



# Madelung's Deformity



Fig. 1 : Patient's wrist showing the characteristic bony deformity with "Bayonet-like appearance" (Madelung's deformity).

A 21-yr old lady presented with primary amenorrhea and short stature for evaluation. She had a wide carrying angle (Cubitus valgus) and a typical deformity at the wrist joint (Fig. 1) which was evaluated. There were no other dysmorphic features. She was found to have high serum FSH and low serum estradiol with streak gonads on ultrasound. Karyotype was 45 XO. She was diagnosed to have Turner's syndrome and the deformity of the wrist is typical of this condition known as *Madelung's deformity* (*Bayonet deformity*). It is a skeletal deformity of the wrist characterized by acute angular configuration and palmar displacement of the proximal row of carpal bones leading to the hand appearing like a 'rifle bayonet'. Its pathogenesis is linked to the haploinsufficiency of the SHOX (short stature homeobox) gene.<sup>1,2</sup> It is associated with Turner's syndrome (10% of cases)<sup>2</sup> and Chondrodysplasia of the distal radial epiphysis, or Leri-Layani-Weill syndrome (dyschondrosteosis).<sup>2,3</sup>

**J Muthukrishnan\*, A Verma\*, KD Modi\*\***

\*Senior Resident, \*\*Senior Consultant and Head, Dept of Endocrinology, Medwin Hospitals, Hyderabad-500001.

Received : 26.6.2006; Accepted : 28.9.2006

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