Pediatr Radiol (2015) 45:1856–1863
DOI 10.1007/s00247-015-3390-0

PICTORIAL ESSAY

Madelung deformity and Madelung-type deformities: a review of the clinical and radiological characteristics

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Received: 29 December 2014 / Revised: 28 March 2015 / Accepted: 8 May 2015 / Published online: 2 July 2015

Abstract Madelung deformity of the distal radius results from premature closure of the medial volar aspect of the distal radial physis, leading to increased volar tilt and increased inclination of the radial articular surface, triangulation of the carpus with proximal migration of the lunate and dorsal displacement of the distal ulna. The deformity is particularly common in Leri-Weill dyschondrosteosis, but it may also occur in isolation. True Madelung deformity can be differentiated from Madelung-type deformities by the presence of an anomalous radiolunate ligament (Vickers ligament). In this article, we will review the imaging characteristics of true Madelung deformity, including the common "distal radius" variant, the less common "entire radius" variant and "reverse" Madelung deformity. We will discuss the role of the Vickers ligament in disease pathogenesis and its use in differentiating true Madelung deformity from Madelung-type deformities arising from trauma or multiple hereditary exostoses. Surgical management of these patients will also be addressed.

Keywords Children · Leri-Weill dyschondrosteosis · Madelung deformity · Multiple exostoses · Physis · Radius · Trauma · Vickers ligament · Wrist

Introduction

In 1878, the German surgeon Otto Madelung described a painful deformity of the wrist developing in adolescents between the ages of 8 and 14 years. Initially asymptomatic, patients often went on to develop pain, loss of grip strength and reduced mobility [1–5]. Both Madelung and Madelung-type deformities stem from premature closure of the volar medial portion of the radial physeal plate, resulting in progressive curvature of the radius, increased volar tilt and inclination of the radial articular surface, and proximal migration of the lunate [6–9]. Associated abnormalities of the ulna, carpal bones, articular cartilage and tendons arise from radial growth failure and resulting deformity. Congenital Madelung deformities are characterized by the presence of an anomalous volar radiolunate (Vickers) ligament. While some reserve the term true Madelung deformity to denote those cases arising in patients with Leri-Weill dyschondrosteosis where it is classically bilateral and symmetrical, this category is generally also extended to include isolated idiopathic cases in which a Vickers ligament is present [2, 3, 10–13]. While the exact incidence is unknown, Madelung deformity demonstrates a 4:1 female predominance and increased prevalence among patients with Leri-Weill dyschondrosteosis. Madelung-type deformities may occur in the setting of isolated or repetitive trauma, Turner syndrome, multiple hereditary exostoses, and Ollier disease when there is an interruption of the medial volar aspect of the distal radial physis [10–12, 14, 15].

In this pictorial essay, we will describe the imaging approach and pertinent findings in Madelung deformity and Madelung-type deformities.

Overview

Patients with Madelung and Madelung-type deformities present with a prominent, dorsally displaced distal ulna, often referred to as a "bayonet" deformity (Fig. 1). Idiopathic cases as well as those occurring in association with Leri-Weill

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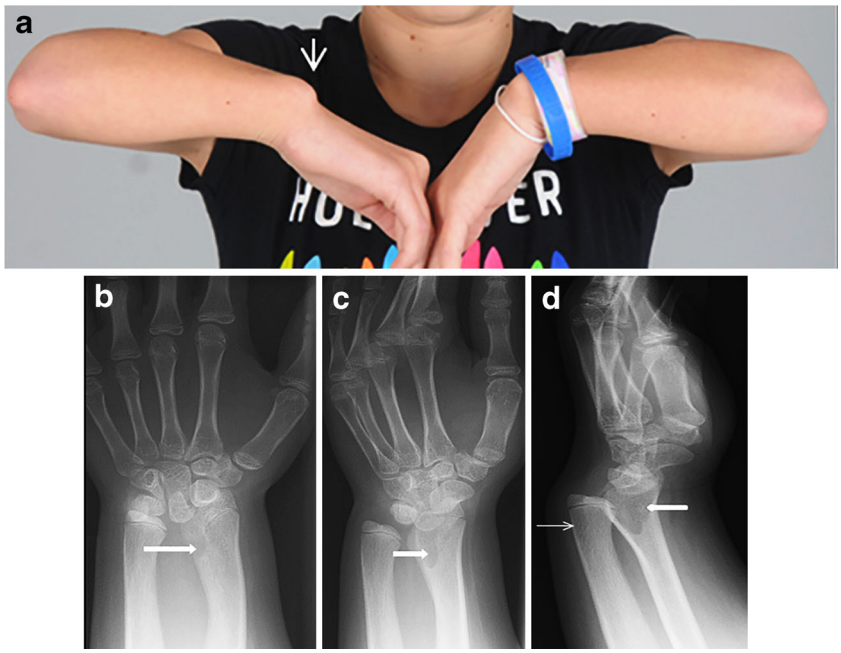

Fig. 1 An 11-year-old girl with Leri-Weill dyschondrosteosis. **a** Bayonet deformity of the wrist with corresponding AP (**b**), oblique (**c**) and lateral (**d**) radiographs. Note the characteristic radial notch at the origin of Vickers ligament (*block arrows*), proximal migration of the lunate with pyramidalization of the carpus, and, allowing for a degree of rotation, the dorsal displacement of the ulna (*thin arrow*)

dyschondrosteosis are characterized by the presence of an anomalous volar radiolunate (Vickers) ligament (Figs. 2, 3, 4 and 5) with a corresponding "notch" seen in the distal medial radius at the ligament origin (Figs. 1, 2 and 6) [16, 17]. The absence of a Vickers ligament distinguishes Madelung-type deformity from true Madelung deformity. The more common "distal radius" variant of true Madelung deformity likely represents a milder form of mesomelia, and the less common "entire radius" variant reflects more severe mesomelia with similar but more pronounced clinical and radiologic features. A reverse Madelung deformity is a rare variant demonstrating dorsal tilt of the distal radius with a dorsal shift of the carpus.

Distal radius Madelung deformity

Radiographic features

The pathognomonic radiographic appearance of Madelung and Madelung-type deformities includes increased inclination

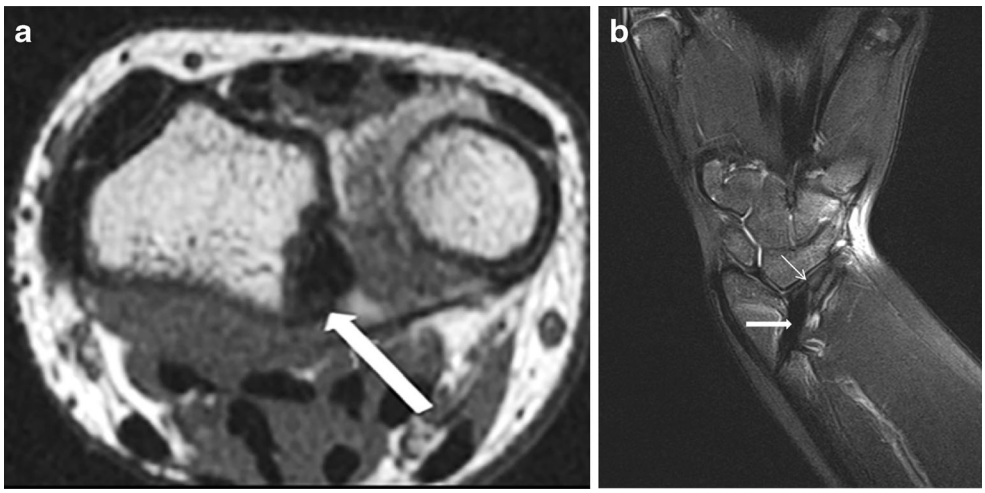

Fig. 2 A 9-year-old girl with Madelung deformity. Vickers ligament. **a** Axial T1-weighted image. *Arrow* identifies the hypertrophic volar short radiolunate (Vickers) ligament with thinning and remodeling of the volar distal radius by the hypertrophic ligament. **b** Coronal fat-suppressed T2-weighted image. A prominent Vickers ligament (*block arrow*) is seen extending from a deep radial notch to the lunate. Incidental lunotriquetral coalition is present. *Thin arrow* demonstrates the oblique orientation of the triangular fibrocartilage

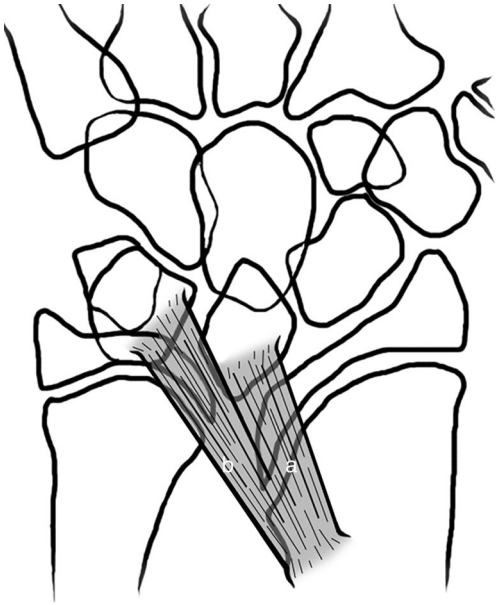

Fig. 3 Illustration shows the orientation of a hypertrophic radiolunate (Vickers) ligament (**a**), and less common radiotriquetral ligament (**b**), both of which can result in tethering of the proximal carpal row to the radius

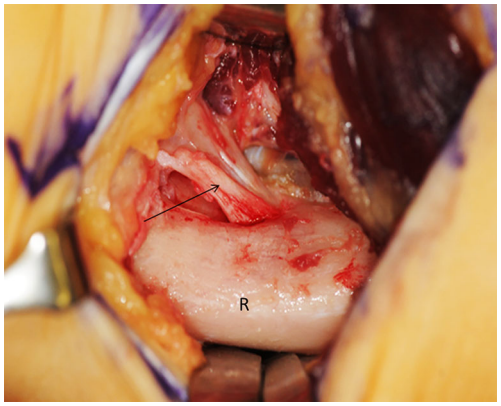

Fig. 5 A 15-year-old girl with Madelung deformity. Intraoperative image of a Vickers ligament (*arrow*), arising from the distal radius (R)

of the radial articular surface and volar tilt, proximal and volar migration of the lunate with triangulation of the carpus, relative ulnar lengthening and dorsal subluxation of the distal ulna. A radial "notch" (Figs. 1 and 6) and bony physeal bar (Fig. 6) can also be seen in the AP projection. The dorsal "subluxation" of the ulna is often not a true subluxation but rather an accentuated dorsal position due to the increased volar tilt of the distal radial articular surface [1]. In long-standing deformity (either Madelung or Madelung-type), the distal ulna may appear irregular and spiculated and associated tendon pathology may become evident (Fig. 7) [18, 19]. Measurement of ulnar tilt, lunate subsidence and palmar carpal displacement may help to determine the severity of the deformity and advise surgical planning [20, 21].

Vickers ligament

The distinguishing feature of true Madelung deformity is the presence of Vickers ligament. Vickers ligament is an anomalous hypertrophic volar radiolunate ligament thought to tether the medial radial metaphysis and triangular fibrocartilage complex (TFCC) to the palmar surface of the lunate. The ligament restricts the medial and volar growth of the radius by exerting a compressive effect on the physis, resulting in the characteristic radiographic features (Figs. 2, 3, 4 and 5) [15–17, 22]. This compressive effect may also cause thinning of the lunate fossa and distal radial epiphysis.

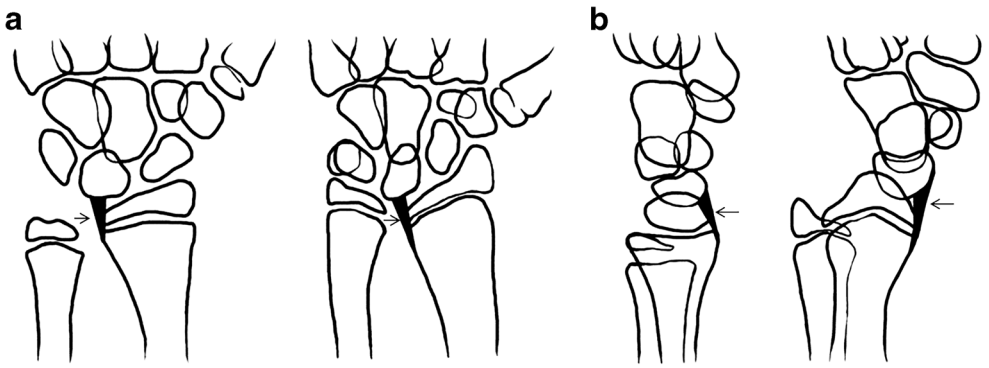

Fig. 4 Illustration in the AP projection (**a**) shows tethering of the radius to the lunate by Vickers ligament (*arrows*), resulting in restricted medial growth and increased inclination of the radial articular surface with pyramidalization of the carpus (*right image*). **b** Illustration in the lateral projection shows similar tethering in the volar wrist by Vickers ligament (*arrows*), resulting in an increased volar tilt of the radial articular surface and mild volar subluxation of the carpus (*right image*). Vickers ligament attaches the radius and triangular fibrocartilage complex to the lunate but also likely has a compressive effect on the growth plate and radial epiphysis, further restricting growth

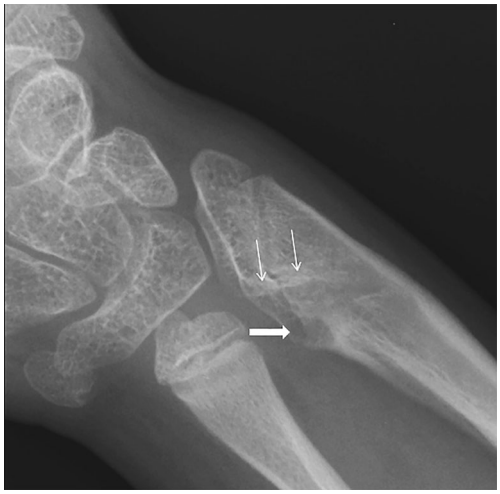

Fig. 6 A 9-year-old girl with Madelung deformity. Coned AP radiograph of the wrist. Characteristic flame- shaped notch (*block arrow*) in the distal radius is at the site of origin of Vickers ligament and a bony bar bridges the epiphysis and metaphysis (*thin arrows*)

Although Vickers ligament is best demonstrated with magnetic resonance imaging (MRI), a characteristic flame-shaped notch at the medial radial metaphysis can be seen on both radiographs and MRI at the origin of the ligament (Figs. 1, 2 and 6) [16, 17]. This notch may have a small bony spur at its proximal margin.

MRI features

Additional features of Madelung deformity can be seen with MRI. A bony physeal bridge resulting from early closure of the medial volar physis will be more readily apparent on MRI but can also be detected on radiographs (Fig. 6) [17]. In addition to Vickers ligament, MRI may reveal an anomalous hypertrophied and elongated volar radiotriquetral ligament (Fig. 3) [16, 22]. The TFCC typically demonstrates a progressively oblique orientation secondary to the radial deformity (Fig. 2) and may become thickened. This abnormal morphology subjects the TFCC to increased stress and may predispose patients to TFCC tears (Fig. 7). Spontaneous extensor tendon rupture is a rare but devastating consequence of long-standing disease that has been reported in elderly women, highlighting the importance of early detection and treatment (Fig. 7) [18, 19, 23]. The TFCC and tendon changes occur in Madelung deformity as well as Madelung-type deformities.

Associations

Although it occasionally presents as an isolated congenital abnormality, Madelung deformity is frequently associated with Leri-Weill dyschondrosteosis, an autosomal dominant inherited skeletal dysplasia arising from mutations in the short stature homeobox-containing gene (SHOX) in the X or Y sex chromosomes [10–12, 20, 21]. Leri-Weill dyschondrosteosis is characterized by Madelung deformity, marked short stature (<25th percentile) and short forearms (mesomelia) (Fig. 1) [20, 21]; all three features must be present for the diagnosis to be made. Several familial cases of isolated Madelung deformity have also been described [6, 13]. All cases of congenital Madelung deformity are characterized by the presence of a Vickers ligament.

Entire radius Madelung deformity

Zebala et al. [20] describe a variant Madelung deformity that involves the entire radius. The imaging characteristics are similar but more pronounced than those of the distal radius variant. In addition to the presence of a Vickers ligament, findings include increased radiocapitellar distance (defined as the distance from the center of the radial head articular surface to the capitellum, with a normal of less than 4 mm), more pronounced radial bowing, shorter radius and ulna, decreased height and a more pronounced deformity (Fig. 8) [20]. Clinically, both subtypes demonstrate loss of supination, pronation and wrist extension. However, those with involvement of the entire radius have greater restriction of movement, loss of elbow extension, and greater foreshortening and curvature of the forearm. This variant is more common in the setting of Leri-Weill dyschondrosteosis and in patients with bilateral involvement. As imaging of Madelung deformity is traditionally confined to the distal radius and wrist, recognition of this variant emphasizes the importance of imaging the entire forearm in all cases of Madelung deformity. Surgical correction is more difficult in this subset, and the results less gratifying when compared to the distal radius variant.

Reverse Madelung deformity

As the name implies, reverse Madelung deformity manifests as dorsal (rather than volar) tilt of the distal radius, suggesting an insult to the dorsal physis (Fig. 9). This results in dorsal shift of the carpus and volar subluxation of the ulna [24]. A distal radial notch was identified in one such case at our institution, suggesting the presence of an anomalous dorsal radiolunate ligament; unfortunately, no confirmatory MRI was obtained.

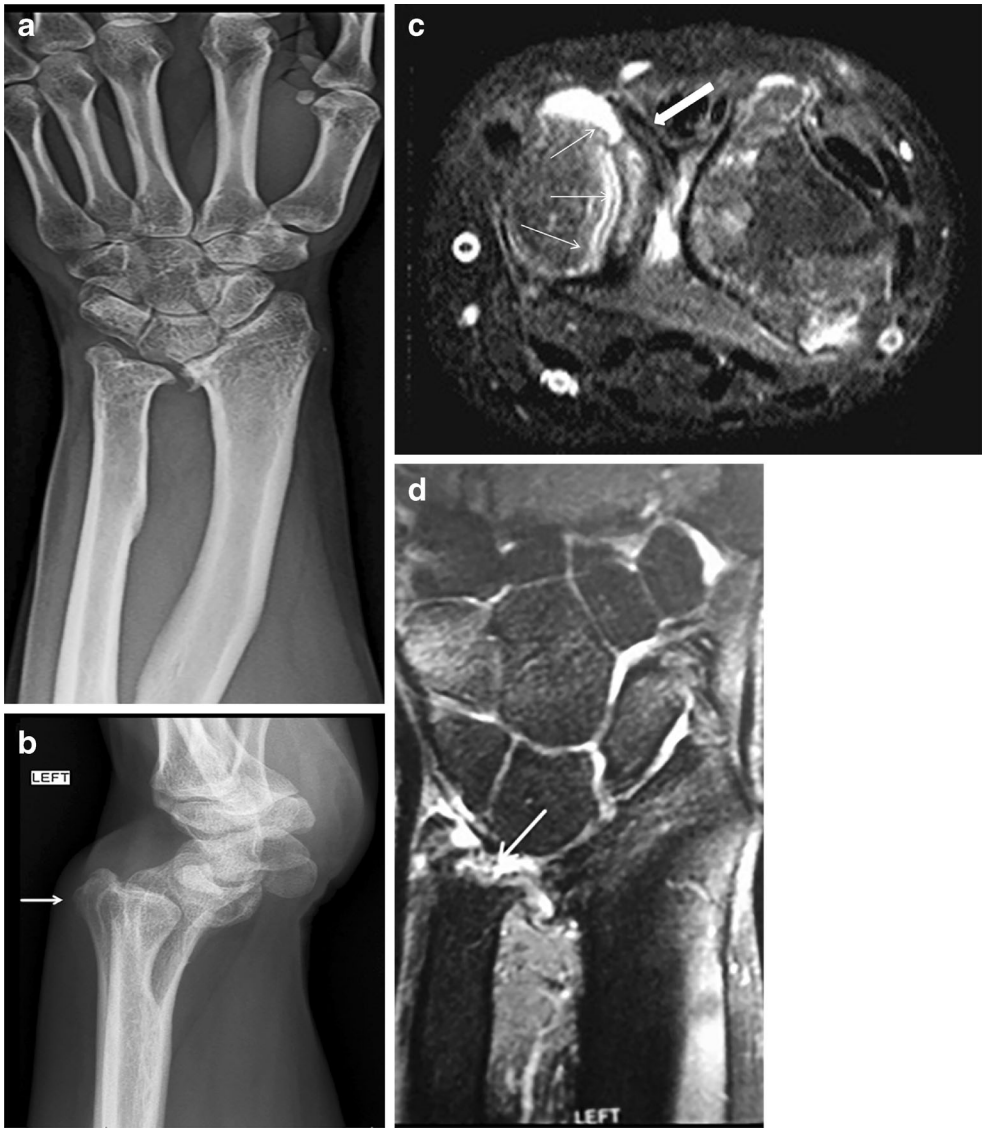

Fig. 7 A 40-year-old woman with advanced Madelung-type deformity. AP (**a**) and lateral (**b**) radiographs show marked deformity. Note irregularity with spiculation of the distal ulna. The absence of a radial notch suggests the absence of a Vickers ligament, likely indicating a post-traumatic etiology. There is increased volar tilt and inclination of the radial articular surface, proximal and volar migration of the lunate with pyramidalization of the carpus, and apparent dorsal subluxation of the ulna (*arrow in b*). **c** Axial fat-suppressed T2-weighted image. Note marked irregularity and spiculations in the distal ulna and radius (*thin arrows*) with associated distal radioulnar joint effusion. The abductor digit minimi tendon (*block arrow*) is displaced by the ulnar deformity, and appears diminutive. **d** Coronal fat-suppressed T2-weighted image. Note marked spiculation of the distal ulna, with associated tear of the triangular fibrocartilage complex (*arrow*)

Madelung-type deformity

Post-traumatic

Madelung-type deformity may be secondary to trauma, infection or infraction (incomplete fracture). Post-traumatic Madelung deformity results from injury to the medial volar physis of the distal radius. It may result from acute or repetitive axial loading trauma, an injury frequently incurred by gymnasts [25, 26]. In these individuals, the medial volar portion of the distal radial physis sustains cumulative microtrauma from repetitive axial-loading maneuvers. The injury is most commonly unilateral but may be bilateral. Initial manifestations include widening and irregularity of the lateral physis and positive ulnar variance, followed by premature closure of the medial growth plate and physeal bridging. MRI may show paraphyseal edema and early physeal bridging. Unlike congenital Madelung deformity, no Vickers or anomalous radiotriquetral ligament is present (Fig. 10). In the absence of Vickers ligament, no corresponding notch should be seen in the distal radius. In early cases, treatment involves resection of the physeal bridge; more severe deformities may require ulnar epiphyseodesis and shortening with radial osteotomy.

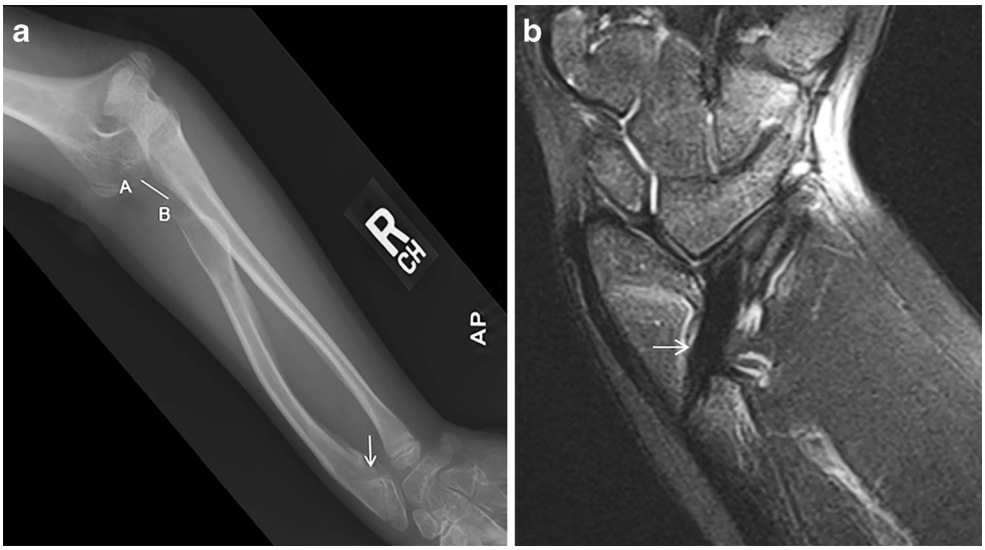

Fig. 8 A 9-year-old girl with entire radius variant. **a** AP radiograph of the forearm shows marked bowing and increased radiocapitellar distance (AB), with a radial notch (*distal arrow*) for the origin of a prominent Vickers ligament. **b** Coronal fat-suppressed T2-weighted image shows the prominent Vickers ligament (*arrow*)

Osseous dysplasias

Patients with osseous dysplasias, including Ollier disease, multiple epiphyseal dysplasia and multiple hereditary exostoses, may present with a Madelung-type deformity, the etiology of which is poorly understood [27, 28]. It is also seen in less than 10% of patients with Turner syndrome [20, 21], where it is thought to be secondary to a hapodeficiency of the SHOX gene. In multiple hereditary exostoses, it is possible that the physeal plate may be injured or tethered by a spanning or adjoining tumor mass (Fig. 11). There is no Vickers ligament to accentuate the pyramidalization of the carpal row. Treatment in these cases is surgical removal of the osteochondromas and possible osteotomy to correct the deformity.

Management

Many patients with Madelung or Madelung-type deformity are asymptomatic or have minimal symptoms; such cases

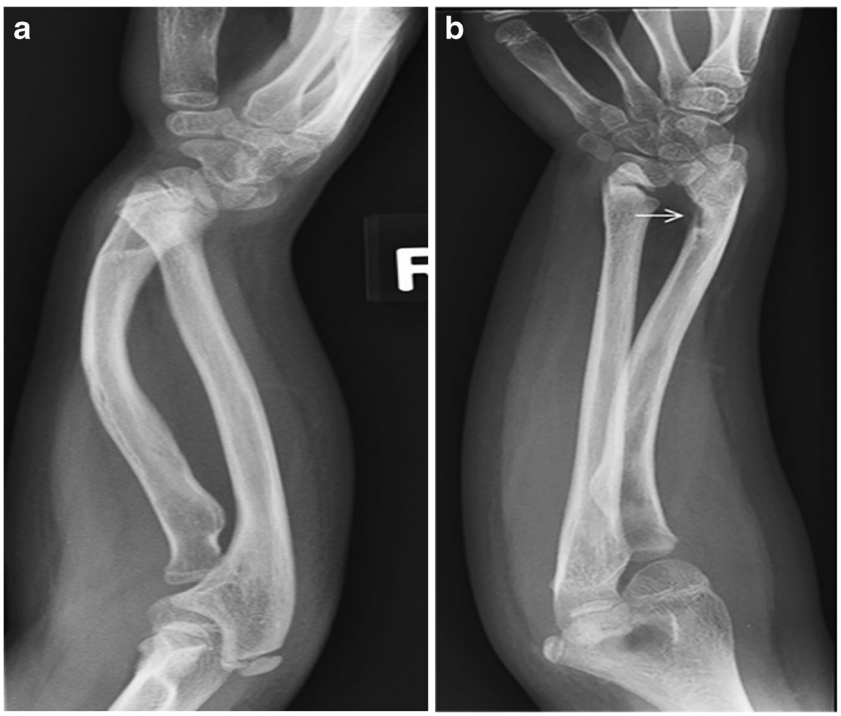

Fig. 9 An 11-year-old girl with reverse Madelung deformity. Lateral (**a**) and AP (**b**) radiographs show dorsal instead of volar tilt of the distal radius, dorsal shift of the carpus and volar subluxation of the ulna. These are the opposite of the classic Madelung deformity. Note the radial notch (*arrow* in b) presumably reflecting the origin of an anomalous ligament

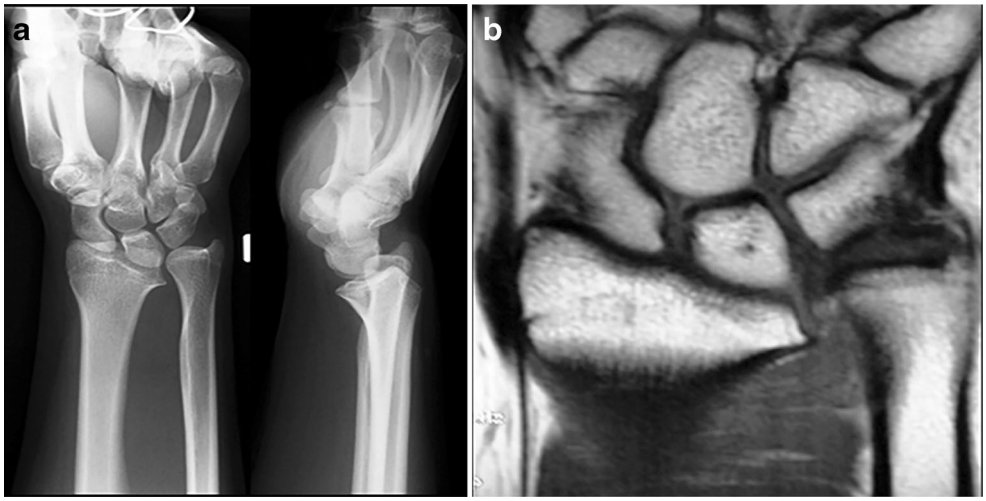

Fig. 10 A 14-year-old girl with "gymnast wrist." **a** AP and lateral radiographs. Growth arrest in the medial and volar direction. No radial notch is present. **b** Coronal T1-weighted image. No Vickers or anomalous radiotriquetral ligament is identified

can be managed conservatively. When symptomatic, treatment for both entities typically includes a dome osteotomy to correct the volar and radial deformity combined with surgical release of Vickers ligament or anomalous radiotriquetral ligament in cases of true Madelung deformity [29]. If significant ulnar positive variance persists, an ulnar-shortening osteotomy and epiphyseodesis is performed to re-level the joint. A Darrach procedure to reduce the length of the ulna may also be performed in adults with degeneration of the distal radioulnar joint [30]. Tendon rupture is treated with tendon grafting or transfer, as the frayed edges of the tendon make primary end-to-end repair difficult. This may be combined with distal ulnar resection to reduce further tendon attrition. Distal ulnar resection may also be performed prophylactically when imaging shows ulnar spiculation and signs of tendon attrition such as tenosynovitis, tendon displacement or partial tearing [18].

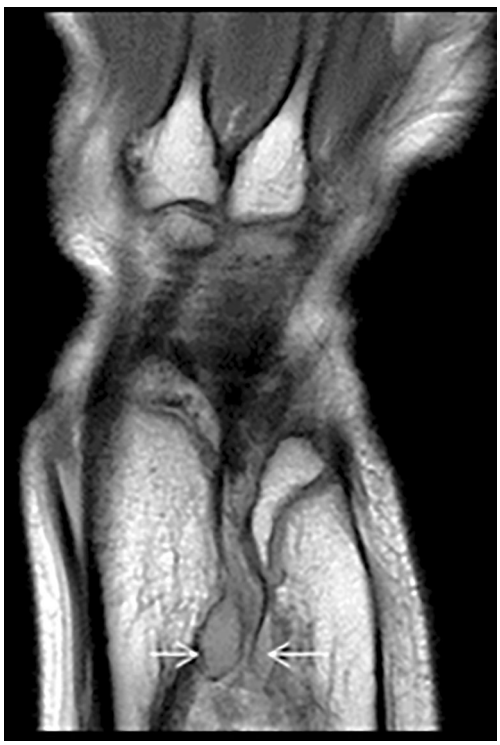

Fig. 11 An 11-year-old girl with multiple hereditary exostoses. Coronal T1-weighted image shows prominent cartilage caps (*arrows*) in the osteochondromas, with associated Madelung-type deformity. A Vickers ligament is not present, and there is no radial notch or physeal bar. The physis may be injured or tethered by the adjacent or spanning osteochondromas

Conclusion

Madelung deformity of the wrist results from premature closure of the volar medial portion of the distal radial physis and may arise from a variety of congenital or acquired etiologies. Patients present with varying degrees of pain, decreased grip strength, and the classic bayonet deformity due to dorsal displacement of the ulna. Radiologic manifestations include increased inclination of the radial articular surface and volar tilt, triangulation of the carpus with proximal migration of the lunate, and relative dorsal subluxation of the distal ulna. Congenital cases may arise in isolation but are often associated with Leri-Weill dyschondrosteosis, an autosomal dominant form of mesometric dwarfism. True Madelung deformity is characterized by a hypertrophic Vickers ligament and/or radiotriquetral ligament, which are not present in acquired Madelung-type deformities. Identification and resection of the anomalous ligaments are fundamental aspects of surgical

management. Two morphological variants of classic Madelung deformity have been described, the common distal radius variant and a more severe entire radius variant. Extensor tendon rupture, a potentially devastating complication of long-standing deformity, is important to recognize, as prophylactic treatment may be beneficial.

Conflicts of interest None

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