A clinical genetic study of familial Dupuytren's disease in the Netherlands.

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Abstract

Hypothesis: In order to elucidate the genetics of Dupuytren's disease, we investigate the mode of inheritance of familial Dupuytren's disease in the Netherlands.

Method: Patients with Dupuytren's disease with two or more affected first-degree relatives were asked to participate in the study together with their family. In five families, all members were clinically examined. In the six other families, the diagnosis of Dupuytren's disease was based on information provided by the proband. All participants completed an extensive questionnaire.

Results: Eleven pedigrees, consisting of 475 family members and including 66 subjects diagnosed with Dupuytren disease, were studied. Of the affected family members, 67% were male and 33% female. Paternal transmission was observed in 45% of cases and maternal transmission in 55% of cases. Fifty-two percent of the offspring of the affected individuals also have Dupuytren's disease.

Summary: Our data suggest an autosomal dominant mode of inheritance for familial Dupuytren's disease, but with a reduced penetrance. This confirms previous studies.