

The Increasing Prevalence of Haemophilia

Dear Sir,

Treatment of haemophilia patients depends on an adequate supply of blood products. As long as clotting factor preparations produced by recombinant DNA techniques are not widely available, changes in the number of patients may threaten the precious balance of blood donations and plasma products consumption.

The observed prevalence of haemophilia is the resultant of the prevalence at birth and mortality. It may be affected by genetic counseling of carrier women and selective abortion and also by the fertility of patients. Until only 20 years ago haemophilia patients had a life expectancy less than 30 years (1), and therefore the prevalence we observe today will be much smaller than the prevalence at birth. Nowadays, life expectancy is only mildly reduced (2), aside from the present AIDS mortality which differs from country to country. The present longevity of haemophilia patients will therefore lead to an increase in their number.

This is illustrated in Fig 1, which shows the prevalence of haemophilia per age group in The Netherlands. These data were obtained by a survey of all registered 1,162 Dutch haemophilia patients. Information was available for 80% of the patients, the prevalence data have been extended to all 1,162. In each age group the number of patients is divided by the total number of men in The Netherlands of that age (3). Up to age 20 (mid interval) the prevalence increases. This is likely to be caused by delays in diagnosis, mainly of mild haemophilia. After age 40 (mid interval), there is a steady decline of the age specific prevalence. This deficit of older patients can only be explained by excess mortality in the past. The plateau of 20.6 per 100,000 males approximates the prevalence at birth. If this prevalence were present at all ages, the number of haemophilia patients would be almost 1,500, i.e. 28% more than the present number of haemophilia patients in The Netherlands.

At the moment the mortality in haemophilia is twice as high as in the general population, aside from AIDS mortality. This leads, as calculated by life table method, to a reduction in the overall prevalence of about 10%. Prenatal diagnosis and selective abortion might affect the prevalence at birth, this effect will be minimal however, since we found in a recent survey of 549 female relatives of haemophilia patients that only a minority of carriers choose for this option (4), which corroborates reports from the United Kingdom (5). Furthermore, we noted that the younger patients were relatively more often married than the older patients. Haemophilia patients had on average less children than men in the general population, which was partly caused by their being less often married, and partly by genetic and physical considerations. The improved physical condition of haemophilia patients is therefore likely to lead to an increase in the number of carrier daughters of haemophilia patients, and in two generations this will cause a further increase in the prevalence at birth.

The number of HIV positive haemophilia patients and the ensuing mortality differ widely from one country to another. In The Netherlands the number of infected patients is relatively low (less than 20%) (6), and therefore the rise in the number of patients will occur in one generation. In countries in which the majority of patients is HIV positive, excess mortality may at first lower the prevalence, but this will be a temporary effect. The attitude towards prenatal diagnosis may also be different in other countries. Nevertheless, we think that the substantial growth in

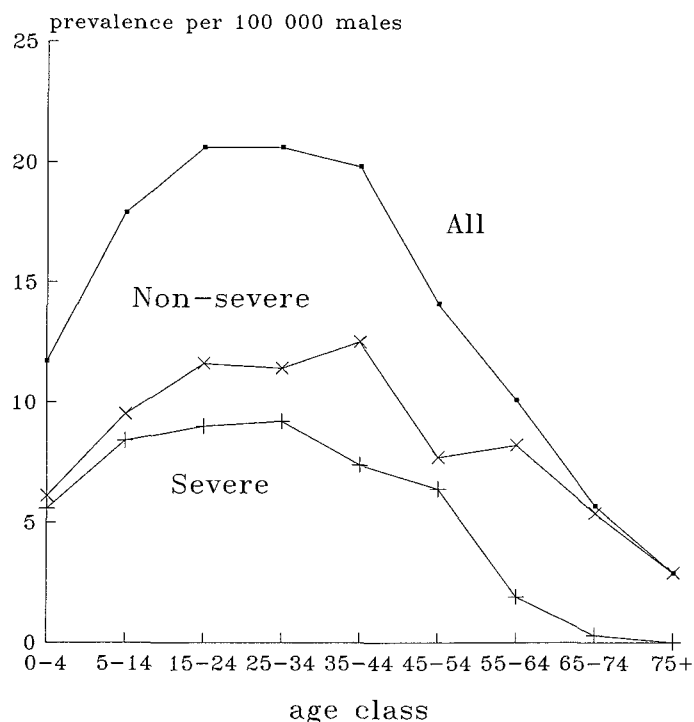


Fig 1 Prevalence of haemophilia by age. For each age group the number of haemophilia patients, corrected for non response, is related to the number of individuals in that age group in the general male population.

the number of haemophiliacs by about 20% we foresee in The Netherlands is likely to occur in many Western countries. The "extra" patients will be mostly those with severe haemophilia, since they showed the largest deficit in the older age groups because of excess mortality in the past.

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