

Mortality from scleroderma in England and Wales 1968-1985

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Abstract

Mortality data for scleroderma derived from death certificates from England and Wales for the years 1968-1985 were analysed. The overall crude mortalities were 0.9 and 3.8 per million per year in men and women respectively, which are similar to comparable data from the United States. There has been a small but statistically significant increase in mortality of 3% a year over this period, possibly reflecting an underlying increasing trend in incidence. Such analyses are subject to the limitation of the accuracy of death certificates, but in a disease as rare as scleroderma the death rates provide useful data on the trends in occurrence and death from this disease.

Population mortalities for diseases such as scleroderma, which are both rare and have a relatively high fatality rate, can provide an alternative to the more costly and time consuming multicentre surveys in yielding estimates of incidence. Additionally, if the natural history and response to treatment are relatively constant, both over time and between different populations, then a comparison of mortalities can provide a guide to both time and geographical trends in incidence. Furthermore, knowledge of both the incidence and mortality for scleroderma within the same population can be used to derive an estimate of patient survival, which otherwise would require the follow up of a large series of patients. Published mortality data from the United States show little change in mortality between 1959 and 1961¹ and between 1969 and 1977² with rates of about 1.5 and 3 per million per year in men and women respectively. In this report mortality data from the United Kingdom for 1968-85 are examined and their use for estimating comparative incidence and survival considered.

Methods

Scleroderma was first identified separately in the eighth revision of the International Classification of Diseases,³ and information on mortality from scleroderma has thus been available since 1968. The total numbers of deaths by sex and year of death for England and Wales were obtained from the Office of Population Censuses and Surveys for the years 1968-1973 and by age and sex from published annual data for the years 1974 to 1985.⁴ The numbers of deaths in any individual year were too small to yield reliable estimates of mortality, and thus the

crude and the age and sex specific annual mortalities were calculated as the average annual mortality over the 12 year period 1974-1985 using million person years at risk in each age sex group over this period as the denominator. Trends during the period of observation were compared by examining three year moving averages—that is, the mean annual mortality during 1968-1970, 1969-1971, etc—to reduce the effect of random variation from a single year's data. These average annual rates were adjusted for age and sex using indirect standardisation taking the 12 year age and sex specific rates calculated above as those of the standard population. Linear regression was used to assess the magnitude and significance of any underlying trend. There were virtually no deaths from scleroderma in children during this period, and thus all rates are based on the adult (aged 15+) population.

Results

The results show overall mortalities of 0.9 and 3.8 per million per year in men and women respectively. Figure 1 shows the age mortalities by age and sex, demonstrating a negligible mortality in early adult life. There is a peak in both sexes in the 65-74 decade and a subsequent decline. There is a marked female excess at all age groups, but the largest relative excess of around 5:1 is seen in the 25-34 and 35-44 age groups compared with a ratio of 2.5:1 in the seventh and eighth decades. When a straight line was fitted to the age adjusted data this suggested a statistically significant average annual increase of about 3% during the 17 years of study ($\beta=3.13$, 95% confidence interval 1.76 to 4.52) (fig 2). The death rates for men varied during this period between 0.6 and 1.6 per million per year and those for women from 3.2 to 4.8 per million per year. There was a

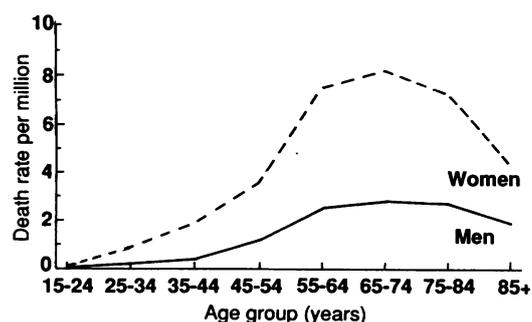


Figure 1 Mortality from scleroderma by age group for England and Wales 1974-1985.

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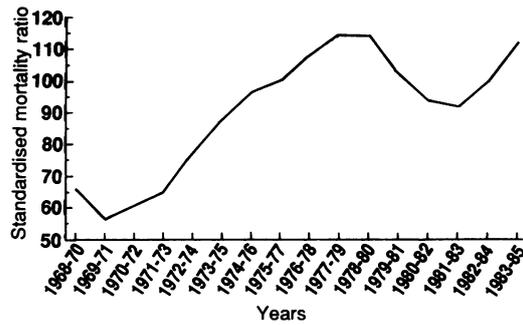


Figure 2 Trends in scleroderma mortality for England and Wales 1968-1985, using three year moving averages. (Standard population rates taken from fig 1.)

weak but statistically insignificant trend for the ratio of women to men to increase in the more recent years.

Discussion

The data used in deriving these estimates are based on the underlying cause of death as entered on the death certification. Inaccuracies in the recording and coding of the true underlying cause of death on death certificates are recognised as the major source of error in such analysis in similar chronic diseases such as rheumatoid arthritis.⁵⁻⁷ It is perhaps reasonable to assume, but this is not tested, that errors in estimates of scleroderma as a cause of death are more likely to lie in the direction of false negatives—that is, failure to record a death as due to this cause, rather than false positives with an inappropriate diagnosis of scleroderma. Thus the rates quoted are possibly underestimates. Further, it is unlikely that there is a selective bias in the error rate between the different age and sex groups. Thus the age and sex effects noted are likely to reflect the true demographic effects on mortality.

The average increase in annual mortality of 3% approximates to a total increase of around 50% during the entire period. It seems unlikely that patient survival has deteriorated; if anything the use of agents such as captopril has improved the outlook in severe renal diseases.⁸ The possibility that the increase in mortality reflects differences in death certificate coding or completion remains unanswered. There is no a priori reason, however, to suspect that this occurred in either direction or specifically towards an increased mention of scleroderma. More probably, perhaps, scleroderma is increasing in incidence in England and Wales, reflecting a similar increase, based on clinically ascertained cases, in the United States.⁹

It is perhaps reassuring that the rates obtained in this study are close to those found by a similarly conducted analysis in the United States.² The total population mortalities in that

population were 1.5 and 3.5 per million per year in men and women respectively, figures which were also similar to data gathered some 10-15 years earlier in the United States.¹

There are few data on the incidence of scleroderma in the United Kingdom. Such data that are available suggest a minimum incidence, based on diagnosed patients attending hospital, of 1.1 and 6 per million per year in men and women respectively.¹⁰ These estimates would suggest therefore that about 60% of women and an even higher proportion of men with scleroderma ultimately die from their disease. These figures seem very high, though they are comparable with survival data from the largest series of 309 patients in the United States, which suggested a nine year corrected mortality of about 62%.¹¹ Many cases of scleroderma are probably undiagnosed in the community¹² and thus would not appear in either incidence or survival data generated from hospital attenders. The 'hospital' incidence of scleroderma, as stated, is apparently increasing,⁹ though this in part might be due to greater ascertainment of less severe cases, and thus future studies of outcome might show a more favourable prognosis.

In conclusion, mortality from scleroderma in England and Wales is broadly similar to that in the United States, and the underlying incidence and increasing trend of the disease in the two countries is probably the same. Further, these mortality data, albeit with their imperfections, reinforce the results from hospital series of a high ultimate fatality from this disease in patients diagnosed in hospital.

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