Percutaneous cholangiography in prolonged jaundice of childhood[1]

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Introduction

Injection of radiographic contrast material into small bile ducts was made easier by the introduction of a thin-bore needle (Chiba needle) with an external diameter of 0.7 mm (Okuda *et al.* 1974), and the technique has been rapidly accepted as a safe method for the investigation of both surgical and medical jaundice. The purpose of this paper is to describe a modification of the technique for the investigation of childhood liver disease and to illustrate some of the results obtained in a group of 22 infants and children who were jaundiced from a variety of causes. The place of percutaneous cholangiography in the investigation of childhood jaundice is also discussed.

Method

Percutaneous transhepatic cholangiography is performed in the diagnostic X-ray department under general anaesthesia and immediately before laparotomy when this is necessary. Coagulation factors are checked preoperatively and vitamin K administered routinely. If the prothrombin time remains elevated by more than 4 seconds, an infusion of fresh frozen plasma is given during the procedure. Prophylactic antibiotics are used only in older children who may have infected bile associated with bile duct strictures or calculi.

The 'Chiba' cholangiogram needle has been modified for use in children by reducing the external diameter to 0.5 mm and by shortening the shaft to reduce flexibility[2] (Figure 1). The

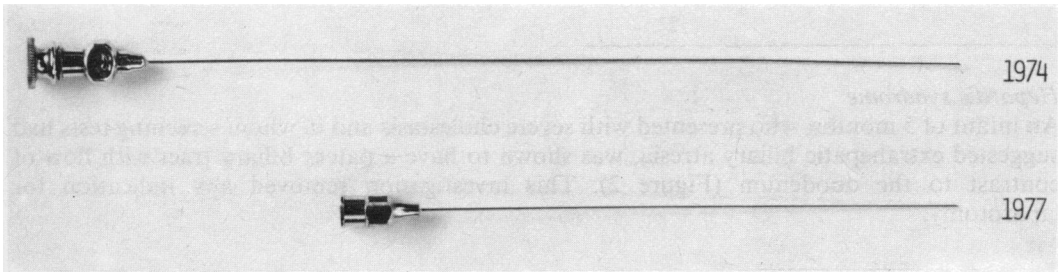

Figure 1. A standard 'Chiba' cholangiogram needle (1974) compared with the modified needle used in children (1977)

needle is inserted in the midaxillary line with respiration arrested in expiration. The point of insertion is determined on fluoroscopy by reference to the level of the hilum of the liver, which is approximately 1 cm above the duodenal bulb on approximately the level of the disc space between the 11th and 12th dorsal vertebrae. The lower level of the right pleural reflection is also avoided. The needle is inserted horizontally into the liver through a small skin incision and is advanced almost as far as the vertebral column to the region of the hilum. Contrast material of low viscosity (Conray 280, May & Baker Ltd) is then injected as the needle is slowly withdrawn

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[2] V Millard, Surgical Instruments, 36 Highgate Hill, London N19

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under fluoroscopic control. Up to six attempts are made to enter bile ducts by making slight alterations in the angle of the needle and by using a second skin entry site if necessary. When a bile duct is entered the injection is continued to fill the common bile duct, and it is sometimes necessary to place the child in an upright or a prone position to achieve this.

Bile ducts are recognized by contrast remaining within the structure and slowly spreading centripetally to fill the hepatic duct system and the hepatic and common bile ducts. In biliary atresia there may be persistence of contrast in isolated segments of ducts. Hepatic veins are easily identified by the rapid clearance of contrast material towards the inferior vena cava and hepatic arteries and portal vein opacifications are also clearly recognizable. Intra- and extrahepatic lymphatics are often visualized in jaundiced children, especially in cases of atresia, and the contrast material drains rapidly to lymph nodes in the porta hepatis.

Complications of intraperitoneal bleeding or bile leakage have not been encountered in this series.

Results

Twenty-two infants and children, in whom previous investigations had suggested a surgically-correctable cause for prolonged jaundice, were investigated by percutaneous cholangiography. The final diagnoses have been listed in Table 1.

Table 1. Causes of jaundice in children investigated by percutaneous cholangiography

	No. of cases
Hepatitis syndrome	1
Extrahepatic biliary atresia:	
(a) Preoperative	12
(b) Postoperative	2
Intrahepatic ductular hypoplasia	3
Hepatic fibrosis	1
Stricture of extrahepatic ducts	2
Tumour of common bile duct	1
	22

Hepatitis syndrome

An infant of 5 months, who presented with severe cholestasis and in whom screening tests had suggested extrahepatic biliary atresia, was shown to have a patent biliary tract with flow of contrast to the duodenum (Figure 2). This investigation removed any indication for laparotomy.

Extrahepatic biliary atresia

(a) *Preoperative:* 12 infants aged 7 weeks to 5 months were investigated before laparotomy. Normal hepatic veins were visualized with rapid flow of contrast to the inferior vena cava (Figure 3). Prominent extrahepatic lymphatic drainage was a striking feature in these infants, the lymph channels appearing dilated and several in number. The contrast material opacified the lymph nodes in the porta hepatis (Figure 4).

Isolated structures in which contrast material persisted were demonstrated in 5 cases and these were interpreted as possibly representing segments of the bile duct system (Figure 4), although histological proof of this interpretation will be difficult to obtain. In one case a larger segment of the duct was identified, which appeared to terminate at the porta hepatis (Figure 5). Surgical treatment of this infant has been satisfactory and she is free of jaundice $2\frac{1}{2}$ years later.

(b) *Postoperative:* 2 infants were investigated after surgery. A 5-month-old child had suffered recurrent attacks of fever and increased jaundice since undergoing surgery 2 months previously. Some bile pigment was present in the faeces and on fluoroscopy contrast material

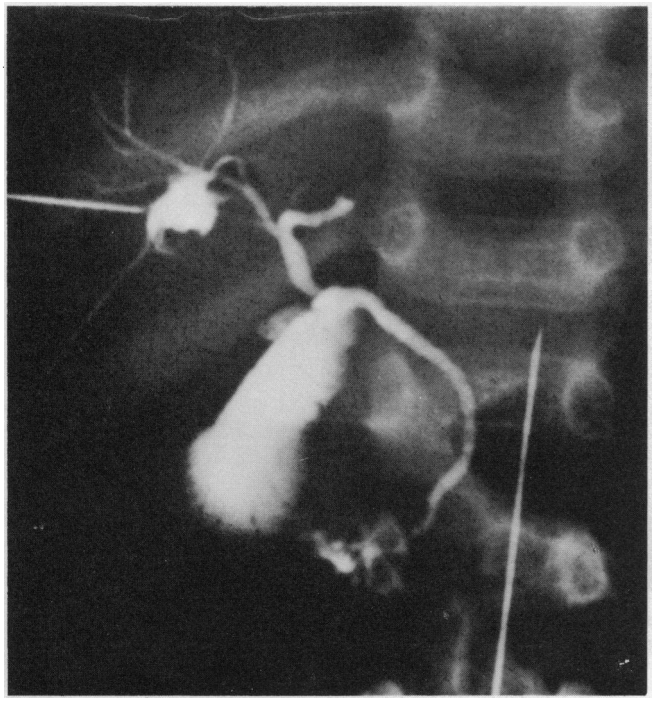

Figure 2. A patent extrahepatic biliary tract in a 5-month infant with hepatitis syndrome. Common bile duct diameter is compared with a 19-gauge needle (overlying spine)

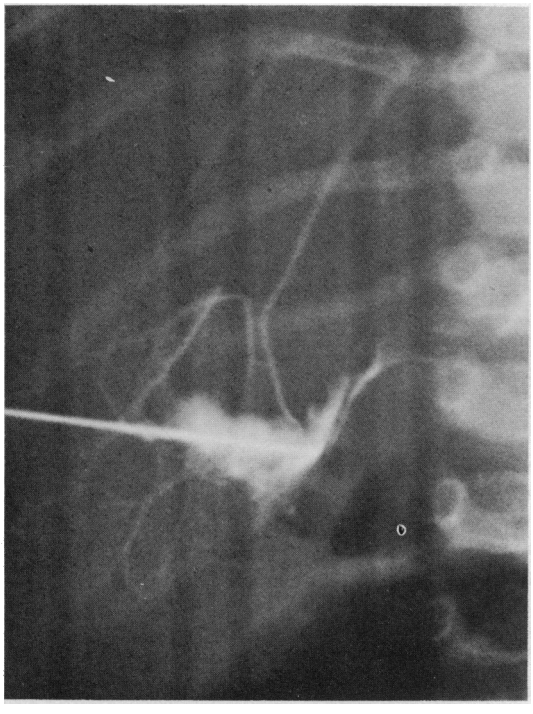

Figure 3. Biliary atresia, age 2 months. Hepatic veins

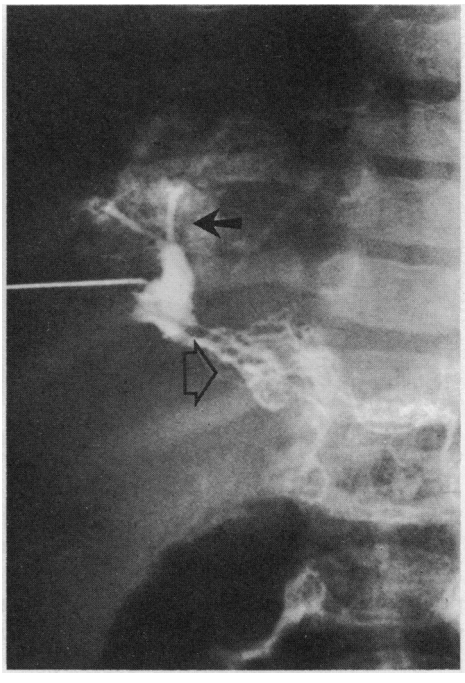

Figure 4. Biliary atresia, age 2 months. Possible isolated segments of bile ducts (upper arrow), and prominent extrahepatic lymphatics (lower arrow)

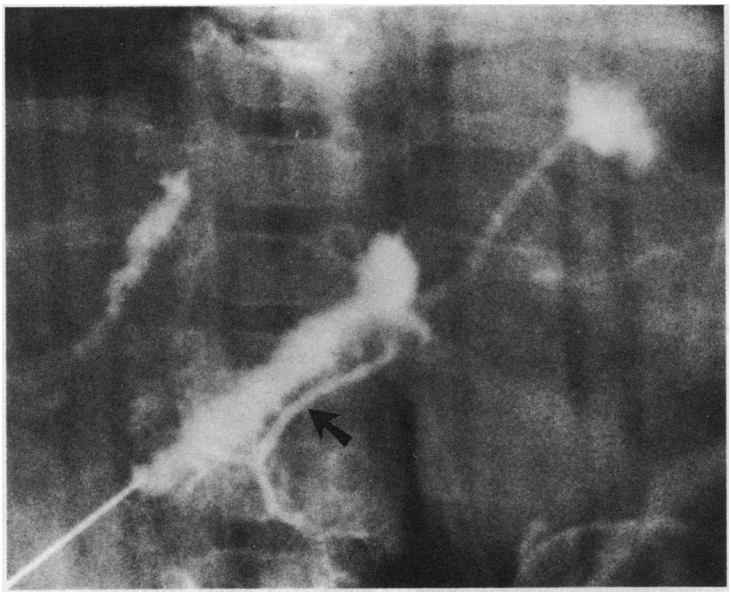

Figure 5. Biliary atresia, age 2 months. Long segment of bile duct (arrow) below intraparenchymal collection of contrast material

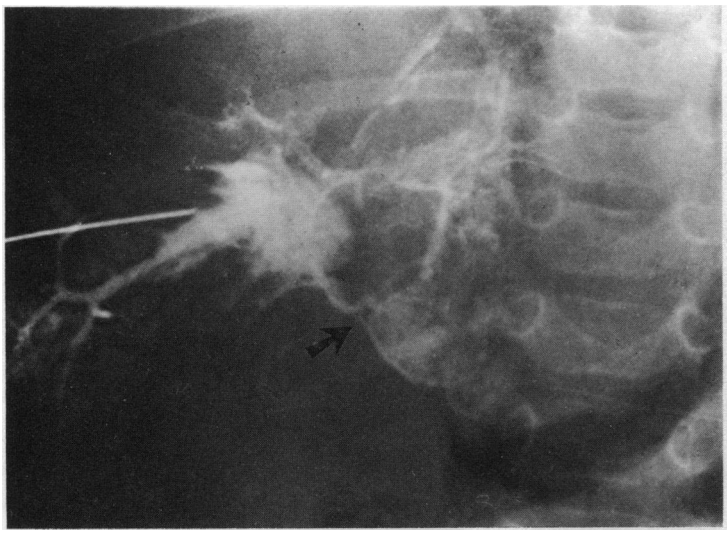

Figure 6. Biliary atresia, age 5 months. Cholangiogram 2 months after surgery. Contrast flowing into loop of bowel (arrow at anastomosis)

was seen to enter a peristalsing loop of jejunum (Figure 6). Jaundice in the second infant, aged 7 months, had failed to diminish after surgery and a cholangiogram was carried out 5 months later. No communication was found between bile ducts and bowel, but opacification of the portal venous system revealed a thrombus in the portal vein and a large varix communicating with the left hemiazygos system (Figure 7).

Intrahepatic ductular hypoplasia

Three infants aged 2, 4 and 16 months were subjected to laparotomy after failure to identify bile ducts by percutaneous cholangiography. Operative cholangiography through the gall

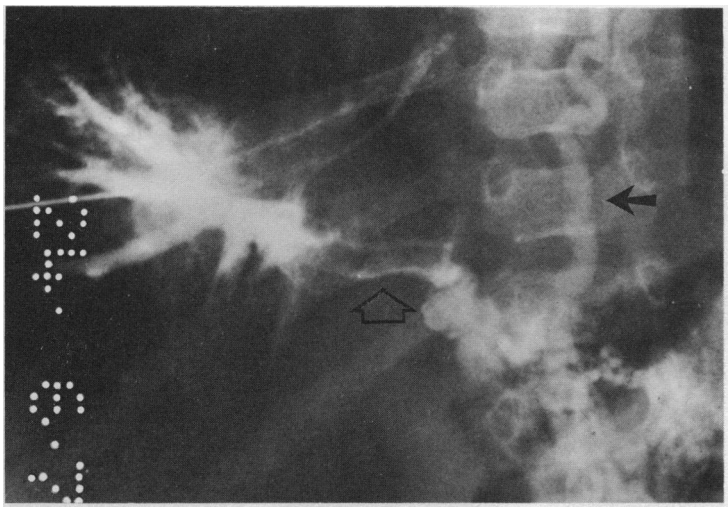

Figure 7. Biliary atresia, age 7 months. Cholangiogram 5 months after surgery. Thrombus in portal vein (lower arrow) and large oesophageal varix (upper arrow), indicating portal hypertension

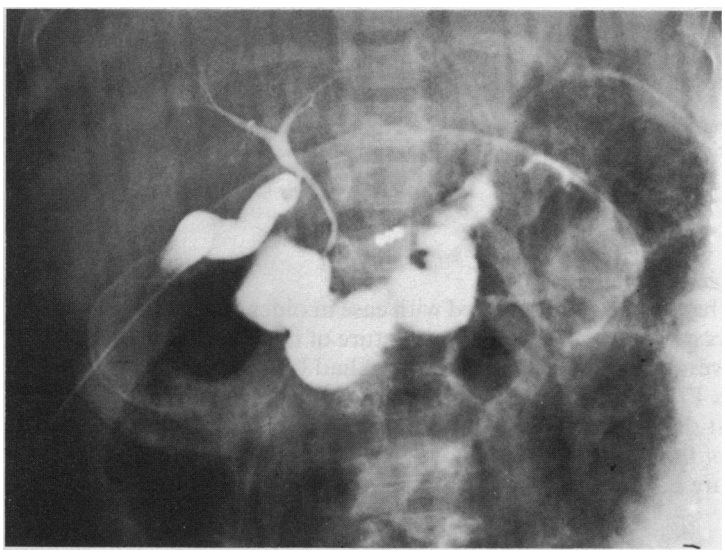

Figure 8. Intrahepatic biliary hypoplasia, age 2 months. Operative cholangiogram shows patent extrahepatic ducts but few intrahepatic ducts

bladder, however, showed hypoplastic but patent extrahepatic ducts, but there was no visualization of peripheral ducts in the liver (Figure 8). Histology of liver biopsies showed absent or hypoplastic bile ducts in the portal triads which gave a diagnosis of intrahepatic hypoplasia. This condition is compatible with survival for a number of years, although the patients may not become free of jaundice (Alagille 1976).

Hepatic fibrosis

An infant of 8 months who had undergone extensive surgery for hepatic haemangiomatosis and portal hypertension developed a persistent jaundice. Extrahepatic bile duct obstruction

was suggested, but fine-needle cholangiography confirmed the patency of the bile duct system (Figure 9). Gross distortion of the intrahepatic ducts was believed to be caused by a severe intrahepatic fibrosis seen on liver histology. A further laparotomy was therefore avoided in this infant.

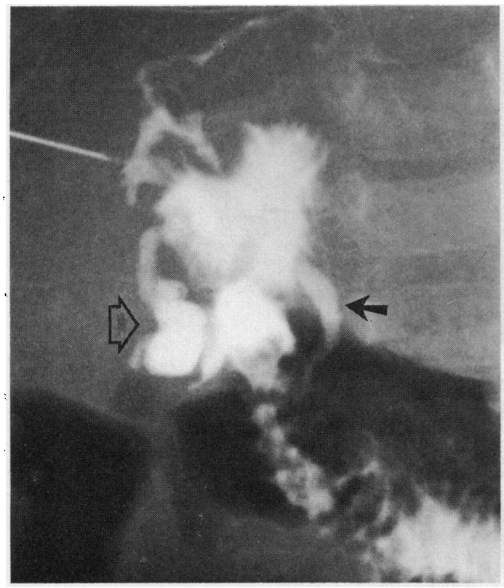

Figure 9. Intrahepatic fibrosis, age 8 months. Intrahepatic ducts distorted and irregular, but gall bladder (left arrow) and common bile duct (right arrow) patent

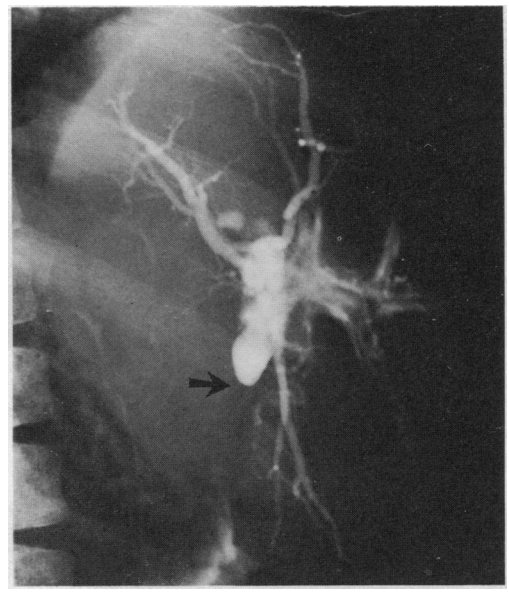

Figure 10. Post-cholecystectomy stricture of common bile duct, age 10 years

Extrahepatic obstruction in older children

Dilated intrahepatic bile ducts have been demonstrated with ease in older children and without complications. Figure 10 shows a post-cholecystectomy stricture of the common bile duct in a boy of 10 years referred for investigation of jaundice and who had been treated for gallstones secondary to sickle cell disease. Figure 11 is a cholangiogram of a 4-year-old diabetic girl who developed a sclerosing lesion at the bifurcation of the common bile duct; the aetiology of this lesion is unknown. A tumour of the common bile duct was clearly defined in a 13-year-old girl with obstructive jaundice (Figure 12) and she remains completely well 17 months after excision of the lesion.

Discussion

The differential diagnosis of prolonged jaundice in infancy usually rests between the neonatal hepatitis syndrome and biliary atresia, although a choledochal cyst can occasionally cause similar signs soon after birth. Many causes of the hepatitis syndrome have been identified, for example infection, metabolic abnormalities and alpha-1-antitrypsin deficiency, and these can be detected by the appropriate screening tests. However, the aetiology remains obscure in approximately 70% of cases and it is this group which must be separated within 2–3 months of birth from those infants with biliary atresia or a choledochal cyst, for recent surgical reports of the treatment of biliary atresia suggest that operative success is more likely in infants under 3 months of age before there has been progression to complete destruction of the bile ducts (Kasai *et al.* 1968). Differential diagnoses depend on specific screening tests and liver biopsy in the first instance, as routine blood biochemistry is of little diagnostic value (Mowat *et al.* 1976). Where there is histological difficulty, the ^{131}I-Rose Bengal faecal excretion test gives further

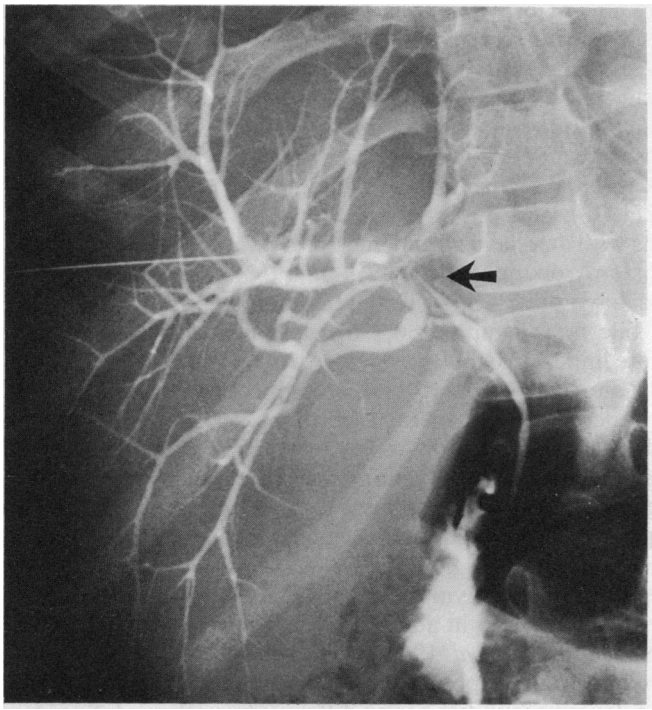

Figure 11. Stricture at confluence of hepatic ducts (arrow) in a diabetic child, age 4 years

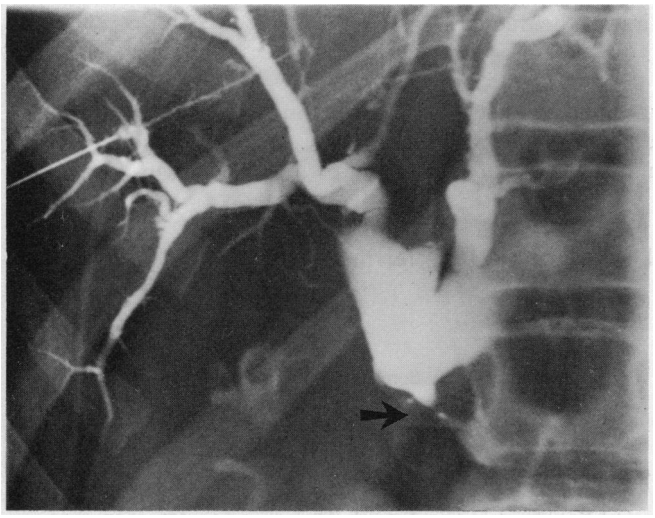

Figure 12. Tumour of common bile duct (arrow), age 13 years

information, and ultrasonography of the right hypochondrium is also useful to exclude choledochal cyst.

We have investigated jaundiced infants by percutaneous cholangiography only when the liver biopsy and ^{131}I-Rose Bengal investigations suggested biliary atresia or remained equivocal, and these children therefore represent a highly selected group of patients. There are few descriptions of percutaneous cholangiography in children. Chaumont *et al.* (1975) reported, without illustrations, the postoperative cholangiograms of 12 children with extrahepatic biliary atresia and described the visualization of hepatic veins, portal vessels, lymphatics, and bile ducts. Recently, Franken *et al.* (1978) used the original 'Chiba' needle to

investigate 8 jaundiced infants, the youngest of whom was $1\frac{1}{2}$ months of age, and achieved visualization of bile ducts in 3 cases of hepatitis and one case of choledochal cyst. Carty (1978) used the same needle in 10 children under $2\frac{1}{2}$ years of age and demonstrated normal biliary tracts in 4 infants and residual segments of bile ducts in cases of biliary atresia.

We have also demonstrated patent common bile ducts in infants with hepatitis and hepatic fibrosis and exploratory laparotomies were avoided in these cases.

The preoperative demonstration of isolated segments of bile ducts in 5 of our cases of atresia confirms the observations of Carty (1978), and one of these children has had an excellent surgical result. However, other infants with atresia in whom ducts were not demonstrated have also drained bile after a portoenterostomy operation (Kasai *et al.* 1968). The best results of surgical treatment for biliary atresia have been achieved in very young infants and recent reports have claimed postoperative bile flow in up to 80% of cases under 60 days of age (Howard & Mowat 1977). It is therefore imperative that preoperative investigations should not delay the operation in these infants and thus we perform percutaneous cholangiography immediately before laparotomy and under the same general anaesthetic.

A failure to identify bile ducts does not prove a diagnosis of extrahepatic atresia, and subsequent operative cholangiography in 3 infants has demonstrated the condition described as intrahepatic hypoplasia, or atresia. It must be emphasized, therefore, that operative cholangiography is still an integral part of the surgical management of these children and is complementary to preoperative percutaneous cholangiography.

The frequent demonstration of large extrahepatic lymphatics in cases of atresia is of interest and similar findings have been described previously in adults with inflammatory liver disease and cirrhosis (Baggenstoss & Cain 1957, Moreno *et al.* 1963, Clain & McNulty 1968).

The indications for percutaneous cholangiography in older children who present with clinical and biochemical features of obstructive jaundice are no different from those defined for adults, although the very fine needle may add to the safety of the procedure, and increase the possibility of achieving a demonstration of the duct system.

Summary

Fine-needle percutaneous cholangiography has been carried out on a selected group of 22 infants and children who were jaundiced from a variety of causes. The technique has aided both the diagnosis and surgical management of the patients and has been free from complications.

Acknowledgment: We wish to thank Dr A P Mowat for referring these patients.

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