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Madelung Deformity: Surgical Correction with Radial Dome Osteotomy

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The data analysis suggested that, in all the cases of this study, the technique described led to pain relief, better wrist function and better aesthetics. The Radiographic outcome showed an improvement in all pathologic criteria.

This study showed promising results for the application of this specific surgical technique.

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I. INTRODUCTION

Madelung deformity is a rare wrist malformation consisting of excessive radial and palmar angulation of the distal radius caused by a growth disorder of the palmar and ulnar part of the distal radius physis. This leads to shortening and angulation (recurvatio) of the epiphysis, causing a pathognomonic inclination of articular surface palmarly and ulnarly.

Shortening of the radius may cause the development of progressive incongruity at the DRUJ and a positive ulnar variance. This carries to a dorsal subluxation of the distal ulna, causing the altered aspect of the wrist with the prominent ulna.

Step-by-step alterations of the distal radius and the DRUJ are important contributor factors of the so-called carpus pyramidalization, a progressive increase of carpus convexity, and of the volar carpus subluxation.

Other anomalies described are:

- Vickers ligament or radio-lunate ligament. It is an accessory volar ligament hypertrophic connecting distal radius metaphysis, TFCC, and volar surface of lunate. It is responsible for a compressive injury at the ulnar side of the distal radius physis and a proximal traction of the lunate, determining its collapse between radius and ulna¹⁻².
- Volar radio-triquetral ligament. It may be present dorsally to Vickers ligament. It is characterized by greater dimension and distal insertion to triquetrum³⁻⁵.
- Anomal insertion of pronator quadratus muscle⁶ and presence of accessory muscles.

Madelung deformity is a rare condition, the incidence is unknown, and there is no described racial predominance.

Usually, patients come to medical observation between 8 and 14 years old, although there are in literature cases of Madelung deformity at birth or childhood. It is more common in females, the M:F rate of the disorder is 1:4. Madelung deformity can be bilateral about in a third of patients. It may be associated with Léri-Weill dischondrosteosis⁷.

There are several types of Madelung deformity⁸:

- Dysplastic type is the more common. It is associated to Léri-Weill dischondrosteosis;
- Idiopathic;
- Genetic or chromosomal. It is associated to genetic pathologies such as Turner syndrome, Hurler syndrome (mucopolysaccharidosis type 1H), mucopolysaccharidosis type IV and VI, achondroplasia, hereditary multiple exostoses syndrome, hereditary osteo-onychodysplasia, pseudohypoparathyroidism type 1a-1b and Ollier disease⁷.
- Post-traumatic. It is a rare form described in gymnastics athletes, and in persons subjected to repetitive microtraumas to the wrist. It may be secondary to infections and osteomyelitis of the distal radial physis.

These types are then subclassified according to the presence or absence of Vickers ligament. The idiopathic and dysplastic may occur in association or not with V. ligament. Therefore, we can classify Madelung deformity as true Madelung deformity in the presence of

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Vickers ligament and Madelung-like deformity if Vickers ligament is absent.

Clinically, patients complain of movement-related pain, altered aspect of the wrist, functional limitations especially in prono-supination.

In literature several therapeutic options are reported⁹⁻¹¹, but because of the limited number of cases, due to low incidence, it is difficult to determine the efficacy of proposed treatment strategies.

- Wait & see for asymptomatic or paucisymptomatic cases;
- Surgery:
 - Surgical correction of the radius: physiolysis with or without release of Vickers ligament; opening or closed wedge osteotomies; reverse wedge osteotomy;
 - Surgical correction of the ulna: epiphysiodesis; shortening osteotomy, Darrach's technique; Sauvè-Kapandji technique;
 - Combined surgery on radius and ulna;
 - Release of Vickers ligament

Harley et al.¹²⁻¹³ in 2006 and then in 2013 described a surgical correction with dome osteotomy of the distal radius. The technique described provides a dome osteotomy with distal concavity for biplanar

correction of deformity, release of Vickers ligament and arthrodesis o shortening osteotomy of the ulna.

In the present study, we report our experience in surgical correction of Madelung deformity with the release of Vickers ligament, distal convexity dome osteotomy of the distal radius, and reverse wedge.

II. MATERIALS AND METHODS

Between 2017 and 2019, four patients (2 males and 2 females) affected by Madelung deformity were treated in our clinic.

Three adolescents aged between 12-14 yearsold and a 30-year-old adult. Three patients had a bilateral Madelung deformity with a mild deformity in the untreated limb (tab.1).

For preoperative planning (fig. 1) we acquired plain radiographs (AP, LL, and oblique projections), MRI to evaluate surrounding soft tissue (in particular presence/absence of Vickers ligament), and CT scans for better analysis of the deformity.

CT scan data were sent to a workstation in standard DICOM format (PACS, Carestream) to create 3D models of the affected wrist. For all patients polyamide templates of the deformed wrists (scale 1:1) were manufactured using a 3D-printer.

Table 1: Sample

Pz	Age	Bilateral	Treated limb	Follow-up (months)	Form	Vickers ligament
1	3	yes	right	15	Idiopathic	Yes
2	12	yes	right	26	Genetic: Léry-Weill	Yes
3	13	yes	left	16	Idiopathic	No
4	13	no	right	28	Idiopathic	No



Picture 1: Pre-op planning. A: X-ray; B: MRI with Vickers Ligament (*). C: CT selection; D: 3D printing model for pre-op planning

Use of 3D-printed polyamide template in preoperative planning allowed to evaluate rotational center of deformity, plan the osteotomy, evaluate needed degrees for triplanar correction, choose implants and simulate preoperatively surgical procedure.

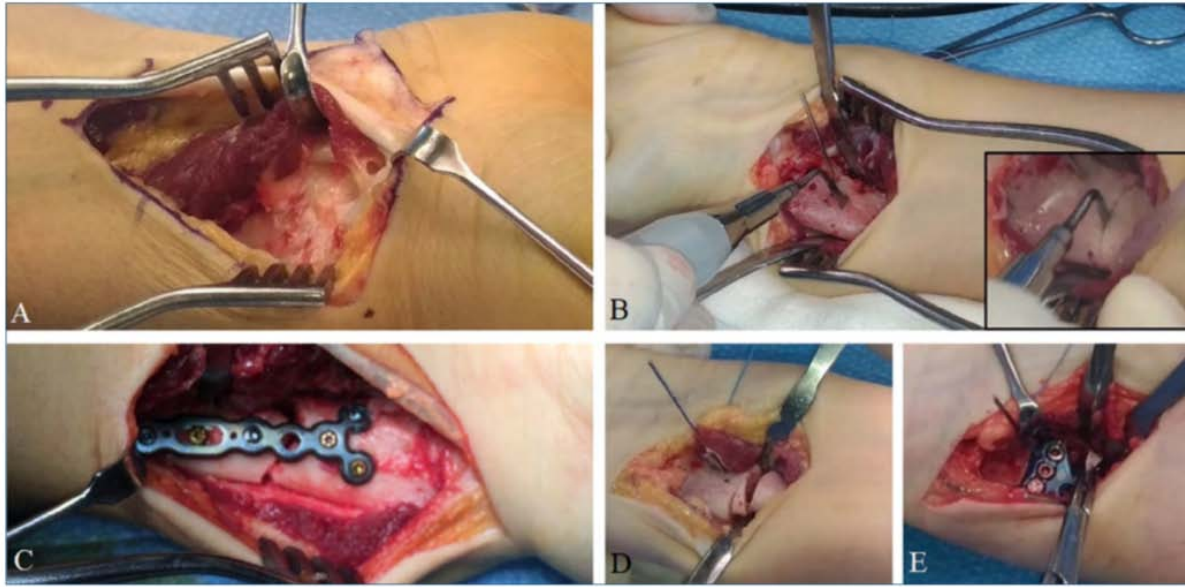
Preoperative planning using 3D virtual planning and 3D-printed templates improves accuracy of surgical correction with dome osteotomy and reduces surgical duration.

It was used a Henry approach, modified according to Orbay, for all the patients in this serie. Release of Vickers ligament in cases where it was present. After identification of rotational center and level of osteotomy, such as preoperative planning, using a piezoelectric saw (piezosurgery), it was possible to

improve the accuracy of the osteotomy and the biplanar correction of the deformity with no risk of thermal necrosis.

In 2 cases it was also needed a bone wedge excision of the radial column for better correction of the deformity on the frontal plane. Then the excised bone fragment was used as auto-graft to fill the gap of dome osteotomy (reverse wedge), allowing faster consolidation and improving stability.

Low-profile mouldable plates were used for fixation because of better adaptability rather than anatomical pre-contoured. At least three screws proximal and three distal to osteotomy were used to fix the plate, avoiding physis and DRUJ.



Picture 2: Sequel of the surgical steps (different patients) A: Release of the V.L. B: Intra-op osteotomy with piezosurgery. C: Correction and synthesis with a volar plate. D: Cut and remove of the bony wedge from the lateral part of the radius. E: Reverse and positioning of the bony wedge in the ulnar side of the osteotomy site.

In one case it was noted the absence of the pronator quadratus associated with the presence of palmaris profundus muscle. The latter anomaly was already described in literature¹⁴.

After surgery, immobilization of the wrist for 3 weeks in a thermoplastic splint was applied to all patients. After this period of immobilization, the wrist and forearm rehabilitation protocol were started for about 6 weeks under the supervision of a hand therapist.

Between 8 and 12 months postoperative plates were removed in order to low the incidence of complication such as tendinitis, infections, and loosening.

The mean follow-up duration was 21 months (range 15-28 months). During this period patients were evaluated like proposed by Peymani et al.¹⁵ in his review. Radiographically McCarroll criteria¹⁶ were used

to judge correction, we evaluated ROM clinically, grip strength via Jamar dynamometer. DASH questionnaires were administered to evaluate functional outcomes, NRS to quantify subjective pain. Finally, the aesthetic defects were estimated using a section of the Michigan Hand Outcome Questionnaire.

These data were related to collected data before surgery.

III. RESULTS

Post-operative radiographs, according to McCarroll criteria, showed a reduction of ulnar tilt, lunate fossa angle, and palmar tilt; whereas no significant difference of palmar carpal displacement was found (Table2).

Table 2: Radiographic McCarrollcriteria

Pz	UT	LS	LFA	PCD	PT
1	*48,7°	*21 mm	*50,8°	*27 mm	*27,8°
	**38°	**9 mm	**33,1°	**27 mm	**18,1°
2	*60,3°	*3 mm	*56,6°	*20,5 mm	*21,3°
	**41,7°	**4,95 mm	**45°	**24 mm	**16,6°
3	*45°	*6,82 mm	*48,3°	*22 mm	*52,6°
	**31°	**6,63 mm	**33,9°	**22 mm	**41,1°
4	*43,6°	*12 mm	*51,3°	*31 mm	*21,5°
	**41°	**6 mm	**47,8°	**27mm	**17,4°

UT = ulnar tilt; LS = lunate subsidence; LFA = lunate fossa angle; PCD = palmar Carpal displacement; PT = palmar tilt.

*preoperative; ** postoperative.

Overall pain reduction after surgery was 4,75/10. Post-operative aROM and pROM were improved near to normal values. There was no

significant difference in postoperative grip strength values comparing the mean value of 3 consecutive tests (Table3).

Table 3: Grip strength, Pain, ROM

Pz	Grip strength	Pain (NRS)	Range of motion					
			F	E	UD	RD	S	P
1	*38 kg	*7/10	*80°	*40°	*30°	*15°	*70°	*70°
	**37 kg	**1/10	**90°	**70°	**40°	**>20°	**>90°	**70°
2	*27 kg	*7/10	*90°	*65°	*40°	*5°	*55°	*80°
	**27 kg	**5/10	**>90°	**80°	**40°	**>20°	**>90°	**90°
3	*21 kg	*5/10	*90°	*80°	*40°	*10°	*70°	*85°
	**19 kg	**0/10	**>90°	**80°	**40°	**>20°	**90°	**90°
4	*34 kg	*6/10	*90°	*70°	*>40°	*10°	*65°	*90°
	**37 kg	**0/10	**>90	**75°	**>40°	**>20°	**>90°	**>90°

F=FLEXION; E=EXTENSION; UD=ULNAR DEVIATION ; RD=RADIAL DEVIATION ; S=SUPINATION ; P=PRONATION
*preoperative; ** postoperative.

There were significant improvements in functional outcomes after surgical correction as suggested by DASH scores, mean score 13/100. It has to be considered that only the adult patient answered

the work-related section of the DASH questionnaire. The MHOQ scores were improved in all patients on average 30/100 (Table 4).

Table 4: DASH and Michigan Hand Outcome Questionnaire (MHOQ)

Pz	DASH	MHOQ
1	*19,4/100	*50/100
	**11,1/100	**75/100
2	*51,62/100	*62,5/100
	**45/100	**50/100
3	*22,4/100	*68,75/100
	**7,76/100	**93,75/100
4	*34,17/100	*18,75/100
	**11,16/100	**43,75/100

IV. DISCUSSION

The purpose of surgical correction in patients affected by Madelung deformity is to relieve pain and improve wrist functions supporting the lunate.

The pain was relieved in all patients, 2 of them were totally pain-free. Analysis of postoperative ROM, grip strength via Jamar dynamometer, and the DASH scores showed good results. Improved MHOQ scores suggested good aesthetic correction.

Corrective dome osteotomy was evaluated on post-operative radiographs, analyzing the degree of correction according to McCarroll criteria. There was a reduction of pathological angles obtaining in some cases a value close to physiological. However, the aim of surgery is not to restore radius but is to improve wrist functions, avoiding lunate collapse. Therefore, it is important that surgery leads to correction of ulnar tilt, lunate fossa angle, and palmar tilt. In our study, we obtained correction of all these parameters.

There was no difference in palmar carpal displacement values between pre and post-surgical correction. PCD measures the palmar-directed

displacement of the carpus (represented by the lunate and capitate) relative to the longitudinal axis of the ulna in the lateral view. There was no correction of PCD values because it was not made any surgical correction of the ulna in contrast with the surgical procedure described by Harley et al. The choice not to modify the deviation of the ulnar longitudinal axis surgically was made so as not to interfere with prono-supination movements since patients showed good ROM pre-operatively. The range of motion may worsen if we modify the delicate equilibrium made by the gradual dysmorphic growth of articular surfaces aiming topophysiological anatomy that is not present in DRUJ.

Eventual surgical correction of the ulna may be considered at the end of the skeletal growth (predominantly for aesthetic reasons).

Some limits to the McCarroll criteria, since x-ray projections are operator dependent, is to always obtain perfect AP and LL x-ray projections. This limit may be overcome using a preoperative CT scan that are improved with 3D reconstructions and/or MRI.

Compared to other techniques reported in the literature for the surgical correction of Madelung

deformities, our technique appeared to be less invasive with shorter recovery time and improved patient's quality of life.

McCarroll and James¹⁷ described a combined technique that includes osteotomies of radius and ulna (very distal radius osteotomy) through a dorsal approach. The same authors highlight an important limitation of the surgical technique in pediatric patients because fixation has to be made very close to the physal growth plate.

Our technique allows to extend the indications to very young patients because the dome osteotomy and the fixation are made more distant to the physal growth plate.

Another limit showed by McCarroll and James is the development of a post-surgical DRUJ instability, often asymptomatic. This is another contributing factor for our choice not to correct surgically the ulna.

Dome osteotomy showed some common points with cylindrical corrective osteotomy for Madelung deformity proposed by Imai et al.¹⁸. Both techniques agree to the fundamental use of CT scan and the case report used a dedicated software for the preoperative planning, for the line of osteotomy, and to create custom-made cutting guides.

Custom-made cutting guide improve surgical accuracy to the preoperative planning, but they limit ulnar column lengthening, using bone wedge excised from radial column osteotomy (reverse wedge).

The use of a pre-op 3D printed model improves surgeons' evaluation of patient-specific anatomy and pathology by way of tactile and visual experience¹⁹. Furthermore, this technology supports the surgeon by selecting the most adequate device for osteosynthesis and helps patients and parents to understand the surgical procedure they will undergo.²⁰

V. CONCLUSIONS

The experience with our patients (even if not statistically significant) showed encouraging results in the use of radial dome osteotomy for the correction of Madelung's deformity.

Our experience suggests that the use of 3D printed bone models in preoperative planning improves accuracy on surgical procedure and on the choice of implants.

In the near future, thanks to a larger sample, our effort will be to standardize this technique as much as possible, providing precise and adaptable indications to each individual case in order to improve the preoperative planning aiming to achieve even better surgical results.

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