Focal Fibrocartilaginous Dysplasia in the Ulna: Report on 3 Cases

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Focal fibrocartilaginous dysplasia is an uncommon, benign bone lesion that causes deformity in young children. It is most commonly encountered in the proximal tibia, with few cases documented in the upper extremity. We report 3 cases affecting the ulna. In each case, the fibrous tissue appeared to anchor itself in the ulnar diaphysis, behaving as a tether that retards growth. All 3 patients had excision of the fibrous tissue. There was concern for radial head subluxation before surgical intervention, but all patients maintained a congruent radiocapitellar articulation. The visible deformity improved in all 3 patients, although the limbs remained shorter than the contralateral side. In our limited series, we believe that early excision of the fibrous tissue tether can prevent radiocapitellar joint dislocation in patients with focal fibrocartilaginous dysplasia of the ulna. (*J Hand Surg 2012;37A:2300–2303. Copyright* © *2012 by the American Society for Surgery of the Hand. All rights reserved.*)

Key words Fibrous periosteal inclusion, focal fibrocartilaginous dysplasia, radiocapitellar dislocation, tether.

Pocal Fibrocartilaginous dysplasia (FFCD) is a rare condition in children and is associated with bony deformity. Described by Bell et al¹ in 1985, the condition occurs most commonly around the knee joint and can cause tibia vara. Other sites that have been reported include the distal femur, proximal humerus, ulna, radius, and phalanx. ¹⁻⁹ Radiographic features include a well-defined, obliquely oriented, lucent defect in the cortex, with sclerosis along the borders of the lesion. It is postulated that fibrous tissue acts as a nonyielding tether, causing asymmetric growth and angulation. ^{1,5,7-9} Jouve et al⁷ expanded this theory and described it as a bony anchor preventing natural sliding of the periosteum during growth and suggested that it had the same effect as an epiphyseodesis.

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0363-5023/12/37A11-0014\$36.00/0 http://dx.doi.org/10.1016/j.jhsa.2012.08.008 Eleven cases involving the ulna have been documented.^{3,5–7,9} We report 3 additional cases. This series adds to the knowledge of this uncommon condition, proposes a mechanism of deformity, and recommends a treatment strategy.

CASE REPORTS

This study was approved by our institutional review board. The parents of the patients studied were informed that data concerning these cases would be submitted for publication. In all cases, no history of trauma was reported.

Case 1

A 30-month-old girl was evaluated for progressive right forearm deformity. At birth, no deformity was noticed, but one became evident over the 6 months prior to presentation. No additional abnormalities were noted. The right forearm appeared slightly shorter than the left and angulated medially. Radiographs demonstrated a relatively short ulna, with bowing of both bones. An indentation in the cortex was evident at the junction of middle-to-distal third ulna (Figs. 1A, 1B). Magnetic resonance imaging (MRI) showed a fibrous band attached to the ulna. Resection of the ulnar fibrous tether was performed through a longitudinal incision along the distal half of the ulna. A thick, taut, fibrous band was

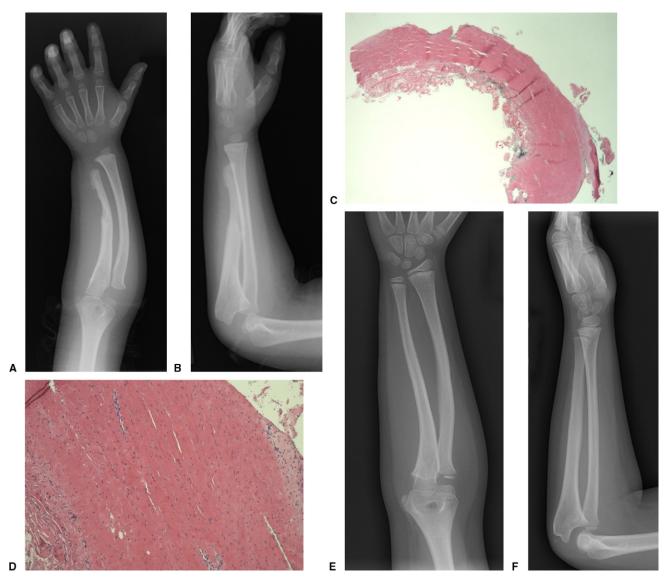


FIGURE 1: Patient 1. **A** Anteroposterior and **B** lateral forearm radiographs with fibrous periosteal inclusion in distal third of the ulna in a 30-month-old child. **C** Histopathologic findings reveal fibroconnective tissue (hematoxylin-eosin \times 10) and **D** dense fibromuscular tissue without areas of cartilage (hematoxylin-eosin \times 40). **E** Anteroposterior and **F** lateral radiographs at age 8 years 9 months.

identified entering into the ulna proximally. The band was excised, and its attachment to the ulna was curetted. Histopathology demonstrated fibromuscular tissue (Figs. 1C, 1D). At the 6-year follow-up, wrist and elbow motion was full. Radiographs demonstrated substantial straightening of the ulna and maintenance of a congruent reduced radiocapitellar joint (Figs. 1E, 1F).

Case 2

A 40-month-old boy was evaluated for right forearm shortening and bowing. The deformity was not present at birth. Over the prior year, the parents noticed gradually increasing asymmetry between the forearms. The right ulna was clinically 2.5 cm shorter than the left. The radial head was prominent at the elbow. He had full

supination but only 45° of pronation. Radiographs demonstrated forearm bowing with an ulna lesion and radial head subluxation. During surgery, longitudinal fibrous bands were defined, extending from the physis, along the ulnar shaft, and through a hole in the ulna. Histopathology revealed fibroconnective tissue and benign bone. Over an 18-month period, ulnar angulation improved from 40° to 15°. Subluxation of the radial head diminished. At 9 years of age, he demonstrated full forearm pronation and supination. The involved forearm was 1 cm shorter than the contralateral forearm.

Case 3

A 42-month-old boy was evaluated because of left forearm deformity and decreased forearm motion. The

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