Hepatic portoenterostomy for biliary atresia

A comparative study of histology and prognosis after surgery

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SUMMARY Specimens of excised tissue from the porta hepatis in 26 infants with extrahepatic biliary atresia undergoing hepatic portoenterostomy were analysed histologically for the presence and size of biliary ductules. No correlation could be found between the establishment of effective biliary drainage and the number or size of biliary ductules. It is suggested that prognosis after surgery may be related to the intrahepatic lesion and age of the child at operation rather than to the histology of the extrahepatic bile duct remnants.

The mean survival for untreated infants with the 'non-correctable' form of extrahepatic biliary atresia is 19 months,¹ and 95% of children die before they reach age 2.² Kasai *et al.*³ showed that if hepatic portoenterostomy is performed before age 3 months (and preferably before 9 weeks) the prognosis is greatly improved. These results have been confirmed in North America⁴ and Europe,⁵ and although most survivors continue to have some disturbance of liver function, they have normal growth and development.⁵⁻⁷

The aetiology of biliary atresia is unknown. That the underlying processes are continuous would seem to be supported by the fact that in some infants postoperative biliary drainage, although initially satisfactory, becomes erratic.

The presence of biliary radicles in the fibrous portal tissue excised at operation is well known,⁸⁻¹⁰ and has been reported to be a major determining factor for effective postoperative biliary drainage.^{6,11,12} Gautier *et al.*¹³ however, disputed this conclusion and, in an analysis of histological specimens from 48 patients, were unable to establish any clear correlation between the presence of biliary ductules at the porta hepatis, and subsequent biliary drainage. There did appear to be a trend towards failure to drain bile in those in whom no ductules were apparent.

In the current study each specimen of tissue excised from the porta hepatis before hepatic portoenterostomy was examined to assess the total number of biliary ductules, and their sizes.

Patients and materials

Between 1973 and March 1979 hepatic portoenterostomy for the 'non-correctable' form of extrahepatic biliary atresia was performed in 30 infants by one of us (E R H). Tissue excised from the porta hepatis was fixed in formalin, set in paraffin, and stained with haematoxylin and eosin. Serial transverse sections were examined by a histopathologist (CT) who did not know the clinical status of the patients. The total number of biliary ductules was counted, and the diameter of each ductule was calculated using a standard mathematical formula for elliptical structures.¹⁴

Results

No histological material was available for 4 of the 30 infants. Postoperative bile drainage had been established in 14 of them and had failed in 12. There were no differences between the successful and failed groups in gestation, birthweight, age at operation, or level of preoperative bilirubin (Table). Representative histological specimens are shown in Fig. 1a, b. Infants were placed in one of three groups according to the total number of ductules present (nil, less than 20, and more than 20), and the relationship between bile drainage and the size of the ductules (greater than or less than 50 μ m) is shown in Fig. 2. The relationship between bile drainage and the number of ductules is illustrated in Fig. 3.

The successful and unsuccessful portoenterostomy

Fig. 1a Transverse section from tissue excised from porta hepatis showing mature fibrous tissue, without ductules. (H and E $\times 16$).

Fig. 1b Transverse section from tissue excised from porta hepatis showing biliary ductules (arrowed) surrounded by mature fibrous tissue. (H and E $\times 16$).

Table Clinical details of 26 infants

	Gestation (weeks)	Birthweight (kg)	Age at operation (weeks)	Bilirubin ($\mu\text{mol/l}$)		Follow-up (months)
				Preoperative	Postoperative	
Drainage (n=14)	39.8 $\pm$ 0.9	3.2 $\pm$ 0.4	12 $\pm$ 4	220 $\pm$ 83	39 $\pm$ 27	24 $\pm$ 18
No drainage (n=12)	39.2 $\pm$ 1.4	3.0 $\pm$ 0.3	12 $\pm$ 4	211 $\pm$ 62	423 $\pm$ 270	17 $\pm$ 12

Data are given $\pm$ 1 SD.Conversion: SI to traditional units—bilirubin 1 $\mu\text{mol/l}$ $\approx$ 0.058 mg/100 ml.

Fig. 2 Size of ductules.

Fig. 3 Number of ductules.

procedures were compared for the numbers and size of biliary ductules, and appeared to show a trend in favour of patients with more and larger ductules at the porta hepatis. However, these differences did not reach statistical significance (Student's *t* test).

Discussion

The majority of patients with the 'non-correctable' form of extrahepatic biliary atresia have patent intrahepatic biliary ducts which extend to the porta hepatis,^{15 16} and the anatomical basis for hepatic portoenterostomy is therefore well known.

The results of this study suggest that there is no clear relationship between the establishment of effective biliary drainage, and either the number or the size of biliary ductules found in residual proximal bile duct. This supports the work of Gautier *et al.*¹³ who found that almost one-third of their patients had no patent biliary structures at the porta hepatis. Nevertheless, 40% of such patients had effective postoperative biliary drainage, although no details of the length of follow-up are given.

The fact that there is no significant difference between patients with fewer or greater than 20 ductules suggests that the quantity of ductules is not the essential prerequisite for biliary drainage as suggested by Altman.⁴

It is perhaps logical to assume that patients with larger biliary ductules (for example greater than 50 μm) should have more effective biliary drainage, but we, like others,⁹ have been unable to confirm this. Although we were unable to confirm the findings of Chandra and Altman¹² that the absence of biliary ductules was related to the age of the patient (that is, over 12 weeks) the numbers in both series are too small to allow such a conclusion to be drawn.

The reason why patients with large numbers of biliary ductules present at the porta hepatis fail to drain bile postoperatively remains unknown, but it seems likely that the eventual prognosis is related to the degree of intrahepatic inflammation and fibrosis⁸ present at the time of surgery and to postoperative complications which may follow portoenterostomy. Cholangitis, for example, may begin soon after operation and a large variety of organisms has been

cultured from liver biopsies during episodes of fever and increasing jaundice.¹⁷ A close relationship between the progression of intrahepatic fibrosis and the frequency of attacks of cholangitis has been described.^{7 10}

Reports of large series of cases indicate that the best results after surgery for biliary atresia have been obtained in children less than 60 days of age,^{3 4} and the timing of surgery appears to be more significant than histology of the excised remnants of the common bile duct.¹⁸

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