Dupuytren Contracture in the Pediatric Population: A Systematic Review

Ali Izadpanah[1] Alex Viezel-Mathieu[1] Arash Izadpanah[2] Mario Luc[1]

[1] Division of Plastic and Reconstructive Surgery, McGill University Health Center, Montreal, Quebec, Canada
[2] Plastic and Reconstructive Surgery, University of Manitoba, Winnipeg, Manitoba, Canada

Address for correspondence Ali Izadpanah, MD, CM, MSc, Division of Plastic and Reconstructive Surgery, McGill University Health Center, 1650 Cedar Avenue, Montreal, Quebec H3G 1A4, Canada (e-mail: ali.izadpanah@mail.mcgill.ca).

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Abstract

Dupuytren contracture of the palm is a relatively common benign fibroproliferative disease of the palmar fascia typically affecting the adult population. There have however been several reported cases of Dupuytren contracture in children. We sought to review the literature for Dupuytren contracture and highlight the main clinical features and management of the disease in children.

Keywords

- Dupuytren contracture
- Dupuytren disease
- Palmar fibromatosis

Introduction

Dupuytren contracture of the palm or palmar fibromatosis is a relatively common benign fibroproliferative disease of the palmar fascia typically affecting the adult population. In particular, this disease primarily affects white males older than 50 years, with a prevalence that increases with advancing age. There have however been several reported cases of Dupuytren contracture in children including two histologically confirmed case in 4-month-old and 6-month-old infants.[1,2] Dupuytren himself described a case of a 6-year-old child presenting with contraction of little and ring fingers in 1832.[3]

Although several authors (listed in ►Table 1) have described Dupuytren disease in the pediatric population, our search of the literature revealed that there has not been a review of published cases done since Urban et al reported their work on Dupuytren contracture in children in 1996.[4] Thus, we sought to review the literature for Dupuytren contracture and the main clinical features of the disease in children.

Materials and Methods

Using the Medline search tool, a search of the National Library of Medicine (PubMed) database was performed using the keywords "Dupuytren contracture" or "Dupuytren disease" or "Palmar Fibromatosis." The search was limited to children ($<$ 18 years) and humans with no limitation on language. All case reports and review articles from the pediatric population were selected. Articles published between 1950 and 2013 were considered.

A second round of searches was performed and involved a citation search of the reference lists of each of the article identified above.

Results

Our literature review revealed 11 manuscripts representing 14 reported cases of histologically confirmed Dupuytren contracture in the pediatric population.[1,2,4–12] Selected manuscripts were analyzed with regards to patient demographics, presenting symptoms, surgery performed, recurrence, and outcome (►Table 1).

Of the 14 cases of pediatric Dupuytren disease, there was a strong male predominance with 11 of the 14 cases (79%) occurring in males. The mean age at the time of presentation was 83 months (6.9 years) (range, 4–168 months).

The most common presenting symptoms included the presence of a nodular mass with a variably present palpable cord or contracture of the affected digit. Range of motion was reported to have been affected in approximately 65% of the cases ($n = 9$), with two cases having no disruption of range of motion, and three cases not specified. There was an

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Table 1 Literature review findings

Author (y)	Sex	Age onset (mo)	Age presentation (mo)	Hand affected	Presenting symptoms	ROM affected	Surgery	Recurrence	Outcome
Bebbington and Savage (2005)	M	Not stated	6	L	• Palmar swelling and cord to D4	Yes	Excision biopsy	None at 21 mo	• No functional disturbances of the hand. • Full active flexion, 10 degrees loss of active extension of D4.
Fernández-García et al (2007)	F	144	Not stated	R	• Subcutaneous indurated tumor on the volar surface of the right palm	Not stated	Excision of the lesion including skin, fat, palmar fascia, and flexor retinaculum	None at 13 mo	• Resolution of all symptoms • No persistent disturbances
Foucher et al (2001)	M	Not stated	10	L	• Flexion contracture of right D3 (DIP [40 degrees] PIP [20 degrees]) • Firm palpable palmar mass	Yes	Cord excision	None at 27 mo	• Full ROM • No recurrence or progression of disease
Goetzee and Williams (1955)	M	132	168	R	• Flexion contracture D4 and D5	Yes	Fasciectomy	Not stated	• Full ROM
Kraus et al (2012)	F	89	95	L	• Palmar swelling and subcutaneous cord	No	Not stated	None at 1 y	• Resolution of all symptoms • No persistent disturbances
Korambayil and Padikala (2011)	M		4	L	• Palmar swelling and associated cord to the thumb	Yes	Excision biopsy + fasciectomy	Not stated	Not stated
Mandalia and Lowdon (2003)	M	108	120	L	• Nodule in palm in line with D4		Nodule excised	Not stated	Not stated
Marsh and Kelly (2008)	M	84	96	R	• Progressive contracture of right D5 • Flexion contracture 40 de grees PIP + hyperextension 20 degrees DIP	Yes	Fasciectomy	None at 1 y	• Full ROM • No recurrence or progression of disease
Rhomberg et al (2002)	F	24	30	L	• Flexion contracture left D2 • 50 degrees extension lag of PIP • Fibrous band from MP to distal phalynx	Yes	Fibrotic band excision	Yes	• Required two subsequent sur geries due to flexion contractures. • Good end result with 10 degrees extension lag
Rhomberg et al (2002)	M	Not stated	120	R + L	• Fibrous band on palmar radial aspect of both thumbs extending from the thenar to IP joint	Yes	Fasciectomy	None at 2 y	• Full ROM • No recurrence or progression of disease

Table 1 (*Continued*)

Author (y)	Sex	Age onset (mo)	Age presentation (mo)	Hand affected	Presenting symptoms	ROM affected	Surgery	Recurrence	Outcome
Rhomberg et al (2002)	M	96	108	R	• D4 and D5 flexion contracture	Yes	Partial fasciectomy and arthrolysis	Yes	• Forearm amputation due to epitheloid sarcoma. • Myoelectric prosthesis
Urban et al (1996)	M	102	108	R	• Flexion contracture D5	Yes	Fasciectomy	Yes	• Recurrence in Rt. hand requiring two subsequent surgeries. • New disease discovered in contralateral (L) hand requiring dermofasciectomy.
Urban et al (1996)	M	120	120	L	• Nodular mass in palm x2	Not stated	Nodule excised	Not stated	Not stated
Usmar and Peat (2010)	M	90	96	R	• 7-mm nodular mass adherent to overlying skin	No	Excisional biopsy	None at 3 mo	Not stated

Abbreviations: DIP, distal interphalangeal; IP, interphalangeal; PIP, proximal interphalangeal.

approximate equal distribution between the hands affected (left $n = 7$, right $n = 6$) with one case affecting both the hands.

In terms of surgical procedure performed, there was an approximately equal distribution between patients reported to have undergone excision of the mass or cord ($n = 6$) versus those who underwent a fasciectomy ($n = 7$). In one case, the exact surgical procedure performed was not specified.[8]

Of the 14 cases described in the literature, only 3 patients were reported to have experienced recurrence of disease following surgical intervention. Of the 14 patients, 50% ($n = 7$) of the patients were stated to have remained disease free, and in 4 cases, recurrence was not specified. The three patients who experienced recurrence underwent subsequent surgeries, and one patient was left with a myoelectric prosthesis following complete loss of function of the affected fingers and forearm amputation was necessary due to epitheloid sarcoma discovered at the time of amputation of the digits.[11]

In terms of outcome, our data show that 50% ($n = 7$) of the patients treated for pediatric Dupuytren disease experienced a positive outcome following surgery. Successful outcome was defined as having no functional disturbance of the affected hand following surgery and rehabilitation. Interestingly, the three patients who experienced persistent limited hand function following surgery were also the same patients who experienced recurrence of disease. Outcome was not specified for the remaining four patients.

Discussion

The etiology of Dupuytren contracture is well described in the adult population and involves the interaction of several well-defined risk factors. Heavy alcohol consumption resulting in liver disease,[13] smoking,[14] diabetes mellitus,[13] and prior hand injury[15] have all been associated with an increased prevalence of Dupuytren contracture in adults.[16] Dupuytren contracture is most common amongst those of Northern European decent,[17] and occurs predominantly in males with a male-to-female ratio of 3:1.[18] It is uncommonly found in African American and Asian patients, and is exceedingly rare in the pediatric population.[18,19]

Although many theories have been proposed to explain the etiology of Dupuytren contracture in the adult population, the factors influencing the development of disease in the pediatric population remains largely unknown. Because of the very low incidence of Dupuytren contracture in the pediatric population, there have only been several risk factors identified. Included in these are previous hand injury, male sex, and a family history of disease, with some authors reporting a positive family history in as many as 53% of the male patients and 33% in the female patients.[16,20] Although the exact mechanism of inheritance remains unclear, one study suggests that a single gene behaving in an autosomal dominant manner is likely responsible.[20]

Although limited by sample size, our data suggest that early age of presentation is associated with lower rate of

recurrence. The mean age of presentation of children who experienced recurrence was 82 months versus 70.5 months in the group that did not experience recurrence.

We were unable to demonstrate a clinically significant difference with regards to the age of presentation of children who experienced a favorable clinical outcome versus those who did not. The mean age of presentation of the patients who experienced a positive functional outcome was 82.5 months versus 82 months for those with persistent limitation.

We were also unable to demonstrate any clinically significant association between surgical procedure performed and rate of recurrence or positive outcome.

There are several other fibrotic hand diseases that can present with similar clinical features, which need to be distinguished from Dupuytren contracture in the evaluation of the pediatric patient presenting with a hand mass.

Infantile digital fibromatosis (Reye tumor) almost exclusively affects the extensor surface of the fingers and toes, and may present as a single or as multiple dome-shaped lesions. These lesions tend to be painless, but they can lead to joint deformity. The presence of intracytoplasmic inclusion bodies on pathological examination is pathognomonic for infantile digital fibromatosis.[21]

A giant cell tumor of the tendon sheath is a firm benign neoplasm that most often affects the flexor surfaces of the fingers, but it can also appear in the hand and wrist. The lesions can be either diffused or localized, arise from the tendon sheath, with occasional intraosseous involvement, and typically cause pain and inflammation.[21]

Calcifying aponeurotic fibroma is a soft tissue tumor most often affecting the hands and feet. The tumors tend to be ill defined and can arise in relation to the skin or underlying tendons. Lesions tend to be solitary and painless.[21]

Plexiform Fibrohistiocytic tumor is a small, slowly growing mesenchymal neoplasm of intermediate malignancy most commonly affecting the upper limbs of children. The lesions tend to be multinodular, firm, and tend to arise from dermal or subcutaneous tissues.[21,22]

Congenital-infantile fibrosarcoma is a rare spindle cell tumor that can be present at birth or diagnosed during the early years of life. It mainly affects the extremities and can present in the hand and wrist. The tumor tends to grow rapidly, is painless, and can invade the underlying bone.[21]

Fibromas of the tendon sheath normally presents as small painless (or occasionally tender) subcutaneous nodules, most commonly affecting the fingers, hand, and wrist. The tumor tends to be well circumscribed and is almost always attached to the tendon sheath.[21]

Conclusion

In conclusion, although rare, Dupuytren disease can affect children and infants, and needs to be considered in the differential diagnosis of the pediatric patient presenting with mass or cord in the hand. Although the etiology of Dupuytren contracture in the pediatric population remains largely unknown, our data suggest that early age of presentation is associated with lower rate recurrence following surgery. Given this, one should not hesitate to operate on these patients to avoid recurrence of disease and long-term functional hand disturbance.

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