

Eighteen years follow-up study of the clinical manifestations and progression of Dupuytren's disease

Kristján G. Gudmundsson¹, Reynir Arngrímsson², and Thorbjörn Jónsson³

¹The Health Care Centre, Blönduós, ²Medical Genetics Unit, Faculty of Medicine, University of Iceland, Reykjavik, Iceland, ³Institute of Immunology (IMMI), The National Hospital, Oslo, Norway

Objective: To evaluate the clinical manifestations and progression of Dupuytren's disease.

Methods: In 1981-82 a total of 1297 men were examined for Dupuytren's disease, and of these 19.2% had the disease. In 1999 those with signs of the disease in 1981-82 were invited for a follow-up study. As controls symptom free individuals from the study in 1981-82 were invited.

Results: A total of 53 individuals from the control group had developed Dupuytren's disease in 1999. Men with palmar nodules/fibrous cord in 1981-82 were more likely to develop contracted fingers than those without Dupuytren's disease. Patients with young age at disease onset more often required operations than those with later onset. Of the men who had been operated 70% still had finger contractures in 1999.

Conclusion: The incidence of Dupuytren's disease is high in elderly men. Dupuytren's disease is progressive in nature and most operated patients have recurrent finger contractures.

Key words: Dupuytren's disease, Dupuytren's contracture, incidence, prognosis

Dupuytren's disease bears the name of a French surgeon who described this condition in the Lancet in 1834 (1). The disease begins with the formation of fibromatous nodules in the palms, usually in the ulnar side. The nodules progress and form a fibrous band or cord lying from the palm to the fingers. Eventually this leads to permanent finger contractures. The ring finger is most commonly affected, followed by the little finger (2). The prevalence increases with age, males are more often affected, and familial predisposition is common (3,4). It is most prevalent in caucasians of North-European origin, especially in Scandinavia and the British Isles (5,6). Several risk indicators of Dupuytren's disease have been reported, including smoking (7), alcohol (8), diabetes mellitus (9), epilepsy (10), and manual work (11,12), although this is debatable (13). Dupuytren's patients complain less frequently of joint swelling and morning stiffness (14) and rheumatoid arthritis seems to be infrequent among them (15). It is relevant in this context that several immunological deviations have been described in patients with Dupuytren's disease (16,17). Most studies have focused on the etiology and surgical treatment of Dupuytren's disease, but little has been done to investigate the natural progression of this common disorder. The aim of the present study was

to evaluate the clinical manifestations and natural progression of Dupuytren's disease.

Patients and methods

Design of the Reykjavik study

Iceland is a 103.000 sq.km island in the North Atlantic Ocean with approximately 270.000 inhabitants. In 1967 a large population based health survey was started in the Reykjavik area of Iceland (The Reykjavik Study), mostly focusing on the epidemiology of cardiovascular diseases (18). The participants of the study were males born in the years 1907-1934 and females born in 1908-1935 with residence in the Reykjavik area. The study cohort was divided into six groups; A, B, C, D, E, and F according to birth dates and these groups were invited to be examined up to six times during the period 1967 to 1997. As a part of the Reykjavik study 1297 men were evaluated for signs of Dupuytren's disease in 1981-82. Of these 249 (19.2%) had clinical signs of Dupuytren's disease (14,19).

The present study

In 1999, as a nested case control study, all Dupuytren's patients alive (n=122) from the 1981-82 study and a matched control group (n=122) were invited to a follow-up examination. The controls had all been symptom free in 1981-82 and were matched in pairs for age and smoking habits. The participants were invited with a letter and those not responding were contacted by telephone. The same medical doctor examined all the participants. The study participants answered a

Thorbjörn Jónsson, Institute of Immunology (IMMI), The National Hospital, Oslo, NO-0027, Norway.
E-mail: thorbjorn.jonsson@rikshospitalet.no

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Table I. Clinical staging of the study cohort in 1981–82 and in 1999.

Clinical staging in 1999	Clinical staging in 1981				
	Not Dupuytren's disease	Palmar nodule or fibrous cord	Finger contractures	Operated	Total
Not Dupuytren's disease	48	8	0	0	56
Palmar nodule or fibrous cord	37	41	2	0	80
Finger contractures	12	14	3	0	29
Operated	4	12	7	5	28
Total	101	75	12	5	193

The incidence per year for developing contracted fingers was approximately 1%, and the incidence for developing nodules in the hand approximately 2% per year.

Risk for contracted finger for men with stage 1 compared to stage 0 in 1981–82: RR=2.2; 95% CI= 1.3–3.8, P=0.004.

structured questionnaire and their hands were evaluated for signs of Dupuytren's disease. The hands were graded as follows: a) normal, b) those with palmar nodules larger than 5 mm or fibrous cord and c) those with contracted finger and those operated on for Dupuytren's disease. The degrees of flexion of the fingers and number of affected fingers were registered. The study was approved by the Medical ethical committee and the Data protection committee in Iceland. The examinations were free of charge and took place at the Heart Preventive Clinic in Reykjavík.

Statistical evaluation

Statistical evaluation was performed using SPSS software. The chi-square test, Student's T-test, and relative risk (RR) with 95% confidence interval (CI) were used for evaluation of the findings. The level of significance was set at $P < 0.05$.

Results

Table I shows the clinical findings from the examination in 1999 in relation to previous findings from 1981–82. **Of 101 men without clinical signs of Dupuytren's disease in 1981–82 a total of 53 (52.5%) had developed clinical signs at the follow-up examination in 1999. These included 4 who had been operated and 12 with contracted fingers. Of the 75 men who had palmar nodules or fibrous cords in 1981–82 a total of 26 (34.6%) had developed contracted fingers or had been operated in 1999.** Thus, men with palmar nodules in 1981–82 were significantly more likely to develop contracted fingers than those with no signs of Dupuytren's disease, (RR = 2.2; 95 % CI= 1.3–3.8). Of the 12 men with contracted fingers in the former evaluation 7 (58.3%) had been operated in 1999. The incidence of Dupuytren's disease in the disease free group was 2.9% per year and the incidence of developing finger contracture was 0.9% per year.

No Nodules: 16/101 contract = 16% ~1%/year

32 Nodules: 26/75 contract = 35% ~2%/year

Of men with Dupuytren's disease in 1999 a total of 58 could give reliable information about the age at onset of the clinical manifestations (Table II). It can be seen that Dupuytren's patients with onset before the age of 50 years were significantly more likely to be operated on for contractures than those with disease onset after the age of 50 years ($P = 0.019$). Furthermore, the number of operations needed was related to the age at first operation. Thus, of 7 men undergoing operation before the age of 50 years 5 (71%) were reoperated and 2 of these were operated 3 times or more. Of 20 men operated for the first time after the age of 50 years only 5 (25%) needed reoperation ($P = 0.029$).

shows clinical manifestations of Dupuytren's diseases in the right and left hands. Most patients had manifestations in both hands. The right hand was slightly more often affected than the left hand, especially if only one hand was affected. Table IV shows that 16 men had two or three fingers in the right hand with 30 degree of flexion or more and in the left hand 10 had the same deformity. The numbers of contracted fingers and operated in right and left hands are shown in Table V. Of the 24 patients being operated in the right hand 19 (79.2%) still had one or more contracted fingers. Similarly 10 (66.7%) of those operated on the left hand still had contracted fingers.

Table II. Relation between disease onset and operation due to Dupuytren's contracture. Reliable information about onset of disease manifestations available for 58 patients.

Clinical staging in 1999	Disease onset	
	≤50 years (n=28)	>50 years (n=30)
Palmar nodule or fibrous cord	8 (29%)	8 (27%)
Contracted fingers	3 (11%)	13 (43%)
Operated	17 (61%) [†]	9 (30%)

[†]P=0.019 compared to patients with disease onset >50 years of age.

Table III. Clinical findings in the 57 men with operated hands or contracted fingers.

	Affected hand	
	Right	Left
Total number of affected hands [¶]	53	44
Only one hand affected	13	4
Both hands affected	40	40
Operated hands	24	15
Contracted fingers [‡] ;		
One	17	18
Two	14	9
Three	17	13
Total number of contracted fingers	96	75
Flexion deformity [‡] ;		
<30°	30	24
30–60°	12	9
>60°	6	7

[¶]Affected hands refers to operated hands or hands with contracted fingers.

[†]19 patients had two or three fingers affected on both hands.

[‡]Flexion deformity refers to the finger with maximal deformity.

Discussion

The study shows that Dupuytren's disease has high incidence and a highly progressive course. The incidence of Dupuytren's disease was almost 3% per year during the 18 year follow-up period. Thus, more than 50% of the men selected as controls, i.e. symptom free in 1981–82, developed clinical signs of Dupuytren's disease during the follow-up period. Most of them only developed mild disease, palmar nodules or fibrous cords, but 16 of the men developed contracted fingers or were operated. Although these figures seem high, they should perhaps not be surprising, as several previous studies have shown high prevalence of Dupuytren's disease among elderly males of Northern-European origin. Thus, up to 40% of elderly men in Norway, Iceland, and Scotland are reported to have clinical signs of

the disease (4,19,20). And this can be compared to 2% incidence found amongst young diabetic patients in Finland (21). Furthermore, smoking is a risk factor for the development of Dupuytren's disease and as the controls were matched for smoking, the men under observation were heavy smokers.

According to the results presented Dupuytren's disease is highly progressive, causing hand deformity that does not only have physical consequences for the person affected, but also social consequences. Thus, an inability to play musical instruments is one of the most obvious handicap of patients with the disease, possibly affecting the career of professional musicians. Dupuytren's patients are facing increasing handicap in the modern technical world, where contracted fingers interfere with work on computers and small instruments. Dupuytren's disease has also cosmetic aspects and the disease can under certain social situations cause embarrassment, for example when shaking hands with people and not being able to straighten the fingers. Due to these reasons it is important to define the etiology and course of the disease in order to be able to find a cure that is simpler and more effective than surgery.

Men with palmar nodules in the previous study from 1981–82 more often developed contracted fingers during the follow-up period than the symptom free men. Thus, palmar nodules/fibrous cords are definite sign of Dupuytren's disease, later causing finger contractures. This indicates that it may become possible in the future to take preventive measures at an early stage and treat palmar nodules or fibrous cords, to prevent further progression to finger contractures.

Eight men were reported to have palmar nodules or fibrous cords in the initial study in 1981–82, but were judged normal at the follow-up examination in 1999,

Table IV. Relation between the number of affected fingers and degree of contraction.

Degree of contracture	Right hand (number of fingers)			Left hand (number of fingers)		
	One (n=17)	Two (n=14)	Three (n=17)	One (n=18)	Two (n=9)	Three (n=13)
<30°	15	9	6	12	8	4
30–60°	2	3	7	3	1	5
>60°	0	2	4	3	0	4

Table V. Number of affected fingers in each hand in men with Dupuytren's disease.

Clinical status in 1999	Right hand		Left hand	
	Contracted (n=29)	Operated [†] (n=24)	Contracted (n=30)	Operated [‡] (n=15)
One contracted finger	11	6	14	4
Two contracted fingers	10	4	9	0
Three contracted fingers	8	9	7	6

[†]5 men operated but without finger contractures in 1999.

[‡]5 men operated but without finger contractures in 1999.

and two patients with contracted fingers in 1981–82 had fibrous cords in 1999. The study performed in 1999 was blind, and the observer did not have any information of the clinical status in the former study. One of the men with contracted finger in the former study and diagnosed with fibrous cord at the examination in 1999 said that he had been cured from contracted fingers by accident, straightening up the finger. Similar results of traumatic release of finger contracture are described in the literature (22,23). The second man with finger contracture in the former study and diagnosed fibrous cord in 1999 had a hand deformity reminding the observer of finger osteoarthritis, possibly affecting the diagnosis. But spontaneous regression of the fibromatous process could explain the difference between the former study and the present study (24). Alternatively it is reflecting an inter-observer variation in the diagnosis of Dupuytren's disease. The method we used to diagnose Dupuytren's disease is similar to those used in several previous studies (4,5,19,20). Contracted or operated fingers are an obvious sign of Dupuytren's disease with high inter-observer diagnostic specificity (20). However, palmar nodules or fibrous cords are more difficult to diagnose and inter-observer diagnostic specificity is reported to be lower (20,25).

Approximately two thirds of the operated patients still had finger contractures at the follow-up examination in 1999. It was not infrequent for these men to have two or three contracted fingers on one hand. This observation reflects the progressive nature of Dupuytren's disease. Furthermore, it shows that surgical correction does not stop the disease process and highlights the need for new treatment.

In conclusion, during an 18 year follow-up period a high incidence of Dupuytren's disease was found. Furthermore, the development of palmar nodules are an early sign of finger contractures. Patients with early onset of disease tend to have an aggressive disease course and often require surgical correction. Dupuytren's disease is progressive in nature and most operated patients develop new finger contractures.

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