Madelung’s Deformity: Diagnosis, Classification and Treatment.

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Abstract

Madelung's deformity is a rare condition characterized by abnormal growth arrest of the distal radial physis. It can cause pain, restricted range of movement and visual deformity. Surgery aims to balance growth in skeletally immature patients and correct residual deformity in skeletally mature patients. Systematic review of the literature reveals a heterogenous and inconclusive evidence base, which makes management decisions difficult. Using the currently available evidence, a classification system is outlined, which is based upon the degree of skeletal maturity and the severity of the deformity and which provides a system for selecting appropriate surgical strategies.

Keywords: Madelung’s deformity, Hand surgery, Congenital hand, Epiphysiodesis, Physeal bar, Vicker’s ligament.

Introduction

Madelung’s deformity is a rare condition first described by OTTO Wilhelm Madelung in 1878 [1]. Madelung’s deformity involves abnormal growth arrest of the distal radial physis at its ulnar and volar aspect. This leads to the development of a characteristic ulnar and volar curvature of the distal radius, a positive ulna variance and proximal subsidence of the lunate [2]. In the majority of patients, the condition arises as part of a congenital syndrome [3, 4, 5], although in a subset the aetiology may be traumatic [6, 7]. It is more common in females, with a female to male ratio of 4:1 [8], and is most often bilateral [9].

The true prevalence of Madelung’s deformity is unknown, owing to a likely asymptomatic subset. It accounted for just 4 cases of 641 patients referred with a congenital upper limb anomaly during a 1-year period in the North-East of America [9]. In our own centre, 6 patients were referred to the tertiary paediatric upper limb service in a case load of 269 patients over a 3-year period between November 2012 and September 2016. The deformity often arises as part of a syndrome and therefore can exist in combination with complex multi-system issues. Surgery therefore might be considered inappropriate and in this respect referral numbers are likely to be a poor surrogate for disease prevalence.

Two anatomical anomalies have a central role in the development of congenital Madelung’s deformity. Firstly, patients have one or two abnormal volar ligaments. The most commonly described is Vickers’ ligament, a fibrous band running from the ulna border of the radius to the lunate [10-12]; however a pathological radio-triquetral ligament has also been identified [10]. Secondly an abnormal bar has been described in the distal radial physis at its ulnar and volar aspect which is thought to restrict growth in this region [10, 11]. A current hypothesis is that the
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ligaments, acting as tethers, cause a compressive force on the distal ventromedial portion of the radial physis, leading to the premature closure of the physis at this site and resulting in the characteristic osseous deformity [10-13].

The deformity typically presents during adolescence following a period of rapid growth [10]. While little is known about the precise natural history of the condition it seems rational that progressive ulno-carpal abutment, distal radio-ulna joint (DRUJ) instability leads eventually to radio-carpal arthritis [14]. Impingement from the prominent ulna head, instability due to stretching of the triangular fibrocartilage complex (TFCC) ligaments and secondary tears to the cartilaginous portion of the complex may contribute to specifically ulnar-sided wrist pain. In severe chronic cases extensor tendon rupture has been reported due to abrasion on the prominent ulna head [15].

The flow diagram in Figure 1 outlines the current understanding of the pathogenesis and long-term consequences of Madelung’s deformity.

**Figure 1:** A summary of current understanding of the pathological evolution and structural consequences of Madelung’s deformity.

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Madelung’s deformity can leave young patients with chronic pain and with varying levels of disability and visual deformity. The condition evolves over time in the skeletally immature subject and therefore the optimum timing and indications for intervening remain controversial.
Multiple operative treatment options have been proposed, principally involving corrective osteotomies, but there is no high-level evidence to support the use of any single surgical strategy. Comparisons between studies are difficult given the marked heterogeneity of the evidence available. As a result, patients with Madelung’s deformity present a considerable challenge to clinicians.

This work aims to review the management of Madelung’s deformity by consolidating the current evidence base. Firstly, the radiographic assessment of disease severity will be considered and secondly the surgical options will be reviewed. A classification system for Madelung’s deformity is proposed which is based on the degree of skeletal maturity and the severity of the deformity. This system of classification advocated as a structured and rational guide to aid clinicians’ decision making when confronted with this challenging problem.

Methods

A literature review was undertaken using the PubMed database and the search term used was “Madelung deformity”. Additional relevant studies were identified by reviewing citations included in these initial papers. The inclusion criteria included those studies which evaluated a surgical treatment and presented interpretable outcome data. Studies were excluded if they were written in a subject other than English or the full text could not be accessed. Forty-three papers were identified and reviewed. These included 30 case series and 13 single case reports. Evidence is no higher than level 4, with authors reporting their experience and outcomes in small case series.

Radiographic Assessment

There are a number of key features on plain radiographs including the plane(s) of deformity and skeletal maturity which guide management. Dannenberg described a number of consistent features, including increased dorsal and radial convexity of the distal radius; increased volar and ulnar tilt of the distal radial articular surface; a widened interosseous space; a relative dorsal position of the ulna head; and pyramiding of the carpus (Figure 2) [2]. However, it should be noted that no correlation has been established between degree of pain and severity of radiographic deformity [16].

Previously, the severity of radiographic deformity was quantified using measurements made against the long axis of the distal radius, but this practice is poorly reliable and reproducible as the landmarks at the distal radius become difficult to identify. McCarroll et al [17] have proposed three measurements based on the long axis of the ulna which is advocated for assessing the severity of the deformity because they demonstrate a high degree of reliability and reproducibility (Figure 3):

1. **Ulnar tilt (A)** is the acute angle between the long axis of the ulna and a line tangential to the proximal surface of the scaphoid and lunate on a PA radiograph. It assesses the angulation of the distal radial articular surface.

2. **Lunate subsidence (B)** is the distance in millimetres between the most proximal point of the lunate and a line perpendicular to the long axis of the ulna at the level of the distal articular surface. Measured on a PA radiograph it indicates the proximal displacement of the carpus that results from the bony deficiency in the lunate fossa. This measurement indicates the degree of ulna overgrowth, and the subsequent requirement for an ulna shortening procedure. Traditionally ulna variance, as opposed to lunate subsidence, was used to quantify the prominence of the ulna head. However, Farr, in accordance with McCarroll, demonstrated superior inter- and intra-rater reliability for lunate subsidence and consequently its use over ulna variance is recommended [17,18]. We use the ulnar tilt and lunate subsidence to assess the coronal plane of the deformity.

3. **Palmar carpal displacement (C)** is the distance in millimetres, on a lateral radiograph, between the longitudinal axis of the ulna and the most volar point on the surface of the lunate or capitate. It describes the palmar directed bow of the distal radius, and indicates the presence of a multiplanar deformity.
In addition to the degree of deformity, it is vital to assess radiographs for skeletal age because this has a fundamental bearing on the treatment approach that can be employed in any specific individual. A variety of methods can be used to assess skeletal maturity. Given that the growth of the wrist in these cases is abnormal, traditional methods of assessment are difficult. In Madelung deformities, assessment of the distal ulna and in particular the thickness of the growth plate and both the presence and degree of definition of the ulna styloid is crucial. A well-defined ulna styloid tends to imply a patient who is within 2 years of skeletal maturity [19]. The potential for remodelling in these patients is therefore likely to be limited after any surgical intervention.

The Use of MRI in Madelung’s Deformity
MRI enables assessment of the abnormal ligamentous structures [12]. Cook et al performed MRI scans of 4 females with bilateral Madelung’s deformity. Vickers’ ligament and a pathological volar radio-triquetral ligament were identified in 7 out of 8 wrists. The abnormal physeal bar was identified in all 8 wrists [11]. The mean length of the bar was 15 mm on sagittal cuts and 16 mm on coronal images.

The role of MRI in management algorithms for patients with Madelung’s deformity has not been fully clarified. It can be used to screen “at risk” patients in which plain radiographic signs are yet to emerge. In our practice it forms part of the pre-operative work-up if release of the abnormal volar ligaments or excision of the physeal bar is being considered. MRI will provide additional information about the size and location of the physeal bar, and it can also define any secondary triangular fibrocartilage complex pathology, although MRI imaging of the TFCC is often difficult to interpret due to the abnormal anatomy at this site.

Figure 2: The bony architecture in Madelung’s deformity. The characteristic osseous deformities are demonstrated, including ulnar and volar curvature of the distal radius, positive ulna variance and proximal subsidence of the lunate.
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Figure 3: Schematic diagrams illustrating the measurement of A= ulnar tilt, B= lunate subsidence and C= palmar carpal displacement.

Figure 4: MRI images demonstrating A= Radio carpal (“Vickers”) ligament, B= Volar subluxation of the lunate and rest of carpus, C= Compressed volar portion of distal radius growth plate.

Classification and Treatment Options

The aim of surgery is to control growth in skeletally immature patients and correct residual deformity in mature patients. Attention should be paid to both radial and ulnar components where necessary. Most reported case series cite either pain [10, 20] or appearance [21] as the primary motivators for surgical intervention. The available evidence to guide decision making in Madelung’s deformity is not comprehensive. Nevertheless, it is the intention of this paper to provide a system to stratify the condition, and to suggest a surgical approach based on these. A summary of this system is proposed and presented in Table 1. Patients are subdivided into 3 groups, based upon the degree of skeletal maturity and the severity of the deformity. The surgical approach to each subtype is discussed in the text below, with reference to the current available evidence base.
Skeletally Immature Patients; Type 1
In Group 1 patients can be subdivided into those who still have adequate growth potential for remodelling, and those in the late phase of growth who do not. Attention should be focused firstly towards releasing any aberrant growth-limiting ligamentous structures.

The next step is to achieve balanced growth of both the ulna and radius, which in practice means promoting normal growth in the radius (the abnormal component), and taking steps to limit growth in the ulna (the healthy component) if this is also necessary.

IA: Pre-Growth Arrest
Group 1A, in which patients are pre-growth arrest, is advocated that only radio-carpal ligament release is required, followed by careful follow up to ensure growth continues as normal. This recommendation is based upon current understanding of the pathogenesis of Madelung’s deformity, although it has no evidence base in the literature to date.

1B: Following Growth Arrest with Potential for Remodelling
In group 1B, in addition to ligament release, the key is to promote normal growth in the radius, and in this regard the most effective step will be to

Table 1: An approach to the treatment of Madelung’s deformity based upon maturity and severity [22].

<table>
<thead>
<tr>
<th>Stage</th>
<th>Patients who are skeletally immature</th>
<th>Surgical Management</th>
<th>Case Series</th>
</tr>
</thead>
<tbody>
<tr>
<td>1A</td>
<td>Pre-growth arrest. Those patients without radiographic deformity, but for whom genetic testing and or family history predict the development of the disease. The “at risk” group.</td>
<td>Release of abnormal radio-carpal ligament</td>
<td>No available evidence</td>
</tr>
<tr>
<td>2</td>
<td>Patients who are skeletally mature AND symptomatic</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Patients with longstanding deformity and sequelae, including Distal radio-ulnar joint instability and osteoarthritic changes</td>
<td>Consideration of ulna excision (Darrach’s procedure) and/or joint fusion for severe pain</td>
<td>Angelini (1996) Coffey (2009) Lluch, (2013) Gibersen-Chen (2020)</td>
</tr>
</tbody>
</table>
carry out physiolysis of the radial physeal bar, with the aim of elevating the epiphysis back into a normal alignment, and then introducing a fat graft to prevent re-ossification.

The procedure was originally described by Vickers and Nielson [10]. Exposure is made using a standard volar approach to the distal radius. The aberrant radio-lunate ligament is identified along the ulnar edge and released to enable mobilisation of the carpus and to perform the osteotomy. The joint is opened to enable direct visualisation of the articular surface and fluoroscopy is then used to identify the portion of the physis that is still open. The osteotomy is made in line with the area of the physis that has prematurely closed on the ulnar side, in the direction of the still open physis on the radial side (figure 5a). The subchondral bone is elevated using the osteotome with care not to damage the bone or cartilage which can be very thin. This is held in place using a 2 mm Kirschner wire passed from the radial styloid along the epiphysis (figure 5b). A dermal fat graft is harvested from the antecubital fossa (removing the epidermis, but preserving the dermis and attached fat), which is rolled up and inserted under the osteotomy.

In Vickers and Nielsen’s study, improvements in range of motion and pain were reported in a cohort of 24 wrists in 17 patients who underwent the procedure at a mean age of 12 years [10]. Mean follow up was not stated but ranged from 15 months to 12 years. As might be expected maximal benefit was seen in patients operated upon early in the course of the disease. Although Vickers originally treated patients in this group with physiolysis alone, subsequent reported outcomes following isolated physiolysis have been mixed. Del Core et al treated 8 wrists with Vickers ligament release and radial physiolysis [23]. Seven of 8 wrists were pain free at 1 year, and 6 of 8 were pain free at final follow up at a median of 10.6 years, although 2 patients had required reoperation, with 1 radial dome osteotomy and 1 ulnar shortening osteotomy having been subsequently performed. Pre-operative range of motion was preserved post operatively, with no significant differences found.

Ogino reported outcomes of 3 wrists in 2 patients with a mean age of 12 years, in which 1 patient developed palmer shift of the carpus and worsened radial inclination [24]. (It should be noted that this patient was 13 years old at the time of surgery, and therefore likely to have fallen into our group 1c as a patient with insufficient growth potential to see benefit from physiolysis alone). Paes described the long-term (12 year) outcomes of 5 wrists in 3 patients who underwent the procedure at a mean age of 12 years [25]. Results were poor, with pain and restriction in range of motion seen at long-term follow up in all 3 patients. In Paes’s series, 3 wrists also subsequently underwent salvage ulna shortening procedure which provided “good pain relief” in two wrists and a “satisfactory” result in the 3rd. In addition, Farr et al. have more recently reported on 41 wrists in which ulnar epiphysiodesis performed at the index procedure (mean age 13.4 +/- 1.5y) may have avoided the requirement for a secondary ulna shortening procedure [26]. Otte’s case series of 12 wrists undergoing Vicker’s ligament release and radial physiolysis included one patient who subsequently required a secondary bilateral ulna epiphysiodesis [27].

With these reports in mind, the senior author recommends proceeding to epiphysiodesis of the ulna in the majority of patients in group 1b, with the aim of preventing or reducing subsequent ulna positive variance which can lead to chronic pain from ulno-carpal abutment and impingement. Epiphysiodesis is achieved by drilling across the epiphysis (figure 5c), and placing a compressing screw to block further growth (figure 5d).

1C: Following Growth Arrest with Inadequate Potential for Remodelling

Those patients in Group 1C, (i.e. those without the potential for further remodelling) require, in addition to the above, a deformity correcting osteotomy. This has the aim of correcting the volar and ulna angulation, and can also be used to
lengthen the distal radius if required. Murphy described an opening wedge osteotomy performed through a volar approach, which enabled a biplane deformity correction [20]. Pain and appearance improved subjectively in all patients, although there was no improvement in range of motion. This procedure needed to be repeated in the two skeletally immature patients in his series, as continued growth overtook the correction. This underlines the importance of ascertaining the potential for further growth and remodelling which separates our groups 1B and 1C.

For patients in group 1C the senior authors’ preference is to perform a radial dome osteotomy which, unlike Murphy’s approach, has the advantage of being able to achieve deformity correction in all three planes. Harley performed a radial dome osteotomy with release of Vickers’ ligament in 26 wrists from 18 patients at a mean age of 13 years. At a mean follow up of 2 years, improvements in wrist supination and extension were seen, as well as improvements in ulnar tilt and lunate subsidence [28]. Steinman reported the long term (11 year) follow up of the above series and concluded that radiographic deformity correction was maintained in addition to a good to excellent functional outcome [29]. In addition to the corrective osteotomy, epiphysiodesis should be considered for whichever of the two bones still has growth potential to limit any further evolution of the deformity.

**Figure 5.** Intraoperative images demonstrating (a) osteotome undertaking radio-carpal ligament release, (b, c) physiolysis of physeal bar, and (d) ulna epiphysiodesis.

**Skeletally Mature Patients; Type 2**

Skeletally mature patients can be further subdivided into deformities with either a single plane (coronal) or with multiplanar (coronal and sagittal) components. With respect to those with a single planar deformity, (i.e. group’s 2A and 2B) the senior author uses thresholds of >5 mm of lunate subsidence and > 40 degrees of radial inclination as a means to differentiate between “mild” and “severe” single planar deformities.

These criteria distinguish those wrists in which the deformity can be adequately corrected by performing a simple ulna shortening procedure,
from those that require a primary osteotomy of the radius.

2A: Coronal Plane Deformity Only
In those patients who only display a coronal deformity, i.e. group 2A, attention should be paid to the degree of both lunate subsidence, and radial inclination. Patients with the milder 2A deformity can be effectively treated with a simple ulna shortening osteotomy. For those patients with ulna sided wrist pain, this procedure has the benefit of reducing impingement and also tensioning the TFCC ligament complex to improve stability [30].

In Bruno et al’s case series, 9 patients undergoing isolated ulna shortening procedures for skeletally mature Madelung’s enjoyed complete resolution of pain and good or excellent functional outcome scores [31]. This finding is supported by an earlier report by Ranawat which included 7 isolated ulna head excisions [32], and by Glard’s subsequent experience with isolated ulna ostetomies of 4 skeletally mature adult wrists [33]. It should be noted that Ranawat reported subsequent ulnar translation of the carpus in all of his 7 patients, although this was not seen in the more recent studies by Bruno or Glard.

2B: Severe Coronal Plane Deformity OR Multiplanar Deformity
Those patients with the more severe single planar deformity will require a radial dome osteotomy (as described for patients in 1C) to correct radial inclination and an additional ulna shortening osteotomy to correct any residual length discrepancy between ulna and radius. Farr et al. reported that >5 mm ulna variance and >22 mm palmar displacement predicted patient selection for an ulna shortening osteotomy at their institution although subsequent patient-centered outcomes were not included in the analysis [26].

Multiplanar deformities require a similar approach, with a degree of extension dialled in to the radial dome ostetotomy to correct the sagittal component of the deformity, followed by ulna shortening ostetotomy as per the severe coronal planar deformity group.

Chronic Madelung’s Deformity: Type 3, Longstanding Deformity and Sequelae
Patients suffering from longstanding Madelung’s deformity present significant management difficulties. Problems usually include degenerative changes and symptoms of pain and stiffness, particularly in relation to the DRUJ. In comparison to types 1 and 2, restoring normal functional anatomy can be even more challenging. Coffey et al performed arthroplasty of the DRUJ in 4 wrists on 3 mature patients using the Schecker prosthesis (APTIS Medical, Louisville, KY, USA) [34]. Three out of four wrists made significant post-operative improvements in the range of supination. Patient reported outcome scores and pain scores also improved.

Another surgical possibility is arthodesis of the DRUJ. The Sauve-Kapandji procedure is a technique involving DRUJ arthodesis and the creation of an ulna pseudoarthrosis [35]. Several studies have employed this technique in the context of longstanding Madelung’s deformity. Angelini et al, in the largest such series, reported improvements in pain for 13 of 15 skeletally mature patients treated with the technique [36]. Recognised patient-reported outcome measures were not included however, and functional results, recorded as single plane improvements in range of motion, were variable. Eid et al treated 13 wrists in 10 skeletally mature individuals with the Sauve-Kapandji procedure for idiopathic Madelung’s deformity, and reported a fall in mean Disabilities of the Arm, Shoulder and Hand (DASH) scores from 41.9 to 18.3 at final follow up (mean 16.2 months) [37].

Lluch reported elimination of pain and instability following the Sauve-Kapandji procedure in a group of 70 patients with a range of wrist conditions, including 3 with Madelung’s deformity, although no subgroup analysis was performed for the Madelung’s patients [38]. A similar study by Giberson-Chen et al also reported significantly improved patient reported outcomes in 57 patients undergoing the procedure, although again the study cohort was mixed, and contained only 3 patients with Madelung’s deformity [39].
Kaempf de Oliveira et al have also recently reported favourable results in one 40 year old patient treated with the Sauve-Kapandji procedure in addition to lunate fossa reconstruction with the healthy distal surface of the degenerate lunate. The patient obtained an excellent functional outcome, with a DASH score of 12, and with complete elimination of pain at 10 year follow up [40].

### Asymptomatic patients with Madelung’s Deformity

There are likely to be patients within the early subgroups that are either entirely asymptomatic or suffer from only very mild symptoms, with no strong indication for surgery. There is no firm evidence demonstrating that prophylactic surgery in these patients is beneficial. Indeed, the prevalence of asymptomatic, untreated Madelung’s deformity in the general population is unknown and could be considerable. Nevertheless, in skeletally immature cases physeal release of the radius and/or epiphysiodesis of the ulna could still be considered as a prudent measure. These are straightforward procedures with relatively low risk and morbidity, and which work to improve normal anatomical relationships around the DRUJ. If the patient is not suitable for either of these procedures, and minimally symptomatic, then we would recommend close observation, with the understanding that these patients may require an osteotomy once skeletally mature.

### Conclusion

A classification system, based on the currently available evidence is presented. The aim is to guide clinicians’ decision making in the management of this challenging problem. The key determinants for classification include the level of skeletal maturity and the severity of the deformity as measured by ulna variance and radial inclination. Surgical strategies in skeletally immature patients rely upon the potential for continued growth and remodelling. In skeletally mature patients the management focuses on corrective osteotomies.

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None

### Funding:

None

### References
