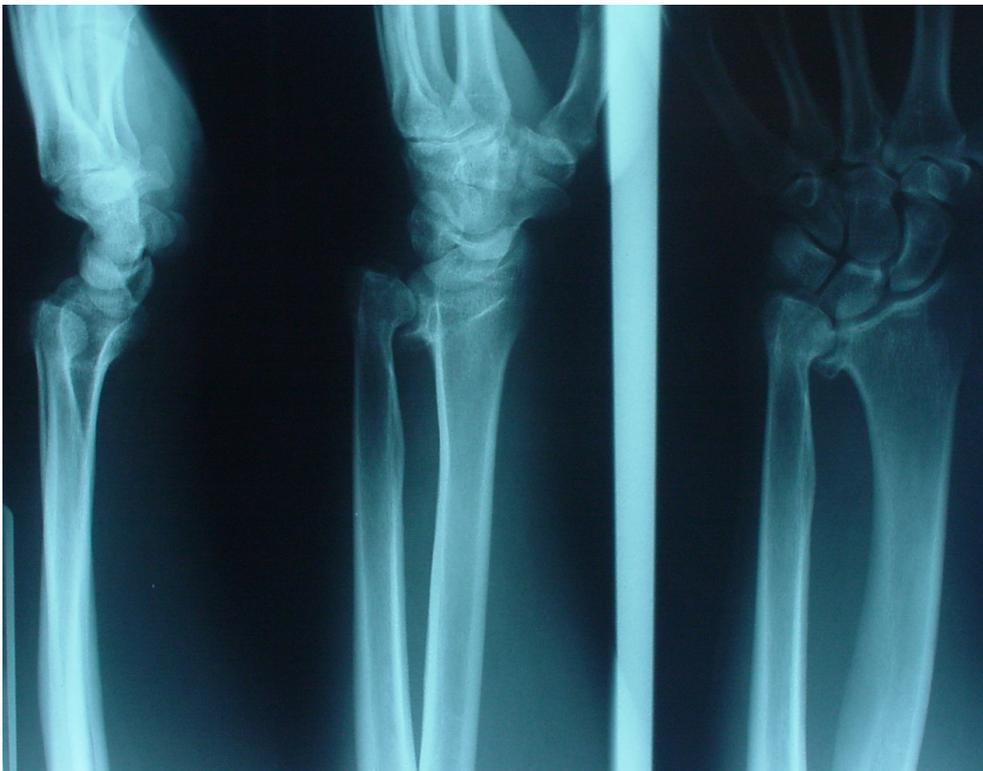


Case 7: A 14-year-old girl with a history of coarctation of the aorta and horseshoe kidney, presented with painful wrist movements.

She was also noted to have a webbed neck and shield chest. Wrist X-rays are shown.

What is this appearance of the distal radius and ulna known as? What are the other possible causes?



Diagnosis Madelung deformity'

The Madelung deformity was first described in 1878 by **Otto Wilhelm Madelung**

Radiographs: - Increase in the normal palmar and ulnar inclination of distal radius.

Triangular epiphysis

Early fusion of the ulnar epiphysis

- Carpus translate palmar and ulnar wards
Lunate is gradually forced to the apex of the V-shaped radioulnaocarpal joint

Pathology Epiphyseal arrest on ulnar and volar half of the distal radius, the articular surface of radius is directed ulnarward & volarward
Leads to shortening of the radius and relative overgrowth of the ulna.

Associated conditions: - generalized dysplasia (AD)

Cause: not known

It is bilateral in two thirds of patients;

AD variable expressivity; females (4:1)

Its incidence is 1:3000 to 5000 live births.

Does not become manifest until late childhood

Bowing of distal radius

Ulna-carpal impingement

Inferior RU dislocation

Wrist motion, particularly extension and supination, is limited.

Pathology

The common mechanism for all causes of Madelung deformity is due to partial closure, or failure of development of the ulnar side of the distal radial growth plate. The underlying cause of this is unclear, with possibilities including

- trauma
- infection
- Dysplasia

Associations

- Leri-Weill syndrome an autosomal dominant dyschondrosteosis
- Turner syndrome
- gonadal dysgenesis
- nail-patella syndrome ⁷
- diaphyseal aclasis (hereditary multiple exostosis)
- Hurler mucopolysaccharidosis ¹
- achondroplasia ¹
- Ollier disease ¹
-

Presentation is with deformity, decreased grip strength and often with pain in the wrist relating to ulnocarpal impaction.

Associated

coarctation, aortic stenosis, horseshoe kidney, primary amenorrhoea, short stature, webbed neck, shield-shaped chest and widely spaced nipples.

Treatment

1. Non Operative Treatment: If patient is asymptomatic;
2. Seen before growth spurt: epiphysiolysis
3. Seen at 12-15 years:
Darrach procedure has provided excellent relief of symptoms, but carries with it the problem of ulnar translation of the carpus;
4. Osteotomy of the distal radius - **Ulnar shortening**

5. SKW: Resection of the ulna proximal to the radioulnar joint synostosis.
6. Arthrodesis considered if the carpus subluxates off the radius;