is fitted for an orthoplast thumb splint. The splint is worn full-time for 4 weeks except for range of motion exercises 3 times a day and bathing. After 4 weeks, night-time splint use is recommended for a further 4 weeks or longer as needed. Occupational and physical therapy is initiated after the K-wires are removed to improve hand function, with resistive exercises delayed until 3 months after surgery.

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