 Failure of Differentiation: Part II (Arthrogryposis, Camptodactyly, Clinodactyly, Madelung Deformity, Trigger Finger, and Trigger Thumb)

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ARTHROGRYPOSIS

The term “arthrogryposis” has traditionally been used to describe various conditions that present with congenital joint contractures.\textsuperscript{1} Under this broad term, more than 150 separate disease entities that share elements of congenital contractures have been described.\textsuperscript{2} The contractures are considered to be the end result of decreased intrauterine movement by the fetus after a period of normal development. This decreased movement may occur because of neuropathies, myopathies, abnormal connective tissue, or decreased intrauterine space.\textsuperscript{2}

Classic arthrogryposis or arthrogryposis multiplex congenita describes a specific disease process. This term was first used by Stern\textsuperscript{3} to describe three children in 1923. To add to the terminological confusion, Sheldon\textsuperscript{4} gave a detailed clinical description of this same entity in 1932 and called it amyoplasia or amyoplasia congenita, emphasizing his belief that the disease was the result of aplasia or hypoplasia of certain muscle groups. This article focuses on the cause and upper extremity treatment considerations of this disorder, which is referred to as amyoplasia.

Distal arthrogryposis refers to a large subgroup of disorders in which the contractures primarily involve the hands and feet. Multiple classification schemes have been used to describe these conditions, but this group is extremely heterogenous and this term does not refer to a specific disease entity.\textsuperscript{5–7}

Children who have amyoplasia present with a characteristic phenotype at birth. Most (84%) present with involvement of all four limbs, although
11% present with only lower limb involvement and 5% with only upper limb involvement. The limbs appear fusiform and cylindric with a pronounced decrease in muscle mass and a lack of flexion creases. There is decreased joint range of motion with a firm inelastic endpoint. Sensation is normal and there are few associated visceral anomalies. The joint contractures are usually bilaterally symmetric. The shoulders are internally rotated and adducted, the elbows are extended, the wrists are flexed and ulnarily deviated, the thumb is adducted, and the fingers are partially flexed. The hips are often flexed and abducted, and one or both hips are dislocated in one third of patients. Knee flexion contractures are the most common presentation, but the child may present with knee extension contractures, and knee joint subluxation or dislocation may be present. Almost all children who have this condition have rigid clubfeet, although congenital vertical talus or another foot deformity may be present. Congenital scoliosis is unusual and should trigger the work-up of other neuromuscular disorders. Neuromuscular scoliosis may develop in 30% of patients, however. Some 90% of patients who have all four limbs involved have a characteristic frontal midline capillary hemangioma.

Amyoplasia is sporadic, with an incidence of 1 of every 10,000 live births. Fifteen identical twin cohorts have been reported in the literature as being discordant for the disease. This finding strongly suggests that genetics do not play a major role in the disease, and that some factors in twins may contribute to the condition. Several authors have primarily attributed amyoplasia to be the end result of damage to the anterior horn cells of the spinal cord. Others believe that a primary myogenic form may exist. Multiple studies have not found a clear environmental cause.

Several authors have commented that children who have amyoplasia are often of above-average intelligence and have the potential to become independent adults despite significant physical limitations. Bevan and colleagues suggest that to help a child who has amyoplasia reach this goal, treatment should be focused on communication, activities of daily living, mobilization, and ambulation, in decreasing order of importance. With this in mind, treatment of the upper extremities must focus on preserving or increasing motion and allowing the upper extremities to be positioned at the tabletop level for self-care and access to computers. Because children who have amyoplasia are extremely limited with regard to upper extremity strength, they depend on bimanual patterns and this must be preserved. Van Heest and colleagues reported one patient who had a loss in function after the development of an elbow flexion contracture. This condition, combined with the patient’s pre-existing contralateral elbow extension contracture, led to a decline in the ability to perform activities of daily living. Occupational therapy is often helpful in assisting with learning adaptive skills and providing assistive equipment.

Most frequently, the child who has upper extremity involvement benefits from treatment of elbow and wrist contractures. The typical arthrogrypotic deformity of the elbows is full extension with some triceps function. The lack of passive elbow flexion is particularly disabling because it does not allow hand-to-mouth function. Children who have amyoplasia who have passive elbow flexion but lack active flexion are often able to use strategies, such as trunk swaying or table top propping, to flex the elbow for self-feeding. Tendon transfers to restore active flexion of the elbow are difficult given the relative lack of donor musculature and carry a significant risk for creating an elbow flexion contracture. They seem to provide little additional functional benefit for most children who have amyoplasia.

Initial treatment of the upper extremities in the infant who has amyoplasia begins with gentle manipulation and splinting. This treatment may be particularly helpful with regard to the wrist flexion and elbow extension contractures. Therapy may also assist in maintaining muscle mass and strength in the child who has amyoplasia. Smith and Drennan evaluated 17 infants who had wrist flexion contractures and found that patients who had distal arthrogryposis were more responsive to early casting, serial casting, and orthotics than patients who had amyoplasia.

Timing of upper extremity surgery is controversial. Mennen advocated an early one-stage corrective procedure between 3 and 6 months of age. He recommended proximal row carpectomy to address the wrist flexion contracture and the triceps-to-radius transfer to achieve active elbow flexion. In his report on 47 limbs, he found that patients who were operated on early gained more active motion and had improved carpal bone remodeling. He also believed that the repositioning of the wrist allowed for improved finger function and noted that creases developed over the finger joints after wrist surgery.

Because amyoplasia frequently involves all four extremities, however, some advocate waiting until the child begins to ambulate before beginning upper extremity surgery. This recommendation may result in a significant time delay, given that clubfeet, knee contractures, and hip dislocations often need to be addressed. In the nonambulatory
or ambulate-with-assist child, there were
corns regarding maintaining elbow extension
to allow transfers and crutch or walker use. Van
Heest and colleagues\textsuperscript{19} reported a series of
patients treated with triceps-to-biceps transfers
and did not find that the associated loss of active
extension strength or the development of elbow
flexion contractures affected mobility.

Van Heest recommended elbow capsulotomy
with triceps lengthening at 18 to 24 months of
age if the child does not achieve 90° of passive
elbow flexion following a minimum of 6-month
stretching program.\textsuperscript{19} Ezaki\textsuperscript{17} reports a preference
for beginning upper extremity operative proce-
dures before age 4 because children often have
developed well-adapted use patterns by age 8
that may be disrupted by surgical changes in the
upper extremity positioning. In addition, opera-
tions may be coordinated with lower extremity
procedures to decrease the total number of
operations.

Van Heest and colleagues\textsuperscript{20} reported their expe-
rience with posterior elbow capsulotomy with
triceps lengthening in their series of 29 elbows in
23 children with a mean follow up of 5.4 years.
The arc of motion improved from an average of
32° to 66°. All children were able to get their hands
to their mouths using passive assistance tech-
niques, such as cross-arm, table-push, and
trunk-sway. A total of 22 of the 23 children were
able to feed independently; the remaining child
was limited by poor hand function. Preoperatively,
only 2 children were able to demonstrate measur-
able biceps function. At final follow-up, 7 children
had some biceps function, suggesting that allow-
ing passive elbow motion may have unmasked
biceps function in some patients.

Restoring active elbow flexion in children who
have amyoplasia is difficult, and as Van Heest
and colleagues\textsuperscript{20} have shown, possibly not neces-
sary. Unless the child has good passive elbow
flexion and a strong muscle available for transfer,
which are unusual conditions in these patients,
the results of elbow flexion transfers are disap-
pointing and if unopposed, tend to cause severe
flexion contractures.

Van Heest and colleagues\textsuperscript{19} also reported their expe-
rience with tendon transfers with and without
posterior elbow releases to restore active elbow
flexion. Although they believed triceps-to-biceps
tendon transfer gave the most predictable results,
patients developed flexion contractures, with an
average flexion contracture of 38°. Five patients
underwent a bipolar pectoralis major and four
underwent a latissimus dorsi transfer. Although
both operations have the theoretic advantage of
adding muscle to the arm, both donor muscles
are difficult to test preoperatively, require exten-
sive surgical dissection, and yield disappointing
results. Van Heest now recommends against
active elbow flexion transfers in this population,
because children are able to passively flex the
elbow against their leg or a table top to perform
activities of daily living.\textsuperscript{20}

Steindler flexorplasty has been considered con-
traindicated by many authors because of a theoret-
ic risk for worsening the typical wrist palmar
flexion and pronation deformity by moving the
flexor pronator origin proximally. Goldfarb and
colleagues\textsuperscript{24} reported promising results in 17
elbows in 10 children treated with Steindler flexor-
plasty, however. The flexor pronator origin was
isolated and transferred to the anterior humerus
approximately 2 to 3 cm proximal to the medial ep-
icondyle with the elbow flexed at 80°. Postoper-
avely, all elbows had active flexion against
gravity to 85°. They did note that patients devel-
oped a mean elbow flexion contracture of 28°
and that the total arc of pronation and supination
decreased from a mean of 88° to 41°. They did
not find an increase in wrist flexion or finger flexion
posture after surgery, however.

The arthrogrypotic flexed and ulnar deviated
wrist often benefits from repositioning in a more
functional neutral position. Proximal row
carpectomy,\textsuperscript{23,25} dorsal radial wedge resection
osteotomy,\textsuperscript{26} and arthrodesis\textsuperscript{27} at maturity have
all been advocated. All of these present difficulties,
however. Proximal row carpectomy may not
provide adequate wrist extension and wrist motion
may not be maintained. Dorsal radial wedge
resection is frequently not at the point of maximum
deformity and may jeopardize growth at the distal
radius along with radiocarpal motion. Arthrodesis
at maturity eliminates all wrist motion.

Ezaki and Carter\textsuperscript{17,28} have described a tech-
nique of dorsal carpal wedge osteotomy that
reduces the deformity while retaining motion at
the radiocarpal joint (Fig. 1). A volar approach is
first performed to release the tight forearm fascia
and the wrist flexor tendons are released or length-
ened. From a separate dorsal incision, the wrist
extensors are isolated, and a biplanar dorsal radial
wedge of carpus is resected taking care to protect
the radiocarpal articulation. The osteotomy is
closed and the extensor carpi ulnaris is transferred
to the extensor carpi radii. In 42 wrists in 24
patients with an average follow-up of 18 months,\textsuperscript{28}
the resting position of the wrist improved from 59°
of flexion to 11° with a more centered arc of
motion.

Although the shoulder has limited motion and
often has an internal rotation contracture, this is
usually not functionally limiting. Patients who
have extreme internal rotation may benefit from an external rotation humeral osteotomy to place them in a resting position of 45° of internal rotation to allow midline function. The thumb-in-palm deformity may require a thenar release in which the origins of the abductor pollicis brevis, the opponens pollicis, flexor pollicis brevis, and both heads of the adductor pollicis are released. Skin deficiency is usually palmar, at the level of the thumb metacarpophalangeal joint; this benefits more from a rotation flap from the radial side of the index finger than from a first web z-plasty.

Fig. 1. Dorsal carpal wedge osteotomy for wrist flexion contracture in child who has amyoplasia. (A) Preoperative clinical photograph. (B) Intraoperative radiograph with wrist in maximum passive extension. (C) Transverse incision over dorsal carpal row. Extensor carpi radialis brevis (in clamp) sectioned proximally. Extensor carpi ulnaris (in vessel loop) prepared for later transfer. (D) Proximal cut made at the level of the capsular attachment of the proximal carpal row perpendicular to long axis of the forearm. Distal cut through the distal carpal row perpendicular to the long axis of the metacarpals. (E) Alignment after closing wedge osteotomy.
Finger motion usually does not benefit from direct surgical treatment, although some have reported improved function after wrist repositioning.23

CAMPTODACTYLY
Camptodactyly is a nontraumatic, often progressive flexion contracture of the proximal interphalangeal joint (PIPJ) that usually affects the little finger. Since Tamplin’s original description in 1846,29 there has been much debate regarding the definition, etiology, and treatment of camptodactyly. The true incidence of camptodactyly is unclear, especially when mild cases may go unreported, but it is estimated to affect less than 1% of the population.30

Benson and colleagues31 combined elements of earlier classification systems and described camptodactyly as falling into three main categories. Type I, or classic camptodactyly, presents in infancy. Frequently, the disorder is confined to one or both little fingers. The ring and long fingers may also be involved, however. Males and females seem to be affected equally.32 This type of camptodactyly is usually sporadic, but it may be inherited as an autosomal dominant trait with incomplete penetrance.29

Type II shares similar clinical characteristics to type I, but presents in adolescence. Females seem to be more often affected than males (Fig. 2). The deformity tends to worsen with the adolescent growth spurt.29 Because infancy and adolescence are normal times of rapid growth, this may account for the bimodal distribution.

Type III is a more severe form of camptodactyly (Fig. 3). In this variant, severe flexion contractions are seen usually involving multiple digits of both hands.33 This form of camptodactyly is associated with other congenital anomalies. Multiple chromosomal disorders, craniofacial diseases, and skeletal deformities have been associated with camptodactyly. The occurrence of type III camptodactyly follows the inheritance pattern of the specific disorder with which it is associated. There is no consensus regarding the causative etiology of camptodactyly, and essentially all structures about the PIPJ have been implicated in the disorder.34 Many anatomic differences have been noted in digits with camptodactyly, and it is difficult to determine which are causative factors and which are secondary changes as a result of the development of the contracture.

Multiple authors have attributed the deformity to a generalized imbalance between the flexor and the extensor tendons leading to secondary contractures.34,35 McFarlane36 argues that camptodactyly is an intrinsic minus deformity condition caused specifically by an abnormal lumbrical insertion (most frequently into the MPJ capsule). Others have noted frequent abnormalities with regard to the flexor digitorum superficialis (FDS) length or an anomalous origin of the FDS from the palmar fascia or transverse ligament.36,37 Still others have argued that the causes are tightness of the skin and connective tissues,38 malposition of the extensor lateral bands,39 or contractures of the joint itself.40 With the development of a flexion contracture, abnormal tension on the extensor mechanism may further contribute to the deformity.41 In addition to soft tissue anomalies, camptodactyly may be associated with changes in bony anatomy with flattening of the proximal
phalanx head and the base of the middle phalanx, and an indentation along the proximal phalanx neck. These are generally regarded to be secondary changes, however. Smith and Grobbelaar\(^41\) took a more global approach and argued that all of the described anatomic factors occur to various degrees in all patients and that in particular, the fascial structures, lumbricals, and FDS tendon are nearly always involved.

The effectiveness of treatment of camptodactyly is difficult to assess from the literature. Some series include different types of camptodactyly, and criteria for success vary among studies. Authors who have identified a specific causative factor have tended to argue for surgical treatment to address the particular anatomic issue. Others who emphasize a generalized imbalance as causative have recommended splinting with good success. Many note that full flexion, especially of the ulnar digits, is more important than extension and have cautioned against aggressive treatment to address the lack of full extension at the risk of compromising the ability to fully flex the digits. All have noted that surgical treatment is associated with decreased motion of the PIPJ.

Some have argued for conservative management with splinting regimens in nearly all cases.\(^{31,42}\) Other have promoted more aggressive operative management,\(^{37,43}\) particularly citing difficulties with compliance with splinting regimens in the adolescent age group.

Smith and Kaplan recommended an FDS tenotomy.\(^{29}\) They reported a decrease in the flexion deformity of at least 33% in 12 fingers that were treated in this manner. McFarlane and colleagues\(^{43}\) believed that it was critical to address the anomalous insertion of the lumbrical and the resultant intrinsic minus hand position by releasing the lumbrical and performing an FDS tendon transfer to the extensor mechanism. In their series of 53 patients with 1 year follow-up, they found that the joint contracture was reduced from a mean of 49° to a mean of 25°. They did note that patients had a slow return of flexion, however, and that only 33% of patients regained full flexion at 1 year with an average residual lag of 1.8 cm from the distal palmar crease. Gupta and Burke\(^{34}\) recommended extensor indicis proprius transfer to the radial extensor expansion to improve intrinsic function.

Smith and Grobbelaar\(^41\) reported on their experience on 18 fingers in 16 patients. They released tight soft tissues and abnormal lumbrical insertions, lengthened the FDS, and splinted the patients in extension for 4 weeks to address the lax extensor mechanism. They reported 15 patients had good to excellent results. Three patients who had preoperative bony changes of the proximal phalanx did poorly. Siegert and colleagues\(^{44}\) reported their results with both nonoperative (38 fingers in 21 patients) and operative (41 fingers in 14 patients) treatment. Their operation was individualized, but most patients underwent a release of the FDS. Additional soft tissue procedures were performed in a smaller percentage of patients. In their operative group, only 7 fingers were rated as good, and there were no excellent results. They noted an average improvement in the flexion contracture of only 10°, and 10 patients were noted to have lost considerable flexion. Of their nonoperatively treated patients, 12 patients had improvements in extension and only two had loss of flexion.

Hori and colleagues\(^{42}\) reported on 34 fingers in 24 patients. They recommended wearing a dynamic Capener-type splint for 24 hours a day for several months, followed by maintenance splinting of 8 hours per day. They reported that 20 fingers showed almost full correction of the flexion contracture, 9 fingers were improved, 3 fingers were unchanged, and 2 fingers worsened. They did note recurrence if the splinting was discontinued and recommended continuing splinting indefinitely until full growth. Benson and colleagues\(^{31}\) reported results on 59 fingers in 22 patients with a mean follow-up of 33 months. This group was divided into 13 patients who had type I deformities, 4 patients who had type II deformities, and 5 patients who had type III deformities. Patients were treated with a nighttime progressive extension splinting program. They found splinting to be particularly effective in type I camptodactyly in which patients improved from an average flexion contracture of 23° to 4°, and 18 of the 24 PIPJs achieved full passive extension. Two of the type II patients requested operative intervention because they were unwilling to participate in splinting and 1 patient was noncompliant with the splinting protocol. The 1 compliant patient had full correction of a 35° flexion contracture. In the type III group, there were variable results. A total of 12 fingers with a flexion contracture of greater than or equal to 15° improved to almost full extension, and 2 patients underwent surgical correction for severe contractures.

**CLINODACTYLY**

Clinodactyly refers to radioulnar deviation of the finger. Minor angulation, especially of the little finger, is so common that it is considered a normal variant. Pathologic clinodactyly is variously described as greater than 10°\(^45\) or 15°\(^46\) of angulation (Fig. 4A).
Clinodactyly of the little finger is inherited in an autosomal dominant fashion and usually presents bilaterally. Estimates regarding the incidence of clinodactyly range from 1% to 20%. The next most common sites of occurrence are the thumb and ring finger, with involvement of the middle and index fingers being relatively uncommon. Clinodactyly may also be associated with other hand anomalies and syndromic conditions.

The deformity is usually caused by an abnormally shaped middle phalanx, which may be triangular or trapezoidal because of a C-shaped physis extending from the normal proximal physis along the shortened side of the phalanx and connecting
to an often aberrantly persistent distal physis. Light and Ogden have further characterized the longitudinal epiphyseal bracket responsible for this and other angular deformities. Early complete ossification of the bracket leads to a very short triangular “delta” phalanx and angular deformity (Fig. 4B), whereas an incomplete or cartilaginous bracket allows some longitudinal growth causing a trapezoidal phalanx and the resultant digit angulation (Fig. 4C).

Most cases of clinodactyly, especially of the little finger, do not require treatment because there is little functional impairment and surgical treatment carries a risk for scarring and stiffness. Splinting and therapy have not been advocated because of lack of efficacy.

Surgical correction has been recommended for severe clinodactyly. Operative treatment consists of either segmental resection of the longitudinal epiphyseal bracket to allow for correction with continued longitudinal growth or an osteotomy to directly realign the digit.

Vickers described physiolysis or resection of the mid-zone of the longitudinal epiphyseal bracket with a fat interposition graft in 6 patients. Postoperative splinting was not required, and he found improvement in length and angulation with growth. Caouette-Laberge and colleagues described their experience with 35 fingers in 23 children and found a mean angular correction of 18° in children who had surgery before 6 years of age. Fingers with more severe preoperative angulation (greater than 40°) were observed to have more significant angular correction (mean 20°) compared with fingers with less than 40° of preoperative angulation (mean 7.5°).

Closing wedge, opening wedge, and combined osteotomies have all been advocated for surgical correction. Closing wedge osteotomies have the advantage of relative technical simplicity, but carry the risk for further shortening an already shortened digit (Fig. 4D). Light and Ogden pointed out that a reverse osteotomy may allow the formation of a bony bridge that crosses both physes and thus limits further growth. Opening wedge and combined procedures allow for length maintenance, but may require extensive dissection or local soft tissue rearrangement.

MADELUNG DEFORMITY

In 1878, Madelung described the wrist deformity that bears his name at the Congress of German Society for Surgery in Berlin. It had previously been noted by other surgeons, including Dupuytren, in the mid-1800s. The clinical manifestation is of a shortened radius and increased palmar and ulnar tilt of the radial articular surface. The relatively normal ulna is prominent dorsally, and the carpus is volar and ulnarly subluxed. Wrist extension and supination are limited. Madelung deformity seems to be the result of the premature closure of the palmar and ulnar distal radial physis. Rarely, a reverse Madelung pattern presents, associated with dorsal-ulnar distal radial physeal arrest with dorsal hand displacement and a volarily prominent ulna. There is currently much discussion regarding the genetics, radiographic assessment, and treatment of Madelung deformity. There is some debate as to whether Madelung deformity represents an isolated disease process. Madelung deformity has been associated with repetitive loading of the wrist leading to a partial physeal arrest.

The relationship of Madelung deformity to Leri-Weill dyschondrosteosis is also debated. Leri and Weill first described the dyschondrosteosis that bears their name in an article in 1929. Leri-Weill dyschondrosteosis is a syndrome that includes short stature, Madelung deformity, and mesomelia or shortening of the middle segment of the extremities. Several authors have argued that Madelung deformity always presents as part of Leri-Weill dyschondrosteosis. Other studies have concluded that patients who have isolated Madelung deformity may represent a discrete heritable condition. Several authors have reported that 17 of 22 of their cases of Madelung deformity fit the description of Leri-Weill dyschondrosteosis and argued that most but not all patients who have Madelung deformity have Leri-Weill dyschondrosteosis. They provided evidence that Madelung deformity presents as a spectrum that may affect only the distal radius, or the entire radius may be bowed and shortened. Madelung deformity is four times more common in females and most patients who have Madelung deformity present with bilateral changes.

Belin and colleagues and Shears and colleagues independently identified loss of function mutations in the short homeobox containing gene (SHOX) resulting in haploinsufficiency to be causative of Leri-Weill dyschondrosteosis. SHOX is located on the pseudoautosomal region of the X or Y chromosome, and has also been implicated in causing the short stature seen in Turner syndrome. Mutations in both SHOX genes seems to result in Langer mesomelic dysplasia. Estrogen may play a role in the development of Madelung deformity because this disease frequently presents in females at the time of adolescence. Madelung deformity is relatively uncommon, occurring only in 7% of patients who have Turner syndrome, a condition in which there
is also SHOX haploinsufficiency and short stature. Although some have postulated that the relative decrease in sex hormones seen in Turner syndrome may be protective, the loss of other modifying growth genes on the X chromosome may also play a role. Genetic testing can be performed on patients to evaluate the SHOX gene. The SHOX gene and its developmental manifestations are continuing topics of ongoing research with many clinical implications.

Munns and colleagues have found disorganization of the chondrocytes and abnormal endochondral calcification in the involved physis. SHOX has also been found to be expressed in the hypertrophic chondrocytes of the growth plate and is believed to modulate cellular proliferation and viability.

Vickers noted the presence of an abnormal ligament between the anterior ulnar metaphysis of the distal radius and the carpus. This condensation of soft tissues begins in the volar metaphyseal region of the radius and attaches to the lunate and triangular fibrocartilage complex. There is sometimes an associated bone spur and fossa at the origin. This ligamentous structure has been noted histologically to have areas of fibrous tissue and fibrocartilage. Murphy and colleagues noted abnormalities in the pronator quadratus insertion. Others believe that these structures may merely represent a soft tissue compensatory response because of the need to support the carpus as a result of the abnormal growth of the radius and progressive loss of lunate facet support (H.R. McCarroll, MD, Sacramento CA, personal communication, August 2008). It is currently unclear if Vickers ligament contributes to the Madelung deformity or whether it is simply a secondary response of the soft tissues.

Imaging of the Madelung deformity can be difficult given that it is a complex three-dimensional deformity. The distal radius articular surface is flexed and pronated. Two-dimensional radiographs merely approximate the disease and must be taken in a uniform fashion to be helpful. Carter and Ezaki describe a standardized fashion of obtaining a true posteroanterior (PA) view with neutral forearm rotation in which the patient kneels with the shoulder abducted and the elbow flexed 90°. They recommend having bilateral images on the same plate for ease of comparison.

Dannenberg and colleagues described 12 radiographic criteria for the diagnosis of Madelung deformity. Several other researchers have attempted to define measurements based on the distal radius but did not present reliability or reproducibility data for these measurements. Given the significant changes in the radius in severe Madelung deformity, the usual bony landmarks are difficult to identify and the bowing of the distal radius makes the longitudinal axis of the radius difficult to determine. This difficulty limits the reproducibility of radially based measurements.

McCarroll and colleagues identified four reliable and reproducible measurements for quantifying the severity of Madelung deformity on radiographs (Fig. 5). Tuder and colleagues further evaluated these criteria and found significant correlations between the measurements. McCarroll and colleagues have recently presented data comparing these measurements in normal wrists and wrists with Madelung deformity. They found that an ulnar tilt of greater than 28°, lunate subsistence of greater than 3.0 mm, and palmar carpal displacement of greater than 19.0 mm only occurred in wrists with Madelung deformity. In addition, they found the lunate fossa angle measurement to have the least overlap with normal wrists, which may represent an early radiographic finding of Madelung deformity.

Treatment of Madelung deformity is controversial. As McCarroll notes, there has been little documentation in studies of treatment of Madelung deformity that the severity of pain, altered range of motion, or visual deformity correlates to the degree of bony deformity. The natural history of the disorder is not clearly understood, and it is not clear if the deformity is always associated with pain or functional limitation. Some argue that the most common reason for surgery is appearance, because most patients dislike the radial bowing and dorsal ulnar prominence. Many adolescents and adults who have Madelung deformity complain of ulnar wrist pain, which may represent a degree of ulnocarpal impingement.

Because the natural history is not clearly understood, it is difficult to recommend a course of treatment in the younger, asymptomatic patient. There is also debate regarding whether intervention can prevent the development of deformity. Vickers reported a reduction in pain and an improvement in the appearance of the wrist with growth in his series of patients who underwent surgical physiolysis, and in fact titled his report “Surgical prophylaxis (physiolysis) during the late growth period by resection of the dyschondroto-tesis lesion.” In contrast, Carter and Ezaki reported that they had limited success with epiphysiolysis. They attributed this in part to their Madelung deformity patient population generally presenting in late adolescence, a point at which they had limited remaining growth potential of the distal radius.
Fig. 5. (A) Ulnar tilt on a PA radiograph is defined as the complement of the acute angle (angle A) between the longitudinal axis of the ulna and a line tangent to the proximal surfaces of the scaphoid and lunate. (B) Lunate subsidence on a PA radiograph (distance A) is defined as the distance in millimeters between the most proximal point of the lunate and a line perpendicular to the longitudinal axis of the ulna and through its distal articular surface. The measurement is positive if the ulna exceeds distal to the proximal surface of the lunate. (C) Lunate fossa angle on a PA radiograph is defined as the complement of the acute angle (angle A) between the longitudinal axis of the ulna and a line across the lunate fossa of the radius. (D) Palmar carpal displacement on a lateral radiograph (distance A) is defined as the distance in millimeters between the longitudinal axis of the ulna and the most palmar point on the surface of the lunate or capitate. (From McCarroll HR, James MA, Newmeyer WL, et al. Madelung’s deformity: quantitative assessment of x-ray deformity. J Hand Surg [Am] 2005;30(6):1213-4; with permission.)
Carter and Ezaki\textsuperscript{69} have described their technique combining a release of Vickers ligament with a dome osteotomy by way of an anterior approach on 23 wrists in 18 patients (Fig. 6). They found that simple release of Vickers ligament may be helpful in alleviating pain; many patients reported improvement in their symptoms even before their osteotomy had healed. In addition, they found that release of this ligament allowed the carpus to assume a more normal alignment on the radius.\textsuperscript{69,80} The same group later reported on their results on 26 wrists in 18 patients treated with a volar Vickers ligament release and dome osteotomy. Patients reported decreased pain and improved appearance, which was associated with radiographic changes in the mean radial inclination and lunate subsidence. There was persistent ulnocarpal and distal radial ulnar joint deformity, however.\textsuperscript{81}

Many other procedures have been described for the treatment of Madelung deformity. Initially, treatment tended to focus on removal of the prominent ulna. Since Ranawat reported that the carpus tended to sublux ulnarily in wrists treated with an isolated Darrach procedure, however, this has fallen out of favor.\textsuperscript{82} More recently the isolated ulnar wedge osteotomy or ulnar shortening osteotomy has been described for the treatment of mild Madelung deformity.\textsuperscript{79,83}

Dos Reis and colleagues\textsuperscript{73} described a dorsal closing wedge osteotomy of the distal radius combined with ulnar shortening, whereas Murphy and colleagues\textsuperscript{70} and de Paula and colleagues\textsuperscript{84} described a radial opening wedge osteotomy. Proponents have reported the successful correction of Madelung deformity using the Ilizarov technique.\textsuperscript{85} Radial osteotomy and the Sauvé-Kapandji technique have also been described for the treatment of Madelung deformity.\textsuperscript{86,87}

A recent English-language literature search found only two studies published on the long-term results after operatively treated Madelung deformity. Potenza and colleagues\textsuperscript{88} published their results on 9 wrists in 5 patients with a mean follow-up of 34 years after surgery and a mean age at follow-up of 53 years. Eight wrists were treated operatively with a dorsoradially based closing wedge osteotomy, and either a shortening osteotomy of the distal ulna (six wrists) or a distal ulna resection (two wrists). All patients were noted to be pain-free and pleased with the cosmetic appearance of their operated wrists at follow-up. One patient who had a bilateral deformity who had elected to only have surgery on one wrist complained of pain in the unoperated wrist. Salon and colleagues\textsuperscript{72} reported a mean 9.7 year follow-up on 11 wrists in 7 patients who were treated with a similar dorsoradial closing wedge osteotomy and ulnar shortening osteotomy. All wrists were pain-free with daily activities and showed improved cosmetic appearance. Eight wrists showed remodeling of the distal radioulnar joint.

**TRIGGER FINGER**

Pediatric trigger fingers are uncommon and seem to represent a different entity from both pediatric trigger thumbs and adult trigger fingers. A unifying cause has not been determined in pediatric trigger finger. Instead, multiple anatomic anomalies, including an abnormal relationship between the FDS and the flexor digitorum profundus (FDP), an unusual proximal decussation of the FDS, nodules in the FDS or FDP, calcific tendonitis, mucopolysaccharidosis, and constriction of the A2 or A3 pulleys, have been identified in the literature.\textsuperscript{89–92} Unlike trigger thumbs, which usually present with a fixed flexion deformity, trigger fingers are more likely to present with locking and catching symptoms.

In contrast to trigger digits in adults, isolated A1 pulley release is unlikely to successfully treat pediatric trigger fingers, because the triggering frequently does not occur at the A1 pulley (Fig. 7A). Tordai and Engkvist recommended additional procedures, such as widening of the FDS chiasm or partial division of the A2 pulley, for persistent triggering after A1 pulley release.\textsuperscript{83} Caridon and colleagues\textsuperscript{89} recommended an extensile approach to examine and also address other elements, including FDS slip resection.

Bae and colleagues\textsuperscript{90} described their experience with 23 trigger fingers in 18 patients. All patients were treated with a standardized approach in which an A1 pulley release was performed by way of a Bruner-type incision and then a single slip of the FDS tendon was resected (Fig. 7B). This combination of procedures successfully resolved triggering in 91% of fingers. Two patients had recurrent triggering. One improved with a second procedure to resect the remaining FDS tendon slip, and the other was found to have an aberrant FDS muscle attachment that was released at the time of reoperation.

In our recent experience with a 17-month-old patient who had bilateral long finger trigger fingers, we found persistent bilateral intraoperative triggering despite a resection of the ulnar slip of the FDS tendon after an A1 pulley release. It was necessary to open the tendon sheath through a transverse incision just distal to the A2 pulley to complete the resection of the ulnar slip of the FDS tendon. After this was performed, there was
no longer any intraoperative evidence of triggering.

**TRIGGER THUMB**

Congenital trigger thumb is a controversial topic with varying opinions. Bae\(^9^4\) recently performed an excellent evidence-based review of the literature and concluded that the natural history and recommended treatment remain unclear.

First, many consider the term “congenital” to be a misnomer. Although Dinham and Meggitt\(^9^5\) reported 19 of 105 patients had a trigger thumb deformity at birth, this was on the basis of parental recall interviews. Several recent studies with physician examination of thousands of newborns
support the conclusion that trigger thumbs are an acquired condition. In the Rodgers and Waters study of 1046 newborns, the Slakey and Hennrikus study of 4719 newborns, and the Kikucki and Ogino study of 1116 newborns, no cases of congenital trigger thumb were identified.96–98

Unlike adults who present with an intermittent locking or catching of the interphalangeal joint,
children usually present with the characteristic Notta nodule (thickening of the flexor pollicis longus tendon at the base of the metacarpophalangeal joint [MPJ] as originally described in 1850\(^9\) and a fixed flexion deformity of the interphalangeal joint (IPJ) (Fig. 8).\(^{100}\) A smaller percentage may present with inability to actively flex the IPJ, when the thickened nodule is trapped distal to the A1 pulley. In a recent review of the literature, Ogino presents the classification scheme developed by Sugimoto and colleagues (originally published in Japanese).\(^{101,102}\) In stage I, Notta nodule is present, but there is no triggering. In stage II, triggering is observed when the IPJ is actively extended. In stage III, the IPJ cannot be extended actively and triggering is observed when the IPJ is extended passively. In stage IV, the IPJ cannot be passively extended.

Although authors have speculated that thumb sucking or the characteristic thumb-in-fist grasp positioning of the infant incites localized trauma that leads to triggering,\(^{103}\) this is not fully supported by pathologic studies. Electron microscopy examination of tendon nodules and sheaths from children who had trigger thumbs has not found evidence of degenerative or inflammatory changes.\(^{104}\)

Trigger thumb occurrence is usually sporadic. Reports of trigger thumbs occurring in twins,\(^{105}\) the frequent bilateral nature of the condition,\(^{95}\) and the association of trigger thumb with congenital conditions, notably trisomy 13,\(^{100}\) support a possible hereditary component, however. Shim and colleagues\(^{106}\) reported a pedigree in which trigger thumbs were inherited in an autosomal dominant mode with incomplete penetrance. Because of the usual infant thumb-in-fist posturing, the precise development of this disorder can be difficult to determine. Kikuchi and Ogino\(^{98}\) reported the prevalence of acquired trigger thumb at 1 year of age to be 3 per 1000 live births. In a retrospective review of 89 surgical trigger thumb releases performed in a 3-year period, Rodgers and Waters\(^{96}\) found that none of the children presented at less than 3 months of age and only 6 presented at less than 6 months of age.

Like the etiology, the natural history of pediatric trigger thumbs is also unclear. In Dinham and Meggitt’s\(^{95}\) review, 30% of patients presenting soon after birth had a spontaneous recovery and no patient who was less than 3 years of age at the time of surgical release had a loss of IPJ motion. More recently, Baek and colleagues\(^{107}\) reported in their prospective observational study of 71 trigger thumbs in 53 patients that 63% resolved spontaneously at an average age of 5 years; resolution was defined as a lack of IPJ flexion contracture. A subset of these patients had less IPJ hyperextension than their unaffected thumb, however.

Nonoperative management of pediatric trigger thumb may include extension exercises and splinting. Watanbe and colleagues\(^{108}\) studied 58 thumbs in 46 children who were treated by daily passive extension exercises and found that 96% had satisfactory results. Final motion was abnormal in 59% of thumbs, however. Lee and colleagues\(^{109}\) demonstrated improvement in

![Fig. 7. Long trigger finger. (A) A1 pulley has been released, able to demonstrate continued triggering. (B) Ulnar slip of FDS resected (in forceps).](image)
flexion contractures in 71% of 31 thumbs using a protocol of full-time hyperextension splinting for 6 to 12 weeks followed by nighttime splinting.

Surgical release of the A1 pulley has been recommended for children who present with trigger thumb after 1 year of age, who fail conservative management, or who manifest rigid deformity. 95 McAdams and colleagues 110 presented long-term results on 30 thumbs in 21 patients with an average follow-up of 15 years. They had no cases of recurrence and no functional deficits; 23% had slightly diminished IPJ motion and 18% had MPJ hyperextension. Abnormal motion was not associated with age at time of surgery. Children who had a longitudinal incision had some concerns regarding scar appearance, and they thus recommended a transverse skin incision.

Recently, there has been some discussion regarding the anatomy of the thumb pulley system and how it may play a role in the pathophysiology of the trigger thumb. Doyle and Blythe described the anatomy of the thumb in 1977 as consisting of two annular pulleys (A1 and A2) with one oblique pulley. 111 They concluded that the oblique pulley was important for normal flexor pollicis longus tendon function and noted decreasing IPJ motion and increasing MPJ motion as pulleys were sectioned. In biomechanical studies, Zissimos and colleagues 112 and Esplin and colleagues 113 determined that either an intact A1 or oblique pulley was necessary for normal kinematics.

Several researchers have noted additional structures, however. Bayat and colleagues 114 noted a distinct annular pulley between A1 and the oblique pulley, which they called the variable annular pulley (Av) and concluded that the oblique pulley did not play a role in the prevention of bowstringing of the flexor pollicis longus. They found that either the A1 or the Av pulley needed to remain intact for normal function. Considerable variation was noted in the anatomy of this pulley, but in its oblique form it was noted to run from proximal ulnar to distal radial.

In contrast to most authors who have recommended division of the A1 pulley 110,115 for the treatment of pediatric trigger thumb, van Loveren 116 determined that this was sufficient to resolve the triggering in only 3 out of 19 thumbs. It was necessary to release an additional structure found distal to the A1 pulley in 69% of cases. In two cases, a completely distinct additional annular pulley structure distal to the A1 pulley was identified and resection of this alone resolved the triggering.
They cautioned that the A1 pulley may not be the main or even causative structure involved in pediatric trigger thumb.

Boretto and colleagues 117 recently performed a prospective clinical study of the A1 pulley in 19 adult trigger thumbs. In 7 thumbs, they found pulleys with a combination of annular and oblique fibers that mimicked those described by Bayat and colleagues 114 as the A1 and the Av pulleys. In contrast to these authors, however, they did not believe that this represented a discrete separate structure. In contrast to van Loveren, sectioning of the standard A1 pulley was sufficient to resolve triggering in the majority (12 of 19) of patients.110,116 This finding may represent differences in the pediatric and adult populations.

It is unclear whether the Av pulley as described by Bayat, the unnamed additional structure and separate annular pulley as described by van Loveren, and the A1 pulley with combined annular and oblique fibers as noted by Boretto represent the same structure.114,116,117 It is possible that variants in pulley anatomy may contribute to pediatric trigger thumb. Variations in the pulley system and differences in which structures are sectioned may account for changes in range of motion after trigger thumb surgery.

REFERENCES


