Life expectancy in osteogenesis imperfecta

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No information is available on life expectancy in osteogenesis imperfecta; it is often assumed that this is impaired. In a recent review this is described as a severe disease in which "some patients reach adulthood."[1] We were stimulated by requests for information for a patient told by an insurer that compensation would be reduced because of "poor life expectancy," and for an affected child who wished to claim from her father's estate. We sought to obtain reliable figures for life expectancy from our large survey of patients with osteogenesis imperfecta.[2]

Patients, methods, and results

We limited our survey to patients in England and Wales. Patients were excluded if they could not, with confidence, be assigned to one of the Sillence types,[3] or if they had the perinatal lethal form (type II). Patients identified only after death were excluded. Patients whose survival to 1 January 1993 was uncertain or whose date of death was unknown were excluded. In all, 743 patients (383 type IA, 77 type IB, 123 type III, 90 type IVA, and 70 type IVB) were included. For each the date of entry was the date of ascertainment; those notified before 1 January 1980 were given that date of entry. The date of exit was 1 January 1993 or the date of death, if earlier. The total number of patient years was 6970; there were 57 deaths.

We carried out separate analyses of type IA, type III, and a composite group of moderate severity (types IB, IVA, and IVB). We determined the number of years at risk for each subject within each age range (0-1, 1-5, 5-10, 10-15, 15-25, 25-35, 35-45, 45-55, 55-65, and 65-75). For each age, sex, or type group we recorded the observed mortality and compared it with that expected from the life tables for 1981.[4] Life expectancy figures were determined by applying the mortality ratios to the life tables for each age group.

In osteogenesis imperfecta type IA the overall mortality ratio was 1·08 (95% confidence interval 0·64 to 1·81). We could not therefore distinguish mortality in these patients from that in the general population. In the composite group the overall mortality ratio was 1·93 (1·17 to 3·13). Figure 1 shows values for life expectancy.

In patients with osteogenesis imperfecta type III the mortality ratio varied significantly with age and sex. We determined the death rate in each age group and smoothed the rates by fitting a linear function of 1/age to determine mortality ratios up to age 45. The number of deaths in older type III patients was small; values for mortality ratio in later life would have been unreliable. To calculate life expectancy, we assumed that the smoothed mortality ratios at age 45 applied at older ages. The figure shows the results.

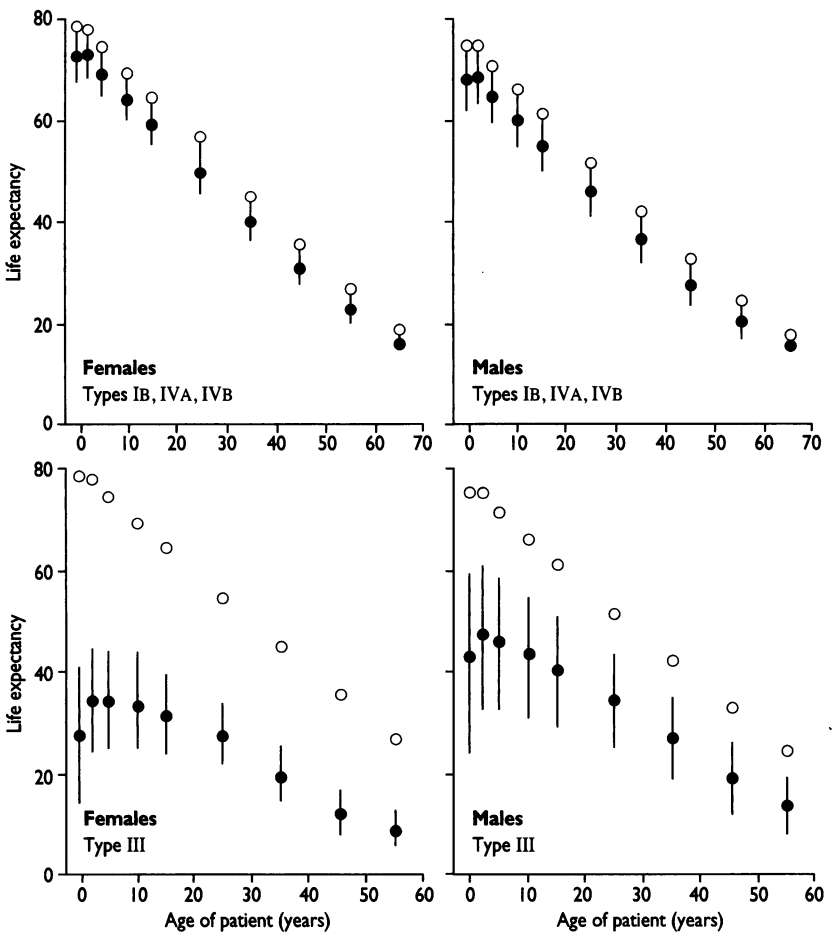

Fig 1—*Life expectancy in osteogenesis imperfecta (●) in comparison with that in the general population (○). Vertical ratios determined with the GLIM statistical package[5] bars represent 95% confidence intervals for mortality*

Comment

We found that life expectancy in osteogenesis imperfecta type IA is similar to that in the general population. In the composite group of patients with types IB, IVA, and IVB, life expectancy was reduced to a modest extent. It was not unexpected that among patients with osteogenesis imperfecta type III life expectancy was impaired. Of the 26 deaths, however, 19 had occurred before the age of 10. Patients surviving beyond this age seem to have a better outlook.

Our findings indicate that a more optimistic account of life expectancy in osteogenesis imperfecta can be given than was hitherto thought possible. Parents of affected children are often given an inappropriately poor prognosis by incautious clinicians. Even in the severe type III disorder life expectancy is better than is often suggested.

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