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Carpal Ligamentous Laxity with Bilateral Perilunate Dislocation in Marfan Syndrome

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Abstract. A case of persistent bilateral perilunate dislocation unrelated to trauma in a patient with Marfan syndrome is discussed. This finding is believed to be a manifestation of the generalized ligamentous laxity occurring in this disorder. Radiographs of eight additional Marfan syndrome patients failed to demonstrate similar carpal instability. Because some carpal derangements are dynamic events, stress views or wrist fluoroscopy may be necessary to demonstrate unsuspected carpal instability in Marfan patients.

Key words: Marfan syndrome – Ligaments, injuries – Wrist, injuries – Joints, carpal

We report an unusual case of persistent bilateral perilunate dislocation in a patient who had no wrist trauma, but who did have Marfan syndrome. As patients with this syndrome have generalized ligamentous laxity and joint hypermobility, the findings may be explained on this basis.

Case History

A 14-year-old girl with Marfan syndrome presented with extreme hypermobility of both wrists. On physical examination, there was a marfanoid habitus, arachnodactyly, mobile wrist deformities, and bilateral subluxation of the first carpometacarpal joint. Past history included posterior spinal fusion for scoliosis two years previously. Radiographs (Figs. 1 and 2) show bilateral dorsal perilunate dislocations. Splinting of the wrists yielded no benefit, and five years later the deformity persisted.

Discussion

With increasing experience and understanding of the functional anatomy of the wrist, it is becoming clear that dorsal perilunate dislocation, volar lunate dislocation, and midcarpal dislocation are all manifestions of similar ligamentous injuries [4, 5, 8]. The difference among these entities reflects the degree to which there is spontaneous reduction of the capitate, with consequent displacement of the lunate into a volar subluxed location [4]. In the typical traumatic dorsal perilunate dislocation, scapholunate ligament complex rupture is followed by rupture of the radiocapitate ligament, dorsal displacement of the capitate, and disruption of the volar and dorsal radiotriquetral ligaments [5]. In our case, the ligamentous instability is caused by generalized connective tissue abnormality rather than trauma.

Prominent among the skeletal manifestations of Marfan syndrome are ligamentous laxity and associated joint hypermobility. Our patient presumably developed bilateral perilunate dislocation as a result of this ligamentous laxity. Although a basic abnormality of collagen has been suspected as the underlying abnormality of Marfan syndrome, there are no structural alterations of collagen in tendons of Marfan patients examined with light or electron microscopy, and there are no detectable biochemical abnormalities [1]. In previous cases of congenital carpal ligamentous laxity, patients who did not have Marfan syndrome developed bilateral palmar flexion instability [6]. A single previous case of bilateral perilunate dislocation of which we were aware resulted from severe wrist trauma [2].

The incidence of atraumatic carpal dislocation or ligamentous instability in patients with Marfan syndrome is unknown. Retrospective evaluation of wrist films in eight additional Marfan patients (age 12–49 years) disclosed no abnormalities. Nevertheless some wrist derangements are dynamic events, and cannot be diagnosed on conventional static

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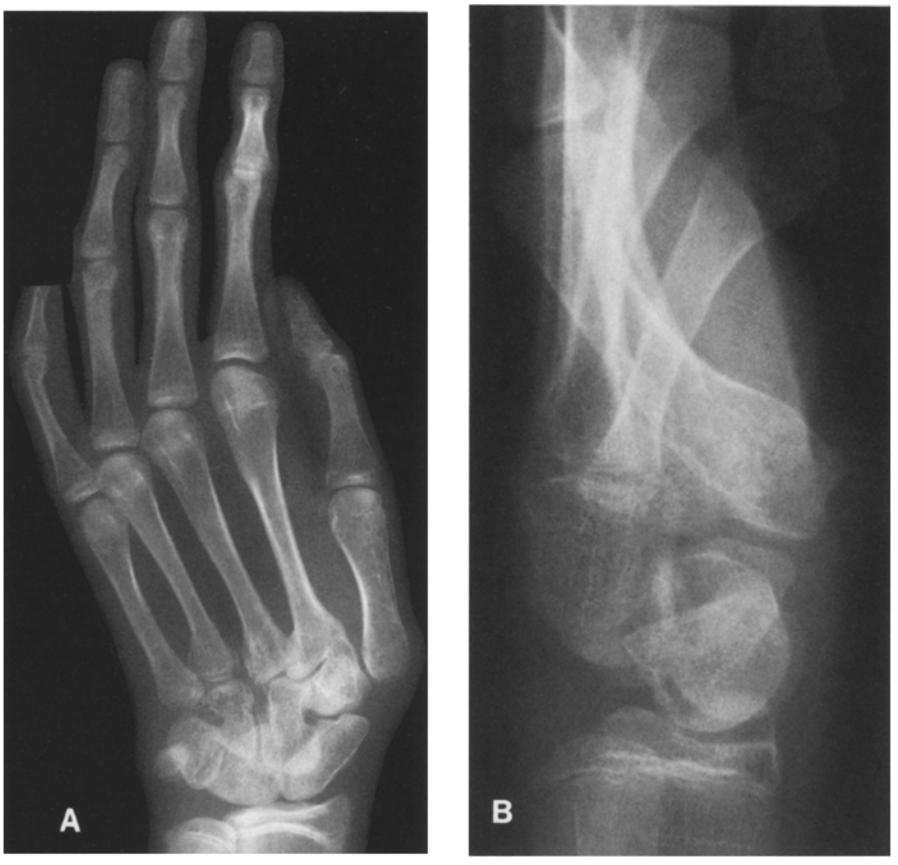

Fig. 1. A Posteroanterior view of the left hand shows arachnodactyly, subluxation of the first carpometacarpal joint, and overlap of the proximal and distal carpal rows. **B** Lateral view demonstrates dorsal perilunate dislocation

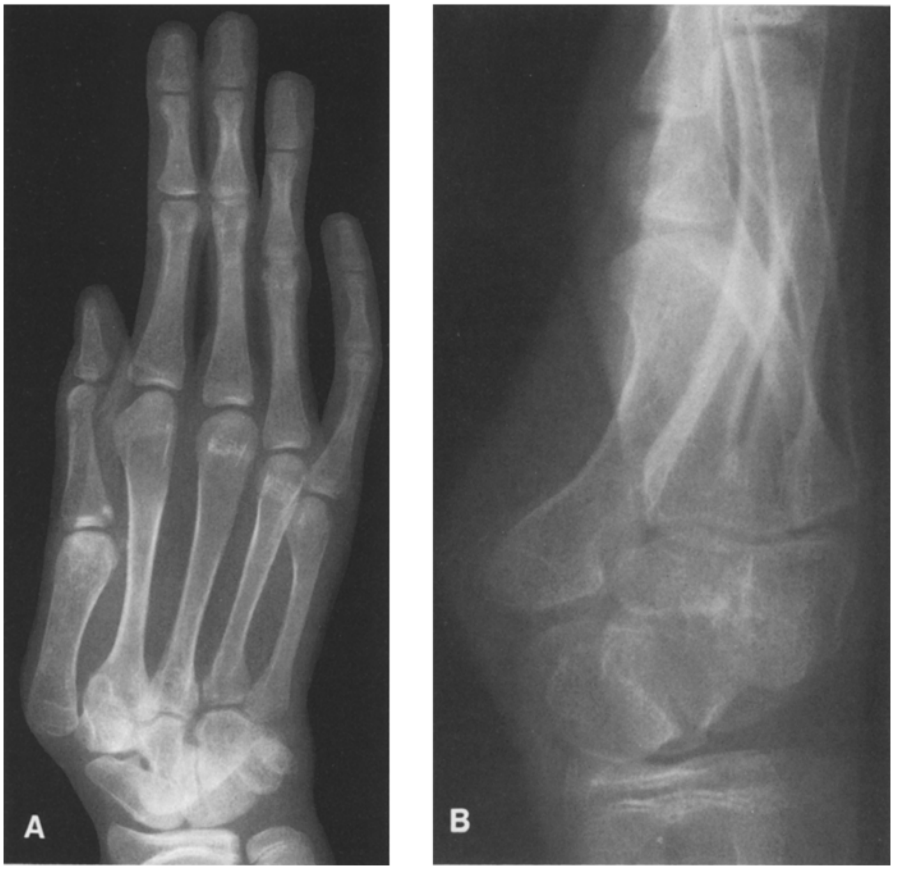

Fig. 2A, B. Right hand. Findings correspond to those in the left hand

radiographs [7]. Prospective evaluation of the wrist in patients with Marfan syndrome using stress views [5], an instability series [3], or fluoroscopy [7] may disclose unsuspected ligamentous laxity.

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