

Original Research Article

Early outcome in Madelung's and Madelung like deformity of wrist in adolescent group

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ABSTRACT

Background: Madelung deformity is the result of bony and ligamentous dysplasia at the wrist that produces characteristic deformity and is usually recognized in late adolescence. Madelung like deformity can occur following trauma, infection or neoplasm. This esthetic deformity is encountered rarely in orthopaedic practice, when surgeon faces the challenging task to correct it using the best available technique. This study has been conducted to analyze the presentation, interventional procedures and their end result.

Methods: We retrospectively analyze the clinical records, radiographs and functional outcome of patients who presented with Madelung and Madelung like deformity. Radiographic and clinical assessment was done. Surgery was considered for patients fitting into the inclusion criteria. Dorsolateral close wedge and palmar open wedge osteotomy of radius were done with or without ulnar procedure. Postoperative protocol was followed and periodic assessment was done. Mayo modified wrist score has been used to evaluate the pre-operative and post-operative scores.

Results: The study enrolled sixteen affected wrists. Average age in our study was 14 years with an average follow-up of 12 months. Statistically significant increase in range of motion of wrist was achieved for in surgically treated patients. Significant change in pain score, grip strength, RAU^o and carpal slip was noted. Postoperative wrist score was also significantly improved.

Conclusions: Our study concludes that there is a definitive role of surgical intervention to improve the range of motion of wrist, grip strength and reduces pain which brings the patient back into the main stream of a functionally independent being.

Keywords: Madelung deformity, Wrist, Early outcome, Adolescent group

INTRODUCTION

Madelung's deformity is the result of bony and ligamentous dysplasia at the wrist that produces palmar displacement of the hand on a short bowed forearm, which is associated with dorsal subluxation and prominence of the ulnar head.¹ Etiology is certainly unknown and despite being categorized as congenital, it lacks evidence for a defect that is present at birth or in early childhood. Guillaume Dupuytren documented first case in 1834 and later Otto Wilhelm Madelung in 1887

first completely described the condition.² Patients usually present in late adolescence with characteristic wrist deformity. There is an abnormal growth at distal radial physis and an abnormal ligamentous structure that tethers the proximal carpal row in relative flexion with respect to distal articular surface of radius.³ The term "Madelung deformity" is now use to describe a variety of conditions at the wrist in which there is to a greater or lesser extent premature fusion at the distal growth plate. Deformity can also occur following trauma, infection or neoplasm.

Various classifications have been proposed but none has been widely accepted.^{4,7} International federation for societies for surgery of the hand classifies Madelung deformity under generalized abnormalities and syndromes at number 7.⁸ We have used Vander and Watson classification in our study as it is the most informative and comprehensive.⁷ Based on this classification Madelung deformity can be classified as – post-traumatic, dysplastic, genetic and Idiopathic. The first category can be expanded to include others besides trauma like infection or neoplasm. Acquired deformities can be differentiated from congenital by lack of physical findings to suggest either dysplastic or genetic causes and being unilateral and having an appropriate history.⁶

This esthetic deformity is encountered rarely in orthopaedic practice, when surgeon faces the challenging task to correct it using the best possible contemporary techniques.⁹ Most surgeons now accept that many of these patients do have significant and prolonged pain in adolescence and on into adulthood besides the deformity which is a significant problem both for men and women. Should this condition be left alone or operated upon, and if the surgeon plans to operate, what is the right age and method to do so. Keeping in mind the above difficulties and controversies, this study has been conducted to analyze the presentation, interventional procedures and their end results.

METHODS

We retrospectively analyze the clinical records, radiographs and functional outcome of patients who presented with Madelung's and Madelung like deformity during 2010 to 2014. The study enrolled 10 patients with 16 affected wrists. There were 7 girls and 3 boys with age group ranging from 12 years to 19 years with a mean age group of 14 years. Radiographic assessment done using Dannenberg's and Ranawat's roentgenographic criteria.^{10,11} MRI was done for only one affordable patient with wrist pain having idiopathic Madelung's deformity. Clinical assessment of range of motion of wrist, grip strength using spring dynamometer and pain score using visual analogue scale (VAS) was done before starting treatment. Criteria considered for surgery were cosmetic deformity, limitation of motion, pain with activity, dorsal ulnar projection and progressive deformity.¹² Our patients and their families were informed that data from the cases would be submitted for publication. Consent was taken from patients and guardians before starting the treatment.

After taking thorough history and detail clinical examination, selected patients were subjected to surgery. Distal radial corrective osteotomy was done using two different techniques. Dorsal-lateral close wedge osteotomy of radius was done through dorsal longitudinal incision over distal forearm. Radial biplane osteotomy was performed parallel to articular surface of the radius. An appropriate wedge of bone was removed from radial and dorsal aspect of proximal cut end of the radius.

Osteotomy was closed and stabilized with k-wires at 0-15° of volar angulation and 60°-70° of angulation of the articular surface of the radius to the long axis of radius.¹¹ Palmar open wedge osteotomy was done through interval between flexor carpi radialis and the radial artery. Using an osteotome, first cut was made parallel to articular surface of radius. Osteotomy was gently wedged open with the help of laminar spreader and trapezoidal iliac crest graft was placed into the wedge and fixed with k-wires.¹² Volar ligament was released through palmar approach. Ulnar osteotomy was performed according to the age of the patient if necessary.

Patients were given above elbow slab post-operatively and were discharged on third post-operative day. Above elbow cast was given after two weeks post-operatively. At 6 weeks following surgery, union at osteotomy site was checked both clinically and radiologically. If union achieved, below elbow splint was given and intermittent mobilization exercises were taught, otherwise immobilization was continued for another 2 weeks.

Clinical assessment of grip strength, range of motion at wrist and elbow was evaluated at 8 and 12 weeks. Repeat X-ray was done 12 weeks post-operatively. Thereafter, patients were followed at 6 months and 1 year post surgery which includes reassessment of range of motion, pain score, grip strength, radiological assessment. Mayo modified wrist score has been used to evaluate the pre-operative and post-operative score.^{13,14}

Data was compiled at the end of study. Analysis was done using SPSS 11.0 statistical package. Intra-operative differences were analyzed using the Wilcoxon signed rank sum test, whereas inter-operative differences were explored using the Mann-whitney non-parametric test.

RESULTS

The study enrolled total of sixteen wrists. Right side was most commonly affected than left side. All three male patients have Madelung like deformity and no true case of Madelung's deformity was seen in male adolescence. In our study unilateral deformity was seen in two patients, one following traumatic Madelung like deformity and another with idiopathic Madelung deformity. These findings were similar to those described in the literature.^{6,15} Most common deformity was idiopathic and least common was genetic type. Female incidence of deformity was seventy percent as compared to male. Average follow up in our study was 12 months ranging from 6 to 21 months. One patient was lost to follow up.

Most common clinical presentation in our study was ulnar sided wrist pain and deformity. Considering the Dannenberg's radiographic criteria, the most common radiological feature is ulnar and volar angulation of the distal articular surface of radius, dorsal subluxation of ulna and wedging of the carpus. MRI was done in one

affordable patient with complaint of pain in wrist having idiopathic Madelung's deformity.¹⁶ The anomalous ligament was demonstrated 4mm in thickness. Seven wrists were treated conservatively (group A) and nine wrists were considered for surgery (group B). We performed 3biplanar open wedge osteotomies, 4 dorsal biplanar closed wedge osteotomies with ulnar shortening, one dorsal close wedge osteotomies with Darrach's procedure and one dorsal close wedge osteotomies with distal matched ulnar resection. All procedures were done without image intensifier.

In conservatively managed group A, pronation at wrist ranges from 10°-85° with a mean of 60.71°, supination

ranges from 80°-90° with a mean of 87.86° and dorsiflexion ranges from 10°-75° with a mean of 59.29°. Pain score on visual analogue scale ranges from 0-5 with a mean of 0.7 which means no pain, grip strength ranges from 9 kgs to 14 kgs with a mean of 12 kgs, RAU° (radioulnar inclination angle) ranges from 20° to 60° with a mean of 45.43° (normal ≥60°), carpal slip (percentage of contact between radius and lunate) ranges from 10% to 70% with a mean of 50% as shown in Table 1. These were functionally good wrists and were kept under constant observation. These patients may be intervened when the deformity progresses and interferes with daily activities.

Table 1: Baseline data of patients included in group A (conservative).

Movement at wrist	Minimum	Maximum	Mean	Standard deviation
Dorsiflexion	10°	75°	59.29°	23.70
Palmar flexion	60°	95°	76.43°	13.45
Pronation	10°	85°	60.71°	23.88
Supination	80°	90°	87.86°	3.93
Radial deviation	0	34°	13.71°	10.27
Ulnar deviation	0	35°	21.29°	10.64
Variables				
Pain (VAS)	0	5	0.71	1.89
Grip strength (Kg)	9	14	12	2.51
RAU° (*)	20°	60°	45.43°	12.47
Carpal slip (%)	10	70	50	19.14

(*RAU° - Radioulnar inclination angle).

Table 2: Preoperative and postoperative analysis of range of motion in wrist in group B patients (operative).

Range of motion of wrist		Minimum	Maximum	Mean	Standard deviation	P value
Dorsiflexion	Pre-op	20°	70°	45.56°	17.93	0.034
	Post-op	20°	75°	63.89°	16.91	
Palmar-flexion	Pre-op	45°	95°	74.44°	20.22	0.068
	Post-op	40°	85°	55°	13.46	
Pronation	Pre-op	20°	75°	43.33°	16.58	0.021
	Post-op	30°	80°	64.44°	15.29	
Supination	Pre-op	10°	60°	40°	14.14	0.007
	Post-op	30°	90°	80°	20	
Radial deviation	Pre-op	5°	40°	16.44°	12.7	0.589
	Post-op	5°	30°	14.44°	8.81	
Ulnar deviation	Pre-op	5°	40°	25°	11.45	0.017
	Post-op	5°	20°	13.33°	5.59	

(Significant level in comparison – Wilcoxon signed rank sum test).

In surgically treated group B, pre-operative pronation at wrist ranges from 20°-75° with a mean of 43.33°, supination ranges from 10°-60° with a mean of 40° and dorsiflexion ranges from 20°-70° with a mean of 45.56°. Pre-operative pain score on visual analogue scale ranges from 0-8 with a mean of 3, grip strength ranges from 4kgs to 16 kgs with a mean of 4.22 kgs, RAU° ranges from 15° to 45° with a mean of 34.44° and carpal slip ranges from 15% to 60% with a mean of 36.67%. Inter group analysis, showed that there was statistically significant difference between supination of group A and

B with p =0.0001. The difference for RAU° with p =0.012 was also significant. There was a borderline significance for preoperative dorsiflexion and pronation. While no statistical significance was noted for radial deviation, ulnar deviation, pain and carpal slip.

Post-operative analysis of range of motion showed statistically significant increase in dorsiflexion from 20° to 75° with a mean of 63.89° (p =0.034), pronation from 30° to 80° with a mean of 64.44° (p =0.021) and supination from 30° to 90° with a mean of 80° (p =0.007).

The mean post-operative palmar flexion and ulnar deviation has decreased the change in statistical significance. A borderline significance was noted for change in palmar flexion ($p = 0.068$) and no significant change was noted for radial deviation ($p = 0.589$) as given in Table 2. Pain and grip strength showed a significant

difference in preoperative and postoperative values with $p = 0.046$ and 0.07 respectively. RAU° and carpal slip showed a significant change with $p = 0.007$ and 0.008 respectively as shown in Table 3. This corroborated clinically with good correction of the deformity and did not interfere with the functional capacity of the hand.

Table 3: Pre-operative and post-operative analysis of parameters being measured for group B patients showing p values with significant change.

Variables		Minimum	Maximum	Mean	Standard deviation	P value
Pain (VAS)	Pre-op	0	8	3.22	3.19	0.046
	Post-op	0	2	0.67	1	
Grip strength (Kg)	Pre-op	4	16	9.22	4.49	0.007
	Post-op	6	19	13	4.58	
RAU°	Pre-op	15°	45°	34.44°	10.44	0.007
	Post-op	50°	70°	56.22°	6.34	
Carpal slip (%)	Pre-op	15	60	36.67	16.77	0.008
	Post-op	60	80	66.67	7.07	

(Significant level in comparison – Wilcoxon signed rank sum test)

Table 4: Table showing comparison of preoperative and postoperative individual parameters of Mayo modified wrist score.

Modified Mayo wrist score		Minimum	Maximum	Mean	Standard deviation	P value
Pain score	Pre-op	15	25	19.44	5.27	0.035
	Post-op	20	25	24.44	1.66	
Functional status score	Pre-op	0	15	10	7.50	0.006
	Post-op	25	25	25	0	
Pronation & supination score	Pre-op	0	15	6.11	4.16	0.026
	Post-op	5	25	13.89	5.46	
Grip strength score	Pre-op	5	5	5	0	0.006
	Post-op	15	25	18.33	5	
Total score	Pre-op	25	50	40.56	9.16	0.008
	Post-op	65	90	81.67	8.29	

(Significant level in comparison – Wilcoxon signed rank sum test)

All patients had cosmetically better wrist specially three patients who presented with chief complaint of deformity. This implies that reconstructive surgery for Madelung deformity can improve cosmetic appearance and relieves pain. Two patients had slightly persistent protrusion of the ulnar head in pronation. Previously restricted activities of heavy work due to their rural background were also resumed.

In surgically treated group, when intra group analysis was run in between the two procedures done i.e. open wedge in 3 cases and dorsal wedge in 6 cases (irrespective of procedure done to ulna) for the variables analyzed (range of motion, pain score, grip strength, RAU°), it was observed that there was no significant change in the end results because the sample size was small.

Results were analyzed using Mayo modified wrist score with a statistically significant increase in postoperative scores. 4 patients had good outcome, 3 patients were excellent and two patients had fair results.

We also compared the preoperative and postoperative Mayo modified wrist score. P values of individual parameters showed significant changes as in Table 4. The standard deviation of postoperative functional status is 0, as all patients returned to their regular work. Preoperative grip strength has standard deviation of -0 because all patients had grip strength <50% compared to normal.

DISCUSSION

Madelung deformity is four times more common in females, and twice as often bilateral as unilateral.¹⁵ In our study, female incidence of deformity is 70% as compared to males. All the three male patients were with Madelung-like deformity and no true case of Madelung's deformity was seen in male adolescents. Because of difference in degree of severity, complaint is often one sided. Unilateral deformity was found in all patients with traumatic etiology and one patient with idiopathic Madelung deformity. Our findings are similar to those described in the literature.¹⁷

Though the deformity is usually recognized in late adolescence, there is no convincing explanation as to why this should appear in adolescence, as growth of child during this period is far less compared to growth in first three years of life, when the deformity is not seen.¹⁶ Due to lack of awareness among people in our community, patients present to us in late adolescence, with a symptomatic hand and visible cosmetic deformity. In our study, the maximum age of presentation is 19 years and minimum was 12 years with a mean of 14 years. The common presentation in this study has been pain and deformity. Patients present with ulnar sided wrist pain, which increases on loading the wrist, leading to restriction in activities of daily living.

An early surgery buys time for remodelling with mechanical loading across the wrist that may prevent abnormal cartilage overload in the wrist especially in radio-carpal joints, which may later reduce long term osteoarthritis in wrist. Bernhard Schmidt followed a patient with asymptomatic Madelung's deformity for 25 years, and reported increase in volar and ulnar angulation of distal radius articular surface, with development of degenerative changes in the wrist.¹⁸ So, early intervention may prevent or slow down the degenerative changes in a mechanically better wrist.

Radiographic criteria as proposed by Dannenberg et al have been used in the analysis of our patients.¹⁰ Previous studies have shown that the most common radiological criteria is increased dorsal and radial bowing of the radius with distal radial articular surface having exaggerated volar and ulnar tilt.^{12,18,19} They also concluded that there is no correlation between radiological features and complaints of the patient. Also, radiographs had limited value in surgical indication but have a role in planning out the procedure. In our study, radiological features reported were, ulnar and volar angulation of distal radial articular surface, decreased length of radius, triangularization of radial epiphysis, dorsal subluxation of ulna and wedging of carpus. Two patients with Leri Weill syndrome, presented with dyschondrosteotic features of thick and bowed proximal radius, dorsal subluxation and tilt of radial head.^{15,20}

Vickers and Neilson described a thick fibrous structure that begins on the ulno-volar metaphyseal region of the radius and attaches to lunate and triangular fibrocartilage.¹ MRI helps in demonstrating this anomalous ligament. MRI was done in one affordable patient with complain of pain in the wrist having idiopathic Madelung's deformity.

Earlier surgeries were performed for relief of pain and painful limitations of wrist, but towards the end of last century, a trend towards accepting the deformity as a primary indication for surgery has been established.¹¹ Murphy et al outlined the criteria for surgery including cosmetic deformity, limitations of wrist supination, pain with activity, dorsal ulnar projection and progressive

deformity. These criteria for surgery were taken up in our study.

In our study we performed 3 biplanar open wedge osteotomy, 4 dorsal biplanar close wedge osteotomy with ulnar shortening, 1 dorsal close wedge osteotomy with Darrach's and one dorsal close wedge osteotomy with distal matched ulnar resection. We found dorsal close wedge osteotomy to be easier to perform with better control of the fragments during fixation with K-wires, as compared to open wedge osteotomy, where precisely contoured trapezoidal iliac crest graft and fixation of osteotomy with K-wires requires technical skills. Volar approach has the advantage of excision of anomalous ligament and better cosmetically acceptable scar.

In our study, postoperative analysis of group B (operative) showed statistically significant improvement in range of motion, pain, grip strength, RAU^o and carpal slip. Postoperative decrease in radial deviation, ulnar deviation and palmar flexion was observed. This corroborated clinically with good correction of the deformity and did not interfere with the functional capacity of the hand. All the patients had cosmetically better looking wrist and reported significant reduction in pain, especially in patients who presented to us with complaints of deformity. Two patients had slight persistent protrusion of ulnar head in pronation. The increase in grip strength seen in our patients may have been due to improvement in biomechanics and positioning of the wrist, resulting in good tendon excursion and also due to significant pain relief seen postoperatively.

Our results corroborated with dos Reis FB et al, who in a prospective study of 18 patients showed statistically significant improvement in grip strength, range of motion and pain in 80% of the patients and 88% were satisfied with the appearance of the hand.²¹ Nielsen JB, showed improvement in pain, subjective function and cosmetic appearance, however, range of motion improved only in one patient.¹⁷ Ranawat et al, used Darrach's procedure with or without radial osteotomy in 13 patients and noted improved range of motion and grip strength.¹¹

Malalignment of the wrist is manifested by increased ulnar inclination altering the carpal kinematics, dynamic overload of the ulnar column of the radius and decrease lunate coverage, leading to a pre-arthrotic wrist. Restoration of proper distal radial orientation by corrective osteotomy restores a normal physiologic range of motion and revert back adaptive carpal changes to normal. Surgeries that do not correct the alignment of the wrist are rarely associated with satisfied patients.

In our study, patients in group A (conservative) had functionally good range of motion in wrist with mild deformity and were kept under observation as recommended in literature. Bruno et al reported patients with Madelung's deformity, who didn't become

symptomatic until adulthood and were treated with ulnar reduction osteotomy alone. They concluded that radial osteotomy may be unnecessary for pain relief and functional improvement.²² We opine that these cases should be constantly observed and intervened only when the deformity is progressive or affecting day to day activities.

CONCLUSION

A malaligned wrist alters the carpal kinematics and increases the dynamic overload over the radio-carpal joint which over the years causes early osteoarthritic changes. Do early surgical intervention of these patients helps in reversing the changes? For this statement to be truly affirmative, we require a longer follow up. But, this study does allow us to conclude that there is a definitive role of surgical intervention to improve the range of motion of wrist, grip strength and reduces pain which brings the patient back in to the main stream of a functionally independent being.

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