

Final Report of Completed Studies

Extrahepatic Biliary Atresia

Background

Extrahepatic biliary atresia (EHBA) is the commonest cause of obstructive jaundice, liver failure and liver related death in children and accounts for greater than 50% of paediatric liver transplants. However, its incidence and prevalence in Australia is unknown. Retrospective studies show outcome is improved by early diagnosis and surgical intervention.

Objectives

- To describe the epidemiology of EHBA
- To determine clinical features at presentation, initial management and short term outcome
- To estimate the potential requirement of liver transplantation for this disease

Case Definition

Any child born in Australia after 1 January 1985 (when paediatric transplantation became available) with surgically defined and microscopically confirmed atresia or fibrous occlusion of the extrahepatic biliary tree.

Newly diagnosed (incident) and previously diagnosed (prevalent) cases seen in the previous month, but not previously reported, should be notified.

Results and discussion

Results are reported for January 1993 to December 1996, inclusive. Information was available for 250 (92%) notified cases. From these, 110 cases of EHBA were confirmed, of which 55 were incident cases (born between January 1993 to December 1996) and 55 were born prior to 1993. 93 duplicate reports were received and were used to supplement clinical information. Of the 55 incident cases, 15 were born in 1993, 19 in 1994, twelve in 1995 and nine in 1996. Between 1993 and 1996 inclusive, the estimated national incidence of EHBA was 5.4/100,000 live births. The geographical distribution of cases is shown in Figure 12. Incidence did not differ significantly between states. Seasonal distribution of cases is shown in Figure 13. The trend toward more cases being born in the winter months (June, July, August) was significant.

Figure 12 Reported incident cases of EHBA by State/Territory (proportion of total cases)

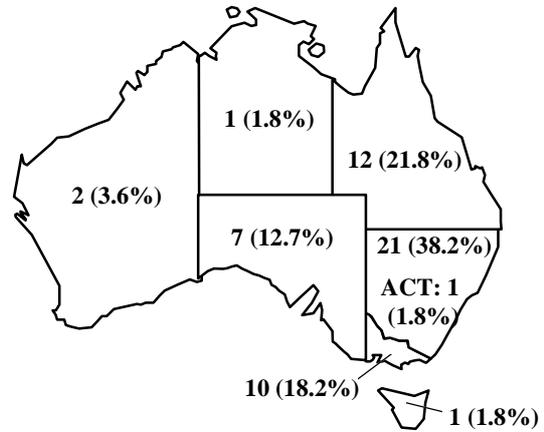
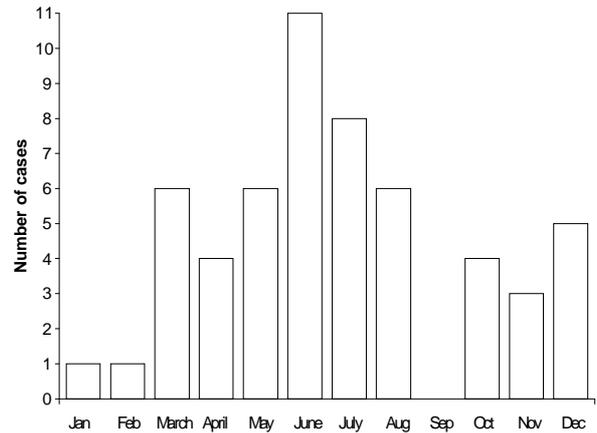
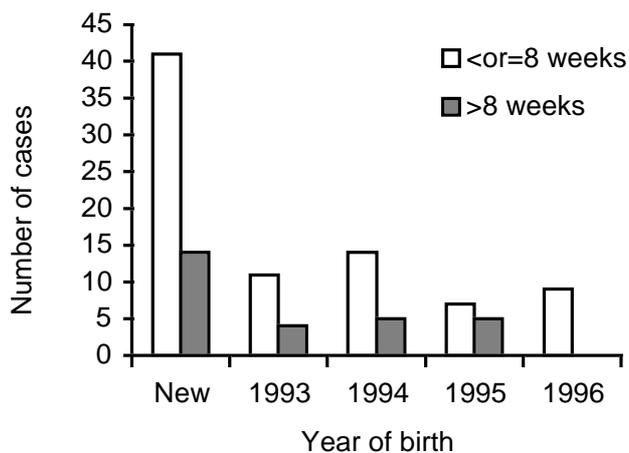


Figure 13 EHBA cases by month of birth



Twenty nine cases (53%) were female. The majority (78%) were Caucasian, three (6%) were of Asian origin and three (6%) were indigenous Australian. Mean age at diagnosis was 2 (SE±0.1) months (range 0.5-6.0 months), which was similar to that of children born prior to 1993 (2.1(SE±0.2) months). One quarter of all cases born after 1993 were diagnosed at greater than eight weeks of age (Figure 14). No cases reported in 1996 had been diagnosed after eight weeks of age. The mean age at diagnosis was similar for cases born before and after 1993, as was the proportion of those diagnosed after eight weeks of age.

Figure 14 Age at diagnosis of EHBA



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All but one of 55 incident cases had a Kasai (portoenterostomy) as the initial surgical procedure at a mean of 2 (SE±0.9) months (range 0.5 - 6) and clearing of jaundice was achieved in 30 (55%). One child had end-stage liver failure on diagnosis at four months of age and is awaiting a liver transplant. By twelve months after Kasai, nine children had been transplanted at a mean of 9.6 (3.7) months and two currently await transplantation. One child died awaiting and one died after liver transplantation. Of cases born prior to 1993, 21 (38%) have been transplanted to date at a mean of 21 (SE±5) months. Preliminary analysis of follow-up data shows increased rates of persistent jaundice and early death, and a trend to increased rates of end-stage liver failure and requirement for early liver transplant in late diagnosed cases compared to children diagnosed before eight weeks.

Surveillance of EHBA has now ceased. Ongoing follow-up of cases during 1997 will allow clarification of these preliminary findings and provide further information about outcome.

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