Madelung's Deformity

82

Scott N. Oishi, Lesley Wheeler, and Marybeth Ezaki

Contents

Introduction 1764
Pathoanatomy and Applied Anatomy 1764
Madelung's Deformity Treatment Options 1765
Nonoperative Treatment 1765
Operative Treatment 1766
Physiolysis and Ligament Release 1767
Volar Ligament Release and Distal Radius Dome Osteotomy 1768
Preferred Treatment 1770
Surgical Pitfalls and Prevention 1770
Management of Complications 1771
Summary 1771
References 1772

Abstract

The exact etiology of Madelung's deformity is not known. In addition, the clinical and radiographic presentation can vary greatly between not only patients but also between forearms in the same patient. In severe cases, the premature closure of the volar-ulnar physis of the distal radius leads to the characteristic appearance of volar subluxation of the hand/carpus as well as a dorsally prominent ulna. In some cases, a genetic component is present in the form of Leri-Weill syndrome in which there is an abnormality in the short stature homeobox gene (SHOX) (Benito-Sanz et al. Am J Hum Genet 77:533–44, 2005). Symptoms can include pain, decreased wrist/forearm ROM, as well as cosmetic dissatisfaction with appearance.

It is important to assess and counsel every patient individually as not everyone will need surgical intervention. If the patient has no symptoms and the deformity is minimal, then careful observation with serial examinations and radiographs is warranted. If the deformity is more severe and symptoms are present, then operative intervention may be warranted. Depending on the age of the patient and deformity present, several types of operative treatment are available. The results following surgery for Madelung's deformity have consistently been shown to be of benefit and should be considered for symptomatic patients with substantial deformity.

S.N. Oishi (✉) • L. Wheeler • M. Ezaki
Department of Hand Surgery, Texas Scottish Rite Hospital for Children, Dallas, TX, USA
e-mail: scott.oishi@tsrh.org; lesley.wheeler@tsrh.org; marybeth.ezaki@tsrh.org

J.M. Abzug et al. (eds.), *The Pediatric Upper Extremity*,
DOI 10.1007/978-1-4614-8515-5_81

Introduction

Otto Madelung first described the deformity that now bears his name in 1878 (Madelung 1878). Although he was not the first surgeon to describe it, he certainly was the first to offer a more comprehensive discussion regarding not only the etiology but also treatment options. His opinion at that time was that the deformity resulted from repetitive joint overloading that led to abnormalities in growth, causing the proximo-volar subluxation of the wrist that is characteristic of the deformity. It was his belief that job modification and splinting would decrease the pain and these were his treatments of choice (Arora et al. 2006).

The etiology of most Madelung's deformity is idiopathic, although it can also result following trauma or infection. Premature arrest of the volar-ulnar distal radial physis leads to the eventual deformity, which usually presents in the adolescent age group and is four times more common in girls. Involvement is usually bilateral, with asymmetry being typical. Most commonly these patients present with complaints of a "funny-looking" wrist as adolescents and radiographs lead to the diagnosis (Fig. 1). Pain is not a usual finding. Some limitations in wrist extension and ulnar deviation along with decreased forearm supination can be present. Clinically, the affected extremities have shortening of the forearm, dorsally prominent ulna, and volar subluxation of the hand/carpus.

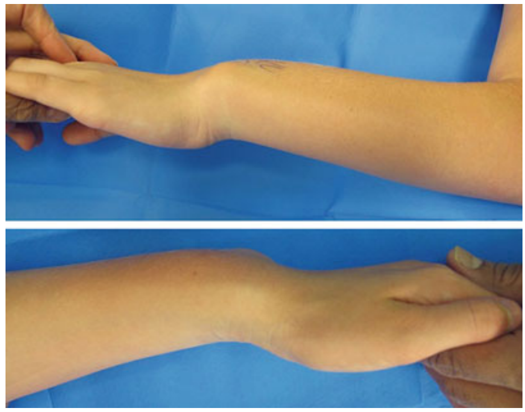

Fig. 1 Typical appearance of a patient with Madelung deformity. Note the obvious appearance of a prominent ulnar with volar subluxation of the hand/carpus

Fig. 2 A patient with Leri-Weill syndrome. Note the short forearms in association with the wrist findings

There is some debate regarding the genetic component of Madelung's deformity, although many cases are thought to be autosomal dominant with incomplete penetrance (Grigelioniene et al. 2001; Hirschfeldova et al. 2012; Benito-Sanz et al. 2005; Rappold et al. 2002). Leri-Weill syndrome (dyschondrosteosis) is characterized by mesomelic dwarfism, short stature, and Madelung's deformity (Fig. 2). These patients have been shown to have abnormalities in the short stature homeobox gene (SHOX) that resides in the pseudoautosomal region of the sex chromosomes. While the SHOX haploinsufficiency was originally thought to be solely caused by gene deletion, more recently investigators have also found it to be the result of mutations within the SHOX gene itself. Madelung's deformity can also be seen as a component in Turner's syndrome, Langer-Giedion syndrome, multiple hereditary exostoses, and multiple enchodromatoses. In addition, there are also patients of normal stature who have no other findings suggestive of underlying syndromes, but who have typical Madelung's deformity.

Pathoanatomy and Applied Anatomy

The patient with Madelung's deformity has a characteristic position of the upper extremity. In most cases, the ulna appears prominent with the carpus

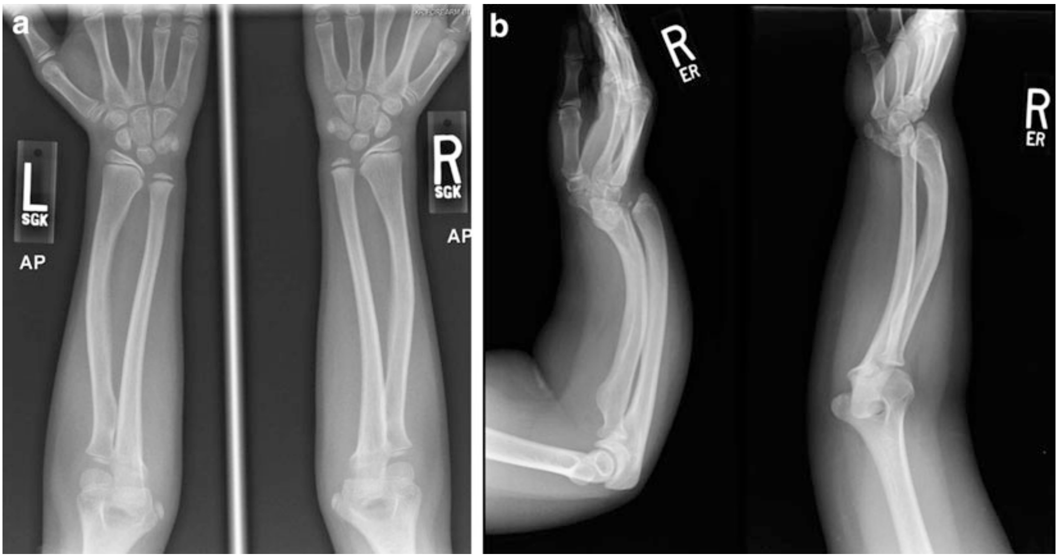

Fig. 3 (**a**) Radiographs of a patient with minimal deformity. (**b**) Radiographs of a patient with severe deformity

and hand volarly translated. Pain over the ulna may be present with some decrease in forearm supination and wrist extension. The forearms themselves may be short. Plain radiographs can show a spectrum of deformity ranging from minimal increase in radial inclination to substantially increased radial inclination along with lunate subsidence (Fig. 3a, b). McCarroll et al. (2010) reported that radiographic criteria for the diagnosis included ulnar tilt of 33° or greater, lunate subsidence of 4 mm or more, lunate fossa angle of 40° or greater, and palmar carpal displacement of 20 mm or more. In the skeletally immature patient, premature closure of the volar-ulnar radial physis is noted. Radiographs of the entire forearm to include the elbows are important as whole-bone involvement of the radius has been documented and has implications on eventual outcomes (Zebala et al. 2007; Fig. 4). CT and MRI imaging can add a three-dimensional view of the deformity, but is usually not necessary to establish or treat the Madelung's deformity patient.

Vickers and Nielsen (1992) described a thick fibrous structure (the so-called Vickers' ligament), which begins on the ulnovolar metaphyseal region of the radius and attaches to the lunate and triangular fibrocartilage. Whether this anomalous structure is truly an additional structure or just prominence of the volar wrist ligamentous structures is subject of debate, but its presence has been reported in up to 91 % of patients (Vickers and Nielsen 1992; Nielsen 1977). Some authors believe that release of this structure during operative intervention is an integral part of the surgery (Vickers and Nielsen 1992; Nielsen 1977; Harley et al. 2006; Steinman et al. 2013). However, it is doubtful that the potential tethering effect of the "ligament" is the primary cause of deformity. Madelung's deformity is not seen in the child less than 3 years of age (a period where rapid growth takes place).

Madelung's Deformity Treatment Options

Nonoperative Treatment

Many patients display asymmetric involvement. Thus, even though one extremity warrants treatment because of symptoms, observation in the less involved extremity is indicated. In some

Fig. 4 A patient with whole-bone involvement of the radius

patients, the physeal involvement on the volar-ulnar portion of the distal radius is minimal, with minimal deformity of the radius and carpus. Many of these patients, in fact, can be carefully evaluated every 6 months during their adolescent growth period until skeletally mature and may never need surgical intervention (Fig. 5a, b, c). Prophylactic release of Vickers' ligament has not been proven to prevent deformity or pain in these patients.

Madelung's deformity	
Nonoperative treatment	
Indications	Contraindications
Minimal deformity	Progressive deformity
No pain	Pain

Operative Treatment

Symptoms that warrant consideration for surgery in most patients include pain and unacceptable deformity. Early reports on operative techniques centered solely on the ulna with varying success (Ranawat et al. 1975; Glard et al. 2007; Bruno et al. 2003). Most surgical treatment now more logically addresses the radius as the deformity appears to be inherent to this bone (Vickers and Nielsen 1992; Harley et al. 2002, 2006; Steinman et al. 2013; Murphy et al. 1996). Age plays an important role with respect to which procedure is chosen. Vickers and Nielsen (1992) have reported excellent success with ligament release and

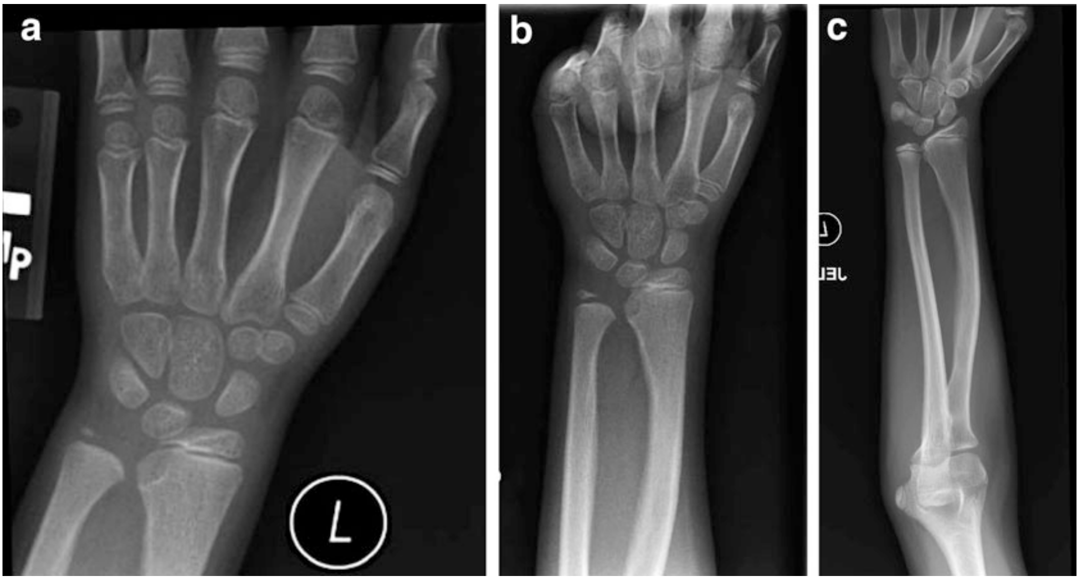

Fig. 5 Serial radiographs of a patient showing no progression of deformity over time

physiolysis in those skeletally immature patients who have considerable growth potential remaining. In the patient with substantial deformity and not much forearm growth remaining, corrective radial osteotomy with ligament release is warranted.

Physiolysis and Ligament Release

After induction of general anesthesia and under tourniquet control, a longitudinal incision is placed over the flexor carpi radialis tendon (FCR) overlying the distal radius. Next, dissection is carried down to the pronator quadratus where the distal portion is divided to allow exposure of the abnormal radio-lunate ligament (Vickers'). It is easily identified in a concavity in the metaphyseal portion of the radius. This is carefully released from proximal to distal until the tethering of the lunate is released. This is usually readily apparent and results in being able to visualize the undersurface of the lunate. Next, physiolysis is performed as described initially by Langenskiold and later by Vickers (Vickers and Nielsen 1992). The downturned metaphysis at the distal volar-ulnar radius is carefully trimmed to expose the remaining growth plate. Using a burr or rongeur, the abnormal physis is removed until more normal-appearing physeal cartilage is identified. Adequate resection is imperative to the success of this procedure. At this point, the area is then packed with a generous amount of autogenous fat that can be harvested from a number of donor sites (Fig. 6). It is important for this fat to make contact with the entire cavity that was created. Layered closure is performed and the extremity is immobilized for 3–4 weeks. After removal of the cast, range-of-motion (ROM) exercises are instituted and serial radiographs are performed to access growth and alignment (Fig. 7a, b, c).

Physiolysis and ligament release for Madelung's deformity
Preoperative planning
Patient in supine position with arm extended out on a hand table
Use of C-arm brought in from end of hand table
Sterile or non-sterile tourniquet
Burr and rongeur
Sterile prep of lower abdomen or similar area for autogenous fat graft harvest

Physiolysis and ligament release for Madelung's deformity
Surgical steps
Longitudinal incision overlying FCR in distal forearm
Exposure and division of distal portion of pronator quadratus muscle, exposing Vickers' ligament
Release of Vickers' ligament from proximal to distal until the tethering of the lunate is released
Exposure of abnormal physis through the metaphyseal region of the distal volar-ulnar radius
Removal of abnormal physis using a burr and/or rongeur
Harvest of autogenous fat and packing of defect in radius
Layered closure and casting

Physiolysis and ligament release for Madelung's deformity
Postoperative protocol
Short-arm cast for 4 weeks
After cast is removed, start range-of-motion exercises
Serial radiographs to access growth and alignment

Volar Ligament Release and Distal Radius Dome Osteotomy

Under general anesthesia and tourniquet control, a longitudinal incision is made overlying the FCR tendon in the distal forearm ending just proximal to the volar wrist case. Dissection is then carried down between the radial vessels and the FCR. After exposure of the pronator quadratus, it is divided near its radial attachment leaving a cuff for repair at the end of the procedure. Next, identification of the abnormal Vickers' ligament is made on the metaphyseal area of the distal radius.

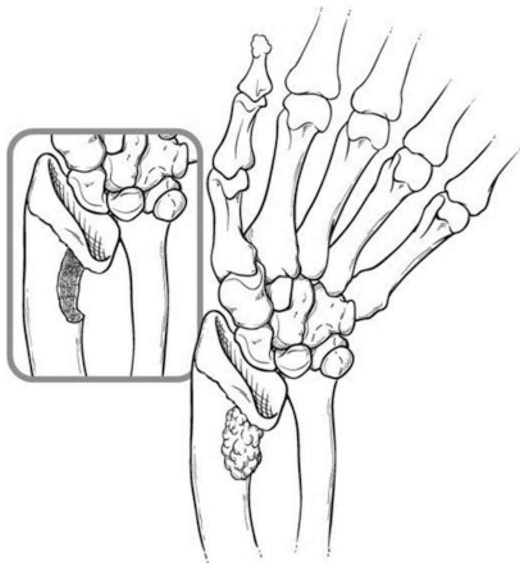

Fig. 6 A schematic drawing of the physiolysis/fat grafting procedure

Fig. 7 Serial radiographs of a patient who underwent physiolysis/fat grafting procedure

This is then released sequentially from its origin until the cartilaginous undersurface of the lunate can be seen. The periosteum overlying the metaphyseal portion of the radius is elevated, and then a crescent-shaped osteotomy is performed. This can be performed with a fine, curved osteotome after first fenestrating the osteotomy location with a k-wire or a dome osteotomy saw (Fig. 8a). Care must be taken to ensure that the osteotomy is proximal to the distal radioulnar joint and that the dorsal periosteum is left intact. After the osteotomy is performed, removal of a bony fragment off the dorsal radial area of the proximal radial fragment is usually required to allow adequate dorsal translation of the distal fragment. Using direct pressure, the distal fragment can then be put into its new position and stabilized using two or more smooth Steinman pins passing through the radial styloid and proximal fragment. Intraoperative fluoroscopy is then used to assess position and alignment of the osteotomy (Fig. 8b and c). Any removed bone can then be placed in the osteotomy area using it as bone graft. The pronator and FCR sheath is then repaired, and the skin is closed.

At times, the ulna needs to be addressed at the same setting. If the ulna physis is still open, then epiphysiodesis is performed through a separate incision. Alternatively, if the ulna physis is closed but a prominent ulna positive variance is present after positioning of the distal radial fragment, then ulna shortening can be performed using the surgeon's technique of choice.

A long-arm cast is then applied and split to allow for postoperative swelling. Pins and long-arm casting are maintained until the 6-week postoperative mark when pins are removed. Short-arm cast is applied until radiographic healing is evident (Fig. 9a, b, and c).

Volar ligament release and distal radius dome osteotomy for Madelung's deformity
Preoperative planning
Supine position with arm extended out on a hand table
Use of C-arm brought in from end of table
Sterile or non-sterile tourniquet

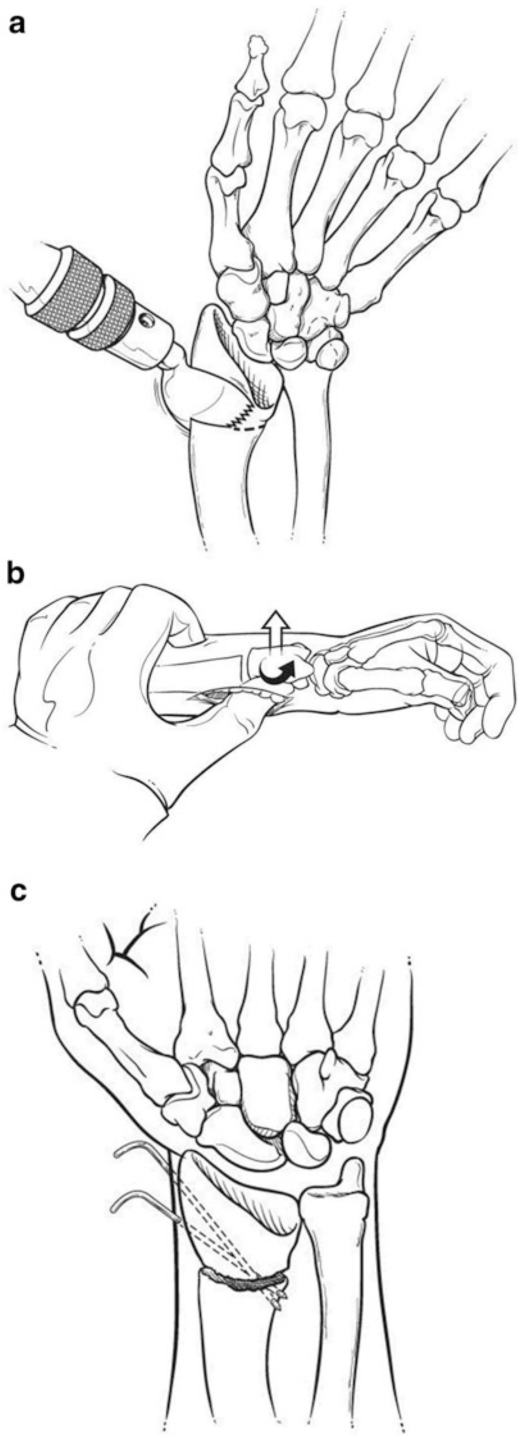

Fig. 8 (**a**) The radial dome osteotomy being performed. (**b**) Manipulation of the osteotomy. (**c**) Osteotomy in its final position with wire fixation

Fig. 9 (**a**) Pre-operative radiographs. (**b**) Osteotomy with pin fixation. (**c**) After healing of osteotomy and pin removal

Volar ligament release and distal radius dome osteotomy for Madelung's deformity
Surgical steps
Longitudinal incision overlying FCR in distal forearm
Exposure and division of pronator quadratus, leaving cuff on radial attachment for later repair
Exposure and release of Vickers' ligament in proximal to distal direction until undersurface of lunate can be seen
Crescent-shaped osteotomy with preservation of dorsal periosteum
Placement of distal fragment in position with stabilization using two or more smooth Steinman pins
Fluoroscopy to assure proper position and alignment
Repair of pronator quadratus and layered closure

Volar ligament release and distal radius dome osteotomy for Madelung's deformity
Postoperative protocol
Long-arm cast that is split to allow for postoperative swelling
Removal of cast and pins at 6 weeks
Placement of short-arm cast until radiographic healing

Preferred Treatment

Many patients with Madelung's deformity present as a result of an incidental finding on x-ray. If minimal deformity is present, then observation alone is indicated until which time progressive deformity and/or symptoms present. In some patients, no treatment may be necessary. If a patient is symptomatic with deformity and assessed to have a reasonable amount of growth remaining, physiolysis and fat grafting is a reasonable option. Most patients, however, present near skeletal maturity and will require dome osteotomy and ligament release. In girls, the start of menarche usually is an indication of limited future growth in the wrist and hand. In the dome osteotomy patient, it is important to assess the ulna at the time of the procedure. If the ulna physis is open, then epiphysiodesis is warranted at the time of surgery. If the patient is skeletally mature and neutral or minimal ulna positive variance is present after stabilization of the osteotomy, then no treatment of the ulna is performed. If, however, significant ulna positive variance exists after stabilization of the radial osteotomy, then ulna shortening is performed.

Surgical Pitfalls and Prevention

Incomplete physiolysis: The surgeon must assure that complete resection of the affected area is performed. Loupe magnification should allow good visualization. Adequate fat graft placement to prevent recurrent closure is also imperative. If not performed correctly, the radius will heal with a new physeal bar.

Improper placement of dome osteotomy: The surgeon must assure that the osteotomy is proximal to the DRUJ. This position can be misleading because of the deformity present. If not absolutely sure about intended osteotomy position, then fluoroscopy should be used to assure adequate position. In whole-bone involvement, correction of a more proximal deformity may be needed.

Improper reduction of the distal fragment: Difficulty translating the distal fragment into desired position is often caused by a bony spike on the dorsal radial aspect of the proximal fragment. This should be trimmed until adequate position of the distal fragment is obtained.

Bony nonunion: This has not been reported as a problem in the osteotomy patients. Pin stabilization and immobilization until radiographically healed is imperative.

Ulna impaction/impingement after osteotomy: If identified, ulnar shortening procedure can be performed at same setting through separate incision.

Madelung's deformity	
Potential pitfalls and preventions	
Potential pitfall	Pearls for prevention
Incomplete physiolysis	Adequate visualization using loupe magnification
	Enough autogenous fat graft to fill the entire cavity
Improper placement of dome osteotomy	Careful assessment of location of intended osteotomy intraoperatively prior to performing saw cut (sometimes using fluoroscopy)
	Careful assessment of entire forearm radiograph preoperatively to assure correct location of osteotomy to correct deformity present
Improper reduction of the distal fragment	Removal of bony dorsal radial bone spike, if present, to allow proper positioning of the distal fragment
	Use of intraoperative fluoroscopy to assess position and alignment

(*continued*)

Madelung's deformity	
Potential pitfalls and preventions	
Potential pitfall	Pearls for prevention
Bony nonunion	Pin stabilization and immobilization until radiographically healed
Ulna impaction/ impingement after osteotomy	Performance of ulna epiphysiodesis based on assessment of remaining growth potential of the radius

Management of Complications

If physiolysis is not successful: Dome osteotomy can be performed to correct the deformity.

If a bony nonunion occurs: Rigid volar plating and bone grafting would be necessary, although this has not been a problem to date.

Persistent ulnar-sided wrist pain with evidence of ulna impaction: Ulnar shortening osteotomy or Darrach procedure can be performed.

Madelung's deformity	
Common complications	Management
Physiolysis is not successful	Dome osteotomy
Bony nonunion occurs (exceedingly rare)	Rigid volar plating and bone grafting
Persistent ulnar-sided wrist pain with evidence of ulna impaction	Ulnar shortening osteotomy or Darrach procedure

Summary

Patients with Madelung's deformity present with a spectrum of clinical and radiographic findings. In asymptomatic patients with minimal radiographic findings, observation is clearly the treatment of choice. Serial examinations and radiographs allow intervention if symptoms or considerable deformity arise; however, some patients may never require any surgical intervention.

In the appropriate patient physiolysis and ligament release have been reported as effective treatments. In 1992, Vickers and Nielsen published their results on 11 patients (15 wrists) who underwent surgery. In their study, all patients had some relief of pain, although only four claimed to be totally pain-free and able to do all activities. Positive metaphyseal growth was observed in 11 wrists, with no progression of deformity noted. Improvement in the angle of the physis and epiphysis was noted in 10 wrists, with gains in wrist ROM and forearm supination (Vickers and Nielsen 1992).

Results following dome osteotomy and ligament reconstruction have been reported both in short-term and long-term follow-up in the same institution. In the first report, Harley et al. (2006) reported on 26 wrists in 18 patients with an average follow-up of 23 months. All patients reported reduction in pain and improved appearance. Improvements in supination and wrist extension were seen, with preservation of pronation and wrist flexion. Improvements were also noted in terms of radiographic parameters of radial inclination and lunate subsidence. Of note, three wrists did require ulnar shortening at a later surgery (Harley et al. 2006). In a follow-up study by Steinman et al. (2013) with average follow-up of 11 years, all patients maintained radial inclination and ROM throughout the follow-up period. The majority of the patients had functional outcomes equivalent to normative data as measured by DASH. As expected, there was a positive correlation between increased DASH score and arthritis grade. In addition, patients with whole-bone deformity also had increased DASH score (Steinman et al. 2013).

In conclusion, patients with Madelung's deformity can have a wide range of clinical presentations. Careful evaluation of each patient on a case-to-case basis will result in the optimal treatment at the ideal time.

References

Arora AS, Chung KC, Otto W. Madelung and the recognition of Madelung's deformity. J Hand Surg Am. 2006;31:177–82.

Benito-Sanz S, Thomas NS, Huber C, Gorbenkodel Blanco G, Aza-Carmona M, Crolla JA, et al. A novel class of Pseudoautosomal region 1 deletions downstream of SHOX is associated with Leri Weill dyschondrosteosis. Am J Hum Genet. 2005;77:533–44.

Bruno RJ, Blank JE, Ruby LK, Cassidy C, Cohen G, Bergfield TG. Treatment of Madelung's deformity in adults by ulna reduction osteotomy. J Hand Surg. 2003;28A:421–26.

Glard Y, Gay A, Launay F, Guinard D, Legré R. Isolated wedge osteotomy of the ulna for mild Madelung's deformity. J Hand Surg Br. 2007;32A:1037–42.

Grigelioniene G, Schoumans J, Neumeyer L, Ivarsson SA, Eklöf O, Enkvist O, et al. Analysis of short stature homeobox-containing gene (SHOX) and auxological phenotype in dyschondrosteosis and isolated Madelung deformity. Hum Genet. 2001;109:551–58.

Harley BJ, Carter PR, Ezaki M. Volar surgical correction of Madelung's deformity. Tech Hand Up Extrem Surg. 2002;6:30–5.

Harley BJ, Brown C, Cummings K, Carter PR, Ezaki M. Volar ligament release and distal radius dome osteotomy for correction of Madelung's deformity. J Hand Surg Am. 2006;31:1499–506.

Hirschfeldova K, Solc R, Baxova A, Zapletalova J, Kebrdlova V, Gaillyova R, et al. SHOX gene defects and selected dysmorphic signs in patients of idiopathic short stature and Léri Weill dyschondrosteosis. Gene. 2012;491:123–27.

Madelung OW. Die spontane subluxation de hand nachvorne. Verh Dtsch Ges Chir. 1878;7:259–76.

McCarroll HR, James M, Newmeyer W, Manske PR. Madelung's deformity: diagnostic thresholds of radiographic measurements. J Hand Surg Am. 2010;35A:807–12.

Murphy MS, Linscheid RL, Dobyns JH, Peterson HA. Radial opening wedge osteotomy in Madelung's deformity. J Hand Surg Am. 1996;21:1035–44.

Nielsen JB. Madelung's deformity. A follow-up study of 26 cases and a review of the literature. Acta Orthop Scand. 1977;48:379–84.

Ranawat CS, DeFiore J, Straub LR. Madelung's deformity: an end-result study of surgical treatment. J Bone Joint Surg. 1975;57A:772–75.

Rappold GA, Fukami M, Niesler B, Schiller S, Zumkeller W, Bettendorf M, et al. Deletions of the homeobox gene SHOX (short stature homeobox) are an important cause of growth failure in children with short stature. J Clin Endocrinol Metab. 2002;87:1402–06.

Steinman S, Oishi S, Mills J, Bush P, Wheeler L, Ezaki M. Volar ligament release and distal radial dome osteotomy for the correction of Madelung deformity: long-term follow-up. J Bone Joint Surg Am. 2013;95:1198–204.

Vickers D, Nielsen G. Madelung deformity: surgical prophylaxis (physiolysis) during the late growth period by resection of the dyschondrosteosis lesion. J Hand Surg Br. 1992;17B:401–07.

Zebala LP, Manske PR, Goldfarb CA. Madelung's deformity: a spectrum of presentation. J Hand Surg. 2007;32A:1393–401.