Summary

Two cases of Reye's syndrome, complicated by acute reversible renal failure, are presented. One case followed an influenza A virus infection.

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S. NICHOLLS,* D. GILL, and J. CRASKE

The Hospital for Sick Children and Guy's Hospital London; and Public Health Laboratory, Poole General Hospital, Dorset.

*Correspondence to Dr. S. Nicholls, Department of Paediatrics, Guy' Hospital, London SE1 9RT.

Life tables for cystic fibrosis

In this study a comparison is made between the survival rates obtained from life tables of 215 children attending The Hospital for Sick Children during the 5 years from 18 January 1969 to 31 December 1973, and the two previous series reported from this hospital (Mantle and Norman, 1966; George and Norman, 1971). The life tables were derived according to accepted statistical methods (Hill, 1966), which have been described in the previous papers. Criteria for acceptance were as in previous studies. Children who were seen purely for confirmation of diagnosis and who were not followed up have not been included.

Cystic fibrosis not presenting as meconium ileus

In the present series 35% were diagnosed before the age of 6 months compared with 26% in the previous 5 years. 46% were diagnosed by one year in the present study compared with 43% in the earlier ones. There was only one death under the age of 7 years in the present (1969–73) series. Mortality was diminished at all ages (Fig. 1) and the gap between the proportion alive at the end of each period when entered at diagnosis, which tends to overestimate mortality, and when entered at birth has narrowed considerably up to the age of 15. After this age the data become less meaningful due to losses from transfer and death.

Cystic fibrosis presenting as meconium ileus

Of the 55 cases in this group the obstruction was relieved by Gastrografin enema in 5 infants, by laparotomy alone in one, and by small intestinal resection usually with a Bishop-Koop anastomosis and ileostomy, in 49. 91% were alive at one month compared with 81% in the preceding survey; 85% at one year compared with 68%; and 79% at 5 years compared with 54% (Fig. 2). However, cases presenting with meconium ileus have a poorer life expectancy than those in the other group, even excluding those dying during the first 6 months (Fig. 3).

Discussion

There has been an improvement in life expectancy in both groups, which is most marked up to 5 years in the meconium ileus group and up to 7½ years in the group with no meconium ileus. Death in the meconium ileus group is usually due to pulmonary sepsis, but this is closely associated with poor early progress and a prolonged stay in an acute hospital ward, as a result of extensive intestinal resection and consequent malabsorption and malnutrition. It is likely that those cases that escape with a Gastrografin enema or simple laparotomy only, fare very much better, but the numbers are too small for statistical analysis.

The results, though confirming that cystic fibrosis remains a serious condition with a limited life expectancy for many of the older children, give good reason to expect that life expectancy will continue to improve for the more recent cases, given earlier diagnosis and effective management with avoidance of serious respiratory infection in the early years. Unfortunately, the present data do not indicate any trend to earlier diagnosis after the first 6 months. Although there is a clear clinical impression that early diagnosis is of major importance in the prevention of serious respiratory infections, the life expectancy of those presenting during the first 12 months is no greater than the average for all cases (Fig. 4). The value of early diagnosis will only be settled when neonatal screening with effective follow-up has been carried out in a co-ordinated manner over a wide region.

Summary

The past 5 years have seen a continuing im-improvement in life expectancy in cystic fibrosis.

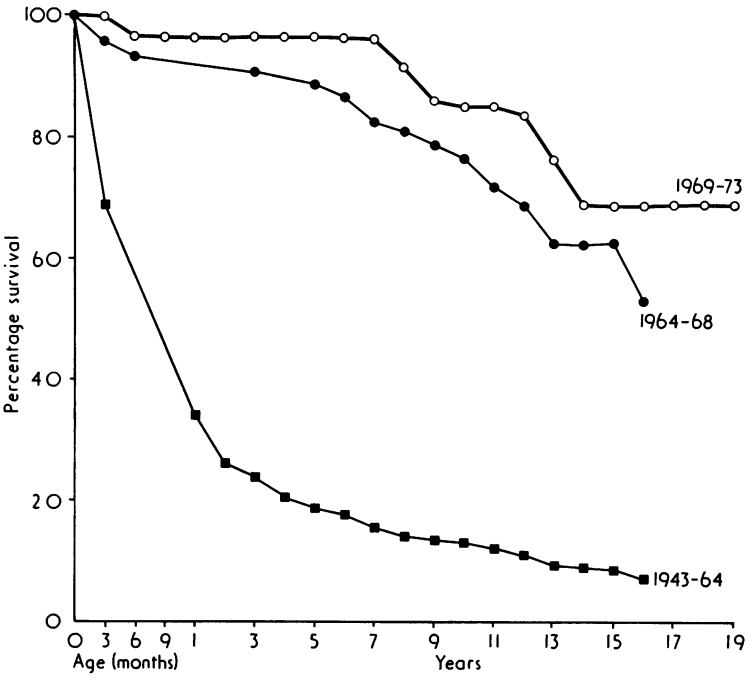

FIG. 1.—*Cystic fibrosis not presenting as meconium ileus, survival rates.*

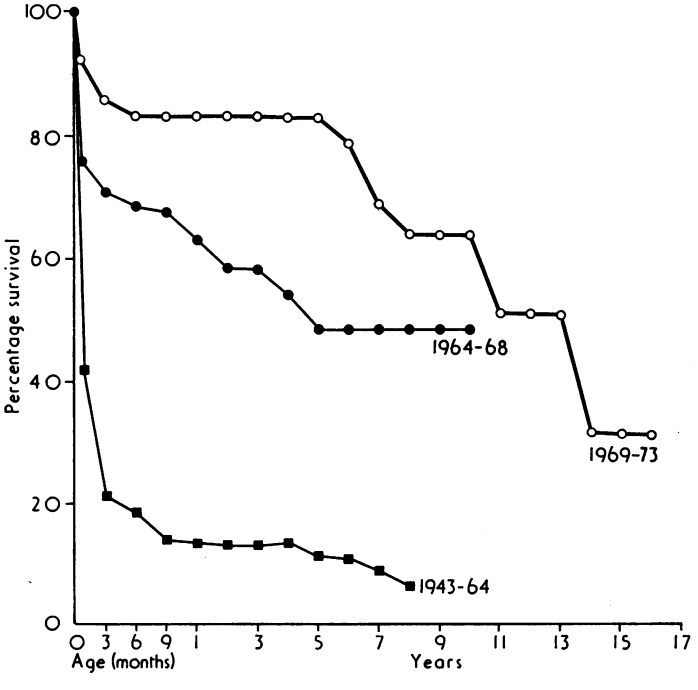

FIG. 2.—*Cystic fibrosis presenting as meconium ileus, survival rates.*

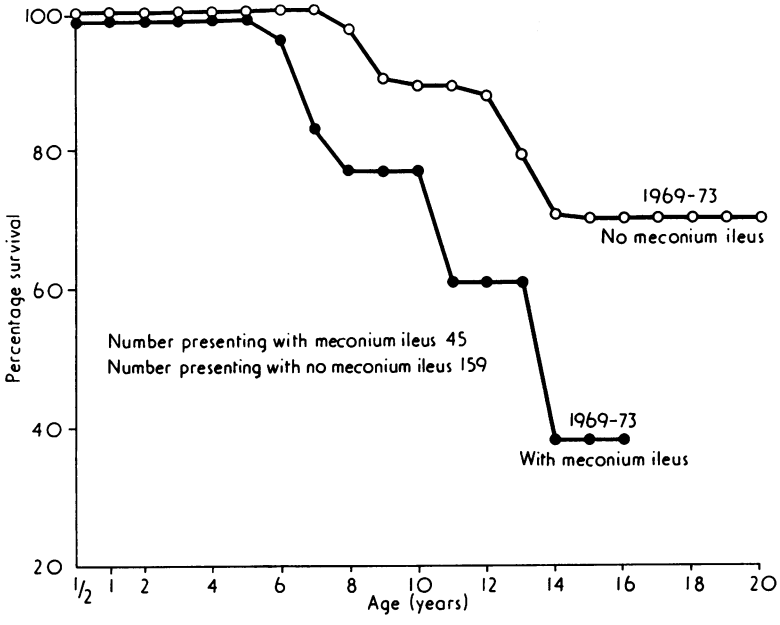

FIG. 3.—*Life expectancy of children alive at 6 months.*

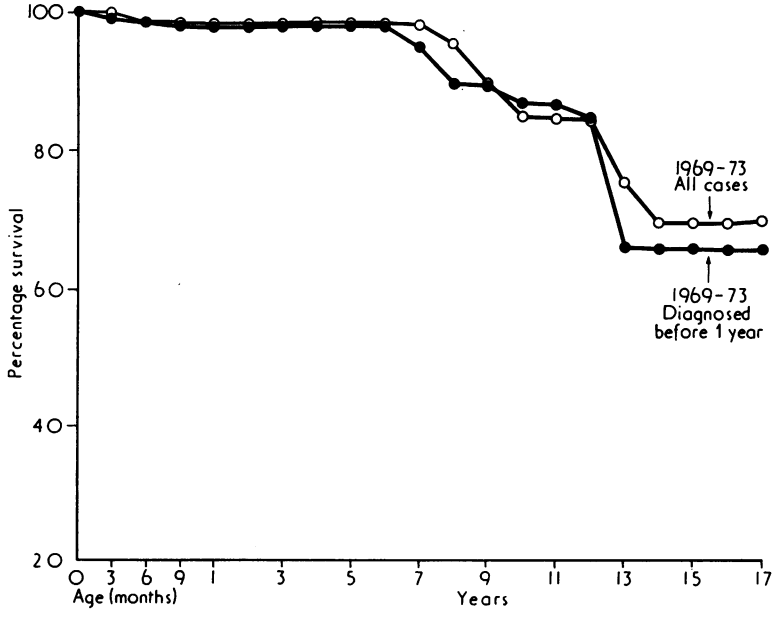

FIG. 4.—*Survival without meconium ileus related to age at diagnosis.*

In the group not presenting with meconium ileus 97% are alive at 5 years compared with 89% during the previous 5-year period and 19% in the earliest survey. Of those presenting with meconium ileus 79% are alive at 5 years, compared with 54% and 13% in the earlier survey. The remarkable improvement in prognosis for those presenting with intestinal obstruction is undoubtedly due to better surgical management. At present, however, the meconium ileus group fares worse than the other group even when deaths during the first 6 months have been excluded. Possible reasons for this are discussed.

There was only one death under the age of 7 years in those presenting without meconium ileus. It appears that the diagnosis is being made with increasing frequency in the first 6 months of life but no sooner at the later ages. The numbers diagnosed before the first pulmonary infection were too few to show what effect this may have on life expectancy.

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M. J. ROBINSON and A. P. NORMAN*
The Hospital for Sick Children, Great Ormond Street, London WC1 3JN.

*Correspondence to A.P.N.