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Progress in the Treatment of Biliary Atresia

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Although the surgical results of biliary atresia have markedly improved following the introduction of hepatic portoenterostomy, further improvement is necessary. In a 31-year period, from 1953 through 1983, we performed corrective surgery on 214 patients. In the first 18 years the disease was successfully treated in only 13% of 96 patients. In the next 7 years (1971 to 1977), jaundice was cleared in 55% of 65 patients, now 29 patients are alive without jaundice. In the last 6 years, the jaundice was cleared in 66% of 53 patients; more than 50% of patients are now without jaundice. Retrospective analysis revealed several important factors contributing to the improvement of operative results: (1) early diagnosis and operation, (b) precise dissection and adequate transection of the bile duct remnant, (c) progress in the postoperative management, (d) prevention of postoperative cholangitis, and (e) early reoperation whenever necessary. Corrective surgery performed within 60 days after birth can achieve sufficient bile drainage to normalize serum bilirubin level in more than 80% of the patients.

The surgical results for biliary atresia have been markedly improved by hepatic portoenterostomy. However, biliary atresia is still an intractable surgical disease in infancy. Although a high rate of postoperative bile drainage has been achieved at many institutions, bile flow after the operation does not always indicate a cure. A real cure is obtainable by persistent active bile drainage and by solving many problems relating to the surgical treatment of this disease.

Two hundred and thirty-eight patients with biliary atresia were treated at our hospital during a 31-year period from 1953 to 1983. Of these, 214 patients underwent corrective surgery and 88 are now alive; 70 cases are jaundice-free but 18 patients including 2 patients who recently underwent surgery still have jaundice. Although serum bilirubin levels returned to normal after the corrective operation in 85 patients, 7 again became jaundiced and 8 died after episodes of cholangitis. Consequently, 70 infants are alive without jaundice.

Our recent advances in the surgical treatment are evident (Table 1). Jaundice disappeared in only 15% of the 96 patients treated between 1953 and 1970. From 1971 to 1977, the disappearance rate increased to 55% in 65 patients. Twenty-nine of these patients are alive without jaundice. In the last 6 years, the rate increased to 66% in 53 patients. More than 50% of these patients are now free from jaundice. Figure 1 represents a survival curve of patients undergoing corrective surgery since 1971. When postoperative bile drainage was not sufficient to clear jaundice, 80% of the infants died before their third birthday. Although jaundice disappeared postoperatively in 77 patients with extended bile drainage, 6 died. Three were from reobstruction due to cholangitis and 1 death each was from ruptured esophageal varices, intestinal obstruction, and unrelated disease, respectively.

Recent surgical results from other institutions inside and outside Japan [1–5] were similar to ours. Retrospective analysis of surgical treatment clarified several important factors contributing to the improvement of operative results: an early operation after birth, precise dissection and adequate transection of the bile duct remnant at the porta hepatis, prevention of postoperative cholangitis, advances in postoperative care, and early reoperation whenever indicated. We will discuss these factors in this paper.

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Table 1. Results of corrective operation for biliary atresia.

Period	No. of pts.	Bile drainage (%)	Jaundice cleared (%)	Surviving jaundice-free (%)
1953–1970	96	65 (68)	14 (15)	13 (14)
1971–1977	65	57 (88)	36 (55)	29 (45)
1978–1983	53	49 (92)	35 (66)	28 (53)
1953–1983	214	171 (80)	85 (40)	70 (33)

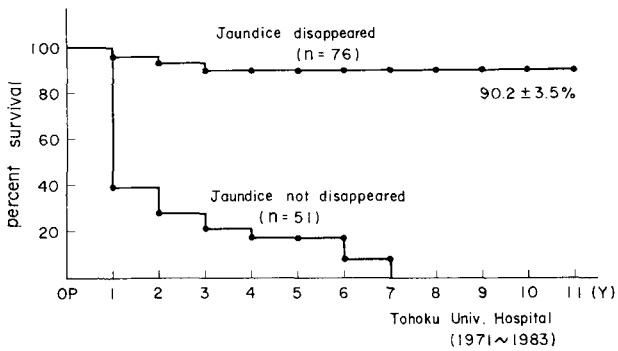

Fig. 1. Survival curve of patients after corrective surgery for biliary atresia at the Tohoku University Hospital from 1971 through 1983.

Table 2. Correlation of age with bile drainage after operation for noncorrectable biliary atresia (1971–1983).

Age at operation (days)	No. of pts.	Bile drainage		
		Good (%)	Poor	None
–60	37	30 (81)	6	1
61–70	28	17 (61)	10	1
71–90	22	11 (50)	10	1
91–120	8	3 (38)	2	3
121–	3	2 (67)	1	0

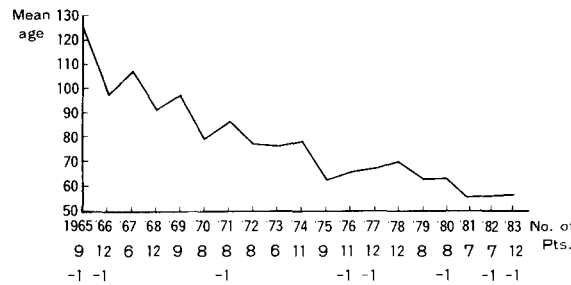

Fig. 2. Chronological change of the mean age at the time of corrective surgery at Tohoku University (1965–1980).

Early Diagnosis and Treatment

Table 2 indicates the relationship between surgical results and patient age at the time of operation. These patients were treated after 1971 when we began using the modified operation to prevent postoperative cholangitis. Bile excretion sufficient to clear jaundice (below 1.9 mg/100 ml of serum bilirubin) was seen in more than 80% of patients receiving corrective operation before the age of 60 days. The frequency of positive bile drainage steadily decreased with age and was 61% at 61–70 days, 50% at 71–90 days, and 38% at 91–120 days of age, respectively. When the operation was delayed 10 days in patients about 60 days old, the success rate was cut by one-fourth. Figure 2 represents the chronological change of the mean age at the time of corrective surgery. The gradual but steady fall of the mean age is obvious.

Consequently, early diagnosis is essential for successful treatment. Various diagnostic means to differentiate biliary atresia from other cholestatic diseases in infancy have been developed recently. At present, in addition to an analysis of the patient's history including examination of the color of the feces and the physical findings of the abdomen, we employ the following examinations: (a) biliary atresia (BA) score test [6], (b) serum lipoprotein-X (Lp-X) determination [7], and (c) histologic examination of percutaneous liver biopsy. When the patient is young, namely, about 1 month after birth, we attempt percutaneous needle biopsy of the liver to make a definite diagnosis. In older patients, to conserve time we disregard the histological examination of the liver and proceed to laparotomy after making a diagnosis by physical findings, BA score, and serum Lp-X determination.

In Japan, infants must undergo a routine medical examination by physicians at the age of 1-month. Utilizing this system, we initiated a survey for possible biliary atresia using Lp-X determination [8]. From 1976 to 1983, specimens from 266 jaundiced babies 4–8 weeks old were tested for serum Lp-X by Tazawa and Konno [8] at Tohoku University Hospital. Of 266 jaundiced infants examined, 102 (38%) were Lp-X positive. Among the Lp-X positive infants, 62 cases were diagnosed as biliary atresia after further diagnostic examinations and were confirmed by surgery. None of infants with biliary atresia were Lp-X negative. Among 82 infants who had intrahepatic cholestasis e.g., neonatal hepatitis, 38 (41%) were Lp-X positive in the screening tests. All blood specimens drawn from infants with prolonged physiological neonatal jaundice had a negative Lp-X. Since biliary atresia does not show a false-negative Lp-X test, this method is considered useful for the screening of biliary atresia patients. Further diagnostic examinations for definitive diagnosis is, however, needed for infants with a positive Lp-X determination.

Improvement of Operative Procedure

After the introduction of hepatic portoenterostomy in 1959 [9], the procedure has been refined each year. Most noncorrectable biliary atresia have a fibrous cone at the porta hepatis. The fibrous cone (representing obliterated hepatic radicles in the porta hepatis) is situated cranial and posterior to the bifurcation of the portal vein. The fibrous bile duct remnant outside of the liver continues to the biliary tracts within the liver without a morphological boundary. In addition, the border of the liver parenchyma is superficial in some and deep in others. Therefore, the liver surface is not reliable as a baseline to determine the transection level during operation. Both the portal vein and its major branches are the most consistent and reliable anatomical landmark for transection of the fibrous cone. According to our histologic studies, the transection should be done at the level of the posterior wall of the portal vein [10]. Although we do not employ histologic examinations by frozen section to confirm the presence of bile ducts during operation, good results have been constantly obtained by this transection.

There are other important surgical aspects. Dissection of the fibrous remnant in the porta hepatis should be carefully advanced by dividing several small veins bridging the fibrous cone and the cranial aspect of the bifurcation of the portal vein. If the acquired theory of the pathogenesis is correct, a normal anatomical situation of the extrahepatic bilary structure at the porta hepatis should be conceived at the time of transection and portoenterostomy. The bile-draining intrahepatic ducts must be close to the area where the preexisting ducts are located. Therefore, we carry out the dissection of the fibrous cone far enough laterally to the entrance of vessels in the hepatic parenchyma and transect the fibrous cone at the level of the posterior wall of the bifurcation of the portal vein. Finally, surgeons must take care not to obliterate the small bile ducts on the transected surface during the anastomotic portoenterostomy.

Progress in Postoperative Management

Progress in postoperative management also contributed to improved surgical results. In addition to routine postoperative care, acceleration of bile excretion and prevention of postoperative cholangitis are required. As antibiotics, 50–80 mg/kg per day of cefalotin (CET) or cefazolin (CEX) for 2–3 weeks and 5 mg/kg per day of aminoglycoside (GM) for 7–10 days are intravenously given after the operation. In patients without signs of cholangitis,

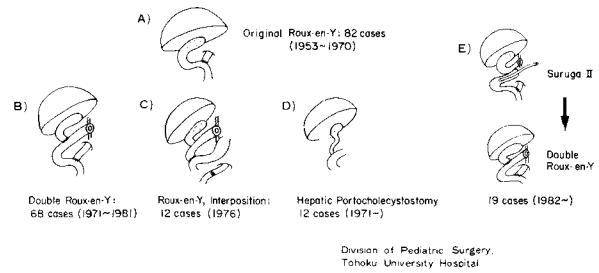

Fig. 3. Modifications of hepatic portojejunostomy at Tohoku University Hospital.

aminobenzyl penicillin (200–400 mg daily) is given orally for several months after cephalosporin and aminoglycoside are discontinued. (Penicillin can be replaced by trimethoprim.)

When patients develop cholangitis, cefoperazone (CPZ) or cefotetan (CTT), which are excreted in bile, should be administered intravenously along with a transfusion. Intravenous injections of choleretics, 3 ml of 10% dehydrocholic acid diluted with 7 cc of 5% glucose, is started immediately after operation and repeated every 12 hours for several weeks to establish an early internal biliary fistula at the porta hepatis by an increase of the water fraction of bile. When bile drainage becomes stable, oral administration of 0.4 mg of urso deoxycholic acid instead of injecting dehydrocholic acid is recommended for 1 or 2 years. Recently, when bile flow is not enough to eliminate jaundice, intravenous glucagon in a dose of 5 mg/m^2 per day [11] and prostaglandin E_2 100μg/kg per week in powder form [12] are also given by mouth twice a week for several weeks. Starting 1 week after operation, we routinely use 20 mg prednisolone to prevent biliary obstruction due to scar formation on the transected surface of the fibrous cone. The dosage of steroid is decreased every 2 days and discontinued within 7–10 days. We administer the same dose of steroid for patients showing a decrease of bile drainage during their postoperative course.

Prevention of Postoperative Cholangitis

Since 1971, we have introduced several modified procedures to replace the simple Roux-en-Y hepatic portojejunostomy to prevent ascending cholangitis (Fig. 3). In an 18-year period, from 1953 to 1970, eighty-two patients underwent hepatic portojejunostomy by the Roux-en-Y procedure (Fig. 3A). Thereafter, a double Roux-en-Y procedure (Fig. 3B) was carried out in 68 patients from 1971 to 1981. Hepatic portojejunostomy by Roux-en-Y interposition between the porta hepatis and the duodenum (Fig. 3C) was performed on 12 patients in 1976. Twelve patients were treated by hepatic portocholecystostomy (Fig. 3D) from 1971 up to the present.

Table 3. Operative results of each modified procedure.

Procedure	No. of pts.	Active bile drainage (%)	Survivors (%)	Surviving jaundice-free
Original Roux-en-Y	82	20 (24)	11 (13)	11
Double Roux-en-Y	68	47 (69)	43 (63)	35
Roux-en-Y interposition	12	9 (75)	7 (58)	5
Portochole-cystostomy	12	7 (58)	5 (42)	3
Suruga II → double-Y	19	12 (63)	17 (89)	11

Table 4. Modified procedures and postoperative cholangitis.

Procedure	No. of pts.	Evaluated case	Cholangitis (%)
Original Roux-en-Y	40	22	15 (68)
Double Roux-en-Y	68	59	33 (56)
Roux-en-Y interposition	12	9	9 (100)
Portochole-cystostomy	12	9	0 (0)
Suruga II → double-Y	19	18	8 (44)

In the last 2 years, we first employed hepatic portojejunostomy by the Suruga II procedure and then converted it to a double Roux-en-Y method after the disappearance of jaundice (Fig. 3E). The purpose of the last procedure is to analyze biologically the totally accumulated bile after the corrective operation and to achieve potential lowering of the incidence of postoperative cholangitis during the immediate postoperative course. This method was performed in 19 patients since 1982.

Operative results of each procedure are shown in Table 3. Postoperative active bile flow was observed in 20 (24%) of 82 patients treated by the original Roux-en-Y, in 47 (69%) of 68 treated by the double Roux-en-Y, in 9 (75%) of 12 treated by Roux-en-Y interposition, in 7 (58%) of 12 by hepatic portocholecystostomy, and in 12 (63%) of 19 by the recent procedure. Patients still alive in each category are 11 (13%), 43 (63%), 7 (58%), 5 (42%), and 17 (89%), respectively. Again by category, number of patients without jaundice were 11, 35, 5, 3, and 11, respectively. Postoperative cholangitis incidences (in patients with bile flow) were 68% in the original Roux-en-Y, 56% in the double Roux-en-Y, 100% in the Roux-en-Y interposition, 0% in the hepatic portocholecystostomy, and 44% in the most recent procedure (Table 4). Comparing the double Roux-en-Y procedures with the original Roux-en-Y, the incidence of cholangitis did not markedly decrease but its medical control became easier with the modified procedures. Consequently, the mortality rate of cholangitis decreased from 47% to 10%.

Other complications in each of the modifications are listed in Table 5. There were 9 cases of intestinal obstruction, 5 of anastomotic leak, and 2 of intestinal bleeding (without evidence of esophageal varices). Although 1 patient who had received a Suruga II jejunostomy experienced shock from excessive fluid loss through the stoma, there were no similar complications in infants undergoing double Roux-en-Y procedure, even though the patients were allowed to go home until the close of stoma. Sudden cessation of bile flow in 3 patients, which was probably due to the obstruction of the small cystic and common bile ducts, was a complication peculiar to patients having hepatic portocholecystostomy.

Table 6 shows the age at the time of closure of the stoma in patients with double Roux-en-Y and Roux-en-Y interposition procedures. Two patients in whom the stoma was closed by 1 year of age (in our early period, employing modifications) had cholangitis after closure of the stoma. Thereafter, in most patients with a stoma, the closure was performed 2 or 3 years after the corrective operation. The incidence of cholangitis was 17% in cases with closure of the stoma at 2 years, 16% at 3 years, and 33% at 4 years of age, respectively. Biliary obstruction from cholangitis was never seen in patients after the closure of the stoma. At present, we close the jejunal stoma according to the following criteria: (a) serum bilirubin level below 1.0 mg/100 ml, (b) no cholangitis for at least 1 year, and (c) no signs of bowel obstruction.

Table 7 represents the results of a questionnaire about postoperative cholangitis compiled by Kimura [13] at the International Symposium on Biliary Atresia and Its Related Disorders held in Sendai in 1983. Suruga and Kasai use an external diversion method, but Tanaka, Akiyama, and Yura do not. Kimura concluded that it is impossible to decide which procedure is better, but that procedures employing external diversion were not always effective in preventing ascending cholangitis. He emphasized that jejunal interposition with an anti-reflux valve, designed by Tanaka et al [14], is effective and that by this method bile can flow physiologically into the duodenum.

Table 5. Modified procedures and complications except cholangitis.

Procedure	No. of pts.	Intestinal obstruction	Anastomotic leakage	Intestinal bleeding	Excessive fluid loss	Obstruction biliary conduit
Double Roux-en-Y	68	9	5	2	–	–
Roux-en-Y interposition	12	2	2	1	–	–
Portocholecystostomy	12	1	–	–	–	3
Suruga II → double-Y	19	1	–	2	1	–

Table 6. Age of patient at time of stomal closure and cholangitis after closure.

Age at closure (years)	No. of pts.	Cholangitis (%)
–1	2	2 (100)
1–	1	0
2–	12	2 (17)
3–	19	3 (16)
4–	6	2 (33)
	40	9 (22)

Reoperation

Thirty-two reoperations for revision of the anastomosis in the porta hepatis were carried out on 28 infants from 1971 through 1983. Twenty-four patients had single reoperations, and 4 patients had 2 reoperations each. There were no operative deaths.

The indications for reoperation were biliary obstruction due to postoperative cholangitis in 16 patients, biliary obstruction after hepaticojejunostomy for the correctable type of this disease in 2, biliary obstruction after hepatic portocholecystostomy in 6, poor bile excretion after the initial operation in 7, and failure of reoperation in 1 patient.

Jaundice disappeared for a certain period of time after the initial operation in 8 of 16 patients followed by a cessation of bile flow after postoperative cholangitis and in 3 of 6 patients receiving hepatic portocholecystostomy. Twenty-three patients underwent a second hepatic portojejunostomy. Two patients with hepaticojejunostomy for a correctable lesion were converted to a hepatic portojejunostomy. Five patients with hepatic portocholecystostomy were converted to a hepatic portocholecystojejunostomy. The anastomosis was between the posterior wall of the gallbladder at the porta hepatis and the jejunum in double Roux-en-Y fashion [15]. One patient had conversion of a hepatic portocholecystostomy to a hepatic portojejunostomy.

Excellent bile drainage after reoperation was obtained in 17 (89%) of 19 patients who had active bile drainage after the initial operation, and jaundice disappeared in 11 of them. On the contrary, good bile drainage was obtained in only 6 (46%) of 13 patients who had poor or absent bile drainage after the initial operation (Table 8).

From a retrospective evaluation of patients who had undergone reoperation, we conclude that reoperation should be done in patients who showed active bile drainage after the initial corrective operation and then had cessation of bile excretion after postoperative cholangitis. Reoperation should

Table 7. Operative procedures and cholangitis (1977–1982).

Procedure	No. of pts.	Bile drainage	Cholangitis	Survivors
Tanaka: Jejunal interposition HPD with antireflux valve	20	17	2	18
Akiyama: Roux-en-Y HPJ	32	17	5	13
Yura: Double tract HPJ	30	27	23	18
Suruga: Suruga II HPJ	59	40%	14	40
Kasai: Double Roux-en-Y HPJ	39	30	18	30

HPD: Hepatic portoduodenostomy
HPJ: Hepatic portojejunostomy.

Table 8. Results of reoperation.

	No. of pts.	Bile drainage (%)	Jaundice cleared (%)	Death
Positive bile drainage after initial operation	19	17 (89)	11 (58)	0
Negative bile drainage after initial operation	13	6 (46)	3 (23)	0
Total	32	23 (72)	14 (44)	0

also be performed for patients without bile drainage after the initial operation if hepatic damage is not too far advanced. Moreover, hepatic portojejunostomy is advisable for patients with correctable lesions who fail hepaticojejunostomy.

Long-term Results

Fifty-two of 88 patients survived for more than 5 years. Forty-eight of the 52 survivors are now leading normal lives. The oldest patient is 28 years old, and the second oldest patient, a 27-year-old, recently married and became a father. Five patients are still jaundiced. Three of them had a recurrence of jaundice following episodes of severe cholangitis. Jaundice in the other 2 patients persisted after the corrective operation. Three patients with Turner's syndrome and with malabsorption due to a blind loop syndrome and extensive intestinal adhesions show retardation of physical growth. Mental retardation was observed in a boy who suffered from craniostenosis. Growth and development were normal in the remaining 49 long-term survivors. The liver was palpable more than one finger breadth below the costal margin in 18 patients, and splenomegaly was evident in 16.

Since 1980, 42 endoscopic examinations were performed in 27 long-term survivors and esophageal varices were found in 13 (48%). All of these patients had episodes of severe cholangitis during their postoperative courses. Four patients underwent surgical treatment for portal hypertension manifested by esophageal varices and hypersplenism. Three splenorenal shunts with splenectomy and 1 splenectomy with devascularization of the left gastric vein were done. No patient has had rebleeding or postoperative encephalopathy.

Discussion

Recently, various institutions not only inside but also outside of Japan have reported successful results of hepatic portoenterostomy. Since most patients with this disease drain bile after corrective surgery, pediatric surgeons have turned their attention to the establishment of extended bile drainage and the satisfactory healing of patients without creating residual liver diseases or physical and mental handicaps.

It is extremely difficult to differentiate biliary atresia and other cholestatic diseases by routine liver function tests. Many diagnostic procedures have been developed. In our experience, a qualitative or quantitative assay of serum Lp-X [7] is useful for the early discrimination of jaundice. Since this examination is less invasive and has never presented a false-negative case of biliary atresia, it is suitable for a screening test of infants with cholestasis. Estimation of the serum bile acid and the ratio of chenodeoxycholic acid to cholic acid (CDC/C ratio) in the serum has been suggested as a possible means of identifying the infant with extrahepatic biliary atresia [16]. In our series, all infants diagnosed as having biliary atresia showed more than 100 μmol/l of serum bile acid, but so did about one-half of the patients with neonatal hepatitis (and some infants with indirect hyperbilirubinemia) [8]. According to Sunaryo and Watkins [17], a large collaborative study failed to substantiate the CDC/C ratio reported by Javitt [16].

Hepatobiliary scintigraphy using recently developed radiopharmaceutical agents, ^{99m}Tc-labeled iminodiacetic derivatives, showed no excretion of radioisotope in patients with obstruction of the extrahepatic bile duct but neither was any excretion apparent in patients with neonatal hepatitis having severe hepatocyte damage [18]. Since hepatocyte clearance was well maintained in the early stage of biliary atresia (patients within 1–2 months after birth), the test identifies those patients with severe neonatal hepatitis in which hepatocyte clearance is severely decreased.

At the present time, histologic study by percutaneous needle biopsy is the most reliable procedure for definitive diagnosis of biliary atresia. Ductular proliferation in the wide portal space with hepatic fibrosis is the most characteristic histologic findings of biliary atresia. The reasons for the most favorable surgical results in patients with early operation are well documented. Irreversible destruction of the intrahepatic bile ducts system and progressive impairment of the intracellular organelle with age have been clearly demonstrated by our previous studies [19, 20]. Since these pathological changes vary in progression and in severity among patients, a few infants older than 3 months have been cured by a corrective operation.

The adequate transection level of the fibrous cone of the extrahepatic bile duct remnant is the key to successful surgical treatment. Several surgeons report a deeper or a more lateral transection with increased bile excretion [21, 22]. Although most authorities believe that postoperative bile flow is dependent on the presence of microscopically patent ducts at the porta hepatis, it is not yet known what biliary structures on the transected surface drain bile after operation. In the correctable type of this disease, bile ducts that are small but can nonetheless be visualized by cholangiography drain bile after hepaticojejunostomy or hepatic portoenterostomy. In the noncorrectable cases, however, controversy still remains as to which biliary structure is crucial for bile drainage. Ohi and Lilly [23]

verified, by histological and histometrical studies, that the type of biliary structures were correlated with postoperative bile flow. The bile duct, which was severely inflammed but appeared to be the preexisting duct, was the only structure that was uniformly present in patients who drained bile after operation.

We have emphasized the dissection of the fibrous cone should be extended laterally to the entrance of the portal vein into the hepatic parenchyma. This is based on the fact that the bile draining routes in biliary atresia follow the normal anatomical geography. Recently, Endo et al. [24] and Ito and associates [25] reported more extended dissection or extensive exploration of the porta hepatis with improvement of the success rate of hepatic portoenterostomy. The recommendation was made on the basis of a histological study of the porta hepatis in the normal infant and in autopsied patients with biliary atresia. Endo emphasized that portoenterostomy should include the space between the bifurcation of the right portal vein. Ito reported that exploration should be extended laterally along the right hepatic artery and the left portal vein into the intrahepatic portal tract.

Adequate bile flow may be the most important factor in preventing postoperative cholangitis. In addition to the surgical procedure, careful postoperative management is also indispensable to obtain good bile flow. The necessity for administration of antibiotics and choleretics for a month or longer were proven by our clinical experience. The stimulation to postoperative bile excretion by the derivatives of bile acid or hormones such as glucagon and prostaglandin has not been studied sufficiently. The establishment of an adequate regimen of choleretics is required for improvement in the treatment of this disease.

Postoperative cholangitis affected not only the immediate results after corrective operation but also the recovery from chronic liver disease in long-term survivors. Prevention of cholangitis is the principal problem in the treatment of biliary atresia at the present time. Since 1968, many different forms of biliointestinal conduit reconstruction and exteriorization have been proposed to prevent postoperative cholangitis. However, there is no modification that prevents postoperative cholangitis completely except for hepatic portocholecystostomy. In our series, the incidence of postoperative cholangitis was not decreased by our introduction of the double Roux-en-Y anastomosis. All patients having Roux-en-Y interposition between the porta hepatis and the duodenum developed cholangitis. It is obvious, however, that the severity of postoperative cholangitis is decreased by employment of these modified procedures. Moreover, the early results of corrective surgery and the quality of life in long-term survivors improved in comparison to patients treated with the original Roux-en-Y procedures.

Hepatic portocholecystostomy carried out on patients with a patent bile duct from the gallbladder to the duodenum prevented cholangitis. Biliary obstruction, however, was frequently encountered in these patients after operation. These experiences prompted us to limit our indications for hepatic portocholecystostomy [26]. That is, this procedure should be performed on patients having fairly large distal bile ducts which have an internal diameter of more than 1 mm on the operative cholangiogram. Patients having evidence of a long common channel (pancreatic and the common bile duct) are excluded. Lilly [27] recommended temporary tube decompression of the biliary conduit in patients having hepatic portocholecystostomy to eliminate complications presumably resulting from impeded bile flow secondary to the hypoplastic distal ductal system.

Tanaka and Satomura [14] recently designed a jejunal antireflux intestinal valve and reported excellent results. Only 2 of 17 patients treated by their procedure had severe postoperative cholangitis and 16 patients are alive without jaundice. Kimura [13] pointed out that procedures with external diversion do not always prevent ascending cholangitis, especially cholangitis within 3 months after corrective surgery. He also stressed that Tanaka's method seemed better than any other procedure not only for preventing cholangitis but also for avoiding the unfavorable effects of biliary diversion. There is also the opinion that no differences exist between external diversion procedures and simple Roux-en-Y procedures with limbs of 50–70 cm. We still use the external diversion method because it lowers the severity of postoperative cholangitis and makes possible an estimation of the amount of bile excretion and permits biochemical analysis of bile.

The postoperative course is often eventful even in patients having active bile drainage. Gradual decrease of bile excretion or cessation of bile flow after cholangitis is most troublesome. The only way to help patients who do not respond to medical treatment is reoperation. Other patients requiring reoperation were those with poor or no bile flow after the initial operation. Our experience showed that a good bile flow was obtained by reoperation in the majority of patients who had once-active bile drainage. When these patients do not respond to vigorous medical treatment for 1–2 weeks after cessation of bile flow, reoperaion should be done. For patients without adequate initial bile flow, surgeons must decide about reoperation by the general condition, clotting functions, and the degree of liver parenchymal damage. As a guideline for deciding

whether to reoperate or not, we examine the findings of a hepatobiliary scintigraphy. When impairment of the hepatocyte clearance and a high degree of ectopic excretion of the isotope are seen, we do not reoperate. Two different reoperation procedures are reported. One is by resection of the scar tissue at the porta hepatis [15], the other is a curettage of the surface on the anastomosed portion [28]. Although resection of scar tissue is a difficult procedure, we still recommend it.

At present when corrective surgery is performed within 60 days after the birth, 80% of the patients with biliary atresia can achieve persistent bile drainage sufficient to return the serum bilirubin level to normal. Moreover, most jaundice-free infants can be completely cured by precise postoperative follow-up care. These patients are expected to grow and develop normally and have healthy lives, as our follow-up studies of long-term survivors demonstrate. In Japan, liver transplantation has not been clinically adopted mainly for religious reasons. Liver transplantation may be the only curative treatment for patients in whom hepatic portoenterostomy fails.

Résumé

Bien que les résultats de la chirurgie pour atrésie biliaire se soient considérablement améliorés depuis la mise en pratique de la portoentérostomie hépatique, des progrès restent à accomplir. De 1953 à 1983, soit pendant 31 ans, nous avons réalisé ce type d'intervention chez 214 malades. Au cours des 18 premières années le taux de succès concernant 98 sujets n'a atteint que 13%. Les 7 années suivantes, de 1971 à 1977, il a atteint 55% chez 65 opérés dont 29 sont actuellement vivants sans aucune trace de rétention biliaire. Au cours des 6 dernières années la rétention biliaire a été traitée avec succès chez 66% d'un groupe de 53 sujets, dont plus de la moitié ne présente aucune trace d'ictère. L'analyse rétrospective de cette série permet de définir les facteurs qui contribuent à l'amélioration des résultats opératoires: (a) diagnostic et intervention précoces, (b) dissection précise et section adéquate du moignon biliaire, (c) progrès du traitement postopératoire, (d) prévention de l'angiocholite postopératoire, et (e) réintervention précoce si nécessaire. La chirurgie correctrice pratiquée au cours des 60 premièrs jours qui suivent la naissance permet d'obtenir un drainage biliaire suffisant, un taux normal de bilirubine étant constaté chez 80% des opérés.

Resumen

Aun cuando los resultados de la cirugía para atresia biliar han mejorado en forma notoria después de la introducción de la portoenterostomía hepática, todavía es necesario un mayor progreso. En los 31 años transcurridos entre 1953 y 1983, hemos realizado cirugía correctiva en 214 pacientes. En los primeros 18 años la enfermedad pudo ser exitosamente tratada sólo en el 13% de 96 pacientes. En los 7 años siguientes (1971 a 1977), la ictericia pudo ser controlada en el 55% de 65 pacientes; actualmente 29 pacientes se hallan vivos y libres de ictericia. En los últimos 6 años la ictericia ha sido controlada en el 66% de 53 pacientes; más del 50% de los pacientes se hallan libres de ictericia en la actualidad. El análisis retrospectivo ha identificado algunos factores de importancia que contribuyeron a la superación de los resultados operatorios: (a) diagnóstico y operación precoces; (b) disección precisa y transección adecuada del cono fibroso del remanente extrahepático del canal biliar; (c) progresos en el manejo postoperatorio; (d) prevención de la colangitis postoperatoria; y (e) reoperación temprana cuando necesario. La cirugía correctiva realizada dentro de los 60 días después del nacimiento puede lograr un drenaje biliar suficiente para la normalización del nivel de bilirrubina sérica en más del 80% de los pacientes.

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