A Rare case of "Reverse" madelung deformity treated with

Radial osteotomy and lengthening.

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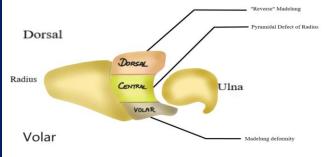
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Introduction

LOCATION OF DYSCHONDROGENETIC LESIONS



MADELUNG DEFORMITY: SURGICAL PROPHYLAXIS (PHYSIOLYSIS) DURING THE LATE GROWTH PERIOD BY RESECTION OF THE DYSCHONDROSTEOSIS LESION

D. VICKERS and G. NIELSEN

From the Royal Children's Hospital, Brisbane, Australia

The majority of cases of Madelung deformity are caused by hereditary dyschondrosteosis at the wrist. The principal lesion in the uhar zone of the distal radial physis retards growth asymmetrically, especially in late childhood. Resection of this zone and its replacement with autologous fat (Langenskiöld procedure, or physiolysis) restores growth and minimizes deformity. The resection of an abnormal ligament tethering the lunate proximally may assist carpal advancement. A series of 17 patients (24 wrists) treated over a 12-year period is presented, with sufficient follow-up for evaluation of 11 patients (15 wrists). The results of this prophylactic procedure are encouraging, and, if it is performed early, the authors believe that Madelung deformity may be preventable, or at least controllable.

Journal of Hand Surgery (British Volume, 1992) 17B: 401-407

Madelung deformity is an Idiopathic condition of distal radius due to premature closure of the medial column and volar aspect of the distal radial physis.

It includes a spectrum of clinic-radiological presentation starting from

- Classical Madelung deformity,
- primary isolated MD (PI-MD),
- MD associated to Leri-Weill's dyschondrosteosis (LWD-MD), and other
- "Madelung-like" deformities" or "pseudo-MD" including post-traumatic and post-infective forms, forms associated with Turner's syndrome, multiple hereditary exostoses, and Ollier disease.

Here we report a rare variant known as "Reverse madelung" characterized by

1. Dorsal tilt of the distal radius

2.Dorsal shift of the carpus

3. Dorso-ulnar arrest of radial epiphysis.

The Madelung deformities in this series were all of primary origin (dyschondrosteosis) and the skeleton was immature at the time of surgery. In no case was a repeat of the surgical physiolysis necessary. There were 17 patients, seven of whom had surgery on both wrists, giving a total of 24 operations. One reverse Madelung deformity is included. There were 15 female and two

Case Report

A 10 year old right-hand dominant girl • progressive deformity of left wrist and forearm presented to deformity clinic at Government Medical College, Kozhikode.

•No history of trauma or other joint involvement.

Clinical examination:

•Shortened radius.

•Mannus varus.

•Increased prominanace of ulnar styloid.

•Terminal supination was restricted.

<u>Radiograph :</u>

•Widening of interroseous space.

•Abnormal radial epiphysis with negative radial height (3.5cm).

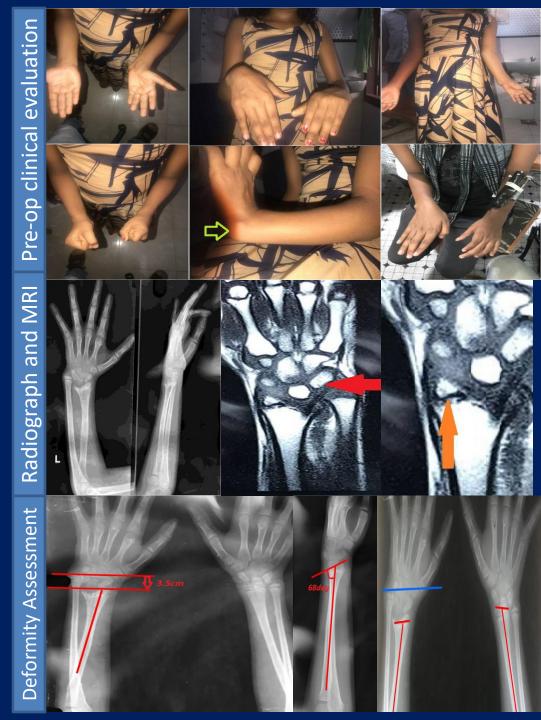
•Dorsal tilt(22degrees).

•Dorsal shift of the carpus.

Triangulation of carpus

MRI:

- 1. Defect in dorso-ulnar part of radial epiphysis .
- 2. Abnormal band of hypertrophied Radiotriquetral ligament.(1.2cm thickness)(Red)
- 3. Lateral half of radial epiphysis is normal (Orange)



MANAGEMENT





Post-op \rightarrow 3weeks



Post-op \rightarrow 8weeks

Post-op \rightarrow 3months

A TWO STAGE PROCEDURE WAS PLANNED.
STAGE 1-RADIO-TRIQUETRAL LIGAMENT
STAGE 2-RADIAL OSTEOTOMY AND LENGTHENING WAS DONE USING LRS(LIMB RECONSTRUCTION SYSTEM).

ATI

•2 years after LRS removal
•Residual dorsal angulation persisting.
•Planned for a distal radial osteotomy and correction in the future

Clinical outcome of the patient at 2 years post-op





Discussion

REVERSE-MADELUNG VS **PHYSEAL INJURY** For For H/o trauma Hypertrophied Radiotriquetral ligament \succ Shortening without bowing. \triangleright Dorsal angulation ,radial shortening and involvement of only ulnar half of radial epiphysis. Widened interrosseous space \geq \triangleright Triangulation or pyramidalisation of carpus. Against Against Absence of posterior bowing. \geq Normal radial half of epiphysis.

Absence of classical vicker's ligament.

>

Absence of physeal bar on MRI.

Conclusion

 \triangleright

1. Reverse Madelung is a rare variant with dorso-radial dyschondrosteotic lesion with abnormal radiolunate or radio-triquetral ligament.

2. Madelung and its variants maybe confused with growth arrest due to physeal injury

3.Management is best done by step wise manner with correction of LLD and dorsal angulation sequentially.

4. Radial osteotomy and lengthening using a LRS is an effective option in such cases.