

Long-term course of the severe haemophilia A or B patients in Sweden: Results from a cohort register study

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AIM

To evaluate long-term incidence, prevalence and survival in Swedish patients with severe haemophilia.

BACKGROUND

The cohort consists of patients enrolled in a large national registry, including 1,431 with haemophilia A or B, born between 1883 through 2008. Registry data were linked to the In- and Out-patient, Cause of Death-, Cancer-, Medical Birth-, Prescription-, Migration- and Multi-Generation registries. Severity of haemophilia was known for 934 out of the 1,431 patients.

Methods

The 384 patients with severe haemophilia were compared to 1,918 age and sex matched controls. Cox proportional hazard regression models were used to estimate hazard ratio and the follow-up was stopped at 60 years after evaluation of the proportional hazards assumption.

Results

The mean follow-up was 30.7 for the patients with severe haemophilia and for the control cohort 31.6 years. 78 of the 384 patients were diagnosed with HIV and 167 with viral hepatitis. The hazard ratio for all cause of death was 6.6, 95% CI: [4.5; 10.0], $p < 0.001$. When HIV positive patients were excluded the hazard ratio was estimated to 3.3, 95% CI: [1.9; 5.6], $p < 0.001$. Exclusion of both HIV and/or viral hepatitis gave a hazard ratio of 8.2, 95% CI: [3.2; 20.8], $p < 0.001$. The most frequent cause of death for the patients with severe haemophilia was found to be HIV/AIDS, 24 (31%) followed by haemorrhage related deaths/cerebrovascular disease 18 (23%) and the comparative numbers for the control cohort 0 (0%) and 6 (6%).

Conclusions

This study shows that patients with severe haemophilia have a higher risk of death both with and without HIV and/or viral hepatitis compared to controls. This unique cohort will provide further valuable insight into co-morbidities and allow evaluation of first degree relatives.

Table. Characteristics of the study cohort for patients with severe haemophilia and for their controls.

Severe haemophilia	Total Study Group		After Exclusion of Patients with HIV Infection		After Exclusion of Subjects with HIV Infection and/or Viral Hepatitis	
	Haemophilia n (%)	Controls n (%)	Haemophilia n (%)	Controls n (%)	Haemophilia n (%)	Controls n (%)
All						
Number of persons	384 (100)	1918 (100)	306 (100)	1529 (100)	196 (100)	979 (100)
Death	78 (20)	102 (5)	31 (10)	63 (4)	14 (7)	24 (3)
HIV-infected	78 (20)	1 (0)	0 (0)	0 (0)	0 (0)	0 (0)
Viral hepatitis	167 (44)	9 (1)	110 (36)	7 (1)	0 (0)	1 (0)
Emigration	2 (1)	129 (7)	2 (1)	90 (6)	2 (1)	42 (4)
Follow up < 60 yrs						
Death	66 (17)	61 (3)	23 (8)	37 (2)	12 (6)	11 (1)
Emigration	2 (1)	128 (7)	2 (1)	89 (6)	2 (1)	42 (4)

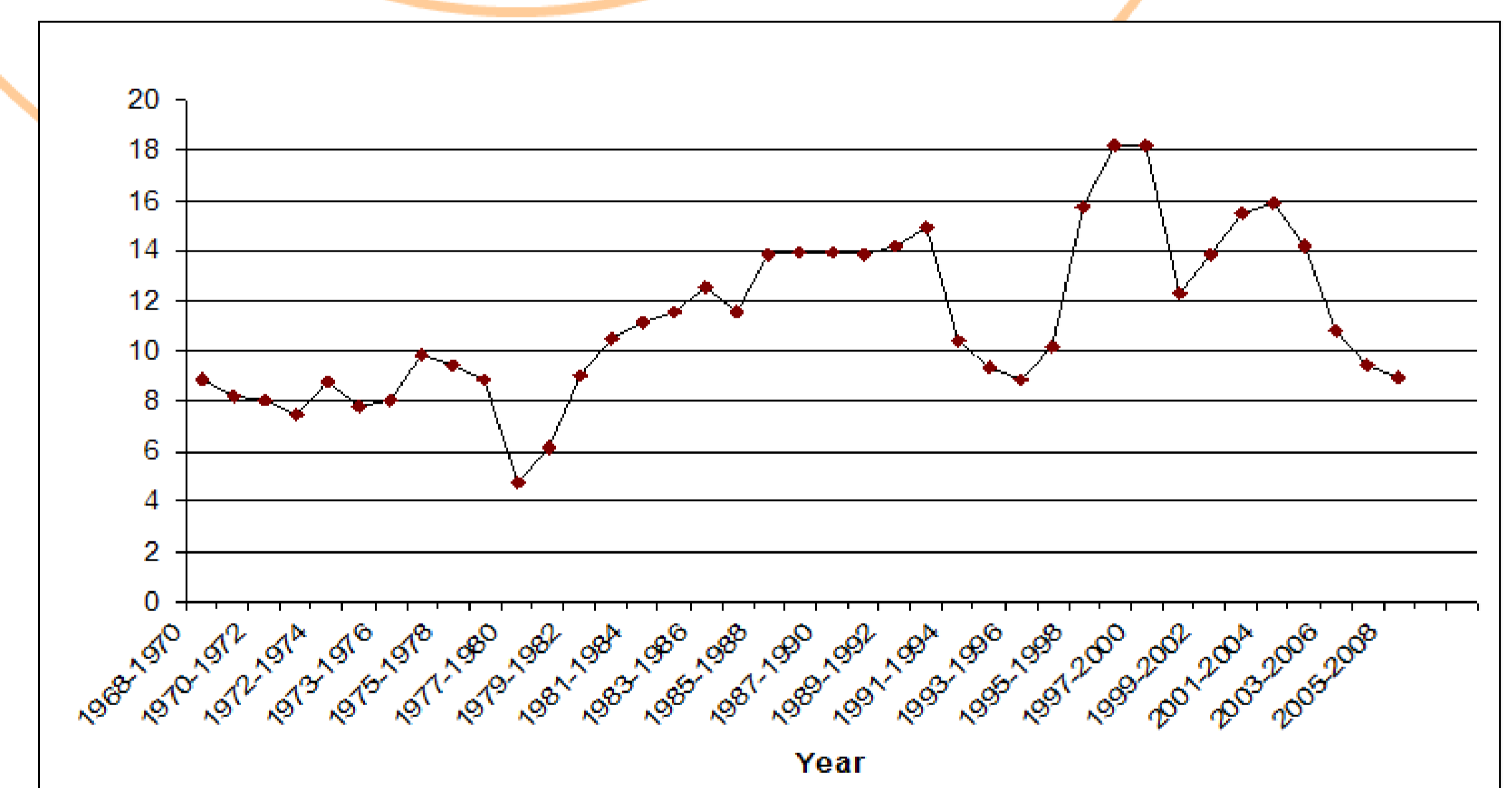


Figure 1. Three year moving incidence (per 100,000) for severe haemophilia patients.

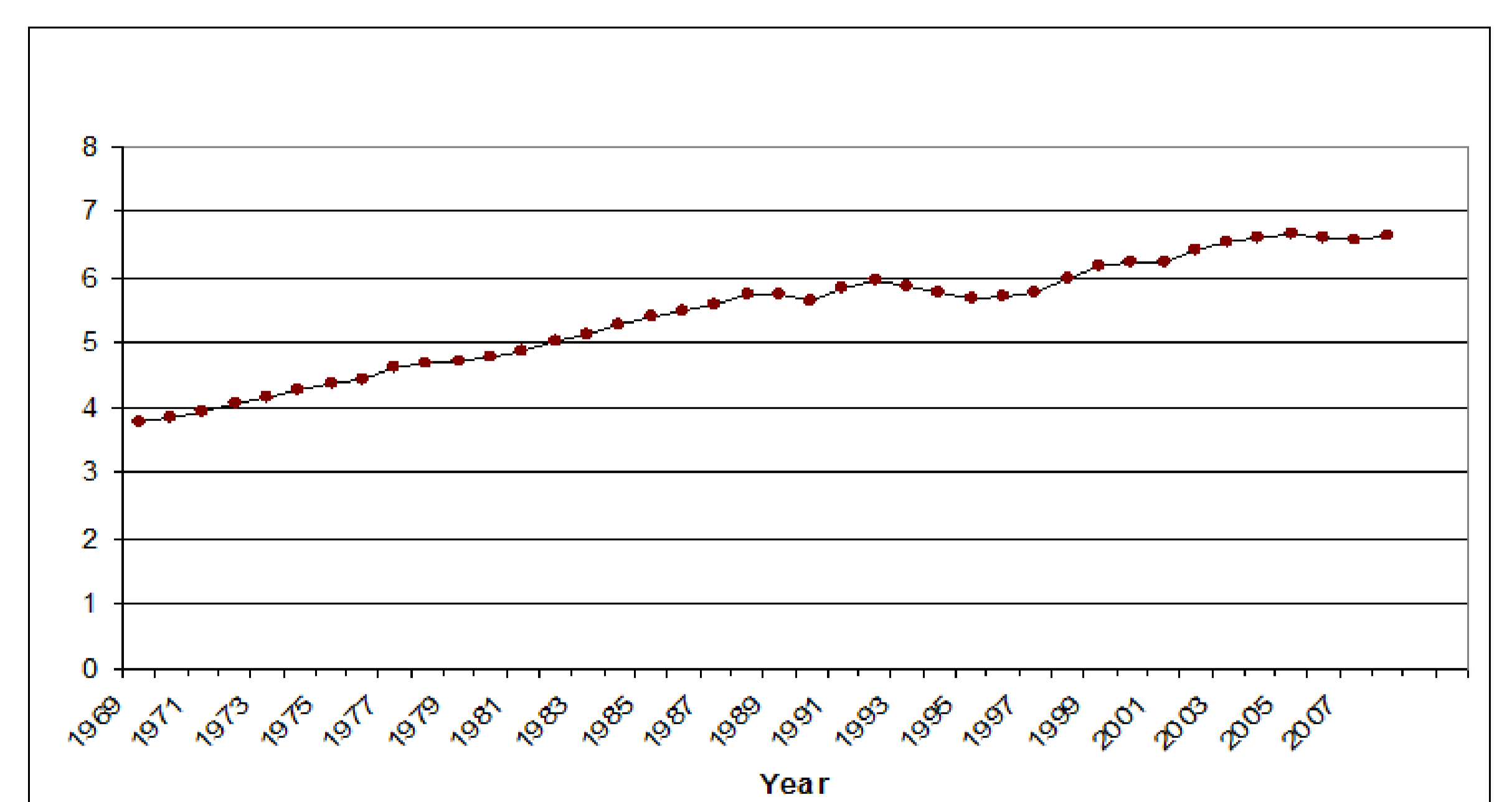


Figure 2. The prevalence (per 100,000) for the patients with severe haemophilia.

