Madelung Deformity: A Case Report

Abstract

This case report presents information about a rare bone deformity of the wrist called Madelung Deformity. It describes the many symptoms and characteristics of the disease as well as its effect on normal physiological function. Also mentioned, are different radiographic methods used in diagnosis of the deformity. The etiology and prognosis are explained as well as the most common and effective ways of treating the deformity. Three case studies are shared to portray the wide variances of this rare deformity.

Introduction

Madelung Deformity (MD) is an uncommon bone deformity occurring in less than 2% of the general population. It was first accurately described by Otto W. Madelung. MD is found in the bones of the wrist and is most common in females, but is not uncommon to also be diagnosed in males. It appears bilaterally 50% of the time. Due to the fact that symptoms don’t usually show up at birth or even during early childhood it is most often diagnosed in mid to late adolescent years; usually between the ages of 8 and 14.

The natural history of this bone deformity is unknown. The etiology of MD is commonly classified into four groups: idiopathic, dysplastic, genetic, and post traumatic. It is assumed that idiopathic MD is any form that does not have a known cause. Dysplastic MD is most commonly linked to dyschondroosteosis, a dominantly inherited skeletal dysplasia that becomes more pronounced during adolescence. Other bone dysplasias that are commonly associated with MD include multiple hereditary osteochondromatosis, Ollier disease, achondroplasia and multiple epiphyseal dysplasias. Genetically, MD can be traced to having association with Mesomelic Dwarfism and a mutation of the X chromosome. "The posttraumatic form of the deformity has been most commonly associated with repetitive trauma or following a single event that disrupts growth of the distal radial ulnar-volar physis." There are other causes of wrist deformity that may mimic MD including sickle-cell disease, infection, tumor and rickets.

Symptoms
MD often progresses from initially mild symptoms to more severe as the individual ages; especially if no intervention takes place. Common symptoms include:

- Pain in wrist/forearm (particularly on the ulnar side)
- Decreased range of motion
- Wrist deformity
- Limitation of pronation, supination, extension and radial deviation
- Decreased strength
- Sensitive to the touch\textsuperscript{1-5,7,8}

### Diagnostic Imaging

A diagnosis of MD is confirmed radiographically by the use of X-ray, Computed Tomography (CT) and Magnetic Resonance Imaging (MRI).\textsuperscript{1-5} X-rays are taken of the wrist and forearm and are usually the first to confirm MD. Most often, both wrists and forearms are x-rayed on the same imaging receptor so that comparisons can be made. Radiographs are extremely important in making pre and post-operational measurements to assist in reconstruction.\textsuperscript{3-4} While X-ray shows detailed bony anatomy, MRI images are extremely helpful in seeing soft tissue anatomy including ligaments, tendons and muscles.\textsuperscript{2} The radioulnar volar ligament is one ligament specifically studied with the use of MRI. CT scans help mainly with pathoanatomy and morphology of the deformity.\textsuperscript{4}

### Radiographic Features of MD

Though MD is known mainly for its symptoms, it can be more specifically described by its many abnormal radiographic features. Many deviations of the anatomy in the region of the distal radius and ulna exist with MD, but two of the most prominent features are a bowing of the distal end of the radius and an extreme prominence of the ulnar head. Radial inclination is often severely increased allowing the carpal bones of the wrist to wedge down into the articulating radioulnar joint.\textsuperscript{1-5} This, in turn, causes an abnormal separation of the radioulnar joint and appearance of a subluxation, or partial dislocation (see Figures 1-3).\textsuperscript{1,4} Often times MD is associated with compromised ligament structure due to tension. This is one of the biggest causes of pain in skeletally immature patients.\textsuperscript{3-4} The abnormalities of the ulna, carpal bones, articular cartilage, ligaments and tendons are secondary to the radial deformity itself.\textsuperscript{2} Along with those
already mentioned, Lamberti\textsuperscript{4} describes several more distinct features of MD including:

- Lateral and dorsal curvature of the radius
- Widened interosseous space
- Premature fusion of the ulnar half of the distal radius
- Focal osteopenia in the area of the ulnar portion of the distal radius
- Exostosis at the distal ulnar border of the radius
- Triangularization of the distal radial epiphysis
- Ulnar and palmar facing distal radial articular surface
- Relative dorsal subluxation of the ulna
- Increased radiodensity of the ulnar head
- Carpal wedging of the lunate at the apex of the wedge
- An arched curvature of the carpal bones in direct continuation of the dorsal bowing of the radius on the lateral radiograph

To compare features of MD with normal wrist features grossly and radiographically (see Figures 4-7)

**Treatment and Management**

*Untreated*

In some instances, MD does not need treated at all. It is not a fatal deformity. If the individual is asymptomatic there is no need for treatment. Even while presenting with the worst of symptoms, a person can still choose to forgo treatment and survive. Treatment for MD is only to rid of symptoms.\textsuperscript{1,4,5}

*Nonoperative*

The severity of MD may vary in every individual. For those who have less severe symptoms, less invasive measures are often taken for treatment. Nonoperative treatments include splinting, nonsteroidal anti-inflammatory drugs, and activity modification.\textsuperscript{6} Splints are commonly used to reduce pain and joint irritability while acting as a stabilization device. Stabilizing the joint allows proper healing and increases strength. Splints contribute to decreasing pain that is caused by damage to the ligaments and soft tissue. Patients may also,
over time, learn to manipulate their manual activity to lessen pain and discomfort.\textsuperscript{4,5}

Nonoperative management is more beneficial to more skeletally mature patients with MD. The younger and more skeletally immature the individual is, the more likely it is that pain is caused by tension within different ligaments due to growth. Splints will not have a satisfactory result. Whereas more skeletally mature patients that have radiocarpal pain or pain within the distal radioulnar joint, splints prove very effective in reducing pain and irritation.\textsuperscript{4}

\textit{Operative}

In contrast, those individuals with more severe symptoms and that are less skeletally mature, operative treatment provides the best improvements.\textsuperscript{5} According to Lamberti\textsuperscript{4}, there are 4 determinants to surgical intervention for MD. These four determinants include 1) the patient’s age and the growth remaining in the distal radius 2) the severity of the deformity 3) the severity of symptoms and 4) the clinical and radiographic findings. Once it is decided that surgery would be an effective solution, an operative treatment must be chosen. They can be divided into 3 areas of focus that include those that are aimed at correction of the primary deformity of the radius, those that attempt compensatory changes to the ulna in hopes to decrease pain and increase range of motion and those that address both.\textsuperscript{3}

There are many operations that address the deformity of the radius. “Conceptually, these can be broken into those that change the growth or anatomy at the physis, those that change the bony anatomy of the metaphysic and those considered salvage-type joint-sacrificing procedures.”\textsuperscript{4}\textsuperscript{(p.1)} Procedures that restore the position of the distal radius include:

- Radial osteotomy
- Distraction histogenesis
- Vicker’s ligament release
- Epiphysiodesis\textsuperscript{4,5,7}

Operative procedures attempting compensatory changes to the ulna are fewer than those of the radius and include:

- Ulnar resection
- Ulnar arthrodesis
- Ulnar shortening osteotomy\textsuperscript{4-7}

Often times, a combination of these surgical approaches are used in order to give the best
restoration to the entire distal radioulnar joint and articulating carpal bones. All procedures are aimed at realigning the position of the joint surface.\textsuperscript{4,7} Laffosse et al\textsuperscript{5} conclude that “Some prefer to wait for skeletal maturity so as to not make existing anomalies worse and to avoid the risk of recurrence. Others suggest operating early to benefit from the remodeling and adaptation potential of the immature skeleton and to prevent the development of irreversible degenerative consequences.”\textsuperscript{5(p.5)}

To ensure proper healing of the bones post surgically, 6-8 weeks of immobilization in a cast is necessary. Also, until skeletally mature, annual follow-up radiographs must be taken to document improvement of the distal radioulnar joint.\textsuperscript{2-3} The length of an individual’s follow up time can be as long as 10 years or more, especially if the patient was a part of a surgical trial.\textsuperscript{3} Some may only last a couple of years depending on the patients age and the severity of the deformity.

**Case Studies**

**Madelung’s Deformity: A Wrist Problem**

Hanel and Krenek\textsuperscript{5} studied a young female patient of 10 years of age who presented with conditions of being short in stature and had odd looking wrists. She did not have any pain at the time, but after being examined, was diagnosed with a skeletal dysplasia called Leri-Weill syndrome and MD of both wrists. Radiographs of her left wrist provided evidence of MD by revealing a severely subluxed ulna and an abnormally bowed distal radius. One year later she was treated with a radial osteotomy and division of the Vicker’s ligament. Volar subluxation of the carpus was corrected and k-wires were used to maintain the position of the corrected radius (see **Figure 1**). A similar procedure was used to treat the right wrist several months later.

This patient was pain free for 2 years after her corrective surgeries. After 2 years she progressively started to feel pain and discomfort along the ulnar side of both wrists. It was especially painful while moving the arm from pronation to supination and vice versa. She complained that the right wrist was more symptomatic than the left. In order to manage pain, she was encouraged to use an immobilization splint for stabilization and anti-inflammatory for pain and swelling. But with activities such as writing, her pain still persisted.

Both wrists were examined once again and both demonstrated prominence of the ulnar heads dorsally. Radiographically, the right wrist presented worse than the left wrist. She
complained of discomfort at the distal radioulnar joint with rotation, especially when this motion was accompanied by holding weighted objects. Radiographs of the wrists showed that the ulna was abnormally longer than the radius by 10mm on the left wrist and 5mm on the right wrist (see Figure 1). Radiographs also indicated other symptoms of MD including:

- Increased volar and ulnar slant of the joint.
- Dorsally subluxed ulna
- Volarly subluxed carpus
- Closed growth plates
- Joint abnormalities of the radius and ulna
- Degeneration of the distal radioulnar joint
- Flattening of the joint on radial and ulnar sides

Because the right wrist showed more severe symptoms, it was recommended to start with treatment on that side first. Advised surgical treatment consisted of an ulnar shortening osteotomy and reconstruction of the distal radioulnar joint. For more ulnar variance, the ulna was shortened by excising the distal 1cm of the ulna. This resulted in less pain and more ulnar variance. “The ulnar head subluxation was addressed by removing the abnormal articular surface and reconstructing the joint by first interposing a spacer of allograft tendon rolled on itself and then using the tails of the graft to reinforce the restraining ligaments of the joint.”

This patient was immobilized in a long arm cast for 6 weeks to ensure proper healing and protection. Unrestricted activities were allowed after the 6 weeks of immobilization. There was no need for further therapy or splinting.

This patient was seen a while after her reconstructive surgery of her right wrist. There was significant improvement. She was not experiencing any pain and had improved function and limitations of her wrist. She was able to perform daily activities with ease and even play musical instruments including the piano. Prolonged writing still caused mild hand pain and limitations of her wrist prevented her from doing physical exercises such as push-ups. Because of the success of the surgery on her right wrist, she opted on having the same surgical procedures performed on her left wrist as well and saw similar results. Radiographs were taken post-operatively at the age of 15 and provided evidence of realignment and stability of the distal radioulnar joint, a resected distal ulna and the allograft tendon filling the space between the radius and ulna (see Figure 1).
Madelung Deformity Diagnosed but Untreated

A 16 year old female presented to local orthopedist with lateral wrist pain of the left wrist after a minor softball injury. She was experiencing significant loss of strength in her wrist. Any lifting of more than 4-5 pounds was extremely painful. She was unable to flex, extend, pronate or supinate her wrist due to pain and tenderness. Hoping to minimize pain until further evaluation could be taken, this patient received a cortisone steroid injection into the wrist joint. As she continued her softball season she saw no evidence that the pain was subsiding. A wrist brace was also utilized but like the steroid injection, had no effect.

When her softball season concluded, this young lady was seen again by her orthopedist for further evaluation. X-rays indicated MD along with an ulnar styloid process fracture. This fracture was from an injury 6 years prior. In regards to the MD diagnosis, radiographs of the right wrist were also obtained for comparison. Her right wrist also showed positive indication of MD. Radiographs of both wrists indicated symptoms of MD. These symptoms included:

- Bowing of the distal radius
- Palmar angulation of distal radius
- Prominence of the ulnar head
- Increase inclination of the articular surface of the radius
- Carpal bones wedging down into distal articulating radioulnar joint

(see Figures 4 and 5)

The orthopedist suggested a corrective radial osteotomy of only the left radius, but did say that surgery might be necessary for the right wrist later on as well. To be completely certain of her diagnosis and options she chose to forgo the surgery and meet with a doctor who specialized in hand and wrist orthopedic surgery at a larger medical facility. With the help of magnetic resonance imaging (MRI), this surgeon was able to confidently agree that this patient did have MD, but found that it was not true source of pain and limitation in regards to the softball injury. The MRI revealed that the source of pain was a torn ligament on the lateral side of the wrist. This patient did decide to have surgery to repair the damaged ligament, and kept with her decision to not intervene surgically to correct her MD malformations. The surgeon mentioned that due to the fact that there were no symptoms of pain, limitation or decreased strength before the softball injury, that her form of MD, though radiographically severe, did not have any significance and therefore would not need corrective surgery.
It has been 5 years since her surgery, and this young lady has not had any symptoms of MD other than the pre-indicated altered bone structure and physical appearance of her left and right wrists (see Figure 7). However, tenderness and sensitivity remain on the lateral side of the left wrist due to surgery that was performed to repair the damaged ligaments.

Madelung Deformity in a Collegiate Gymnast: A Case Report

Wrist pain is extremely common in young gymnasts due to the intense amount of compressive forces exerted on the wrist joints. “Epiphyseal plate changes at the distal radius have been reported in more than 42% of male and female preadolescent and adolescent gymnasts.”¹(p.170) This type of injury usually presents in gymnasts between the ages of 6-13. Changes to the distal radial epiphyseal growth plates have been reported in many athletes that have not fully developed skeletally. In some cases, growth plates have undergone complete premature closure. It is highly possible that these skeletal changes are directly related to the force placed on these joints during gymnastic events. Interestingly, symptoms and findings to these changes mirror those of MD. Due to the fact that these skeletal changes appear to be MD but cannot be proven to have originated from the actual deformity, the nickname Madelung-like deformity has been adopted. Although it has been found that some gymnasts are in fact diagnosed with MD.¹

Brooks¹ performed a study of a 21 year old female complaining of wrist pain in her left wrist after 17 years of competitive gymnastics. Her symptoms included pain, instability and sensitivity to her left wrist. She was first diagnosed with a dorsal capsular wrist sprain, her symptoms being managed 3 times a week consisting of ice and manual joint traction. Her symptoms subsided and treatment was discontinued. Soon after, not being able to recall a particular injury, pain returned during typical gymnastic swings. She was further examined by the team’s athletic trainer. Examination revealed bilateral palmar subluxation with associated radial and ulnar styloid process prominence. She also demonstrated a loss of about 5º pronation and 10º supination compared with the contralateral side.

The athletic trainer advised this young woman to participate in regularly scheduled practices, but not to impact load or grip with her upper extremity. A week later she was referred to an orthopaedist. At this time, her wrist had become extremely swollen and tender to palpation. Treatments for pain and inflammation were instituted. They included ice-bag application, cold
whirlpool immersion, transcutaneous electric nerve stimulation therapy and oral nonsteroidal anti-inflammatory medication.

Radiographs were obtained and did indeed confirm MD. Her radiographs showed a significant widening of the radioulnar joint which had allowed the lunate to move down in between the articulating surface of the distal radius and ulna (see Figure 8). Carpal instability tests were negative. Due to the gross and radiographic appearance of the distal radioulnar joint, the team orthopaedist then referred this gymnast to a hand surgeon. The hand surgeon was in agreement with the orthopaedist and suggested that this young lady be placed in a short arm cast for 3 weeks to stabilize the wrist and allow healing. However, the immobilization made insignificant improvement. Constructive surgery was discussed but denied due to the surgeon’s conclusion that future participation in gymnastics would not cause further injury. If she were to continue gymnastics, however, she would need pain management, stability bracing and symptomatic treatments previously mentioned.

**Conclusion**

MD is a rare bone malformation described by distinct changes/deformities in the distal radius and ulna. Two most common radiographic indications of MD are bowing of the distal radius and a prominently subluxed distal ulna. MD occurs most often in adolescent females. Symptoms of MD vary from mild to severe. Most common symptoms of MD include pain, decreased range of motion, wrist deformity and decreased strength. The severity of symptoms and bony abnormalities dictate whether or not to intervene surgically. Surgical treatments have proven to be effective in corrective reconstruction of MD, but treatment is not always necessary.
References


Figures and Captions

**Figure 1.** Image A represents PA and Lateral views of left wrist of an 11 year old female with Madelung Deformity. The Radius is completely abnormal and bowed. The articulating radioulnar joint is widened. The carpal bones are wedged down into the distal radioulnar joint. Image B represents the post-operative radiographs of the first and unsuccessful surgery of the same individual. Palmar subluxation of the carpus has been corrected and the distal radius is more normal. The K-wires are maintaining the osteotomy in place while it heals. Image C represents the final post-operative radiograph of this patient at the age of 15. It shows the realignment and stability of the distal radioulnar joint, a resected distal ulna and the allograft tendon filling the space between the radius and ulna. Images courtesy of Seattle Children’s Hospital Research Foundation. Hanel D, Krenek L. Madelung’s deformity: a wrist problem. Seattle Children’s Hospital Research Foundation website.http://www.seattlechildrens.org/healthcare-professionals/resources/orthopedics-case-of-the-month/madelungs-deformity-wrist-problem-2-12. Accessed October 24, 2013.
Figure 4. PA radiograph of patient’s wrist with MD. This patient was a 17 year old female. Notice the increased inclination of the articulating surface of the radius, the prominent styloid process and slight bowing of the distal radius. Image courtesy of a local hospital.
Figure 5. Image A represents a PA radiograph of a 17 year old female with MD. You can see the fractured styloid process of the ulna and the inclination of the articulating surface of the distal radius. Image B represents a Lateral radiograph of the same individual. It demonstrates the dorsal subluxation of the ulna. Image courtesy of a local hospital.
Figure 6. Image A represents a PA radiograph of a normal wrist. There is no bowing of the radius, the carpal bones are not being wedged into the distal radioulnar joint, the inclination of the articulating surface of the distal radius is normal. Image B represents a Lateral radiograph of a normal wrist. The radius and ulnar are midline along with the carpals and metacarpals. There is no dorsal subluxation of the ulna. Image courtesy of http://emedicine.medscape.com/article/1260002-overview. Updated July 12, 2012. Accessed October 25, 2013.
Figure 7. Five years ago this patient was diagnosed with MD. The gross appearance shows the prominent subluxation of the distal ulna in both wrists; more prominent in the left (top) wrist. There is a scar present from a surgery that was not related to MD. This patient has not suffered from any pain or limitation due to the deformity since she has been diagnosed; merely just the bony malformation creating a deformed physical appearance.