

A Congenital Hand Deformity: Dupuytren's Disease

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A 10-month-old child presented with a lack of extension at the distal interphalangeal joint. Despite the absence of trauma, a provisional diagnosis of mallet finger led to treatment using a short dorsal splint. Four months later the flexion had increased and included the proximal interphalangeal joint. Palpation revealed a palmar cord on the lateral aspect of the finger. Surgery disclosed a typical Dupuytren cord and the histology supported this diagnosis. There was no known family history of the disease. There was no sign of recurrence 27 months after surgery in this case of Dupuytren's disease that was present at birth. (*J Hand Surg* 2001;26A: 515-517. Copyright © 2001 by the American Society for Surgery of the Hand.)

Dupuytren's disease is considered by most to be a disease that occurs in adulthood. "Congenital" Dupuytren's disease has been reported, but the youngest Dupuytren's disease patient reported in the English literature was 8 years old. Dupuytren himself reported a case in a 6-year-old.¹ We report a case of histologically proven Dupuytren's disease in an infant.

Case Report

A 10-month-old male infant presented to his pediatrician with a "crooked" long finger. The deformity had been present since birth. The pediatrician referred the child to our clinic for further evaluation. Examination in our department revealed a distal interphalangeal (DIP) joint fixed flexion deformity of 30°. No other anomalies were noted. Our initial

impression was that the child had a chronic mallet deformity of the long finger secondary to occult birth trauma. This diagnosis was made in spite of the report of a normal and uneventful vaginal delivery and a history absent of any trauma. An extension splint was applied. The child was examined 3 months later; despite good compliance with the splint, the contracture of the long finger DIP joint had increased. Four months later (at the age of 17 months) the flexion contracture of the DIP joint had increased to 40°, a proximal interphalangeal (PIP) joint contracture of 20° was present, and a firm palpable palmar mass was noted. Flexion of the PIP joint permitted more extension at the DIP joint. The same "tenodesis effect" was seen at the PIP joint when the DIP joint was flexed.

In view of the increasing digital contracture and palmar mass, we elected to proceed with excision of the "tumor." The surgery was performed 1 month later (at the age of 18 months) through a Bruner incision. Two well-defined lateral cords typical of Dupuytren's disease were found. The thickest was ulnar; its relationship with the neurovascular bundle was identical to that encountered at this level in Dupuytren's disease. The tendon sheath was intact. The nerves and vessels were carefully dissected and preserved and a cord identical to a Dupuytren cord was excised (Fig. 1). The absence of skin involvement allowed a simple closure incorporating a "Y" to

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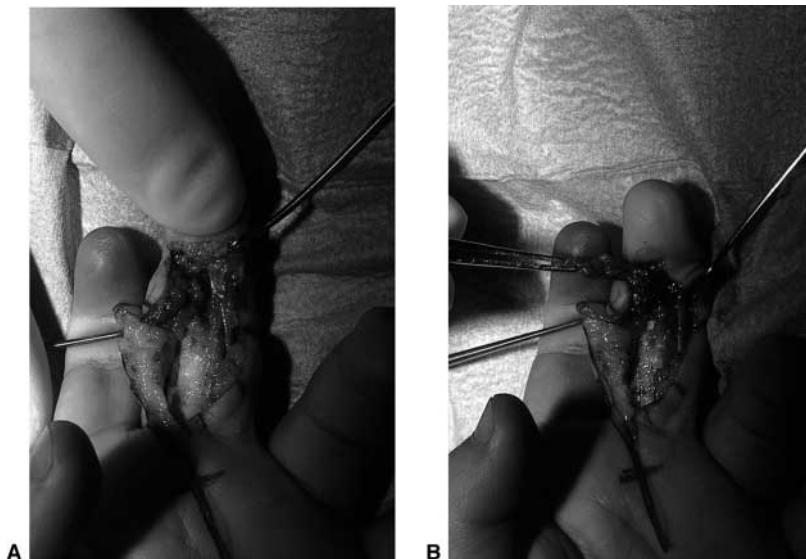


Figure 1. Preoperative view of the laterodigital cord and the neurovascular bundle before (A) and after (B) elevating the cord.

“V” plasty. The histology of the specimen was reviewed by an independent histologist and was typical of Dupuytren's disease (Figs. 2, 3). There was a marked increase of fibrous tissue with fibroblasts encircling fat lobules and nervous branches. There was no granulomatous tissue or foreign body. There were no mitoses in the fibroblasts or intracytoplasmic inclusions as seen in digital infantile fibromatosis. There were no perioperative complications. At 27 months after surgery no recurrence was palpable and full range of motion was present at the PIP and DIP joints of the long finger. Palpation of the other fingers was normal. There was no family history of Dupuytren's disease.

Discussion

In our case trauma, infantile fibromatosis, and camptodactyly were ruled out based on history, clinical examination, and macroscopic and microscopic presentations. The intraoperative observations were identical to a Dupuytren cord as seen in adults.

In his “leçon” of October 16, 1832, Dupuytren¹ described a 6-year-old boy with a permanent contracture of the ring and small fingers. The boy's grandmother also had the condition. Since then other investigators have reported cases in children. In 1954 Goetzee and Williams² reported the first histologically proven case of Dupuytren's disease in an adolescent and in 1963 Hueston³ reported a case of histologically confirmed Dupuytren's disease in a 12-year-old boy. The largest series reported to date

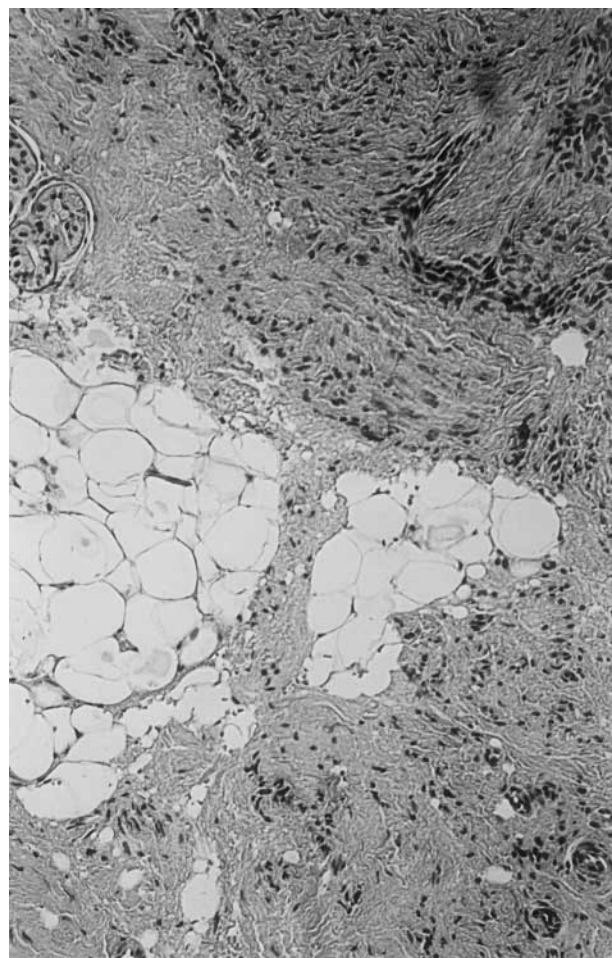


Figure 2. Histologic aspect of the excised cord with fibrous proliferation and fibroblasts surrounding fat lobules. (Magnification $\times 145$.)

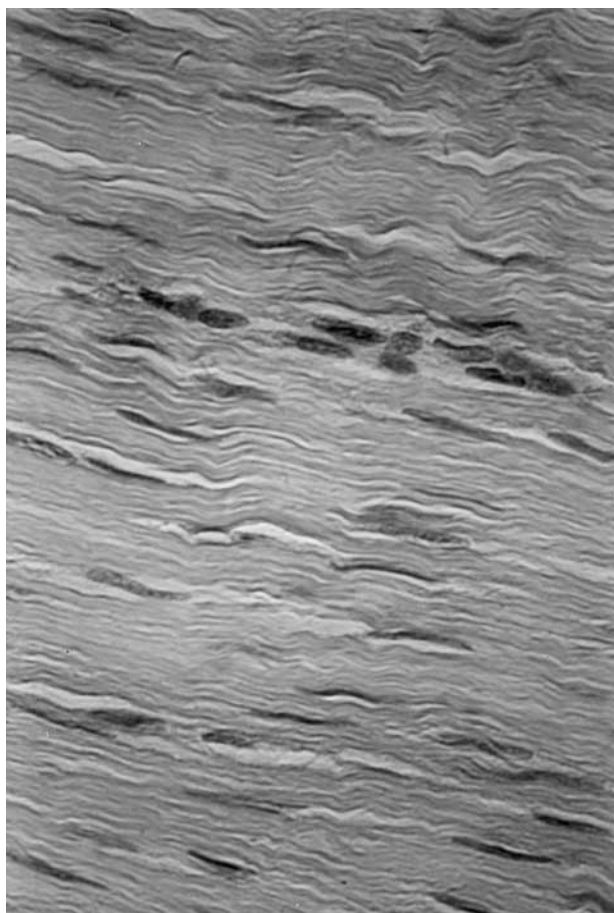


Figure 3. Uniform proliferation of fibroblasts. (Magnification $\times 400$.)

reviewed 8 patients younger than 13 years.² There are, however, no reports of Dupuytren's disease occurring in children younger than 8 years. In reviewing the natural history of the disease in 647 men and 254 women, Mikkelsen⁴ found only 1 woman and 2 men reporting their first clinical sign of Dupuytren's disease before the age of 10 years and 2 women and 10 men noting signs of the disease between 10 and 20 years of age. Coffin and Dehner,² reviewing 900 soft tissue tumors in patients younger than 20 years, found fibromatosis in 72% but only 3% were found to have Dupuytren's diseases (2 boys aged 8 and 11 years).

In 1897 Hutchinson⁵ reported "juvenile forms of Dupuytren's [disease]" in a group of patients with the disease "present at birth," frequently "in associ-

ation with other malformations," and with "little tendency to increase." Careful review of the case reported reveals that it was in fact an inherited unilateral mirror hand with 8 fingers (with a central syndactyly) and a bilateral "contracted" fifth finger seemingly due to a camptodactyly. In 1917 Grieg⁶ reported a 5-week-old boy with bilateral clenched fists with overlapping of the long fingers as seen in congenital anomalies such as the Hall syndrome (distal arthrogyriposis type 2). This explains the bilaterality and the absence of progression of the disease. Another cause of "congenital" Dupuytren's disease was studied by Zancolli and Zancolli,⁷ who reported a congenital disease in which the skin and subcutaneous fascias are severely contracted. The disease entities discussed in these reports should be considered as differential diagnoses when examining infants with contracted digits.

We treated our case with simple excision of the diseased fascia. It could be argued that a dermofasciectomy should have been considered to prevent recurrence; however, the skin was not involved and the literature is insufficient to draw any conclusion regarding the recurrence rate of Dupuytren's disease in this age group. Urban et al² noted a recurrence 2 years after the excision of Dupuytren's disease in a 9-year-old boy.

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