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DUPUYTREN'S DISEASE IN CHILDREN

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Although Dupuytren's disease of the hand has been reported in teenagers, it is generally considered to be a disease of adults. A series of nine children who developed Dupuytren's disease of the hand before the age of 13 years is presented. Eight had surgical removal of the diseased tissue and histological confirmation of the diagnosis before the age of 13 years and one at 14 years of age. The presence of the condition in young children and teenagers is discussed and the literature summarized.

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In young children, Dupuytren's disease of the plantar fascia, or Ledderhose's disease (Ledderhose, 1897), is well recognized (Pickren et al, 1951; Goetzee and Williams, 1954–1955; Aviles et al, 1971; Rao and Luthra, 1988). Children with this problem are seen not uncommonly in areas of the United Kingdom with a high incidence of Dupuytren's disease. Goetzee and Williams (1954–1955) excised histologically confirmed Dupuytren's tissue from the hand of a 14-year-old who had had the disease since the age of 11. In 1963, Hueston described one boy of 12 as the youngest case of "proven" Dupuytren's disease of the hand, implying that histology was available, although this was not stated in so many words (Hueston, 1963). Other than this case, we have been unable to find previous evidence in the literature of histologically confirmed Dupuytren's disease of the palmar fascia in children under teenage.

This paper presents a series of nine children, all of whom developed Dupuytren's disease when under 13 years of age. Eight of them had material which proved on histology to be Dupuytren's disease excised from the hand before the age of 13 (Table 1). The literature on the occurrence of the disease in young children and teenagers is also reviewed and summarized.

PATIENTS

Case 1

A 9-year-old boy presented in 1990 with a 6-month history of flexion contracture of the right little finger (Fig 1). There was no history of acute trauma and his past medical history was unremarkable. However, his father, who was aged 34 years, had significant Dupuytren's disease (Fig 2) and his maternal grandmother also had the condition.

Table 1—Dupuytren's disease before the age of 13 years

Authors	*Year*	*No. of cases*	*Age of onset (years)*	*Age presented (years)*	*Surgery reported*	*Histology reported*
A. Histologically confirmed						
Goetzee and Williams	1954–1955	1	11	14	Yes	Yes
Hueston	1963	1	—	12	Yes	Yes
Fletcher	1995	5	—	4	Yes	Yes *
			—	6	Yes	Yes *
			—	9	Yes	Yes *
			—	11	Yes	Yes *
			—	12	Yes	Yes *
Urban et al (this series)	1995	2	9	9	Yes	Yes
			10	10	Yes	Yes
B. Histologically unconfirmed						
Dupuytren	1832	1	birth	6	No	No
Todd	1927–1928	1	12	15	No	No
Kanavel et al	1929	1	11	36	Yes	No
Maurer	1936	2	0–10	—	No	No
Skoog	1948	1	12	—	No	No
Winkler	1958	1	12	15	Yes	No
Dahmen and Kerckhoff	1966	1	11	20	Yes	No
Mikkelson	1977	3	0–10	Adults	No	No
Rosenfeld et al	1983	1	10	13	Yes	No
Hueston	1991	1	8	8	No	No *

* Unpublished data.

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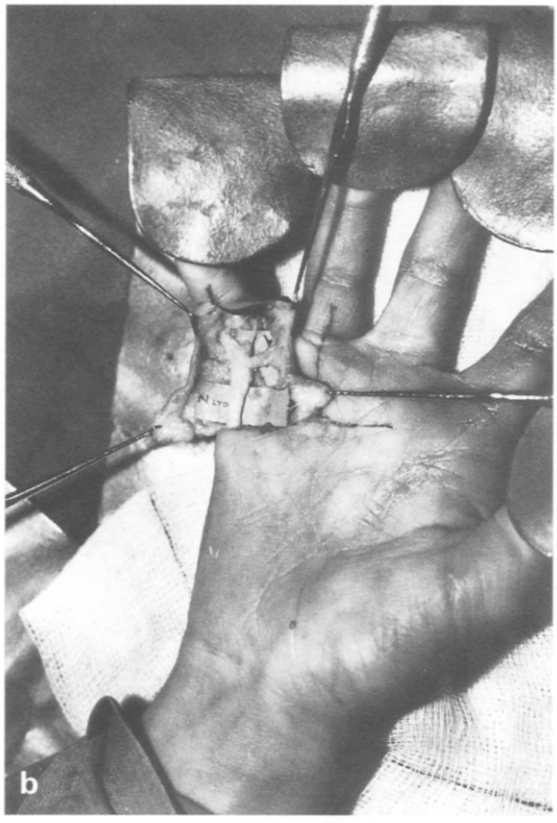

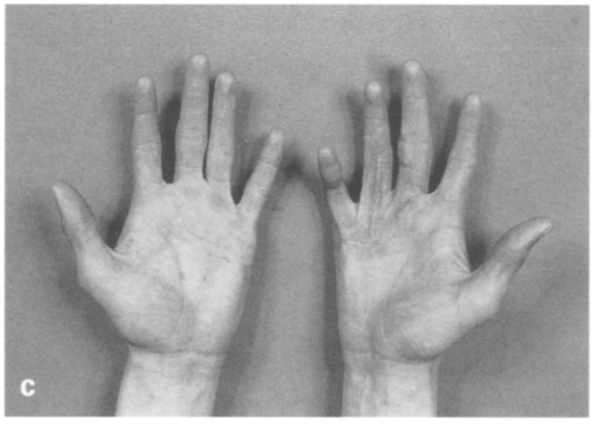

Fig 1 (a) The right little finger of case 1 on presentation at the age of 9 years, with a classical abductor band of Dupuytren's disease, subsequently excised and confirmed histologically. (b) Intra-operative view of the same. (c) Most recent views of the hands of case 1, now 14 years old, after dermofasciectomy to three digits of the right hand. There is a pit visible on the ulnar border of the left hand associated with early formation of an abductor band.

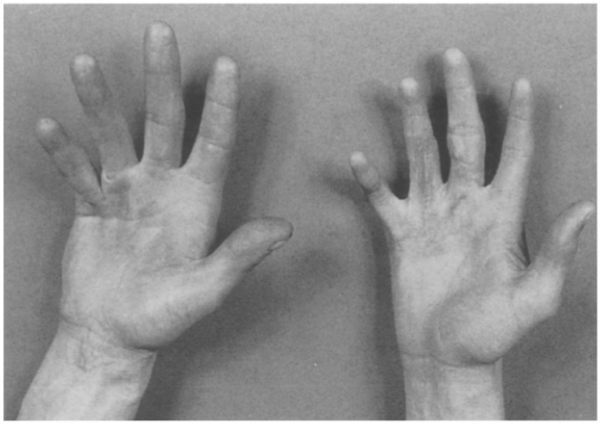

Fig 2 The right hand of case 1, age 14 years, (shown on the right) alongside that of his father, age 39 years, who has untreated Dupuytren's disease (shown on the left).

On examination, he had a typical abductor band of Dupuytren's disease of the right little finger extending to the middle phalanx with flexion contracture of the PIP joint, and extending through the natatory system to the fourth web and into the ulnar side of the base of the ring finger. Surgery was carried out in October 1990. In view of his young age and the absence of any personal experience or advice from the literature or from various experts on how to deal with the disease in children, a routine fasciectomy, and not a dermofasciectomy, was carried out. The disease densely involved both the skin over the proximal part of the little finger and the ulnar aspect of the PIP joint. After fasciectomy, a 10° lag in extension of the PIP joint remained. Post-operatively, extension of the PIP joint deteriorated during splinting. The child's co-operation was poor but the increasing flexion contracture may have been due to early recurrence of disease and not failure of splinting. In retrospect, we feel that a dermofasciectomy should have been carried out as the primary operation. His disease recurred in the pretendinous band of the little finger and the natatory system of the fourth web, with extension into the ring finger extending across the PIP joint to the middle phalangeal level. In May 1992, a dermofasciectomy of the ring and little fingers was carried out. Intraoperatively, he was found to have extensive disease of both digits. Excision of disease and overlying skin from the mid-palm to the middle phalanges was carried out. 2 months later, he was noted to have developed a nodule at the base of the opposite (left) little finger.

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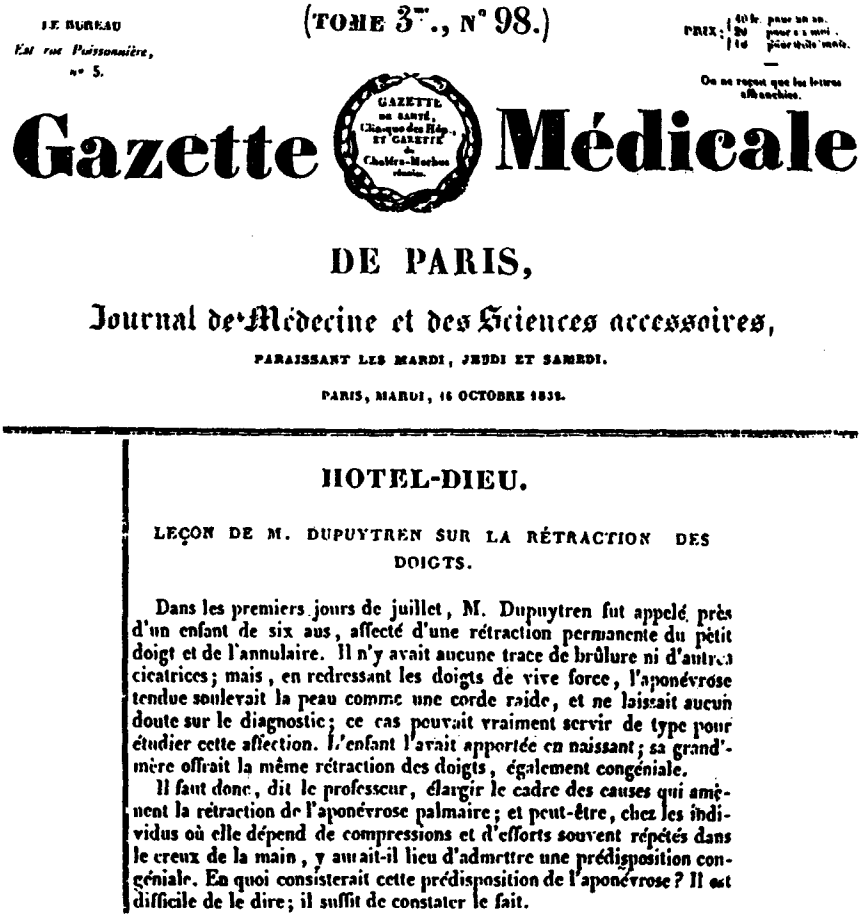
The disease of the right hand began to reappear alongside the graft in the palm 1 year after surgery. Within 6 months this had extended into the third web space and the ulnar border of the middle finger with a 20° flexion contracture of the MP joint. 9 months later (4 years after his first presentation), a further dermofasciectomy of the right hand was carried out to excise this disease which had now extended to the PIP joint of the middle finger. 6 months later, at the time of writing this paper, the boy is 14 years old and has just had surgery to the little finger of the left hand, in which a well-defined abductor band had developed. In view of the experience with the right hand, a dermofasciectomy was carried out.

Following each operation, histology has confirmed Dupuytren's disease. The first pathological specimen was sent for a second opinion with confirmation of the diagnosis.

Case 2

A 10-year-old keen musician was routinely admitted for circumcision. His mother drew attention to 2 nodules in the palm of the left hand, in the line of the index and middle fingers. These were asymptomatic. He gave no history of acute trauma to this hand and his past medical history was unremarkable. There was no known family history of Dupuytren's disease. Ultrasound confirmed that these nodules were within the subcutaneous tissues. The nodules were excised and histology confirmed them to be Dupuytren's disease. The specimens were re-examined by an independent pathologist who also



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DE PARIS,

Journal de Médecine et des Sciences accessoires,

PARAISSANT LES MARDI, JEUDI ET SAMEDI.

PARIS, MARDI, 16 OCTOBRE 1832.

HOTEL-DIEU.

LEÇON DE M. DUPUYTREN SUR LA RÉTRACTION DES DOIGTS.

Dans les premiers jours de juillet, M. Dupuytren fut appelé près d'un enfant de six ans, affecté d'une rétraction permanente du petit doigt et de l'annulaire. Il n'y avait aucune trace de brûlure ni d'autres cicatrices; mais, en redressant les doigts de vive force, l'aponévrose tendue soulevait la peau comme une corde raide, et ne laissait aucun doute sur le diagnostic; ce cas pouvait vraiment servir de type pour étudier cette affection. L'enfant l'avait apportée en naissant; sa grand'-mère offrait la même rétraction des doigts, également congéniale.

Il faut donc, dit le professeur, élargir le cadre des causes qui amènent la rétraction de l'aponévrose palmaire; et peut-être, chez les individus où elle dépend de compressions et d'efforts souvent répétés dans le creux de la main, y aurait-il lieu d'admettre une prédisposition congéniale. En quoi consisterait cette prédisposition de l'aponévrose? Il est difficile de le dire; il suffit de constater le fait.

Fig 3 The original record of the 6-year-old boy seen by Dupuytren in 1832. A translation of the report is given below.

Gazette Médicale de Paris, Tuesday 16th October 1832, Hôtel-Dieu. Lesson of Mr Dupuytren on Contracture of the Fingers. In the first days of July (1832), Mr Dupuytren was called to see a child of 6 years with a permanent contracture of the little and ring fingers. There was no trace of burn, or other scars, but, on straightening the fingers forcibly, the taut aponeurosis lifted the skin like a tight cord, leaving no doubt of the diagnosis; in fact, this case could have served as a classical example of the condition. The child had had this from birth and his grandmother had an identical contracture of the fingers, also of the same nature.

Professor Dupuytren said that it would be necessary to enlarge the list of causes of contracture of the palmar fascia; and, perhaps, in those individuals in whom the condition occurred as a result of compression and repetitive strains in the hollow of the hand, one would have to consider a predisposition of some kind. Of what would this predisposition consist? It is difficult to say; except to state that it existed.

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confirmed the specimens to have the appearance of Dupuytren's disease which had been present for a sufficient period of time to have matured beyond the proliferative phase.

Further histologically proven cases under teenage

In 1954–1955, Goetzee and Williams presented a case report of a 14-year-old boy who had surgery to Dupuytren's disease which had appeared in the foot at the age of 10 years and in the hand at the age of 11 years. Histological confirmation of the diagnosis, both in the hand and the foot, was available at 14 years. The degree of certainty of the age of onset of disease is unknown but it is likely that this child had Dupuytren's disease before reaching teenage. In 1963, Hueston reported the case of a 12-year-old-boy with histologically confirmed Dupuytren's disease. Seven other cases of histologically confirmed disease of the hand in children under teenage are known to the authors. These children had operations elsewhere and we have no clinical information about them. The operative specimens of these children were referred to a specialist oncological unit and all five specimens proved to be Dupuytren's disease (Fletcher, 1995).

Further unproven cases under teenage

In July 1832, a 6-year-old child was seen by Dupuytren himself (Fig 3). Although the clinical findings were said to have been pathognomonic of Dupuytren's disease, this child was reported as having had the condition from birth. This feature and the fact that there was, of course, no histology available leaves the diagnosis of this case in doubt (Elliot, 1988). The same is true of an 8-year-old with palmar disease shown to Hueston by another surgeon in Australia in 1991, although it could fairly be assumed that both Dupuytren and Hueston were more than capable of diagnosing the condition on clinical grounds.

Several other authors have recorded flexion contracture of the fingers in young children which they believed to be Dupuytren's disease in whom the diagnosis must remain in doubt because there was no histological confirmation. The exact age of onset is also in doubt in some of these cases as authors seeing patients at a later age were presumably reporting ages of onset given to them by the patients.

These cases have been summarized in the second half of Table 1. As can be seen from the Table, the group contains a mixture of children who were seen by surgeons before the age of 13 with a clinical diagnosis of Dupuytren's disease and older individuals whose onset of disease was, or was claimed to be, before this age.

Teenage patients and patients of unspecified age under 20 years old

The literature contains a number of references to patients presenting for the first time with Dupuytren's disease as

Table 2—Dupuytren's disease in teenagers and in patients of unspecified age under 20 years old

Authors	*Year*	*No. of cases*	*Age of onset (years)*	*Age presented (years)*	*Surgery reported*	*Histology reported*
Keen	1882	7	0–30	—	No	No *
Costilhes	1885	2	0–20		No	No
Nichols	1899	1	0–20		No	No
Krogius	1921	2	10–19	—	No	No
Kanavel et al	1929	1	19	27	Yes	No
Scholle	1930	1	10–20	—	No	No
Davis and Finesilver	1932	1	18	—	No	No
Maurer	1936	7	10–20	—	No	No
Stapelmohr	1947	2	0–19	—	No	No
Skoog	1948	1	13–19	—	No	No
Kohler	1960	1	18	25	Yes	Yes
Jayes	1969	1	16	19	No	No
Early	1962	1	—	15–24	No	No
Ling	1963	1	—	0–14	No	No
		3	—	15–24	No	No
		10	5–14	—	No	No **
		49	15–24	—	No	No
Dahmen and Kerckhoff	1966	15	0–20	—	Only 3	No
Clarkson	1974	2	—	11–20	No	No
Mikkelson	1977	12	10–20	adults	No	No
Fletcher	1995	2	—	16	Yes	Yes ***
			—	16	Yes	Yes ***

* 2, possibly 3, of the 7 cases reported by Keen were described as congenital, making the diagnosis unlikely.
** Ling's paper contains his own series then a combined series of his own work combined with unpublished data from Early's work of 1962 which is included above in brackets.
*** Unpublished data.

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teenagers. Some authors have given the exact ages of the patients; other papers only record that the patients were within the teenage group. In addition, there are a number of papers which either record only that the patients were under 20 years old or have banded the ages in such a way that it is not possible to determine whether the patients were teenagers or under 13 years of age. All of these papers have been included in Table 2. Only one of these papers includes histological confirmation of the diagnosis. We have obtained histological evidence in a further two cases in which specimens taken from the palms of teenagers proved to be Dupuytren's disease (Table 2). Clinical details of these patients are not known (Fletcher, 1995).

DISCUSSION

Flexion contracture of the fingers in children may be due to a number of causes. The most common is camptodactyly which was recognized by Landouzy in France and by Adams in England at the end of the last century (Adams, 1890; Herbert, 1898). Less common, and more recently described, is congenital ulnar drift (Zancolli and Zancolli, 1984). These conditions make a diagnosis of Dupuytren's disease in childhood or in the teenage years only absolutely tenable where histological confirmation is available. Cases in which histology was not available but which may have been Dupuytren's disease in childhood are included in the second half of Table 1. Early references to congenital or very early onset of Dupuytren's disease which are more likely to have been one of the above conditions have not been included (Hutchinson, 1897; Bunch, 1913; Grieg, 1917).

This small group of cases indicates a rare but definite presence of Dupuytren's disease in children under 13 years of age. Our very limited clinical experience suggests that these children should be treated aggressively by dermofasciectomy if they develop progressive finger contracture.

Acknowledgement

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