Madelung Deformity

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Madelung deformity of the wrist is more common in females and is often associated with Leri Weill dyschondrosteosis, a mesomelic form of dwarfism. Patients with Madelung deformity often report wrist deformity resulting from the prominence of the relatively long ulna. The typical Madelung deformity is associated with a Vickers ligament that creates a tether across the volar-ulnar radial physis that restricts growth across this segment. The distal radius deforms in the coronal (increasing radial inclination) and the sagittal (increasing volar tilt) planes. There is lunate subsidence and the proximal carpal row adapts to the deformity by forming an upside-down pyramid shape or triangle. Treatment depends on the age at presentation, degree of deformity, and magnitude of symptoms. Mild asymptomatic deformity warrants a period of nonsurgical management with serial x-ray examinations because the natural history is unpredictable. Many patients never require surgical intervention. Progressive deformity in the young child with considerable growth potential remaining requires release of Vickers ligament and radial physiolysis to prevent ongoing deterioration Concomitant ulnar epiphysiodesis may be necessary. Advanced asymptomatic deformity in older children with an unacceptable-appearing wrist or symptomatic deformity are indications for surgery. A dome osteotomy of the radius allows 3-dimensional correction of the deformity. Positive radiographic and clinical results after dome osteotomy have been reported.

Key words Madelung deformity, Vickers ligament, osteotomy, epiphysiodesis.

In 1878, OTTO MADELUNG[1] DESCRIBED his namesake deformity of the wrist in adolescents aged 8 to 14 years. The deformity was initially asymptomatic but patients frequently developed pain, decreased strength, and reduced mobility. Madelung deformity is more common in females and is often associated with Leri Weill dyschondrosteosis, a mesomelic form of dwarfism. These patients have abnormalities in the short stature homeobox gene that resides in the pseudoautosomal region of the sex chromosomes.[2]

Madelung-like deformities are seen in a variety of other diagnoses including multiple hereditary exostosis, Ollier disease, growth plate arrest, and gymnast wrist. Any condition that limits the growth of the volar-ulnar corner of the distal radius can yield a similar clinical picture. A Vickers ligament is not seen in Madelung-like deformities and serves to distinguish between these 2 entities. In this current concept review, we will focus on Madelung deformity and describe the clinical presentation, radiographic findings, and treatment strategies.

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CLINICAL PRESENTATION

Patients with Madelung deformity most often report gross disfigurement at the wrist caused by the prominence of the relatively long ulna and are often unaware of the deformed radius or the volar subluxation of the hand/carpus. Patients with bilateral deformities often present with more advanced deformities because there is no normal side for comparison. In children with short stature and bilateral wrist deformity, Leri Weill dyschondrosteosis should be suspected. The hand surgeon needs to broach this topic carefully because the family is often unaware of the genetic implications. A genetic consultation is warranted to confirm the diagnosis and to discuss the concepts of variable

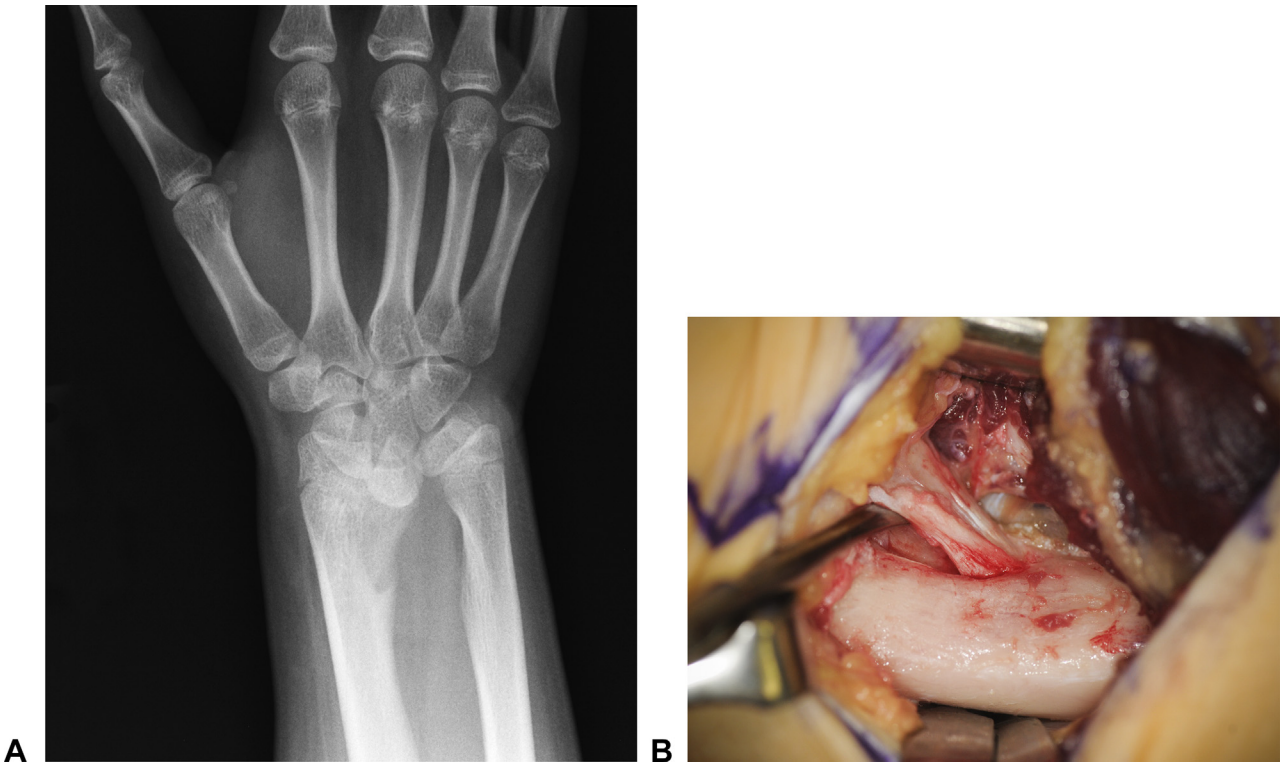

FIGURE 1: A 17-year-old girl with Madelung deformity and prominent ulna. (Courtesy of Shriners Hospital for Children, Philadelphia, PA.) **A** X-ray with increased radial inclination and ulnar positive variance. Note radiolucency in the metaphysis of the radius, consistent with the presence of a Vickers ligament. **B** Operative findings with a markedly thickened Vickers ligament.

phenotype and variable penetrance. The family should be discouraged from an immediate Internet search that will result in a compendium of misinformation.

In some cases, the presenting problem can be limited range of motion or pain. Loss of wrist extension is more common and is proportional to the sagittal deformity of the radius. Despite the prominent ulna, restriction of forearm rotation is uncommon. Pain can occur at the distal ulna or the radiocarpal joint. Paradoxically, distal ulnar pain from ulnar abutment is more common in milder deformities before the ulna escapes the deforming sigmoid notch. Radiocarpal joint pain often occurs with forced extension, such as during upper extremity weightbearing.

ETIOLOGY

The precise etiology remains speculative and elusive. The typical Madelung deformity is associated with a Vickers ligament, which can be considered an abnormal short volar radioulnar ligament.[3,4] Instead of originating from the epiphysis, the ligament originates from the metaphysis of the distal radius, creating a tether across the volar-ulnar physis that restricts growth across this segment. As the remaining physis grows, the radius deforms with increasing radial inclination and increasing volar tilt. The unaffected ulna continues to elongate, yielding a progressive ulnar positive variance. Ultimately, the distal radioulnar joint fails to form normally and the distal ulna subluxates or dislocates in a dorsal direction.

RADIOGRAPHIC FINDINGS

Plain x-rays are the mainstay of diagnosis because the findings are pathognomonic (Fig. 1). The origin of Vickers ligament creates a radiolucent flame-shaped notch at the medial radial metaphysis. The x-ray findings vary with the amount of deformity. Milder deformities can be missed as the coronal lunate fossa begins to settle and the sagittal distal radius begins to flex. Moderate and severe deformities demonstrate classic findings.[5] The distal radius deforms in the coronal (increasing radial inclination) and the sagittal (increasing volar tilt) planes. There is lunate subsidence and the proximal carpal row adapts to the deformity, forming an upside-down pyramid shape or triangle.[5] The wrist and hand translate in a volar direction following the curvature of the distal radius. Severe deformities obscure visualization of the lunate fossa and exhibit frank dislocation of the distal ulna. Patients who maintain a reduced distal radioulnar

joint despite restricted radial growth will distract the radial head away from the capitellum (Fig. 2).

Children with Leri Weill dyschondrosteosis can present with extreme and bizarre deformities (Fig. 3). The entire radius can be bowed with distortion of the radiocarpal joint and frank dislocation of the distal radioulnar joint.[6] This cohort of patients has greater restrictions in forearm movement and more pronounced aesthetic issues with a short and curved forearm. Despite the impressive clinical and x-ray findings, most of these patients do not have pain.

Advanced imaging studies are unnecessary for diagnosis; however, a magnetic resonance imaging is frequently obtained before the initial evaluation. This imaging better delineates the 3-dimensional deformity, the extent of growth plate involvement, and the presence of a Vickers ligament (Fig. 4).[7,8] The triangular fibrocartilage complex typically demonstrates a progressively oblique orientation resulting from the radial deformity.

TREATMENT

Treatment depends on the age at presentation, degree of deformity, and magnitude of symptoms. Mild asymptomatic deformity warrants a period of nonsurgical management with serial x-ray examinations because the natural history is unpredictable. Many patients can be carefully evaluated every 6 months until skeletally mature and never require surgical intervention. Progressive deformity in the young child with considerable growth potential remaining requires intervention to prevent ongoing deterioration. Release of Vickers ligament and physiolysis of the radius provides an opportunity for the growth plate to recover.[4] Although physiolysis is not uniformly successful, the operation is relatively minor and straightforward to perform. The principles of guided growth have also been applied to the Madelung deformity. This technique uses the growth of the physis to correct deformity over time. In addition to the release of Vickers ligament and physiolysis, staples can be placed across the unaffected radial growth plate to temporarily curtail its growth. Guided growth is more commonly used in Madelung-type deformities such as multiple hereditary exostoses.

Advanced asymptomatic deformity usually presents near skeletally maturity. As long as the deformity is acceptable, advanced deformity does not require treatment. An unacceptable-appearing advanced asymptomatic deformity is a relative indication for surgery. The patient and family must understand the surgical risks and benefits. The surgical procedure and potential complications should not be minimized with respect to anesthetic risks, intraoperative error, and postoperative problems.

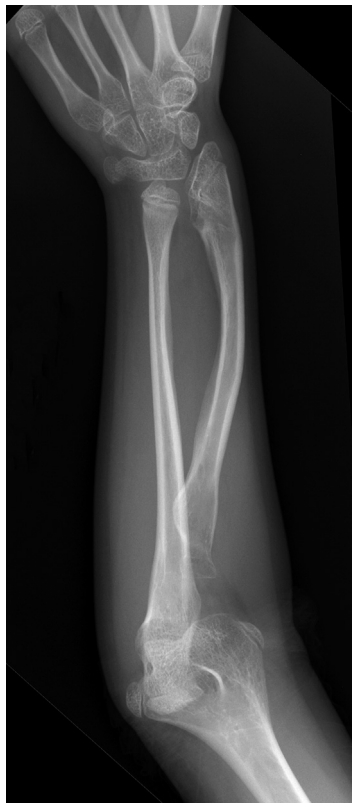

FIGURE 2: A 12-year-old girl with unilateral Madelung deformity, maintenance of a reduced distal radioulnar joint, and distraction of the radial head away from the capitellum. (Courtesy of Shriners Hospitals for Children, Philadelphia, PA.)

Symptomatic deformity requires a careful assessment to determine the precise cause of the pain. Milder deformity yields an ulnar-positive variance and ulnar impaction. Ulnar shortening osteotomy with or without distal ulnar epiphyseodesis can alleviate impaction and pain without having to address the primary radial deformity. Symptomatic deformity is initially treated with nonsurgical measures such as placement of orthosis and activity modification. Recalcitrant symptoms may warrant a radial osteotomy. A dome osteotomy is preferred to allow 3-dimensional correction of the deformity without the need for bone graft.[9,10] For complex deformities, 3-dimensional radiographic modeling may be helpful to plan correction using multiple osteotomies. This technique is still being refined, but it provides a more precise correction than standard osteotomy techniques.[11]

Vickers ligament release

The patient is placed supine on the operating room table. The entire extremity is prepped and draped. The limb is exsanguinated and a tourniquet is used. A

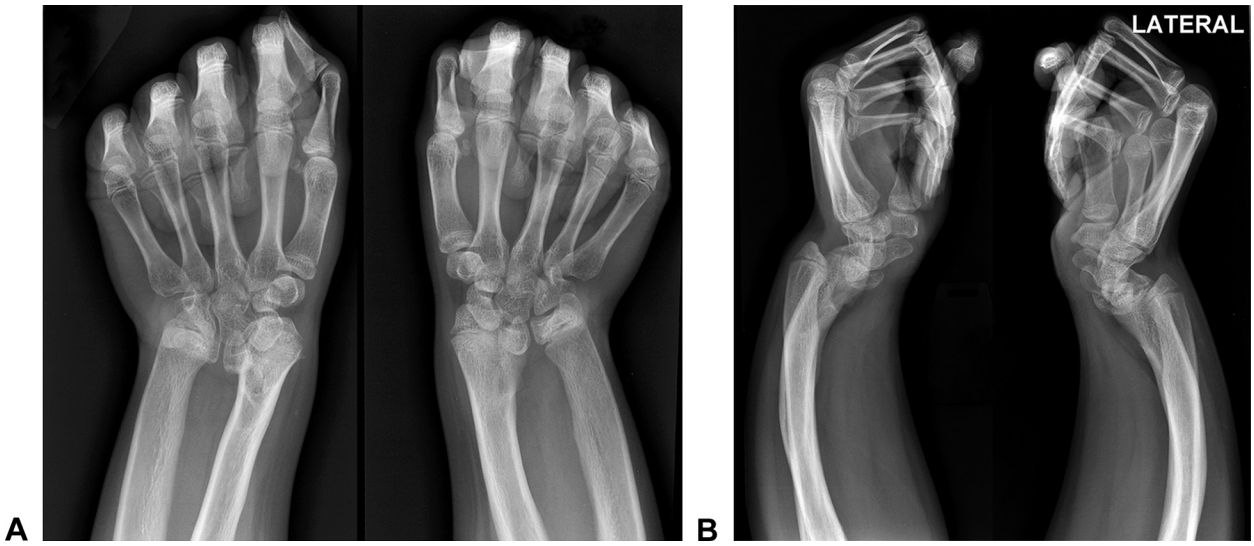

FIGURE 3: A 12-year-old girl with Leri Weill dyschondrosteosis and bilateral Madelung deformity. (Courtesy of Shriners Hospitals for Children, Philadelphia, PA.) **A** Anteroposterior x-rays with severe distortion of the distal radius and carpus. **B** Lateral x-rays with frank dislocation of the distal radioulnar joint.

trans-flexor carpi radialis (FCR) approach is performed. The distal edge of the pronator quadratus muscle is identified and retracted in a proximal direction. With the aid of mini-fluoroscopy, the origin of Vickers ligament is isolated. A scalpel blade is used to cut across the origin of the ligament into the metaphysis of the radius. The ligament is carefully elevated from proximal to distal until the physis is untethered.[4] This is usually apparent as the undersurface of the lunate is visualized. The growth plate is inspected and any bony bar is resected using a burr or rongeur until normal-appearing physeal cartilage (bluish tint) is identified. The resection is filled with autogenous fat that can be harvested from within the surgical field.

The subcutaneous tissue and skin are closed with absorbable suture. A short arm orthosis or cast is applied for 3 to 4 weeks followed by active and active assisted range of motion. Formal therapy is usually unnecessary and routine activities are quickly resumed. Serial radiographs are performed to assess growth and alignment over time.

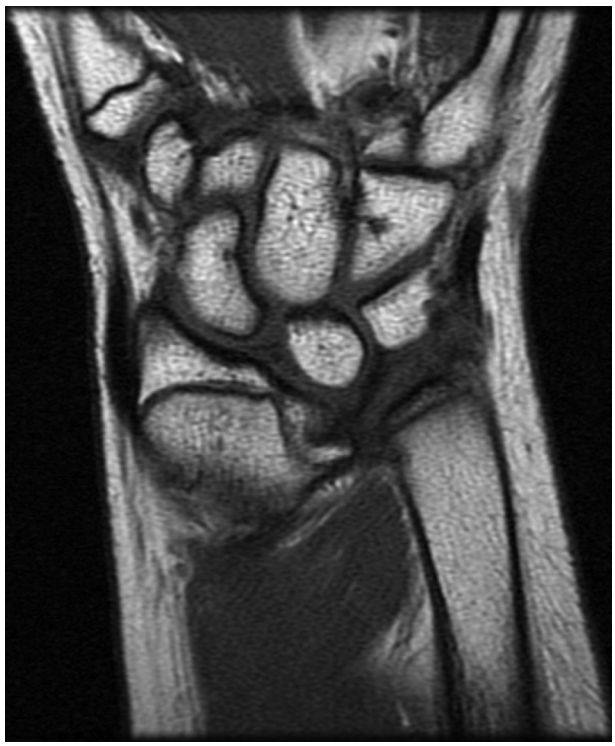

FIGURE 4: Magnetic resonance image showing Vickers ligament originating from the metaphysis of the distal radius, creating a tether and growth disturbance across the volar-ulnar physis. (Courtesy of Shriners Hospitals for Children, Philadelphia, PA.)

Ulnar shortening osteotomy

The patient is placed supine on the operating room table. The entire extremity is prepped and draped. The limb is exsanguinated and a tourniquet is used. An incision is made along the ulnar border of the forearm. The inter-nervous interval between the flexor carpi ulnaris and FCR muscles is used to expose the ulna in an extraperiosteal fashion. We prefer to place the plate along the volar aspect of the ulna. As long as the ulna is of adequate caliber, an ulnar shortening osteotomy system is used to remove the necessary amount of ulna to achieve ulnar-neutral variance. The plate and screw construct and ulnar length are checked with mini-fluoroscopy before closure. The subcutaneous tissue and skin are closed with absorbable suture. A long arm

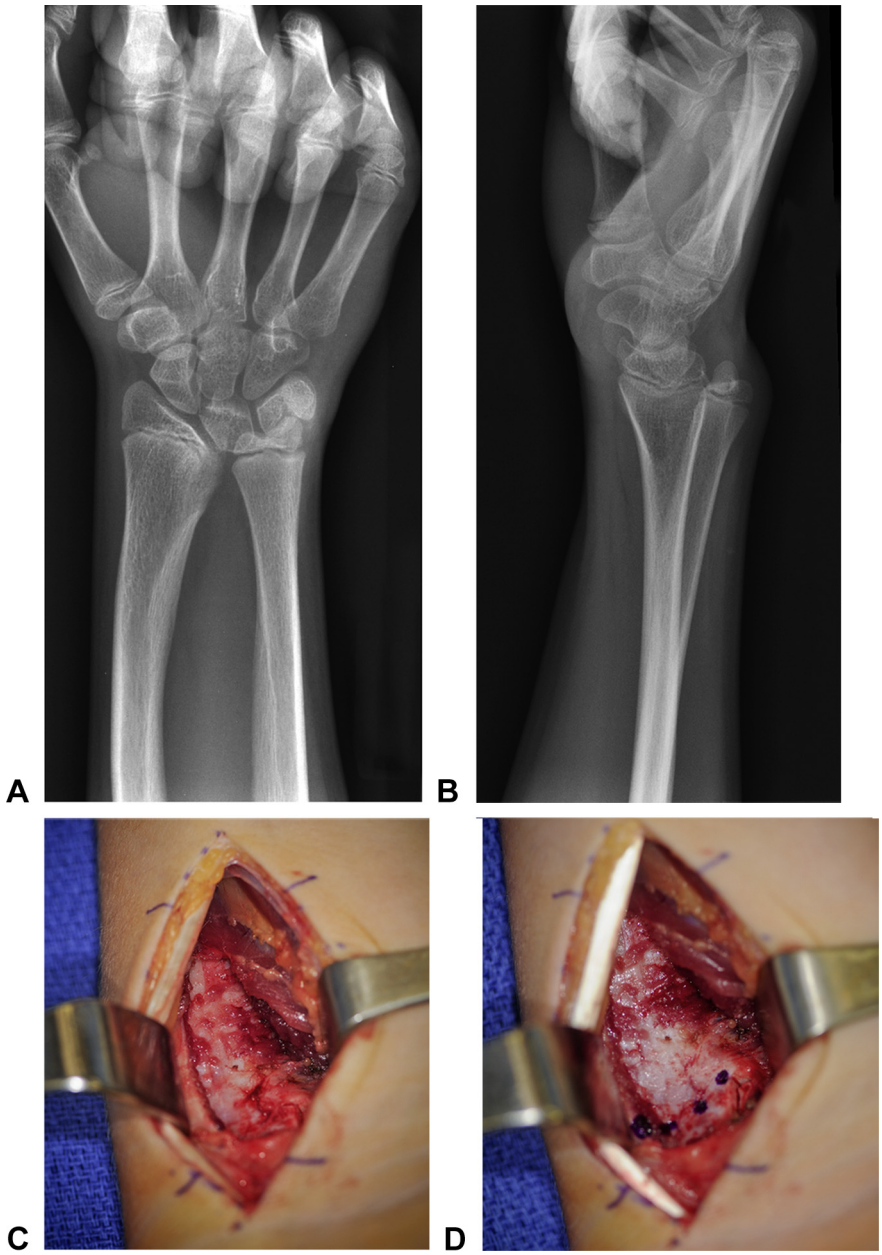

FIGURE 5: An 11-year-old with Madelung deformity. (Courtesy of Shriners Hospitals for Children, Philadelphia, PA.) **A** Anteroposterior x-ray. **B** Lateral x-ray. **C** Exposure of volar radius. **D** Markings with dome configuration. (Continued.)

orthosis is applied for 2 weeks. The orthosis is changed to a well-molded short arm cast that is removed at 6 weeks from surgery. A short arm orthosis is fabricated and active range of motion is started. Formal therapy is often unnecessary. Routine activities are resumed. Weightbearing is avoided until 3 months from surgery.

Radial dome osteotomy

The patient is placed supine on the operating room table (Fig. 5). The entire extremity is prepped and draped. The limb is exsanguinated and a tourniquet is used. A trans-FCR approach is performed. The pronator quadratus muscle is elevated in a radial to ulnar direction to expose the metaphysis of the radius (Fig. 5C). Vickers ligament is released as described above. With the aid of mini-fluoroscopy, the extent of deformity is defined and the osteotomy is planned. The convexity of the dome can be oriented in a proximal (smile) or distal (frown) direction.[9,10] We have found no difference with respect to difficulty or degree of correction. We prefer convexity distal to avoid iatrogenic violation of the radiocarpal and/or distal radioulnar joint (Fig. 5D).

The osteotomy is started with multiple 1.6-mm (0.062-in) Kirschner wire fenestrating holes to define the configuration (Fig. 5E). Curved osteotomes are used to complete the osteotomy through the dorsal

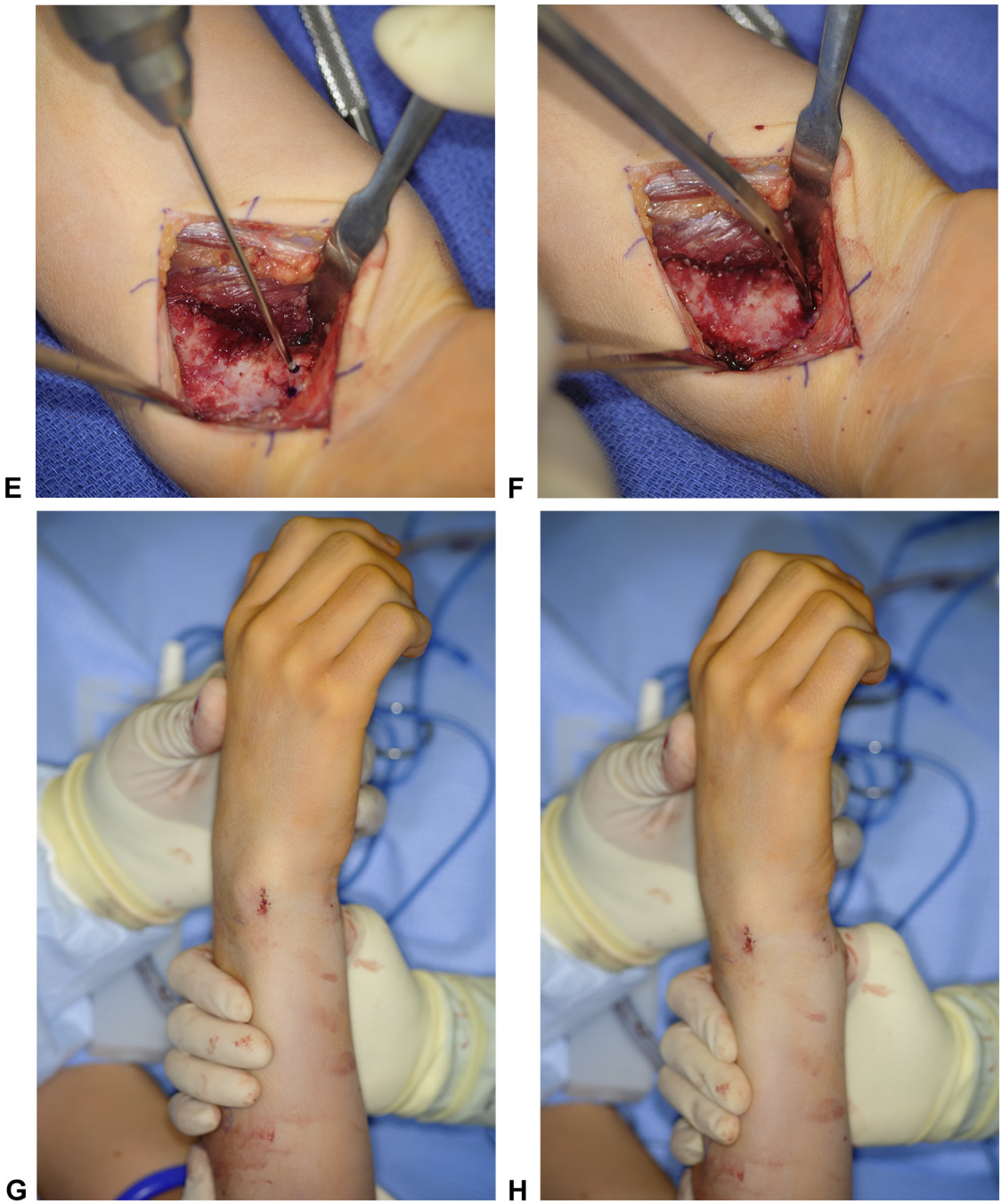

FIGURE 5: (Continued.) **E** Kirschner wire perforation. **F** Osteotome at the Kirschner wire perforations. **G** Before correction. **H** After correction. (Continued.)

cortex (Fig. 5F). The distal piece is pushed in a dorsal direction and rotated to lessen the radial inclination (Fig. 5G, H). An osteotome placed into the osteotomy helps pry the bony fragments apart to allow correction. In addition, some proximal radial bone may have to be removed to allow the dome to rotate and some proximal volar bone will have to be removed once the sagittal correction has been achieved. Marked clinical and radiographic improvement is readily evident. Once acceptable correction has been achieved, provisional fixation is accomplished by 1 or two 1.6-mm Kirschner wires drilled from the radial styloid across the osteotomy site. In the past, these wires were maintained for definitive fixation. We now favor internal fixation with a plate and screw construct (Fig. 5I). Because of the deformity, anatomic plates usually do not conform to the distal radius. Therefore, more modular plates that can be contoured are

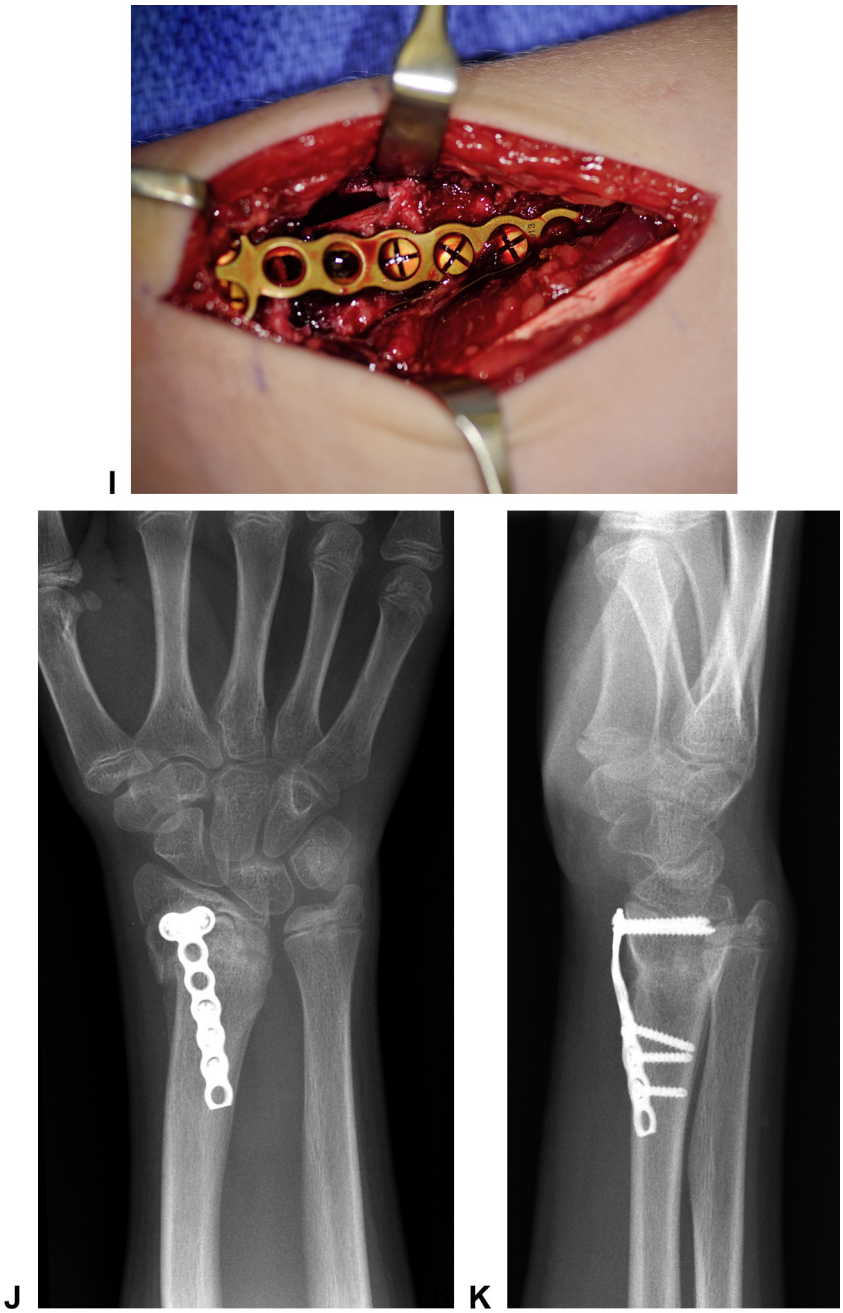

FIGURE 5: (Continued.) **I** Plate and screw fixation. **J** Postoperative anteroposterior x-ray with improved coronal alignment. **K** Postoperative lateral x-ray with improved sagittal alignment.

available. In addition, true rigid fixation is unobtainable and supplemental cast immobilization is required. After plate and screw fixation, the provisional Kirschner wires are removed.

At the time of dome osteotomy, assessment of remaining ulnar growth is necessary. If the ulnar physis is open and ongoing growth is likely, a concomitant epiphysiodesis should be performed (Fig. 6). Epiphysiodesis can be performed with the aid of mini-fluoroscopy and a small 1.7-mm cannulated drill. The ulnar physis is identified and the guide wire is placed, directed into the growth plate (Fig. 6A). A small stab incision is made about the Kirschner wire. The drill is placed over the wire and drilled into the growth plate (Fig. 6B). The Kirschner wire must be removed at this time (Fig. 6C). The drill is then passed back and forth and translated distal and proximal to ablate the physis (Fig. 6E). Cartilage will extrude from the stab incision and the physis will widen on the fluoroscopic images. The drill is then removed. Additional ablation can be accomplished with a curette or small burr to ensue complete epiphysiodesis (Fig. 6F).

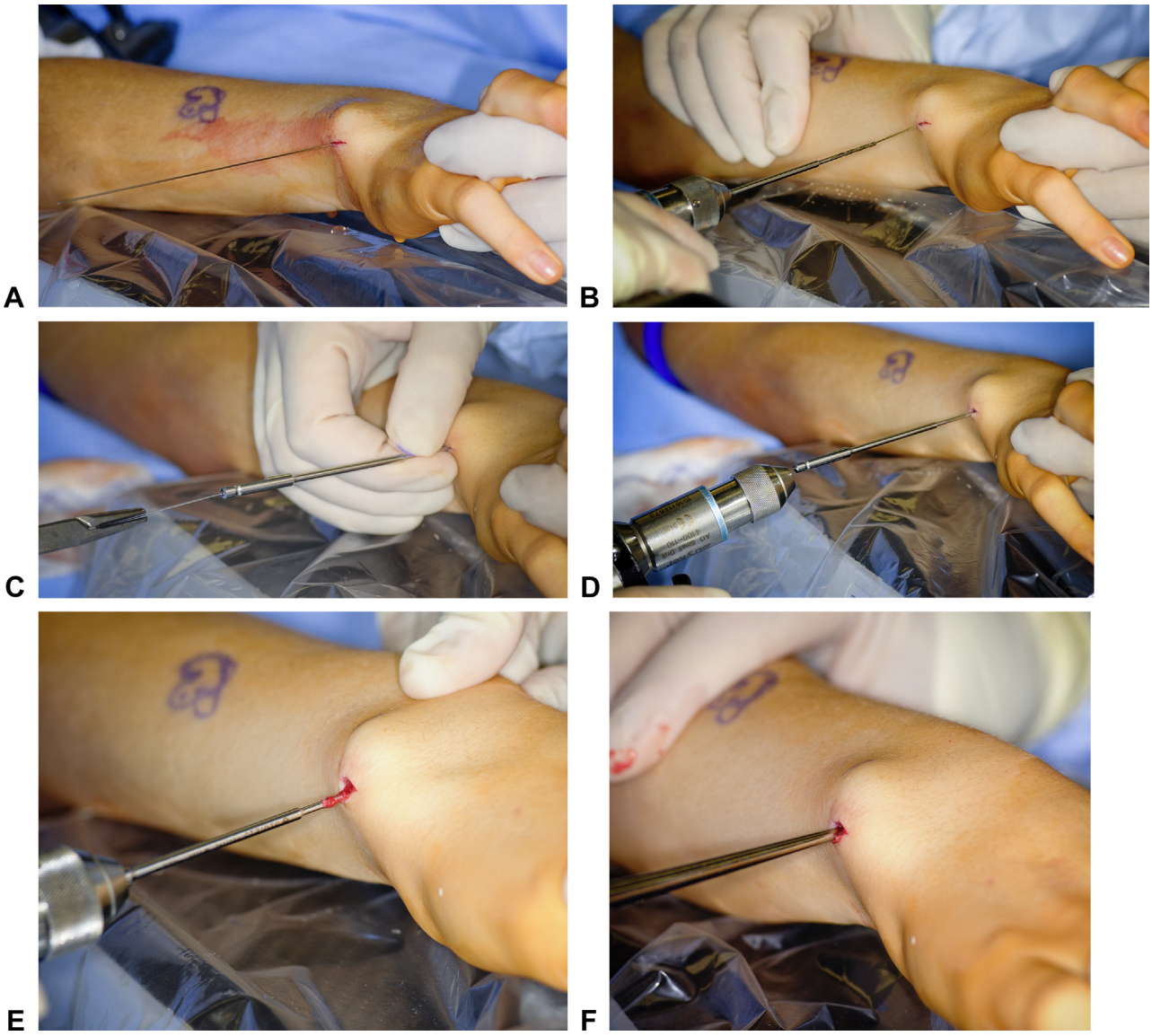

FIGURE 6: Epiphysiodesis of the distal ulna for Madelung deformity. (Courtesy of Shriners Hospitals for Children, Philadelphia, PA.) **A** A Kirschner guide wire is placed into the growth plate. **B** Cannulated drill is placed over the guide wire. **C** The guide wire is removed after the drill is driven across the physis. **D** The drill is reapplied to the bit. **E** The drill is driven back and forth and translated distally and proximally to ablate the physis. Cartilage is seen extruding from the stab incision. **F** Additional curettage to ensure epiphysiodesis.

The subcutaneous tissue and skin are closed with absorbable suture. A long arm orthosis is applied for 2 weeks. The orthosis is changed to a long arm cast that is removed at 6 weeks from surgery. A short arm orthosis is fabricated and active range of motion is started. Formal therapy is often unnecessary. Routine activities are resumed. Weightbearing is avoided until 3 months from surgery.

COMPLICATIONS

Surgical pitfalls include incomplete physiolysis or failure of the volar-ulnar corner to grow, improper placement of the dome osteotomy resulting in violation of the radiocarpal or ulnocarpal joint, insufficient reduction of the distal fragment resulting in suboptimal deformity correction, nonunion of the osteotomy site, and persistent ulnar impaction. Most of these surgical complications can be avoided by careful preoperative planning and strict adherence to the surgical plan. The judicious use of mini-fluoroscopy, especially in severe deformities, will diminish the chances of error with regard to osteotomy configuration and placement.

OUTCOME

The outcome of treatment for Madelung deformity is difficult to decipher because there is a wide spectrum of clinical presentations and radiographic findings. In the patient with mild to moderate deformity and

considerable growth remaining, physiolysis and ligament release has been successful. Vickers and Nielsen[4] reported results of 11 patients (15 wrists) who underwent surgery. All patients had some relief of pain; however, only 4 were completely pain-free and capable of performing all activities. Metaphyseal growth was documented in 11 wrists with radiographic improvement in 10 wrists. No progressive deformity was seen.

Short-term and long-term results after dome osteotomy and ligament release were reported by Harley et al[9] and Steinman et al.[10] Harley et al reported on 26 wrists in 18 patients with an average follow-up of 23 months. All patients reported pain reduction and improved appearance. Clinical improvements in supination and wrist extension were reported, with maintenance of pronation and wrist flexion. Radiographic improvements were encountered in radial inclination and lunate subsidence. In this cohort, 3 wrists ultimately required an ulnar shortening osteotomy at a second surgery. In 2013, a follow-up study by Steinman et al was published with average follow-up of 11 years. All patients maintained radial inclination and motion. Patient-reported outcomes using the Disabilities of the Arm, Shoulder, and Hand outcome measure noted that most of the patients reported outcomes equivalent to normative data.

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