A Valved Hepatic Portoduodenal Intestinal Conduit for Biliary Atresia

KOICHI TANAKA, M.D., ISAO SHIRAHASE, HIROFUMI UTSUNOMIYA, TETSUO KATAYAMA, SHINJI UEMOTO, KATSUHIRO ASONUMA, YUKIHIRO INOMATA, M.D., and KAZUE OZAWA, M.D.

Forty-six consecutive patients with biliary atresia were operated on at our institution during the 11-year period between 1978 and 1989. Their ages at operation ranged from 18 to 153 days (mean, 59 days). After dissecting the porta hepatis structures by Kasai operation, a biliointestinal anastomosis was constructed with a valved hepatic portoduodenal intestinal conduit in all cases. The intestinal valve is an intussuscepted muscular valve. Valvular function was examined radiologically. The upper gastrointestinal series demonstrated no reflux of contrast material into the conduit proximal to the valve and liver scintigraphy demonstrated that radioactive isotope drained readily into the duodenum through the valve. Thirty-nine of the forty-six patients (85%) had bile drainage after initial operation. At present 30 patients (65%) are alive without jaundice, 6 (13%) are alive with jaundice, and 10 (22%) are dead. The 5-year jaundice-free survival rate was 64%. Cholangitis occurred in 9 of 39 patients (23%) who had obtained apparent bile drainage: 5 had tractable cholangitis and 4 had refractory cholangitis. Reoperation restored bile flow in 2 of 8 patients who abruptly ceased to have bile drainage without cholangitis. In conclusion, with a valved hepatic portoduodenal intestinal conduit, the incidence of cholangitis was decreased, its medical control became easier, and the survival rate was improved.

From the Second Department of Surgery, Kyoto University, School of Medicine, Kyoto, Japan

HEPATIC PORTOENTEROSTOMY FOR biliary atresia permits bile drainage[1] and immediate postoperative bile drainage is achieved in more than 80% of patients.[2,3] After obtaining bile drainage, ascending cholangitis is the most serious complication, leading to biliary reobstruction and/or progressive liver fibrosis.

Many types of biliointestinal reconstruction have been performed to prevent ascending cholangitis. In most procedures the incidence of postoperative cholangitis is more than 48%.[2,4] Hepatic portocholecystostomy is the only procedure producing excellent results with regard to postoperative cholangitis.[2,5] This procedure uses the valve mechanism of Vater's ampulla to prevent reflux of duodenal contents into the porta hepatis. However the indication for this method is limited by the types of complication and the anatomic circumstances.

At our institution a newly developed intestinal valve, an intussuscepted muscular valve, has been used as an isolated jejunal conduit for biliary atresia.[6] Its valvular function has been examined clinically and experimentally. In this communication we describe the key surgical features of the procedure and outline our long-term results.

Materials and Methods

Forty-six consecutive patients with biliary atresia were operated on between May 1978 and August 1989 at our institution. Of the 46 patients, 28 were female and 18 were male. Their ages at operation ranged from 18 days to 153 days (mean, 59 days). Seven of the forty-six patients were younger than 30 days, 18 were between 31 and 60 days old, and 21 were more than 61 days old. Forty-three cases were type III and three were type I, according to the classification of the Japanese Society of Pediatric Surgeons.[7] The follow-up period ranged from 5 months to 11 years.

The diagnosis of postoperative cholangitis was made from the tetrad of unexplained fever (temperature > 38 C), elevated serum bilirubin level, acholic stools, and leukocytosis. Cholangitis was graded according to the severity of inflammation: (+) tractable cholangitis controlled by antibiotic therapy in an ambulatory setting; and (++) refractory cholangitis treated with hospitalization.

To evaluate the effectiveness of the valvular function in preventing the reflux of intestinal contents, upper gas-

Supported by a Grant for Pediatric Research (63-A-05) from the Ministry of Health and Welfare.

Address reprint requests to Koichi Tanaka, M.D., Second Department of Surgery, Kyoto University, School of Medicine, 54 Kawara-cho, Shogoin, Sakyo-ku, Kyoto 606, Japan.

Accepted for publication March 23, 1990.

trointestinal (UGI) series were performed in 15 patients with biliary atresia more than 1 year after the operation. As a control UGI series were performed in 10 patients with choledochal cyst more than 1 year after hepaticojejunoduodenostomy without valve. On the other hand, the anterograde bile passage through the valve was examined with hepatobiliary scintigraphy in nine patients with biliary atresia more than 10 years after the operation.

Bacteriologic study was performed in eight patients who had sufficient bile drainage to examine bile after initial operation. Bile for culture was obtained by a sterile catheter inserted into the conduit proximal to the valve during operation. Culture results were expressed as logarithmic colony counts per milliliter.

Operative Technique

After dissecting the porta hepatis structures by the Kasai operation, a biliointestinal anastomosis was constructed with a valved hepatic portoduodenal intestinal conduit in all patients (Fig. 1). An intestinal conduit measuring about 15 cm long was made from the jejunum. The proximal end of the segment was closed and the side of the segment was anastomosed to the porta hepatis. The distal end was anastomosed to the descending portion of the duodenum.

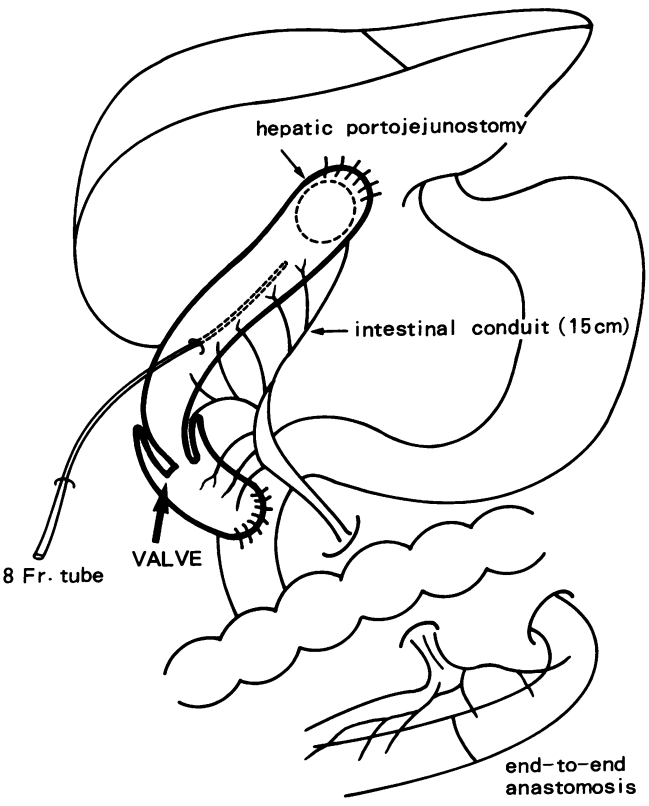

FIG. 1. A valved hepatic portoduodenal intestinal conduit.

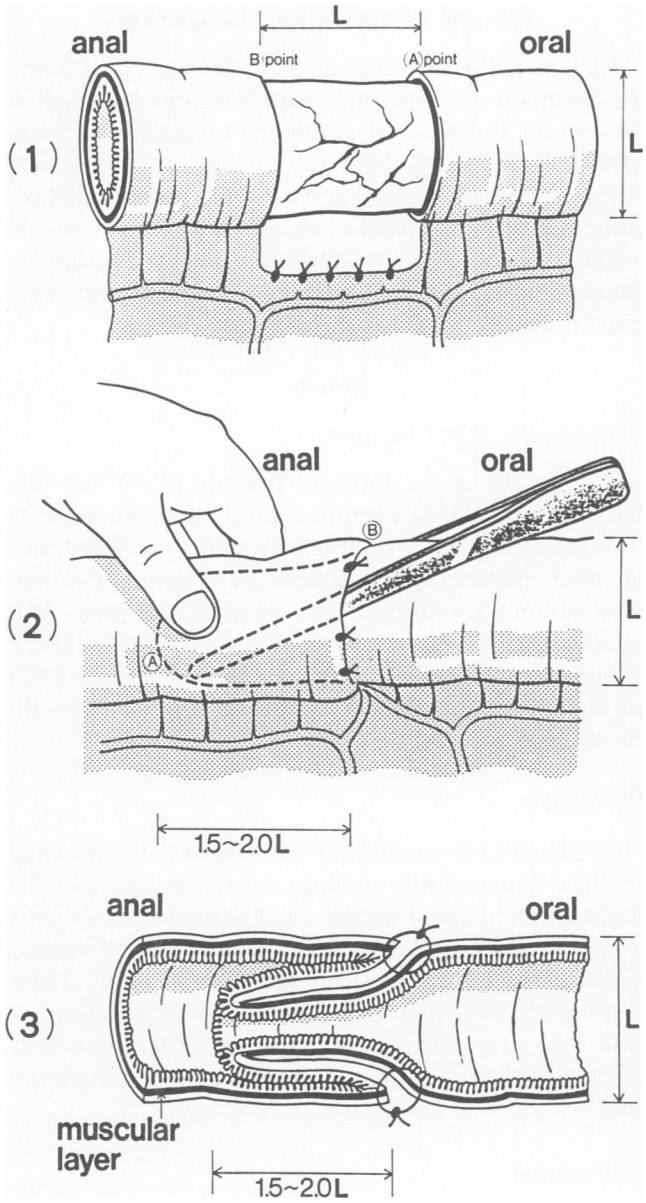

FIG. 2. Construction of an intussuscepted muscular valve. (1) Removal of the seromuscular layer. (2) Intussusception and valvular fixation. (3) Cross-section of the valve.

A valve was established at the distal third of the conduit. An 8 French polyethylene catheter was inserted into the conduit proximal to the valve to decrease the pressure in the conduit and examine the bile flow for 1 week. A muscular valve was constructed in the following manner (Fig. 2). The vasa recti of the conduit were ligated and divided by the width equal to the intestinal diameter. The seromuscular layer of the devascularized intestine then was removed. The denuded segment and an equivalent length of its proximal portion of the intestine was intussuscepted into the distal segment. The intussuscepted valve was fixed with 8 to 10 interrupted sutures.

Pre- and Postoperative Management

Kanamycin syrup was given orally for 2 or 3 days before the operation. As antibiotics, cephalosporin or penicillin for 4 weeks and aminoglycoside for 1 week were administered intravenously after the operation. Ursodesoxycholic acid, glucagon, and adrenocorticosteroid (prednisolone) were administered as choleretic agents for several weeks. Multivitamin compound and vitamin D (Alfarol®, Chugai Pharmaceutical Co., LTD., Tokyo, Japan) were given routinely.

Results

Postoperative Bile Drainage

Thirty-nine of the forty-six patients (85%) had bile drainage after initial operation (Fig. 3). Seven patients (15%) had no bile flow, one of which obtained bile drainage after reoperation. At present 30 patients (65%) are alive without jaundice, 6 patients (13%) are alive with jaundice, and 10 patients are (22%) dead. Sixteen of the twenty-five patients who were operated on before 1985 are alive without jaundice. The 5-year survival rate with the absence of jaundice is 64%.

Cholangitis

Cholangitis occurred in 9 of 39 patients (23%) who had obtained apparent bile drainage: 5 patients had tractable cholangitis (+) and 4 patients had refractory cholangitis (++) (Fig. 4). All of the tractable patients are alive without jaundice and have almost normal liver function. Three of the refractory patients are alive without jaundice but have mild liver dysfunction. The other patient has liver cirrhosis with jaundice and is awaiting liver transplantation.

Reoperation

Eight of the thirty-nine patients (21%) abruptly ceased to have bile drainage without cholangitis. All patients underwent reoperation for scoring of the liver hilus and revision of the hilar anastomosis. Laparotomy revealed that the porta hepatis was covered with thick granulation in all cases. Two of the eight patients became jaundice free again and have regained almost normal liver function. Two patients have liver cirrhosis with jaundice and are awaiting liver transplantation. The remaining patients died of liver failure, pneumonia, disseminated intravascular coagulation, and complication after liver transplantation.

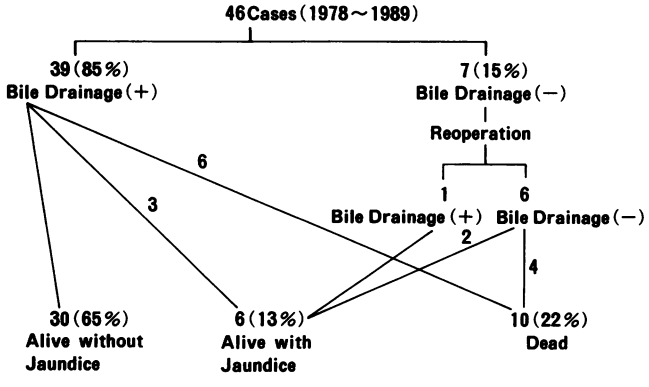

FIG. 3. Overall results after surgery.

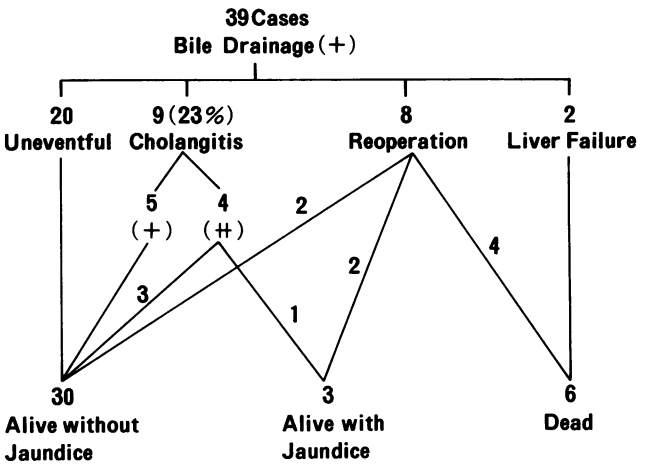

FIG. 4. Results after obtaining bile drainage.

Seven of the forty-six patients (15%) had no bile flow after initial operation and underwent reoperation (Fig. 3). One of these patients obtained bile drainage and is alive with jaundice.

Valvular Competence

In 15 patients with biliary atresia, UGI series demonstrated no reflux of contrast material into the conduit proximal to the intussuscepted muscular valve (0 of 15, 0%) (Fig. 5A). In contrast 5 of 10 patients with choledochal cyst had reflux of contrast material into the intrahepatic bile duct (5 of 10, 50%) (Fig. 5B). Hepatobiliary scintigraphy demonstrated that radioactive isotope drained readily into the duodenum through the valve in all nine cases examined (Fig. 6).

Both qualitative and quantitative cultures of bile drainage confirmed enteric bacteria in six of eight cases (Table 1). Colonization of the intestinal conduit began within the first few weeks after the operation.

Discussion

The pathogenesis of cholangitis after Kasai operation is conjectured to be due to pre-existing cholestasis and bacterial contamination.[3,8,9] That is, because the intrahepatic bile ducts are to some extent hypoplastic and stenotic in all patients with biliary atresia, cholestasis arises

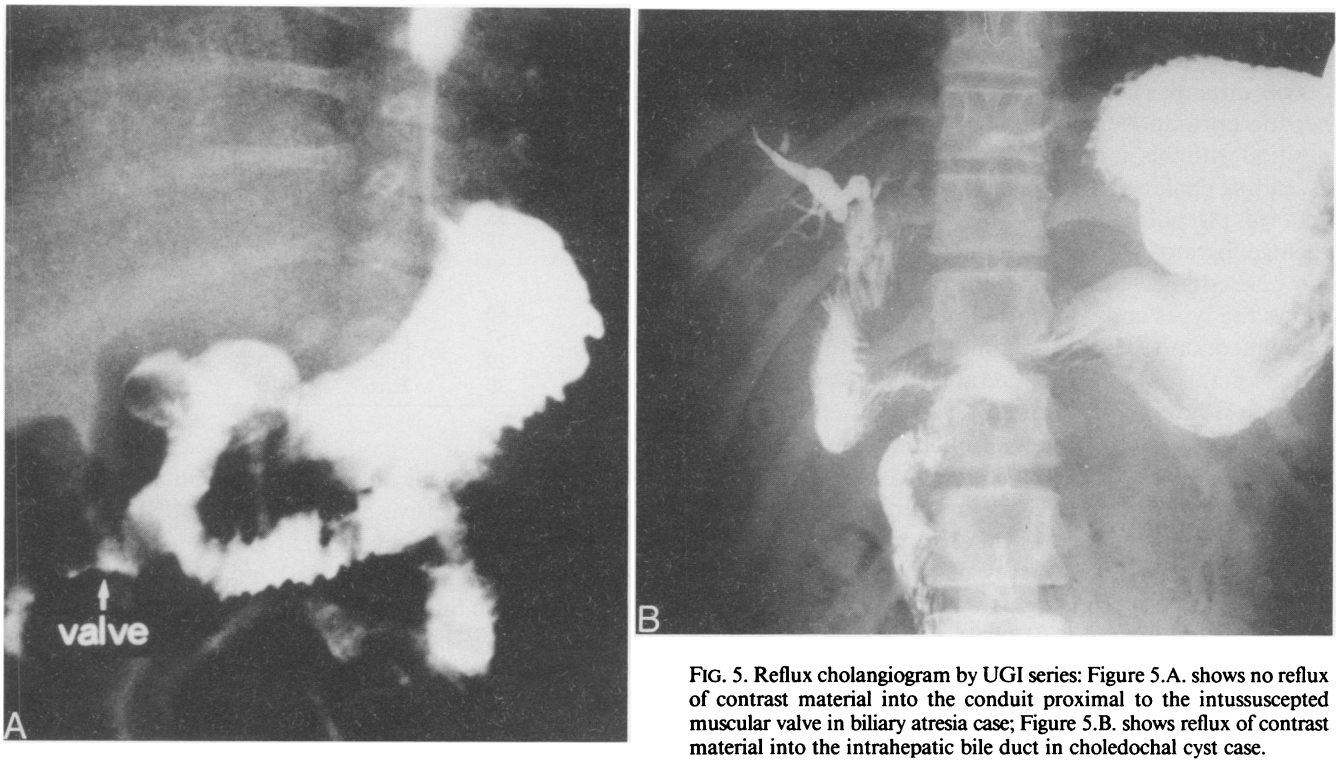

FIG. 5. Reflux cholangiogram by UGI series: Figure 5.A. shows no reflux of contrast material into the conduit proximal to the intussuscepted muscular valve in biliary atresia case; Figure 5.B. shows reflux of contrast material into the intrahepatic bile duct in choledochal cyst case.

as a consequence and persists for months after operation.[3] Cultures of fluid in the conduit from the area of the anastomosis at the porta hepatis *via* catheter reveal that the intestinal conduit is colonized by enteric flora within the first few weeks after the operation.[6,10–13] Both factors are present in most instances of hepatic portoenterostomy for biliary atresia and cholangitis after the Kasai operation merely compounds the pre-existing liver damage.

Another postoperative complication with high incidence and often serious results[14,15] is biliary obstruction caused by the granulation formed at the porta hepatis. Occurring in the early postoperative period, the features characteristic of biliary obstruction caused by granulation, as compared with those of ascending cholangitis, are the absence of infectious signs and the lack of response to medical treatment. In a survey of our patients, its incidence was 21% (8 of 39), and its mortality rate was 50% (4 of 8). Although bile flow was restored after the operative removal of the granulation in all cases, one half of the patients died of liver failure and other related complications. A reoperation to remove the granulation or scar tissue at the porta hepatis also is effective for intractable ascending cholangitis.[2] However reoperation for ascending cholangitis usually is done as a last resort because medical treatment is generally sufficient.

In hepatic portoenterostomy there is no mucosal continuity between the bile ducts and the intestinal wall, and the bile ducts open onto the cut surface of the remaining fibrous mass and hepatic parenchyma. Autoanastomosis occurs between the major intrahepatic duct epithelium and the coapted intestinal mucosa, a process that takes about 6 weeks.[15] If complications such as cholangitis and biliary obstruction caused by the granulation at the porta hepatis occur in this early postoperative phase, the transected microscopic ducts are prone to inflammatory closure. To prevent these complications, especially during this phase, the most effective means seems to be to prevent reflux of the intestinal contents to the porta hepatis. In the presence of reflux of intestinal contents, ascending cholangitis develops due to the exacerbation of bacterial contamination along with the rise in the biliary intraluminal pressure, and biliary obstruction caused by the granulation at the porta hepatis develops due to disturbances in healing of the hepatic portoenterostomy. Accordingly we took advantage of the valve mechanism of Vater's ampulla to prevent reflux of intestinal contents.

Hepatic portocholecystostomy for biliary atresia often has been performed to prevent postoperative cholangitis using the valve mechanism of Vater's ampulla. Ohi, Okamoto, and Kasai[16] reported that none of nine patients treated by this procedure developed postoperative cholangitis. Lilly also reported that none of four patients developed cholangitis after this procedure.[5] Although this procedure is simple and postoperative cholangitis is prevented, complications frequently occur, such as obstruction of the biliary conduit, bile leakage at the porta hepatis, and so on.[1,2,5] Furthermore the indication of hepatic portocholecystostomy is limited in number because it must

be carried out in patients with a patent bile duct from gallbladder to duodenum.

Our attempt is to construct an antireflux valve in the hepatic portoduodenal intestinal conduit. Jejunal interposition with this antireflux valve is effective in preventing ascending cholangitis and bile flow into the duodenum is enabled physiologically by this method. Postoperative roentgenograms showed no reflux of contrast material in the cases studied by upper GI series (Fig. 5A). Table 1 shows the types and concentrations of bacteria in the intestinal conduit. This finding indicates that the presence of bacteria is only one of several factors related to cholangitis. Although this antireflux valve may not be sufficiently tight to prevent flow of microorganisms in a retrograde manner, it prevents both reflux of intestinal contents and increase in the biliary intraluminal pressure and enables smooth bile flow in an anterograde manner through the valve. In a survey of our patients, the incidence of ascending cholangitis was 23% (9 of 39), and the mortality rate was zero (0 of 9). Compared with other procedures, our procedure had a decreased incidence of cholangitis and medical control was facilitated: the incidence of postoperative cholangitis in patients with bile drainage was 68% in the original Roux-en-Y, 56% in the double Roux-en-Y, 100% in the Roux-en-Y interposition, and 48% in the Suruga II.[2,4] Similar attempts to prevent cholangitis have been made. Kaufman and coworkers[17] used an isolated hepatic portoduodenal jejunal conduit with a 1-cm intussuscepted valve in seven cases with biliary atresia. Endo[18] used an ileocecal segment with a plicated ileocecal valve with good results.

TABLE 1. *Bacteriologic Study*

Patient No.	Age at Operation (days)	Bacteria	Icterus	Cholangitis
1	43	*E. coli* 10^6 Pseudomonas 10^5	−	−
2	55	—	−	−
3	78	—	−	−
4	80	*E. coli* 10^6 Pseudomonas 10^7	+	−
5	50	Pseudomonas 10^7	−	+
6	52	Pseudomonas 10^6	−	−
7	19	Enterococcus 10^7	−	−
8	30	*E. coli* 10^5	−	−

FIG. 6. Hepatobiliary scintigraphy, performed 45 minutes after injection of radioactive isotope, shows good hepatic uptake, normal excretion into the interposed jejunal conduit, and readiness of drainage into the duodenum through the valve.

At our institution we used an isolated jejunal conduit without valve in 45 patients with choledochal cyst and none of the patients developed postoperative cholangitis. X-ray studies showed the reflux of contrast material into the biliary tree in one half of the choledochal patients with hepatoduodenal intestinal conduit without valve (Fig. 5B). The difference in the incidence of postoperative cholangitis between biliary atresia and choledochal cyst suggests that adequate bile flow may be one of the most important factors in preventing cholangitis. However it takes considerable time to establish sufficient postoperative bile flow for biliary atresia. Therefore, in the early phase after obtaining bile drainage, it is necessary not only to prevent reflux of intestinal contents to the porta hepatis but also to increase the bile flow with choleretic agents.

Our 5-year jaundice-free survival rate of 64% is comparable to the 20% to 55% 5-year survival rates reported from other centers.[2,3,19–21] In conclusion our valved hepatic portoduodenal intestinal conduit decreased the incidence of cholangitis, facilitated medical control of cholangitis, and improved the survival rate.

References

1. Kasai M, Suzuki S. A new operation for non-correctable biliary atresia—hepatic portoenterostomy. Shujutu 1959; 13:733–739
2. Ohi R, Hsnamatsu M, Mochizuki I, et al. Progress in the treatment of biliary atresia. World J Surg 1985; 9:285–293.
3. Lilly JR, Karrer FM, Hall RJ, et al. The surgery of biliary atresia. Ann Surg 1989; 210:289–296.
4. Miyano T, Ohya T, Kimura K, et al. Current state of the treatment of congenital biliary atresia. J Jpn Soc Surg 1989; 90:1343–1347.
5. Lilly JR. Hepatic portocholecystostomy for biliary atresia. J Pediatr Surg 1979; 14:301–304.

6. Tanaka K, Satomura K, Ohnishi S, et al. A new operation for treatment of biliary atresia—jejunal interposition hepatic portoduodenostomy with intestinal valve. J Jpn Soc Pediatr Surg 1980; 16:227–235.
7. Kasai M, Sawaguchi S, Akiyama H, et al. A proposal of new classification of biliary atresia. J Jpn Soc Pediatr Surg 1976; 12:327–331.
8. Lilly JR, Hitch DC. Postoperative ascending cholangitis following portoenterostomy for biliary atresia: measures for control. World J Surg 1978; 2:581–587.
9. Rothenberg SS, Schroter GPJ, Karrer FM, et al. Cholangitis after the Kasai operation for biliary atresia. J Pediatr Surg 1989; 24: 729–732.
10. Hitch DC, Lilly JR. Identification, quantification and significance of bacterial growth within the biliary tract after Kasai's operation. J Pediatr Surg 1978; 13:563–569.
11. Hitch DC, Lilly JR, Reller B, et al. Biliary flora and antimicrobial concentrations after Kasai's operation. J Pediatr Surg 1979; 14: 648–652.
12. Leblanc A, Lambert-Zechowsky N, Binge E, et al. Bacteriological analysis of jejunostomy fluid after surgery for extrahepatic biliary atresia. J Pediatr Gastroenterol Nutr 1983; 2:307–310.
13. Lilly JR, Hitch DC. Postoperative ascending cholangitis following portoenterostomy for biliary atresia: measures for control. World J Surg 1978; 2:581–587.
14. Takemoto H, Inomata Y, Matsukawa Y, et al. Icteric flare-up in patients with biliary atresia after hepatic portoenterostomy. Z Kinderchir 1988; 43:92–94.
15. Takemoto H, Tanaka K, Inomata Y, et al. Granulation at the porta hepatis following hepatic portoenterostomy for biliary atresia: the healing of experimental hepatoenterostomy. J Pediatr Surg 1989; 24:271–275.
16. Ohi R, Okamoto E, Kasai M. Some considerations of hepatic portocholecystostomy for the treatment of biliary atresia. Jpn J Pediatr Surg 1978; 10:983–989.
17. Kaufman BH, Luck SR, Raffensperger JG. The evaluation of a valved hepatoduodenal intestinal conduit. J Pediatr Surg 1981; 16:279–283.
18. Endo M, Katsumata K. Nonrefluxing draining conduit for biliary atresia—an intussuscepted ileocecal conduit and conduit duodenostomy. Jpn J Pediatr Surg 1985; 17:41–49.
19. Grosfeld JL, Fitzgerald JF, Predaina R, et al. The efficacy of hepatoportoenterostomy in biliary atresia. Surgery 1989; 106:692–700.
20. Houwen RHJ, Zwierstra RP, Severijnen RSVM, et al. Prognosis of extrahepatic biliary atresia. Arch Dis Child 1989; 64:214–218.
21. Kobayashi A, Itabashi F, Ohbe Y, et al. Long-term prognosis in biliary atresia after hepatic portoenterostomy: analysis of 35 patients who survived beyond 5 years of age. J Pediatr 1984; 105: 243–246.