Review Article

Madelung Deformity

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Abstract

Madelung deformity is a rare congenital anomaly of the wrist caused by asymmetric growth at the distal radial physis secondary to a partial ulnar-sided arrest. The deformity is characterized by ulnar and palmar curvature of the distal radius, positive ulnar variance, and proximal subsidence of the lunate. It more commonly occurs in females than males and typically affects both wrists. The deformity can occur in isolation or as part of a genetic syndrome. The pattern of inheritance varies, with some cases following a pseudoautosomal pattern and many others lacking a clear family history. Nonsurgical management is typically advocated in asymptomatic patients. Few studies exist on the natural history of the condition; however, extensor tendon ruptures have been reported in severe and chronic cases. Stiffness, pain, and patient concerns regarding wrist cosmesis have been cited as indications for surgery. Various techniques for surgical management of Madelung deformity have been described, but clear evidence to support the use of any single approach is lacking.

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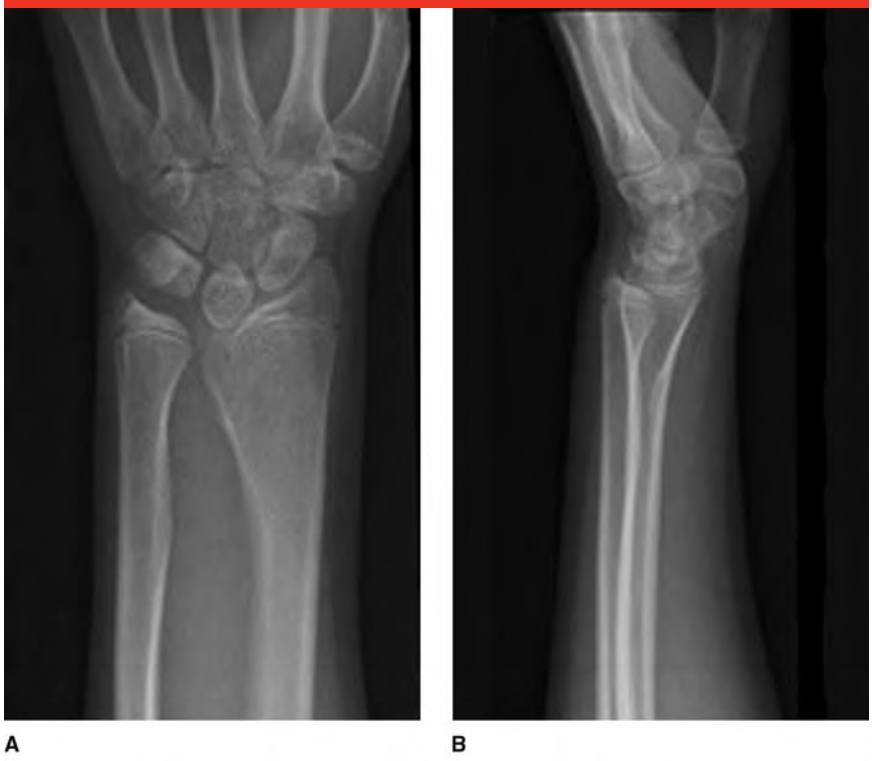
Madelung deformity is caused by partial arrest of growth at the volar and ulnar aspects of the distal radial physis and is frequently associated with Léri-Weill dyschondrosteosis (LWD). Cases without a known genetic association have traditionally been considered "isolated." However, novel genetic defects associated with this deformity have been recently identified, and future studies may better characterize the genetic origins of so-called idiopathic cases. Although many patients are asymptomatic, wrist pain, stiffness, and concerns regarding cosmesis can lead affected persons to seek orthopaedic care. Citing these symptoms and concerns as indications for surgery, the use of various surgical techniques has been advocated, including physiolysis, soft-tissue releases, osteotomies of the radius or ulna, ulnar shortening, ulnar head resection, and distal radioulnar joint (DRUJ) fusion or arthroplasty in various combinations. Although good surgical outcomes have been achieved in terms of pain relief and restoration of motion, few studies have included objective data that validate these outcomes. DRUJ arthroplasty has recently been advocated for management of Madelung deformity, but results are limited to a single study.[1] Future study should focus on gathering objective outcomes data and providing guidance on the management of symptomatic Madelung deformity.

Anatomy

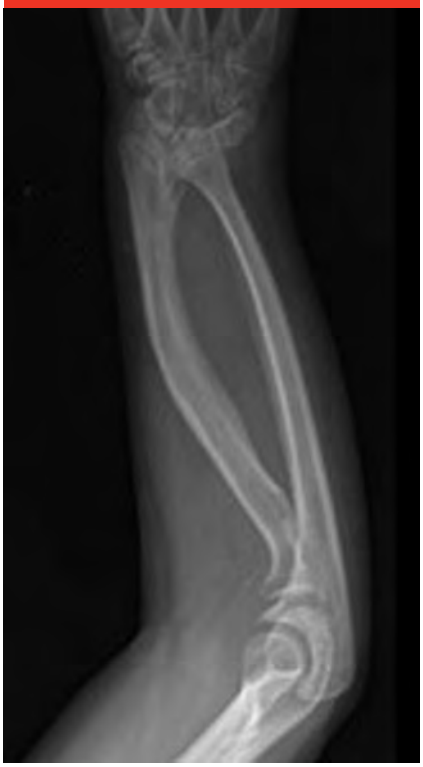
Madelung deformity is a collection of anatomic variances of the wrist that arise from a disturbance in growth at the ulnar and volar aspects of the distal radial physis. Asymmetrically retarded growth at this physis is the essential lesion of this condition. Both wrists are typically af-

Figure 1



PA (**A**) and lateral (**B**) radiographs of a 14-year-old girl with Madelung deformity in both wrists. Note the characteristic pyramidal configuration of the carpus, with the radial physis narrowed ulnarly and the prominent ulnar head dorsally.

Figure 2



Lateral radiograph of the forearm demonstrating bowing of the radial diaphysis in a 13-year-old girl with Madelung deformity.

fected. Anton et al[2] reviewed 171 cases of Madelung deformity from numerous published sources and found that the deformity was bilateral in 127 (74%). The hand appears to be translated volarly and ulnarly relative to the wrist, and dorsal prominence of the ulnar head is a distinguishing feature. Although the deformity is often characterized by its bony irregularity, an anomalous ligamentous structure is also involved in the development of Madelung deformity. Vickers and Nielsen[3] described an abnormal volar ligament that tethered the lunate to the volar distal radius (the Vickers ligament), resulting in the characteristic appearance of lunate subsidence between the ulna and radius. The authors suggested that this ligament contributed to radial deformity by asymmetrically slowing growth at the compressed section of the physis.

Radiographic Definition

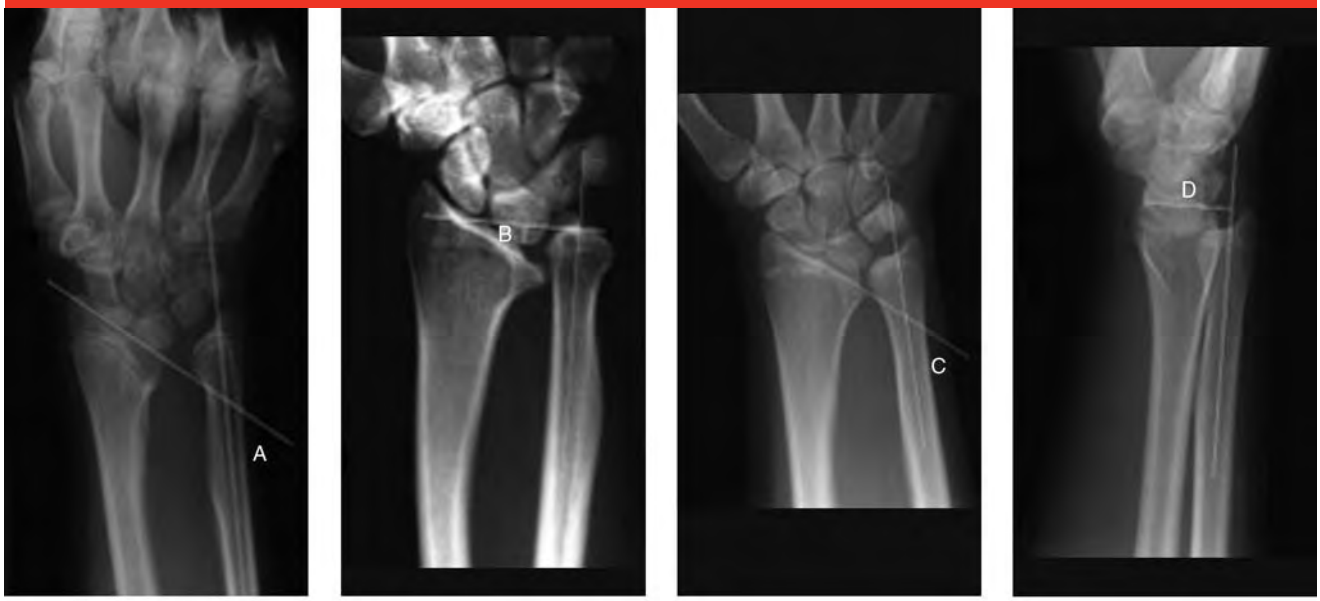
Radiography

The earliest reports on the radiographic appearance of Madelung deformity described features such as "pyramidalization" of the carpus as a result of proximal subsidence of the lunate, the absence or narrowing of the ulnar aspect of the distal radial physis, anterior bowing of the radial shaft, and dorsal subluxation of the ulnar head[2] (Figures 1 and 2). Later studies sought to establish criteria for measurement of the deformity and objective thresholds to guide diagnosis. Initially, measurements were based on the longitudinal axis of the radius measured on an AP radiograph.[3] Recently established criteria are based on the ulna instead of the radius and may be more reliable because the axis of the radius may be angulated as part of the deformity. McCarroll et al[4] identified four radiographic parameters for Madelung deformity, including ulnar tilt, lunate subsidence, lunate fossa angle, and palmar carpal displacement. The authors validated these measures by establishing threshold values at which they could predict a unanimous diagnosis of Madelung deformity among raters (Figure 3).

MRI

Little has been written on the role of MRI in the evaluation of Madelung deformity. Vickers and Nielsen[3] were among the first to advocate its use,

Figure 3



Radiographic parameters for predicting Madelung deformity on PA (**A** through **C**) and lateral (**D**) radiographs of the forearm. **A,** On a PA radiograph, ulnar tilt is defined as 90° minus angle A, the angle between a line drawn along the longitudinal axis of the ulna and a second line drawn tangential to the proximal ends of the scaphoid and lunate. The threshold value for diagnosis is ulnar tilt ≥33°. **B,** On a PA radiograph, lunate subsidence is defined as the distance between a line drawn perpendicular to the longitudinal axis of the ulna and a second line drawn through the distal articular surface and the proximal end of the lunate (B). The threshold value for diagnosis is ≥4 mm. **C,** The lunate fossa angle is defined on a PA radiograph as 90° minus angle C, the angle between a line drawn along the longitudinal axis of the ulna and a line drawn along the lunate fossa of the radius. The threshold value for diagnosis is ≥40°. **D,** Palmar carpal displacement is defined on a lateral radiograph as the distance D from the longitudinal axis of the ulna to the most volar point on the lunate or capitate. The threshold value for diagnosis is 20 mm or more of palmar carpal displacement. (Reproduced with permission from McCarroll HR Jr, James MA, Newmeyer WL III, Manske PR: Madelung's deformity: Diagnostic thresholds of radiographic measurements. *J Hand Surg Am* 2010;35[5]:807-812.)

suggesting that early identification of the thickened volar radiolunate ligament in children with a family history of Madelung deformity might allow for prophylactic excision. In a study of three patients with Madelung deformity who underwent MRI with a high resolution 3.0 Tesla MR scanner, Stehling et al[5] confirmed the presence of the Vickers ligament and an anomalous thickened volar radiotriquetral ligament. The clinical usefulness of MRI for diagnosis of Madelung deformity has not been evaluated critically and is theoretical at best based on the available literature. MRI is not part of the senior author's (D.P.H.) typical workup for evaluation of Madelung deformity.

History

Madelung deformity is named for Otto W. Madelung, a German surgeon who, in 1878, was the first to present a detailed report on the anatomic differences and possible causes of a rare congenital wrist deformity that he believed was analogous to other pediatric growth deformities such as genu varum.[6] Madelung was not the first to report on this condition, and controversy exists as to who deserves that distinction. Some authors, including Madelung himself, credited Guillaume Dupuytren, the French anatomist and surgeon who, in 1834, reported on a wrist deformity observed in adult male laborers.[7] Critics argue that because the deformity that Dupuytren described was acquired through occupational trauma, it could not be the same entity to which Madelung referred.[2,8] Credit is given to others instead, including Smith (1847), Adams (1854), Malgaigne (1855), and Jean (1875), all of whom independently contributed to a growing recognition among surgeons of the distinct clinical entity that was eventually described by Madelung.[2,9]

Epidemiology

Madelung deformity is a rare condition of unknown prevalence. Subtle

deformity is likely present for years before it is noticed clinically because the magnitude of the deformity tends to worsen with skeletal growth. Females are more commonly affected than males, with a ratio of approximately 4:1.[2,9,10] According to several published sources, age at presentation varies but is typically cited as before age 20 years.[2,11-13] Madelung deformity has been associated with numerous other skeletal dysplasias and genetic syndromes (eg, LWD, Turner syndrome [TS]).

Genetic Syndromes

Léri-Weill Dyschondrosteosis

LWD is a skeletal dysplasia that is frequently associated with Madelung deformity. Normal intelligence, short stature, and mesomelic limb shortening are characteristic features of LWD. Mesomelic shortening is the impaired longitudinal growth of the "middle" bones, including the radius, ulna, tibia, and fibula and is a feature seen in several skeletal dysplasias. The prevalence of this condition is unknown. In a study of 43 patients with genetically confirmed LWD, Ross et al[10] found that 32 patients had Madelung deformity (74%). Both conditions are more common in females, perhaps due to the role estrogen plays in the development of the dyschondrosteosis phenotype and the effect of estrogen on the physis.[10]

Haploinsufficiency of the short stature homeobox (*SHOX*) gene, which maps to the pseudoautosomal region 1 of chromosomes X and Y, results in the genetic defect that causes LWD. This defect has been implicated in nearly 70% of cases of LWD; in the remaining 30%, the genetic defect remains unknown.[14] Because the *SHOX* gene is located in the pseudoautosomal region of the sex chromosome, the defect is passed to offspring in a pseudoautosomal pattern; thus, instead of following a sex-linked pattern, recombination between sex chromosomes during meiosis allows the trait to be passed in a manner typical of autosomal dominant inheritance. Therefore, the children of a person carrying a SHOX haploinsufficiency disorder such as LWD have a 50% chance of acquiring that genetic defect.[15]

Turner Syndrome

TS, a genetic disorder in which nearly all affected persons are haploinsufficient for the *SHOX* gene, is also associated with Madelung deformity.[10] Patients with TS have a 45 XO karyotype and carry only a single X sex chromosome. Like other SHOX haploinsufficiency conditions, short stature is a characteristic of TS; however, Madelung deformity occurs in less than 10% of patients with TS.[10,16] This relatively low incidence may further implicate the role of estrogen in the development of the deformity because patients with TS notably lack normal levels of endogenous estrogen production as a result of gonadal dysgenesis.

Other Genetic Syndromes and Implications for Idiopathic Madelung Deformity

Mutations in the *GNAS* gene, which bears no relation to the *SHOX* gene, have recently been implicated in the development of Madelung deformity.[17,18] Cases of Madelung deformity also have been reported in the setting of nail-patella syndrome, an autosomal dominant condition of variable penetrance.[19] These examples demonstrate the complex nature of the genetic origins of Madelung deformity. Ongoing identification of previously unknown mutations that cause the deformity suggests that further study will reveal the genetic origins of this condition. It is also reasonable to assume that so-called idiopathic cases may represent de novo and yet uncharacterized genetic mutations. Further research on Madelung deformity may reveal more about the complex genetic origins of this condition.

Posttraumatic Pseudo-Madelung Deformity

Several authors have reported cases of Madelung-like wrist deformities caused by trauma; however, this matter has been a source of controversy for decades. As early as 1938, Anton et al[2] argued against trauma as a potential etiology of Madelung deformity, pointing out that most reported cases were in young girls, a population the authors described as "certainly less exposed to trauma than the general population." In a more recent study, a Madelung-like deformity in female gymnasts has been noted and was believed to be the result of physeal trauma experienced from repetitive loading of the wrist during intense and prolonged training.[20] Isolated trauma has also been implicated in patients who developed wrist deformity after distal radial physeal separation as a result of a fall.[21] Posttraumatic cases should be distinguished from Madelung deformity because any perceived association implied by the name pseudo-Madelung or Madelung-like is related only to a shared disturbance of growth at the distal radial physis. However, pseudo-Madelung conditions are not related to Madelung deformity. Their etiologies are entirely disparate.

Nonsurgical Management

Little has been written on the natural history of untreated Madelung defor-

mity. Patients can be asymptomatic well into adulthood.[22] Progressive arthrosis and instability of the DRUJ, radiocarpal arthrosis, or ulnocarpal abutment may eventually occur as the deformity progresses but, because the prevalence of asymptomatic Madelung deformity remains unknown, it is impossible to say how often these associated conditions occur. When the deformity is severe enough to cause symptoms before skeletal maturity, the amount of growth remaining should be considered. We consider an open distal radial physis to be indicative of skeletal immaturity. Patients with more growth remaining have more potential for progressive deformity than those who are skeletally mature. In the senior author's (D.P.H.) experience, surgery should be considered in these patients, with annual radiographic follow-up performed until skeletal maturity is reached. In contrast, Nielsen[11] advocated postponing surgery until skeletal maturity, noting that pain often decreased after a mean 10 years of observation in a cohort of skeletally immature patients. In the absence of long-term prospective cohort studies, the natural history of untreated Madelung deformity remains poorly understood.

In cases of severe and symptomatic deformity, attritional rupture of finger extensor tendons has been reported as a rare complication of nonsurgical management of Madelung deformity.[23-25] This complication, which is encountered in patients with rheumatoid arthritis, is caused by repeated mechanical irritation of the digital extensor tendons over the dorsal prominence of the subluxated ulnar head. The ulnar-sided digital extensor tendons are most commonly involved. This complication occurs in the setting of severe and chronic cases, with most cases reported in persons older than age 70 years. In a series of six cases of spontaneous rupture of the extensor tendon of the fingers associated with Madelung deformity, the largest published series on the topic, Ducloyer et al[23] suggested early surgical management of the deformity to prevent this complication. Tendon transfers, tendon grafting, and direct repair have all been advocated to manage this complication; however, no studies have compared different treatment methods.[23-25]

Surgical Indications

Indications for surgical management of Madelung deformity include wrist pain, limited wrist range of motion (ROM), and deformity. Wrist pain is often progressive in nature and is typically experienced in the ulnar aspect of the wrist. Although restricted wrist motion associated with Madelung deformity is widely reported, agreement on which planes of motion are most restricted is lacking. However, loss of extension and ulnar deviation are commonly described.[1,3,12,13,26-28] Substantial disagreement exists with regard to the range of pronosupination, with some authors describing greater restriction in pronation than supination in patients with Madelung deformity.[22,26]

A lack of consensus with regard to the primary indications for surgical management of Madelung deformity exists, with some authors citing the appearance of the wrist, and others citing wrist pain or loss of motion. Several studies cite the appearance of the wrist as the primary reason that some patients seek surgery.[3,28-30] In a study of 18 patients with Madelung deformity treated with wedge subtraction osteotomy of the radius and shortening of the ulna, dos Reis et al[28] found that a main complaint of 14 of 18 patients was dissatisfaction with the appearance of the deformity. In a cohort of 18 patients with a mean age of 12 years, Harley et al[29] reported that concern for increasing deformity was the primary complaint in 14, with a secondary complaint in the remaining 4 patients.

Other authors consider wrist pain or loss of motion to be the primary indications for surgery, with concerns about appearance alone considered an inadequate indication for surgery.[1,11,22,27,31-34] However, some have argued against loss of motion as an indication for surgery, citing no improvement in ROM after surgical correction.[11] These conflicting opinions demonstrate the lack of consensus on the indications for surgical management of Madelung deformity.

Surgical Management

Physiolysis

Vickers and Nielsen[3] identified two distinct lesions that are central to the pathogenesis of Madelung deformity: partial growth arrest at the distal radial physis, which causes volar and ulnar bowing of the distal radius, and the Vickers ligament. Because this ligament tethers the lunate to the distal radius, the carpus assumes a characteristic triangular arrangement (Figure 1, A). This ligament may also contribute to partial growth arrest of the radial physis secondary to compression. Vickers and Nielsen[3] recommended addressing both lesions through a transverse volar approach. Others have modified this approach by using a standard longitudinal volar incision.[13] An osteotomy is performed 5 mm ulnar to the radioulnar joint. A local fat graft is placed in the area of the physeal defect to prevent formation of a bony physeal bar, a procedure sometimes called a Langenskiöld physiolysis. The Vickers ligament is also excised.

The efficacy of prophylactic release

of the Vickers ligament for management of early, mild deformity in the skeletally immature patient remains unknown. The senior author (D.P.H.) excises the ligament in conjunction with an ulnar shortening osteotomy or distal ulnar epiphysiodesis in patients with mild deformity. The shortening osteotomy palliates the symptoms of ulnar abutment. The benefits of ligament release are largely conjectural; however, release of the Vickers ligament is believed to prevent thinning of the radial physis caused by compression, thereby preventing further radial deformity with continued growth. Of 15 wrists (11 skeletally immature patients) treated by Vickers and Nielsen[3] with prophylactic release of the Vickers ligament, the authors reported no progression of deformity.

Radial Dome Osteotomy and Physiolysis

Although Vickers and Nielsen[3] reported successful improvement of pain and wrist ROM with release of the Vickers ligament, the technique does not correct preexisting deformity, which is a drawback in management of severe cases of Madelung deformity. This led others to develop techniques that provided the deformity correction afforded by radial osteotomy in addition to the release of the Vickers ligament. Harley et al[29] described a technique in which the volar (Henry) approach is used to perform a radial dome osteotomy with curved osteotomes in addition to a release of the thickened volar radiolunate ligament. Correction of the radial deformity is then achieved via longitudinal traction followed by radial deviation and pronation of the hand and dorsal displacement of the distal fragment. The distal fragment is fixed internally across the osteotomy site with 2.4-mm Steinmann pins. The arm is immobilized in a long arm cast for 6 weeks, at which time the pins are removed, and the cast is replaced with a short arm splint for an additional 4 weeks.[29,35]

Combined Radial and Ulnar Osteotomies

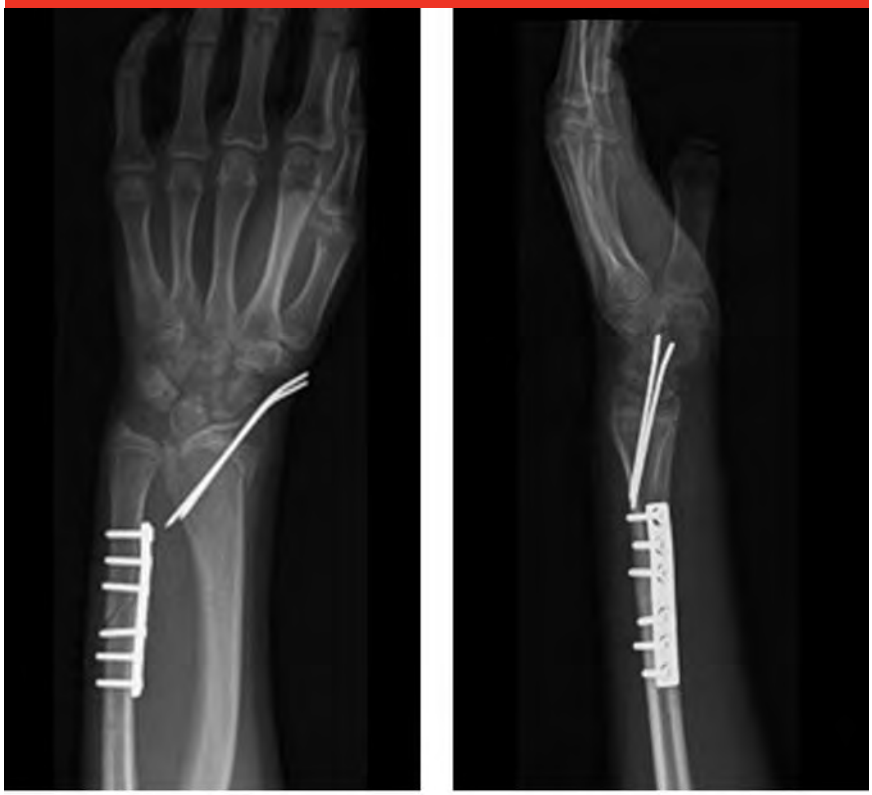
Corrective osteotomy of the radius with DRUJ arthrodesis has been advocated by several authors for management of symptomatic Madelung deformity.[36,37] The goal is to achieve simultaneous correction of the distal radius deformity and arthrodesis of the unstable and painful DRUJ. Both opening and closing wedge osteotomies have been used in this combined technique. DRUJ arthrodesis involves segmental resection of a portion of the distal ulnar shaft just proximal to the ulnar head (Sauvé-Kapandji procedure) to create a pseudarthrosis.[38] Schroven et al[36] believed that, rather than performing a Darrach resection of the ulnar head, preserving the ulnar head and stabilizing the DRUJ via arthrodesis would restore the radiocarpal architecture and avoid progressive ulnar subluxation of the carpus. Similarly, dos Reis et al[28] and Laffosse et al[33] both described the use of a dorsolateral closing wedge osteotomy of the radius combined with ulnar shortening osteotomy to preserve the ulnar head while alleviating ulnocarpal abutment. Salon et al[34] advocated the use of radial closing wedge osteotomy with ulnar shortening osteotomy to correct ulnar variance and relocate the dorsally prominent ulnar head. Avoiding ulnar carpal subluxation may be particularly important in the setting of increased radial inclination associated with Madelung deformity (Figure 4).

Ranawat et al[31] advocated the use of Darrach resection of the ulnar head either alone or with radial osteotomy to manage ulnar-sided wrist pain in patients with Madelung deformity. A total of 12 wrists in eight patient were treated with Darrach resection alone (7 procedures), Darrach resection with radial osteotomy (4 procedures), and isolated ulnar shortening (1 procedure). No statistical analysis was provided; however, improvements in pain and wrist motion were seen in seven of eight patients at a mean 8-year follow-up. Asymptomatic ulnar translation of the lunate occurred in all patients who underwent Darrach procedure alone. No patients performed "heavy work." The authors suggested that people pursuing heavy work might not tolerate Darrach resection because the lunate subluxation could become painful later in life.

Isolated Radial Osteotomy

Isolated osteotomy of the radius has been advocated to correct Madelung deformity. This technique focuses on correcting the volar and ulnar angular radial deformity that is characteristic of the condition. In the European literature, management of the deformity with transverse distal radial osteotomy and Ilizarov external fixation has been described.[27,32] A perceived advantage of external ring fixation is the ability not only to correct the angular deformity of the radius but also to lengthen it, thereby obviating the need to perform a separate ulnar osteotomy to address resultant ulnar positive variance caused by a closing wedge radial osteotomy. In a study of 11 patients with Madelung deformity, Murphy et al[30] performed an isolated radial opening wedge osteotomy in 8, with a second more proximal radial shaft osteotomy performed in 3 patients to address "significant radial bowing." Isolated radial osteotomy afforded correction of the primary radial deformity, improved congruence of the DRUJ, and improved ulnar variance by lengthening the radius. In three

Figure 4



Postoperative PA (**A**) and lateral (**B**) radiographs of the forearm in the patient shown in Figure 1, following radial dome osteotomy and ulnar shortening osteotomy. Ulnar shortening was deemed necessary when intraoperative examination of the distal radioulnar joint (DRUJ) demonstrated instability that was concerning for the eventual development of DRUJ arthrosis. At 26 months postoperatively, the patient was active in sports and had no reports of wrist pain.

patients, radial osteotomy alone left >3 mm of ulnar positive variance, and an ulnar shortening osteotomy was performed.

Isolated Ulnar Osteotomy

Because dorsal prominence of the ulnar head and ulnocarpal abutment are two sources of substantial disability and pain in patients with Madelung deformity, some authors have recommended the use of surgical techniques that focus exclusively on the ulna. Bruno et al[22] and Glard et al[39] recommend isolated ulnar shortening osteotomy for management of the symptomatic Madelung wrist. In both studies, patients had relatively mild bony deformity, symptoms of ulnocarpal abutment with ulnar positive variance, and were skeletally mature at the time of treatment. Bruno et al[22] performed an ulnar shortening osteotomy using multiple fixation techniques, including tension band wiring and dynamic compression plating. Glard et al[39] used a dorsal approach to the ulna, performing an osteotomy 6 cm proximal to the ulnar styloid. The size of the osteotomy was determined preoperatively on radiographs in an attempt to achieve neutral ulnar variance. A contoured dynamic compression plate was then applied to produce volar angulation to reduce the DRUJ.

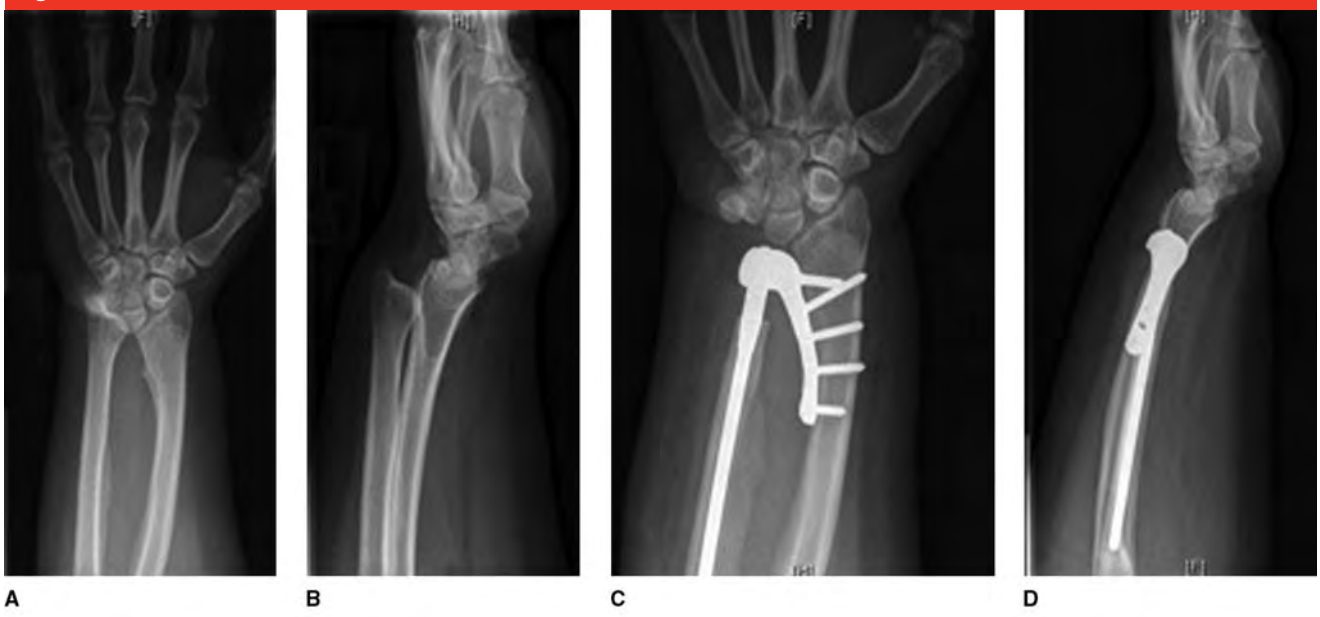
Arthroplasty

Recently, total DRUJ arthroplasty has been used for surgical management of symptomatic Madelung deformity. However, only a single report specifically cites the deformity as an indication for this procedure.[1] In this small series, three women with Madelung deformity presented with painful DRUJ instability, limited wrist pronosupination range of motion and dorsal prominence of the ulnar head. Two of these patients had previously undergone surgical treatment without improvement in symptoms. Coffey et al[1] treated the patients with DRUJ arthroplasty with a semi-constrained device implanted using the technique described by Scheker et al.[40] The main body of the prosthesis was affixed to the distal radius. A press-fit stem and an ultra-high-molecular-weight polyethylene ball comprise the ulnar component, which articulates with a hemi-socket on the radial plate (Figure 5). Substantial radial deformity may require staged management, with corrective radial osteotomy preceding arthroplasty to allow proper distal placement of the radial plate. Coffey et al[1] believed that DRJU arthroplasty was indicated for management of persistent painful DRUJ instability and limited pronosupination following other prior surgical procedures in skeletally mature patients.

Outcomes

A paucity of objective data is available to guide management of symptomatic Madelung deformity. Reported surgical outcomes vary but are generally positive (Table 1). Few of the studies that we reviewed in-

Figure 5



Preoperative PA (**A**) and lateral (**B**) radiographs of the forearm in a 24-year-old woman with significant Madelung deformity, disabling wrist pain, and limited wrist range of motion. Postoperative PA (**C**) and lateral (**D**) radiographs of the forearm after distal radioulnar joint arthroplasty was performed with a semiconstrained implant.

cluded statistical analysis, often because of the small cohort size. All of the studies reported postoperative improvement of pain in most patients, although few studies provided objective data to quantify these improvements.[1,22,27,28,34] With regard to improved ROM, results were mixed. Although many studies did not perform statistical analysis on or found no differences in postoperative ROM, several larger studies consistently reported improvement in supination and wrist extension.[28,29,33] In contrast, deformity correction outcomes were inconsistently reported; many authors compared preoperative and postoperative radiographs to quantify deformity correction.[22,27-30,34,36,39] However, some authors assessed the statistical significance of reported changes.[28,29,33] Because patient dissatisfaction with the appearance of the deformed wrist is often considered an indication for surgery, some studies reported on patient satisfaction with the appearance of the wrist following surgery as a subjective measure of outcome.[11,27-30,32,39]

Interest in the use of implant arthroplasty for management of disorders of the wrist and DRUJ has increased; however, results of DRUJ arthroplasty for management of Madelung deformity are currently limited to a single report.[1] At a 2-year follow-up, three patients (four wrists) were available for evaluation. Two patients had undergone procedures prior to DRUJ arthroplasty; one patient had undergone bilateral corrective radial osteotomy and another had undergone ulnar shortening osteotomy, synovectomy, and dorsal wrist capsulodesis. Following arthroplasty, the patients achieved substantial improvement in supination ROM, with a mean increase of 58.8°, the largest improvement in ROM reported by any study that we reviewed. Improvements in the Disabilities of the Arm, Shoulder, and Hand score and visual analog pain score were also reported, although the authors did not state whether these gains reached statistical significance. Recognizing the limitations of their study, the authors ultimately recommended that this procedure be used in patients who have previously undergone surgical intervention for symptomatic deformity but continue to have pain and DRUJ instability. Because patients with Madelung deformity often present at a young age with pain and disability, additional long-term studies are needed to establish the durability of DRUJ arthroplasty in this patient population.

Author's Preferred Treatment

In the senior author's (D.P.H.) experience, pain and limited wrist motion rather than deformity are appropriate indications for surgical management of Madelung deformity. In the asymptomatic growing child, annual clinical and radiographic examina-

Table 1

Comparison of Surgical Techniques for Management of Madelung Deformity

Study	No. of Wrists	Surgical Technique	Follow-up	Mean Difference in ROM	Outcomes
Bruno et al[22]	9	Isolated ulnar shortening osteotomy	42 mo (mean)	No significant difference	Mean postoperative objective Jiranek pain score 96/100 (7 excellent, 2 good results). Cosmetic result NR.
Coffey et al[1]	4	Total DRUJ arthroplasty (Scheker prosthesis, Aptis Medical)	24 mo	+58.8° supination, –2.3° pronation	Mean VAS pain score decreased from 4.5 preoperatively to 1.5 postoperatively. Cosmetic result NR.
de Billy et al[32]	5	Radial osteotomy, Ilizarov technique	NR	30° improvement in extension	Pain resolved and "aesthetic appearance" improved in all patients. Data not provided.
De Smet and Fabry[37]	1	Closing wedge radial osteotomy, Lauenstein (Sauvé-Kapandji) procedure	18 mo	+65° supination, +10° pronation, full flexion/extension at baseline	No pain at follow-up. Cosmetic result NR.
dos Reis et al[28]	25	Dorsal closing wedge radial osteotomy, ulnar shortening osteotomy	53 mo (mean)	+15.4° supination, +18.2° pronation, +19.3° extension	"Improved" pain in 80% of patients and 88% satisfied with cosmetic appearance.
Glard et al[39]	4	Isolated shortening and apex dorsal ulnar osteotomy	24 mo	+28.5° supination, +13.75° pronation, +5° extension (loss in 3 of 4 wrists)	10.5 mean pain score improvement on scale of 0 to 30 (30 signifies no pain, 0 signifies daily narcotics required). All patients satisfied with cosmetic appearance.
Harley et al[29]	26	Radial dome osteotomy, release of volar radioulnar ligament; ulnar shortening performed in one wrist to correct residual deformity	23 mo (mean)	+13° supination, +17° extension, no significant difference in pronation/flexion	"Markedly improved" pain in all, but 3 wrists required ulnar shortening for residual pain. Radial inclination improved a mean 13°.
Houshian et al[27]	8	Transverse radial osteotomy, Ilizarov technique	30 mo (mean)	+34° supination, +9° pronation, +15° flexion	Mean VAS score 0 at follow-up. Persistent but improved deformity in 4 of 8 wrists.
Laffosse et al[33]	14	Dorsal closing wedge radial osteotomy, ulnar shortening osteotomy	61 mo (mean)	+28.9° supination, +19.6° pronation, +25.5° extension; no difference in flexion	Pain present in 2 of 11 patients at follow-up. Cosmetic results NR. The deformity was not listed as an indication for surgery.
Murphy et al[30]	12	Volar radial osteotomy, excision of volar radioulnar ligament; proximal radial osteotomy and ulnar osteotomy (when indicated)	48 mo (mean)	No significant difference	All patients "generally pleased with pain relief," mild ulnar wrist pain in one patient. Patients were "pleased" with improved forearm appearance.
Nielsen[11]	15	Various	102 mo (mean)	Signficantly improved (1); no change (10); worsened (2)	Pain improved in 9 of 13 patients. 9 of 13 patients satisfied with cosmetic appearance.
Ranawat et al[31]	12	Dorsal closing wedge radial osteotomy and Darrach (5), Darrach alone (7)	96 mo (mean)	+13.8° supination, +5.4° pronation, +15.8° extension, –11.7° flexion	Improved pain in all wrists, no objective data provided. Cosmetic results NR.

DRUJ = distal radioulnar joint, NR = not reported, ROM = range of motion, VAS = visual analog scale

Table 1 (continued)

Comparison of Surgical Techniques for Management of Madelung Deformity

Study	No. of Wrists	Surgical Technique	Follow-up	Mean Difference in ROM	Outcomes
Salon et al[34]	14	Closing wedge radial osteotomy, ulnar shortening osteotomy. No surgery in 3.	116 mo (mean)	+48° pronosupination arc	All patients "pain-free during daily activities." 8 wrists with "complete" resolution of cosmetic concerns, 3 with "slight" residual ulnar head protrusion.
Schroven et al[36]	3	Closing wedge radial osteotomy, Lauenstein (Sauvé-Kapandji) procedure	Range, 5–36 mo	+50° supination, +20° pronation, +43° extension, +38° flexion	NR
Vickers and Nielsen[3]	24	Radial physiolysis, excision of volar radioulnar ligament	15 mo (minimum)	+23° supination; flexion, extension, and pronation range "improved" with no data given.	"Significant" reduction in pain in all wrists. Cosmetic result NR.

DRUJ = distal radioulnar joint, NR = not reported, ROM = range of motion, VAS = visual analog scale

tions are performed. Progressive deformity, DRUJ instability and arthrosis, and ulnocarpal abutment warrant surgical intervention. In patients with an open distal radial physis, release of the Vickers ligament is combined with corrective radial osteotomies. If ulnar height is neutral, then a distal ulna epiphysiodesis is performed at the time of the radial osteotomy. If ulnar height is positive, then the ulna is brought to neutral height with a shortening osteotomy. Adult patients with Madelung deformity are advised that increasing wrist pain and loss of motion are associated with this condition. Removable splints are used for intermittent pain and surgical intervention is recommended for chronic, unrelenting symptoms.

Three clinical scenarios are encountered in patients with Madelung deformity: predominant radiocarpal pain, predominant DRUJ pain and, most commonly, a combination of both. Radiocarpal pain and arthrosis are managed with fusion. If the midcarpal joint is preserved, then a radiolunate or radioscaphoid-lunate fusion is recommended, otherwise a pancarpal fusion is performed. Management of DRUJ pain and arthrosis is controversial. Resection arthroplasty (eg, Darrach procedure) can palliate pain, but it is not eliminated. In addition, the procedure does not allow repetitive forearm pronosupination activities.

DRUJ arthroplasty with a semiconstrained implant has yielded promising early results, with relief of DRUJ pain and improved repetitive forearm motion. In the typical setting of pancarpal and DRUJ arthrosis and pain, the senior author (D.P.H.) chooses to address both issues simultaneously except when the radial diaphysis is excessively bowed and an osteotomy to realign the hand relative to the forearm is required. In that setting, the radial osteotomy is allowed to heal before proceeding with reconstruction of the DRUJ and radiocarpal fusion.

Summary

Madelung deformity is a topic of intense interest among surgeons who seek to understand how best to follow and treat this relatively rare condition. The literature is replete with conflicting opinions on the best treatment for patients with this deformity. Future research should focus on longitudinal studies and objective outcome measures to establish guidelines for the surgical and nonsurgical care of patients with Madelung deformity.

References

Evidence-based Medicine: Levels of evidence are described in the table of contents. In this article, references 4 and 10 are level I studies. References 16 and 20 are level III studies. References 1-3, 5, 6, 8, 11, 12, 17-19, and 21-39 are level IV studies. References 7 and 40 are level V expert opinion.

References printed in **bold type** are those published within the past 5 years.

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