

Dupuytren's Contracture

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This article was prompted by the frequency of Dupuytren's contracture found in the small community of Sandur (Population 31.12.1965: — 571) in the Faroe Islands.

The percentage of the affected people seemed in excess of the accepted figures and further studies showed that all the recorded cases are descendants of the same family.

The first cases appear after the arrival in Sandur of a Danish Priest in 1783. He married on arrival with a local girl and all subsequent cases are his descendants.

History.

The condition was first described by Platter in 1610, but has been known as Dupuytren's contracture since Dupuytren described the actual pathology in 1832 (*Mercer 1955*).

This is a localized condition with thickening of the palmar fascia which involves the overlying skin and which shows a strong tendency to contract and draw the affected fingers into rigid flexion. The condition is essentially bilateral, although the manifestation on two sides may not be simultaneous.

It may be associated with a similar condition of the plantar fascia. It is a progressive condition, although the rate of progress may be infinitely variable, affecting principally the

fingers of the ulnar side. The ring fingers are more often affected followed by the little, middle, and forefingers.

It is a disease whose manifestations become most apparent during the middle and latter period of life, although it has been described at almost any age. There appears to be a predominancy in men of about 7 to 1.

Pathology.

In Dupuytren's contracture part of the palmar fascia becomes thickened by fibrosis and undergoes contraction. The short fibres from the fascia to the skin proliferate and contract and obliterate all the subcutaneous fat, the sweat glands and the blood and lymph vessels so that a continuous layer is formed between the epidermis and the palmar fascia.

Histologically in the slower and more chronic condition, the skin shows great thickening of the cornified layer, flattening of the stratum mucosum and obliteration of the papillae of the corium. At a deeper level there is a dense layer of cicatricial tissue. This tissue is more cellular and vascular in the early stages but becomes thick, dense and fibrous.

The fibrous masses are poorly demarketed and fade out into the surrounding fascia. These masses are composed of small spindle shaped cells with elongated nuclei.

The cytoplasm is scanty and poorly outlined. The cells are embedded in a matrix of fibrous tissue whose weavy pattern is suggestive of normal fascia. Mitoses are not seen and the cells are all alike. These fibrous masses are identical with the benign fascial fibromas seen elsewhere.

In the more rapidly extending and cellular types it may be difficult to differentiate the tissues from that of fibrosarcoma on purely histological grounds.

Aetiology.

Innumerable theories have been advanced since the condition was first described. It has been thought to be congenital and familiar, neoplastic, inflammatory, traumatic, as result of con-

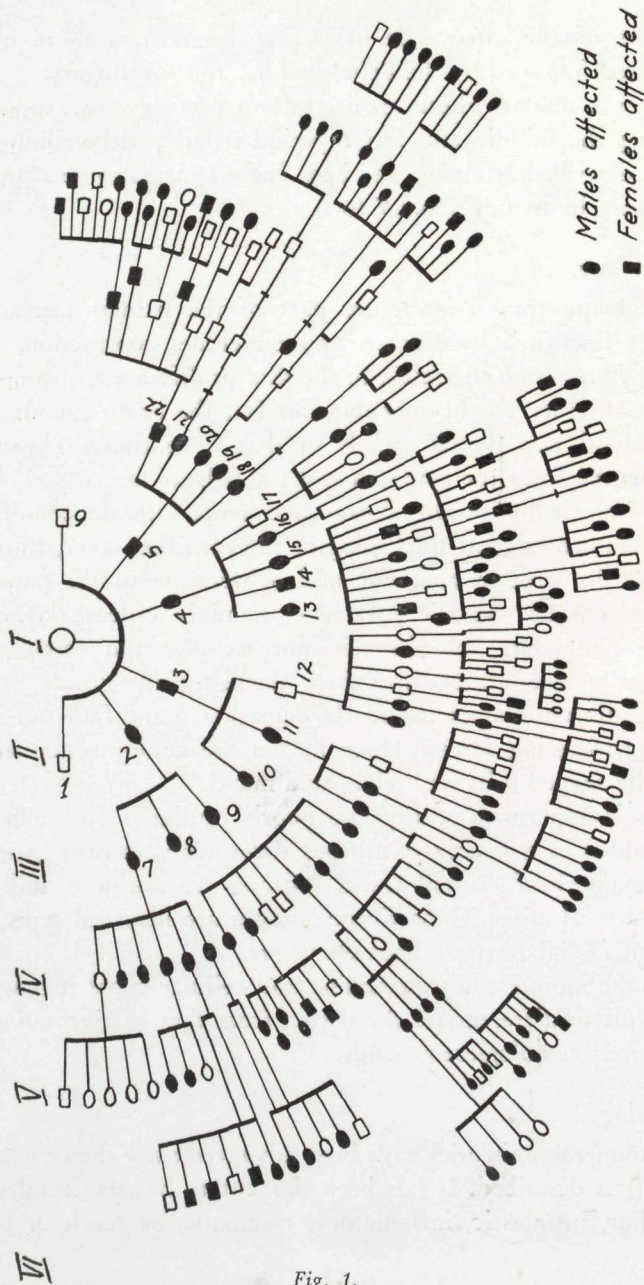


Fig. 1.

stitutional inherent disease or collagen disease associated with rheumatic disorders.

There appears to be no end to the possible aetiological factors which have been discussed by various authors. However there are many aspects for consideration. The condition is prevalent in males. The condition becomes apparent mostly during the middle and latter period of life. It is a condition which may be multicentric in origin and may affect any part of the fascia of the hand, although commoner to the ulnar two fingers. It is a progressive disease. In a number of patients a strong family history has been obtained. Although heredity is thought to be a factor the degree of its importance and its mode of action remains unknown.

While the information is suggestive of a familiar factor this does not contribute much to the understanding of the inheritance of Dupuytren's contracture.

Family History.

The family history is summarized in the family pedigree Fig. 1, based on an investigation of all the inhabitants of the islands of Sandoy and Skúvoy (population 31.12.1965: — 1699) and of the parish register for these islands since 1783, 6 successive generations of affected individuals, including males and females suggest autosomal dominant inheritance. The members of the sixth generation below the age of 25 were not included, neither were children of the previous generation who died before they reached the same age. Two (2) of the female members of the second generation 1 and 6 married and left for elsewhere and it was not possible to trace the descendants.

On the average in this type of inheritance, one half of the children of the affected parents can be expected to be similarly affected. However, it seems that more than the expected number of the affected family members are found in this pedigree. The finding of unaffected children from affected parents might be considered as evidence of a reduced or low expressivity of the mutant gene which has led to Dupuytren's contracture in others.

Symptoms.

The first sign is the appearance of a small hard nodule in the palmar fascia followed by a progressive flexion contracture, most commonly of the ring finger.

The nodule enlarges to become a cord like band of contracted fascia and the overlying skin becomes puckered and bound to the contracted fascia. The patient occasionally complains of aching although actual pain is uncommon. Paraesthesia as a result of the distortion of the digital nerves by the thickening bands may appear rarely.

The affected fingers are flexed at the metacarpo-phalangeal joints and proximal interphalangeal joints and extended at the terminal interphalangeal joints.

The flexion of the fingers is due to the contracture of the prolongations of the palmar fascia to the digit. These bands do not extend as far as the terminal phalangs so they remain extended.

Treatment.

A variety of conservative methods have been tried including cortisone and vitamin E. (Steinberg 1946), without benefit to the patient.

Operation is the only procedure to influence the rate of progress. It is the only weapon to relieve contracture and prevent permanent fixation of the joints and crippling.

The surgery (Bunnell 1944, Einarson 1946) of Dupuytren's contracture has ranged from a conservative multiple subcutaneous fasciotomy to the ultraradical fasciectomy. It is of great importance to consider the age of the patient in relation to the expectation of life and in relation to his work.

The operation is not curative. It only relieves existing contracture for a varying period of time.

An important feature of the operation is the management of the overlying skin which is usually adherent to the contracted band of fascia. In severe cases skin grafting may be necessary for closure of the operation wounds.

SUMMARY

A large number of people affected by Dupuytren's contracture, all members of the same family in Sandoy, Faroe Islands, in whom the incidence of Dupuytren's contracture suggests an autosomal dominant mode of inheritance for this condition, is reported.

A brief account of the history, pathology and treatment is given.

ÚRTAK

Sagt verður frá stórum tali av fólki við kropnaðum fingrum — contractura fasciae palmaris (Dupuytren) — öll í somu ætt í Sandoyar læknaðömi. So sum kropnaðir fingrar koma fyri í hesi ætt, bendir tað á, at hetta brek hevur eina autosomala, ráðandi (dominanta) arvgongd.

Í stuttum verður sagt frá søgu og sjúkugongd hjá hesum breki og viðgerðini.

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