Pancreaticobiliary Maljunction Without Bile Duct Dilatation in Children: Distinction From Choledochal Cyst

Yasuyuki Ono, Kenitiro Kaneko, Takahisa Tainaka, Wataru Sumida, and Hisami Ando

Department of Pediatric Surgery, Nagoya University Graduate School of Medicine, Nagoya, Japan

ABSTRACT

Objectives: Pancreaticobiliary maljunction without bile duct dilatation (nondilated type) is rare in children, and its definition remains unclear. There is controversy over treatment between pediatric and adult patients. We reevaluated our previous definition of the nondilated type in children (common bile duct diameter ≤6 mm on cholangiography).

Patients and Methods: Of 150 children with pancreaticobiliary maljunction, 14 fulfilled the definition of the nondilated type. Clinical and cholangiographic findings were reviewed and compared with cholangiograms of 17 children without biliary diseases (controls). The age-related normal range of the choledochal diameter was defined in combination with previous data.

Results: All of the patients had symptoms and signs similar to those of choledochal cysts. Cholangiographic features of choledochal cysts were detected in all of the patients (dilated common channel, 8; distal bile duct stenosis, 5; dilated cystic duct, 4; filling defect in the common channel, 8). The choledochal diameter in control children increased with age. Only 4 patients (29%) had a choledochal caliber within the normal range.

Conclusions: Most pediatric cases reported as the nondilated type are slightly dilated (forme fruste) choledochal cysts and differ from the nondilated type seen in adults. The nondilated type in children should be defined on the basis of the age-related choledochal diameter.


Key Words: Pancreaticobiliary maljunction—Nondilated type—Choledochal cyst.
FIG. 1. Cholangiograms of choledochal cyst (upper left) and nondilated type pancreaticobiliary maljunction in children. The nondilated type also exhibits features of choledochal cyst (arrows). NDS, narrow distal segment.
children, which we have defined previously (8), is really different from choledochal cyst. We reviewed our pediatric patients with pancreaticobiliary maljunction without bile duct dilatation and compared their cholangiographic features with those of children not having biliary diseases.

PATIENTS AND METHODS

Between 1971 and 2006, 150 children (age 15 or younger) with pancreaticobiliary maljunction underwent excision of the extrahepatic biliary tract as a primary operation. Pancreaticobiliary maljunction was diagnosed by cholangiography when the main pancreatic duct and common bile duct joined above the narrow distal segment, a thin portion before the outlet of the papilla of Vater created by the action of the sphincter of Oddi (Fig. 1, NDS, upper left). The maximum transverse diameters of the common bile duct were measured from the cholangiograms without correction for radiographic magnification. Pancreaticobiliary maljunction without bile duct dilatation (nondilated type) in children was defined by a maximum diameter of the common bile duct of 6 mm or smaller (8). The clinical features of the nondilated type were reviewed. The radiographic features of the nondilated type were examined regarding whether they showed the characteristic features of choledochal cysts: stenosis of the distal common bile duct (Fig. 1, 2 upper left), dilated common channel (Fig. 1, 3 upper left), and filling defects by protein plugs (7,13–15). The cholangiographies of patients were compared with cholangiograms (6 operative and 11 endoscopic retrograde cholangiopancreatograms) performed in 17 children without biliary diseases (control children). They consisted of 8 boys and 9 girls with an age range of 1 month to 14 years. Five children had been suspected of having biliary diseases but no such disease was identified, 6 had neonatal hepatitis, 3 had chronic pancreatitis, 2 had pancreas divisum, and 1 had a pancreatic tumor. The maximum diameters of the common bile duct were compared between patients and control children together with previous data from Witcombe et al (16).

RESULTS

Fourteen patients (9.3%) were assigned to the nondilated type. The mean and median ages were 4.8 and 4.0 years, respectively (range 1–15 years). All 14 had clinical symptoms and signs, including abdominal pain in 13 (92%), vomiting in 10 (71%), jaundice in 5 (36%), elevated serum transaminase levels in 7 (50%), and elevated serum amylase levels in 11 (79%). All of them had cholangiographic features of choledochal cysts, including a dilated common channel in 8 (Fig. 1, lower right), distal stenosis in 5 (Fig. 1, lower left), and dilated cystic duct in 4 (Fig. 1, upper right). A filling defect was observed in the common channel in 8 patients (57%) (Fig. 1, lower right). All of the patients showed a resolution of symptoms after excision of the extrahepatic biliary tract with Roux-en-Y hepaticojejunostomy. The mean follow-up period was 10.5 years, ranging from 4 months to 25 years.

The cholangiographic figures of control children varied from a very thin to a relatively thick bile duct, but none showed features of choledochal cyst (Fig. 2). The diameters of the common bile duct of control children increased with age (Fig. 3). Although a comparison was

FIG. 2. Cholangiograms of children without biliary diseases. A very thin (left) and relatively thick (right) bile duct case.
not possible below 2 years of age because of a lack of data, the present measurements in children older than 2 years were within the range of the data of Witcombe et al (16). With these 2 datasets combined, the normal range was obtained (Fig. 3). Judging from the range, only 4 (29%) of the 14 patients had a nondilated caliber (Fig. 4). However, all 4 patients showed cholangio- 
graphic features of choledochal cyst, including a dilated cystic duct in 2, dilated common channel in 2, and filling defect in 1.

**DISCUSSION**

Information concerning the normal caliber of the pediatric common bile duct is limited. To our knowledge, only 2 measurements using intravenous cholangiography and ultrasound have been published to date (16,17). However, the diameters of the common bile duct are much smaller on ultrasound than on cholangiography (18,19). Therefore, ultrasonographic measurements of patients’ bile ducts would be underestimated. The diameters obtained via endoscopic and operative cholangiographies possibly differ from those using intravenous cholangiography in the absence of injection pressure (18,19). Our results on cholangiography in control children showed that the effect of the contrast medium injection pressure may be negligible (Fig. 3). Apparently, the age-related influence cannot be ignored.

So far, 48 pediatric patients with nondilated type pancreaticobiliary maljunction have been reported, and the exact diameters were described in 36 (Table 1). Only 7 (19%) of them and 4 (29%) of our 14 patients showed a truly nondilated caliber according to the present age-related normal range (Fig. 3). Most children with a nondilated-type diagnosis should have been classified
as showing slightly dilated choledochal cysts, which Okada et al (6) called common channel syndrome in 1983 and Lilly et al (7) called forme fruste in 1985. During childhood, these forme fruste choledochal cysts cause symptoms similar to those of ordinary choledochal cysts, through the same mechanism of transient occlusion by protein plugs (8,15,33). The treatments of forme fruste and ordinary choledochal cysts should be identical (7).

Truly nondilated type pancreaticobiliary maljunction seldom causes symptoms during childhood. Protein plugs hardly form, probably owing to smooth bile flow. Additional anomalies such as a dilated cystic duct or common channel may be necessary for stasis. Almost all patients grow up without symptoms, and some of these conditions are incidentally discovered in adulthood by a thickened gallbladder wall on ultrasonograms (10,34). This gallbladder mucosal hyperplasia is caused by the regurgitation of pancreatic juice and may be related to carcinogenesis (5). In the majority of patients the condition goes undetected, and many patients experience gallbladder cancer. Some patients, however, must remain asymptomatic throughout their lifetime. Therefore, truly nondilated type pancreaticobiliary maljunction is different from forme fruste choledochal cysts. Pediatric surgeons have confused these two entities, which has caused controversy over the treatment.

The treatment of truly nondilated type pancreaticobiliary maljunction should be modified according to each situation. For asymptomatic adults, prophylactic cholecystectomy alone is reasonable (9,10,12). Mild symptoms in adults, probably resulting from associated gallbladder diseases, were relieved by cholecystectomy (10). Excision of the extrhepatic biliary tract seems appropriate for symptoms produced by pancreaticobiliary maljunction. This led to good results in our 4 children and in other reported pediatric cases (11). Some asymptomatic pediatric patients will be found incidentally, in a similar way to adults: detection of hyperplastic gallbladder on ultrasonography. Treatment for these children is difficult to determine. From the experience with biliary cancer cases, cholecystectomy may be necessary until the age of 40 (12,35). However, it is unknown whether carcinogenesis in the bile duct may not necessarily occur in pediatric patients. Many pediatric surgeons have insisted on bile duct excision to eliminate the regurgitation of bile and pancreatic juice in view of carcinogenesis, although this argument was based on symptomatic cases, most of which were in fact forme fruste choledochal cysts (4,8,11). The accumulation of cases remains necessary to elucidate its natural course and determine appropriate treatment.

### REFERENCES
