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

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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 $\leq$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. ***JPGN 46:555–560, 2008.*** **Key Words:** Pancreaticobiliary maljunction—Nondilated type—Choledochal cyst. © 2008 by European Society for Pediatric Gastroenterology, Hepatology, and Nutrition and North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition

Pancreaticobiliary maljunction is defined as an aberrant union of the pancreatic and biliary ducts located outside the duodenal wall (1). Because the sphincter of Oddi does not functionally affect the union, 2-way regurgitation of bile and pancreatic juice occurs and damages these systems (1). This regurgitation was once considered to cause biliary dilatation of choledochal cysts (2); however, the results of animal experiments and the existence of patients without bile duct dilatation showed that pancreaticobiliary maljunction is an unlikely cause of dilatation (3–9). Pancreaticobiliary maljunction is known to have a close association with biliary malignancies. The incidence and location of cancer is different between patients with and without biliary dilatation (9). According to a nationwide survey in Japan, in 1,239 patients with pancreaticobiliary maljunction with biliary dilatation (ie, choledochal cyst), cancer occurred in 131 (11%), of whom 85 (65%) had gallbladder cancer and 44 (34%) had extrahepatic bile duct cancer. In 388 patients without biliary dilatation, cancer occurred in 147 (38%) of patients, of whom 137 (93%) had gallbladder cancer.

In patients with pancreaticobiliary maljunction without bile duct dilatation (nondilated type), the clinical characteristics differ between children and adults. The condition is much rarer in children than in adults (9). Pediatric patients always have symptoms similar to those seen in choledochal cysts, whereas adult patients seldom have symptoms (8,10–12). The treatment widely accepted for pediatric patients is excision of the extrahepatic biliary tract, the same procedure as for choledochal cysts (8,11). In adult patients, prophylactic cholecystectomy alone is generally accepted because of the close relationship with gallbladder carcinoma but weak relationship with bile duct carcinoma (9,10). These differences raise the question whether pancreaticobiliary maljunction without bile duct dilatation is actually the same entity in children and adults. This confusion partially depends on the vague definition of ''without bile duct dilatation'' in children (6–8,11).

The aim of this study was to elucidate whether pancreaticobiliary maljunction without bile duct dilatation in

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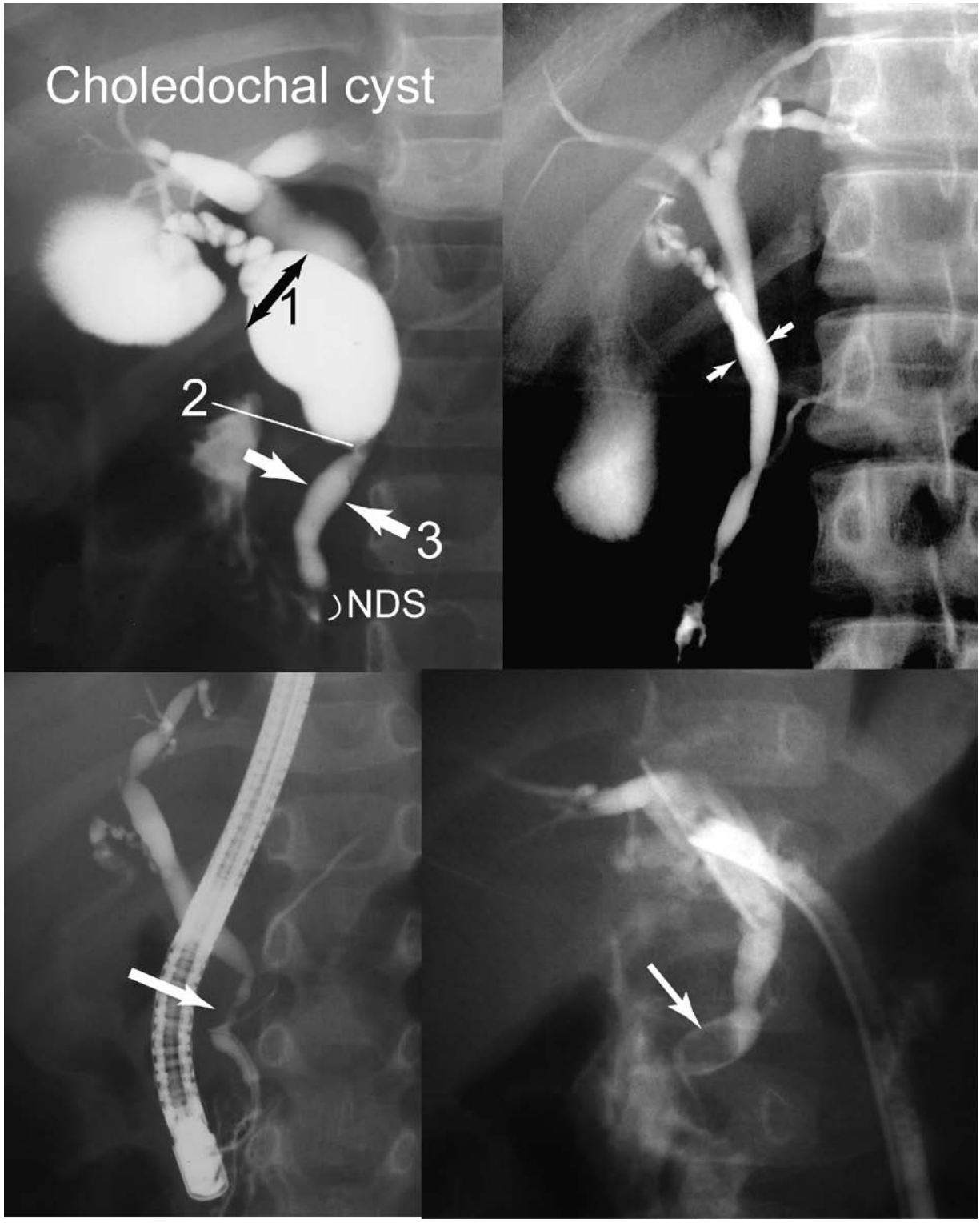

FIG. 1. Cholangiograms of choledochal cyst (upper left) and nondilated type pancreaticobiliary maljunction in children. The nondilated type also exhibits features of choledochal cyst (arrows). NDS, narrow distal segment.

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), dilated cystic duct (Fig. 1, 1 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

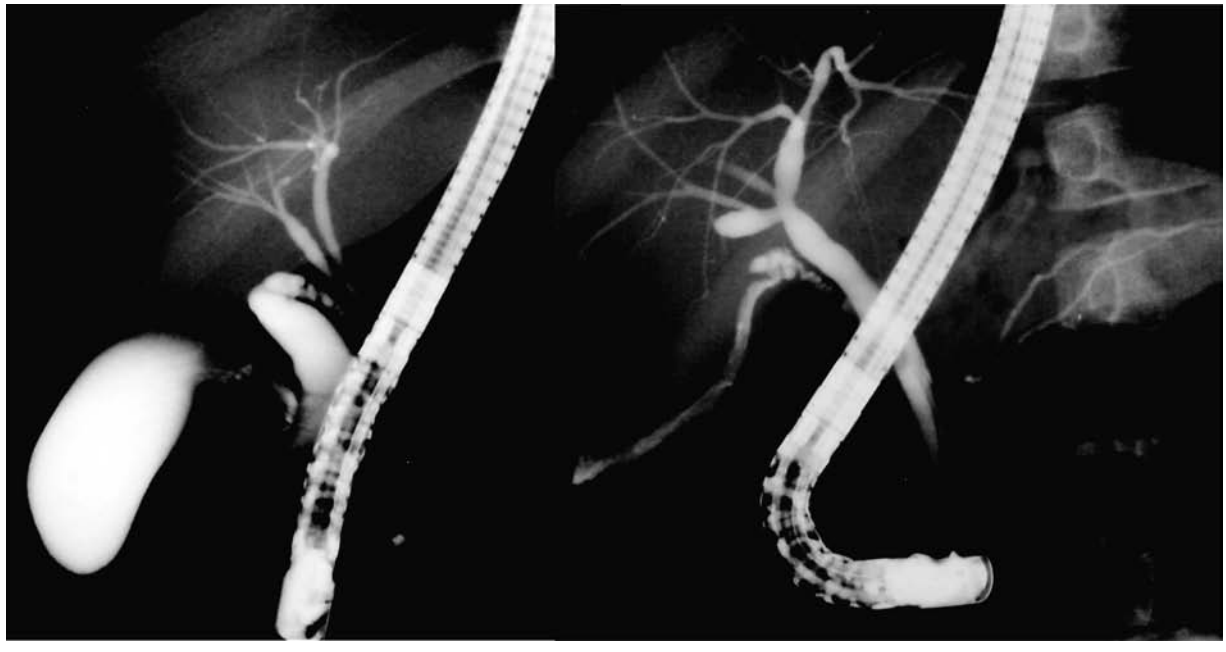

FIG. 2. Cholangiograms of children without biliary diseases. A very thin (left) and relatively thick (right) bile duct case.

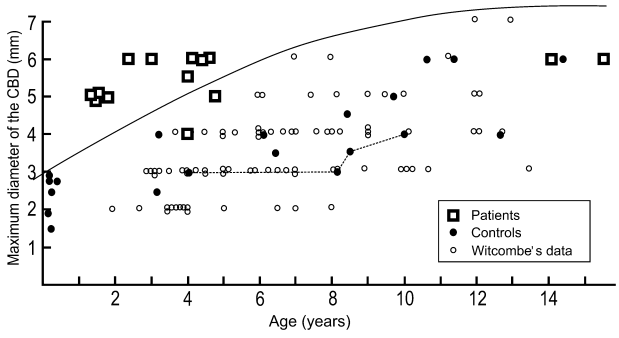

FIG. 3. Scatter diagram showing age-related distribution of common bile duct (CBD) calibers measured on cholangiograms with contrast medium injection in 14 patients with nondilated type pancreaticobiliary maljunction (patients) and 12 children without biliary diseases (controls), superimposed on the intravenous cholangiographic data of normal CBD calibers from Witcombe and Cremin (16). Continuous line, provisional normal range of the CBD caliber. Dotted line, measurements from the same person. (Reprinted with kind permission from Springer Science and Business Media (16) and the author).

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 cholangiographic 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

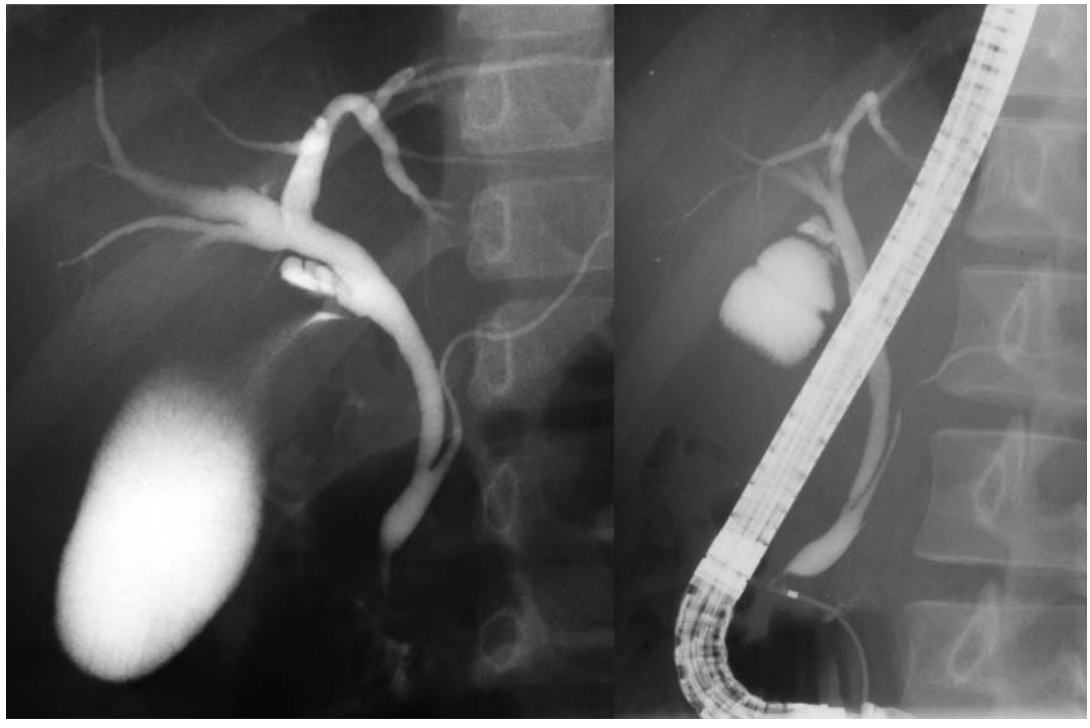

FIG. 4. Cholangiograms of the nondilated type with a CBD diameter within the normal range in a 4-year-old girl (left) and a 15-year-old girl (right). Both patients had a dilated common channel.

TABLE 1. *Reported pediatric cases of nondilated type pancreaticobiliary maljunction*

Year	Author	No. of patients	No. of patients with CBD diameters within normal range
1983	Okada et al. (6)	2	0
1985	Lilly et al. (7)	4	0
1992	Kawamitsu et al. (20)	2	0
1995	Kitatani et al. (21)	2	2
1996	Miyano et al. (22)	8	1
1996	Ito et al. (23)	1	1
1996	Azuma et al. (24)	1	0
2000	Pushparani et al. (25)	1	0
2002	Thomas et al. (26)	6	1*
2003	Ladd et al. (27)	1	0
2003	Shimotakahara et al. (28)	17†	NA
2004	Iwai et al. (11)	6	1
2005	Sarin et al. (29)	2	NA
2005	Kusano et al. (30)	1	NA
2005	Miyano et al. (31)	1	0
2006	Moriuchi et al. (32)	1	1
		36‡	6

CBD = common bile duct; NA = not assessed.
* Ultrasonographic measurements; this case may be of the dilated type.
† Includes 8 cases of Miyano (1996).
‡ Excluding cases not assessed.

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 extrahepatic 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.

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