Endoscopic Retrograde Cholangiopancreatography (ERCP) in Biliary Tract Disease of Infants Less than One Year Old

Naomi Ohnuma, Hideyo Takahashi, Masahiro Tanabe, Hideo Yoshida and Jun Iwai

Department of Pediatric Surgery, Chiba University, School of Medicine, Chiba 260

Ohnuma, N., Takahashi, H., Tanabe, M., Yoshida, H. and Iwai, J. Endoscopic Retrograde Cholangiopancreatography (ERCP) in Biliary Tract Disease of Infants Less than One Year Old. Tohoku J. Exp. Med., 1997, 181 (1), 67-74 —— We performed a total of 75 examinations with endoscopic retrograde cholangiopancreatography (ERCP) in 73 infants aged from 8 days to 300 days (mean 71 days) between 1977 and 1995. ERCP examination was performed with a prototype duodenoscope and was successful in 47 of 52 examinations in biliary atresia, 9 of 11 in neonatal hepatitis, all 4 in paucity of intra-hepatic bile duct, 4 of 5 in congenital biliary dilatation, 1 of 2 in duodenal stricture, and 1 case of in postoperative jaundice of hepatoblastoma. In 46 infants with biliary atresia, excluding one in whom the findings could not be evaluated due to poor x-ray image quality, we distinguished the following four patterns of ERCP findings: Pattern 1, only the pancreatic duct could be demonstrated and no bile duct was visualized (76%); Pattern 2, only a part of the distal common bile duct with the pancreatic duct was visualized (2%); Pattern 3, the entire length of the common bile duct with the pancreatic duct was visualized without the gallbladder and the common hepatic duct (4%); and Pattern 4, the common bile duct and the gallbladder with the pancreatic duct were visualized without the common hepatic duct (18%). In all of these 46 patients, laparotomy, an operative cholangiogram, and histological evaluation of the biliary duct were performed. In 9 neonates with neonatal hepatitis, the biliary tract was opacified and biliary atresia was excluded. Laparotomy was thus avoided in these neonates. There was no complication caused by either ERCP or by anesthesia. ——— ERCP in infant; biliary atresia; infantile cholestasis; common channel of biliary atresia

It is well known that endoscopic retrograde cholangiopancreatography (ERCP) is an useful diagnostic instrument, even in the field of pediatric surgery,
for establishing the diagnosis of anomalous arrangement of the pancreatobiliary ductal system, including the dilatation of the common bile duct (Urakami et al. 1977; Cotton and Laage 1982; Allendorph et al. 1987; Weidmeyer et al. 1989; Putnam et al. 1991).

However, ERCP has rarely been attempted in infants with suspected pancreatobiliary disease because no duodenoscope that is entirely suitable for infants has been developed. The recent development of a pediatric lateral view endoscope has made ERCP examination more satisfactory in infants. (Guelrud et al. 1987, 1991; Heyman et al. 1988; Wilkinson et al. 1991; Derkx et al. 1994).

We performed 75 ERCP examinations in 73 infants as a diagnostic procedure to evaluate bile duct patency and to define the anatomical abnormality of the pancreatobiliary ductal system and we report our experience in the evaluation of infantile cholestasis.

MATERIALS AND METHODS

Between March, 1977 and March, 1995, 75 ERCP examinations in 73 infants were performed at our institute. The infants were 30 boys and 43 girls whose ages ranged from 8 days to 300 days (mean, 71 days). The indications for ERCP are summarized in Table 1; they included 52 examinations of biliary atresia, 11 of neonatal hepatitis, 4 of paucity of intra-hepatic bile duct, 5 of congenital biliary dilatation, 2 of duodenal stricture, and 1 of postoperative jaundice of hepatoblastoma. Informed consent for ERCP was obtained from the parents of all patients. All of the ERCP examinations were performed under general anesthesia with endotracheal intubation to prevent airway obstruction and dyspnea due to abdominal distension during the procedure. We used the Machida FGS-PEII with the diameter of 8.0 mm before 1979 and the Olympus PJF with the diameter of 8.8 mm since 1980, both of which are of small enough design to be used in infants without hazard.

ERCP examination was performed with the patient in the left lateral position on a fluoroscopic table. After the recognition of the papilla of Vater, the scope was moved to a position at which the papilla was in the center of the visual field. A cannula was inserted into the orifice of the papilla and a small amount of 60% Urographin injected slowly under fluoroscopy to obtain the cholangiopancreatography image. After the examination oral intake was withheld for 24 hr and antibiotics were given. Blood and urine amylase levels were monitored.

RESULTS

The papilla of Vater was identified and cannulated in 66 (88%) of the 75 ERCP examinations (Table 1). The 66 successful ERCP examinations comprised 47 in biliary atresia, 9 in neonatal hepatitis, 4 in paucity of intra-hepatic bile duct, 4 in congenital biliary dilatation and 1 each in duodenal atresia and postoperative jaundice after resection of hepatoblastoma. In 9 examinations, (5
Table 1. Frequency of successful ERCP examination in infants

<table>
<thead>
<tr>
<th>Condition</th>
<th>Numbers of examination</th>
<th>Numbers of successful ERCP</th>
<th>Success rate (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Biliary atresia</td>
<td>52</td>
<td>47</td>
<td>90</td>
</tr>
<tr>
<td>Neonatal hepatitis</td>
<td>11</td>
<td>9</td>
<td>82</td>
</tr>
<tr>
<td>Paucity of intrahepatic duct</td>
<td>4</td>
<td>4</td>
<td>100</td>
</tr>
<tr>
<td>Congenital biliary dilatation</td>
<td>5</td>
<td>4</td>
<td>80</td>
</tr>
<tr>
<td>Others</td>
<td>3</td>
<td>2</td>
<td>67</td>
</tr>
<tr>
<td>Total</td>
<td>75</td>
<td>66</td>
<td>88</td>
</tr>
</tbody>
</table>

in biliary atresia, 2 in neonatal hepatitis, 1 each in congenital biliary dilatation and duodenal stricture), the papilla could not be cannulated due to technical problems with the endoscope and catheter. The ERCP findings in 46 patients with biliary atresia, excluding one whose ERCP findings could not be evaluated, were classified into 4 patterns according to observations such as filling by contrast medium of portions of the biliary duct and the pancreatic duct, and the diameter of the common bile duct (Fig. 1). All of the 46 patients underwent laparotomy and an operative cholangiogram was performed. The classification based on the ERCP findings was compared with the Kasai’s classification as assessed using the intraoperative findings. In 35 patients (76%) only the pancreatic duct could be demonstrated and no bile duct was visualized (Pattern 1): (Fig. 2).

These 35 patients included 5 with Kasai’s type I cyst, 28 with type IIIb, and 1 each with type IIIc and type IIId. In one (2%) patient only part of the distal common bile duct with the pancreatic duct was visualized (Pattern 2). This patient was confirmed to have Kasai’s type I cyst. In 2 (4%) patients the entire length of the common bile duct with the pancreatic duct was visualized but without depiction of the gallbladder or the common hepatic duct (Pattern 3). These patients had Kasai’s type IIIa. In 8 (18%) patients both the common bile

![ERCP findings](image)

Fig. 1. The ERCP findings in 46 patients with biliary atresia.
duct and the gallbladder with the pancreatic duct were visualized but without depiction of the common hepatic duct (Pattern 4): (Fig. 3). These 8 patients all showed Kasai's type IIIa. In 9 (82%) of 11 examinations in neonatal hepatitis, biliary ductal system opacification was seen (Fig. 4).

During the follow-up period, cholestasis diminished in all 9 neonates. In 4 infants with increased cholestasis and development of splenomegaly, the extrahepatic biliary tree was visualized as normal but the intrahepatic bile ducts were shown to be sparse and very narrow.

Intrahepatic bile duct paucity was confirmed on the liver biopsy. The entire anatomy of the junction of the pancreatobiliary ductal system was visualized in 4 of 5 patients with a cystic type of congenital biliary dilatation who were also diagnosed on ultrasonography (Fig. 5).

One boy with duodenal stricture underwent ERCP at 10 months of age due to detection of anomalous orifice of the biliary duct on the intraluminal membrane of the duodenum. One girl with hepatoblastoma was examined with ERCP at 9
Fig. 3. A 34-day-old boy diagnosed with biliary atresia. The gallbladder, the common bile duct and the pancreatic duct are visualized with ERCP, and the Kasai classification is type IIIa.

months of age as monitoring for obstructive jaundice following right hepatectomy. Neither of these two patients showed anomalous anatomical findings on ERCP.

The urinary and serum amylase levels were monitored after ERCP examination and the patients were observed for signs of infection, pain and vomiting. There was no complication in any of these infants caused by either ERCP or anesthesia.

**DISCUSSION**

ERCP has a well-established role in the investigation of biliary and pancreatic disease in the adult, but there is relatively little information about the use of this technique in children. Since 1987, five prospective studies have been published on the use of ERCP in cholestatic infants. Guelrud et al. (1987) reported the usefulness of a new pediatric duodenoscope PJF in the diagnosis of infantile cholestasis. Cannulation of the biliary tree was successful in 20 of 23 consecutive infants aged from 19 to 150 days. They were able to diagnose bile duct obstruc-
Fig. 4. A 99-day-old girl diagnosed with neonatal hepatitis. The common bile duct, the gallbladder, the intrahepatic bile duct and the pancreatic duct are visualized.

tion in five infants with biliary atresia, normal biliary tree in 13 with neonatal hepatitis, and a choledochal cyst in one patient. Heyman et al. (1988), also using PJF, were successful in cannulating the biliary tree in 3 of 11 infants with an age range of 3 to 16 weeks. Wilkinson et al. (1991), using PJF and XPJF, later reported nine ERCP examinations in cholestatic infants ranging in age from 7 to 42 weeks and Guelrud et al. (1991) described its usefulness in the diagnosis of biliary atresia. They performed ERCP in 32 infants with prolonged cholestasis (Guelrud et al. 1991). The cannulation succeeded in 20 of 22 infants with biliary atresia, and the authors distinguished three types of anatomical abnormalities: type 1, no visualization of biliary tree; type 2, opacification of the distal common bile duct and the gallbladder without visualization of the main hepatic duct; and type 3, opacification of the distal common bile duct, the gallbladder, and a segment of the main hepatic duct with biliary lakes at the porta hepatis. Derkx et al. (1994) reported successful ERCP in 18 of 20 infants with cholestasis of
Fig. 5. A 3-month-old girl with the cystic type of congenital biliary dilatation. The anomalous junction of the pancreaticobiliary duct, a pigtail-like narrowed distal end of common bile duct, and the pancreatic duct are visualized.

uncertain etiology.

In our study, the usefulness of ERCP in differentiating biliary atresia from neonatal hepatitis was confirmed. In 46 infants with biliary atresia, we could distinguish 4 patterns of anatomical abnormalities, in contrast to the 3 types described by Guelrud et al. (1991). There were few differences, however, between the frequencies of ERCP findings in the two studies. In 76% of our patients there was no visualization of the biliary tree compared with 35% in the report by Guelrud et al. (1991). We found opacification of the distal common bile duct and the gallbladder without visualization of the main hepatic duct in 18% of our patients compared to 35% in the previous study (Guelrud et al. 1991). We did not see opacification of the distal common bile duct, the gallbladder, and a segment of the main hepatic duct with biliary lakes at the porta hepatis, whereas Guelrud et al. (1991) reported this observation in 30%. On the other hand, Guelrud et al. (1991) did not report the pattern we observed of opacification of the
distal end of the common bile duct or the pattern of opacification of almost the entire length of the common bile duct without visualization of the gallbladder and common hepatic duct.

In congenital biliary dilatation, ERCP is an important examination in the evaluation of an anomalous junction of the pancreaticobiliary ductal system including the length of the common channel, the presence of common channel enlargement, and the connection between the main pancreatic duct and the accessory pancreatic duct. The congenital biliary dilatation in our infants were of the cystic type, and ERCP was very useful in appraising the pancreaticobiliary ductal system before operation.

In conclusion, our evaluation of ERCP examination in infants indicates that it is particularly useful in the early diagnosis of biliary atresia, the diagnosis of non-surgical cholestatic disease without exploratory laparotomy, and the morphological assessment of pancreaticobiliary ductal disease.

References