Second Operation for Repair of Biliary Atresia

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● **In our experience with biliary atresia, there are few cases amenable to reoperation for recurrent jaundice. All authors would agree that specific conditions such as complete bile flow recovery from the first operation followed by early recurrence should be an unquestionable case for revision of the anastomosis, inasmuch as no biologic signs of ongoing cholangitis can be traced. The same decision would apply to the problem of bile leakage after hepatoportocholecystostomy. In other cases, however, one should be aware that these reoperations expose the child to ascitis, poor healing of the abdominal wound, liver failure, and also bring with the decision to reoperate undue hopes to the parents of the child. Moreover, if the child should be a future candidate for liver transplantation, it may be wiser to avoid useless laparotomies and abdominal dissections that are known to complicate the task of hepatectomy.**

INDEX WORDS: Biliary atresia; hepatoportoenterostomy.

FROM 1968 to 1984, 353 children with biliary atresia (BA) have been submitted to surgical repair of the anomaly, by the same surgical team, first at Saint-Vincent of Paul's Hospital, and during the last years at Bicêtre Hospital. Among these patients, 21 were reoperated because of persistence or recurrence of jaundice with the hope of giving them a second chance by revising the anastomosis at the porta hepatis. The recommendation of a second-look operation in cases of failure of hepatoportoenterostomy (HPE) has been advocated by many operators, even in cases when the first operation was not a success. In our experience, different situations have led to the decision to reoperate, with unequal results.

MATERIALS AND METHODS

Among the 353 children treated for BA in our series, a second operation was undertaken for 34 of them. Of these, ten had been submitted elsewhere to only exploratory laparotomy, and three were referred after failure of HPE, but were only submitted to exploratory laparotomy at second look, so that the remaining 21 cases were collected for review (Table 1). The various situations in which a decision for reoperation was made led to the following classification in five groups: Group I (2 children). Early recurrence of jaundice, within 6 months after the first operation considered a success. Group II (6 children). Recurrence of jaundice occurred after incomplete recovery from the first operation with persistent mild biliary retention. Group III (3 children). Biliary peritonitis after hepatoportocholecystostomy (HPC). Group IV (6 children). Late recurrence of jaundice, more than 6 months after the first operation, which had been considered a complete success with disappearance of jaundice. Group V (4 children). The first operation brought no improvement. As it was performed elsewhere, a new attempt was felt justified in case the HPE had not been constructed with a correct technique.

Technique

Depending on the condition of the patient and the technique of the first operation, reoperation was done following different procedures. A HPE was established as a second operation in 18 cases. Eleven children had been previously treated with HPE; two others had a previous duodenal anastomosis on a cystic structure near the porta hepatis; the last five children had been treated by HPC. Three other patients who had been submitted to HPC as a first operation underwent the following secondary surgery: (1) anastomosis to the duodenum of the free gallbladder wall in one child aged 9 years where biliary lithiasis had developed with stenosis of the cystic duct; (2) anastomosis to the jejunum of the gallbladder in one case following a routine transcutaneous cholecystogram, which had been complicated with bile peritonitis and blood clotting within the gallbladder; and (3) re-HPC for a child operated elsewhere, and in which the gallbladder had been misplaced, close to the porta hepatis.

RESULTS

Results were assessed by clinical appreciation of jaundice and level of serum bilirubin. The result was considered good if jaundice cleared completely and serum bilirubin was under 17 μmol/L. Mild improvement was noted in cases with slight residual jaundice and serum bilirubin between 17 and 50 μmol/L.

In group I, both patients had good results and have remained anicteric respectively 2 and 8 years postoperatively. In group II, five of the six children subsequently died. One death occurred postoperatively with septicemia; one never recovered bile flow and progressively developed liver failure; the three others did temporally improve, but with episodes of cholangitis, their condition deteriorated leading to death between 15 months and 3 years postoperatively. The only patient surviving in that group is a girl aged 3 years who was submitted to a third HPE and whose serum bilirubin is presently 36 μmol/L. In group III, three children are surviving between 18 months to 5½ years postoperatively, two of them without jaundice, and one with elevated serum bilirubin at 80 μmol/L after episodes of cholangitis. In group IV, three patients have a good result respectively, 1, 2, and 11 years after the operation. One case has a mild improvement after

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Table 1. Clinical Features and Outcome

	Type of First Operation	Age at First Operation (d)	Type of Second Operation	Age at Second Operation	Results at 6 Months	Results at Last Examination	Follow-up
Group I							
1	HPE	72	HPE	131 d	Good	Good	2 yr
2	HPE	120	HPE	179 d	Good	Good	8 yr
Group II							
1	HPE	89	HPE	113 d	Mild	Mild	1 yr, 2 mo
2	HPE	63	HPE	114 d	Good	Died	
3	HPE	78	HPE	156 d	Mild	Died	
4	HPE	67	HPE	101 d	Poor	Died	
5	HPC	96	HPE	217 d	Mild	Died	
6	HPE	67	HPE	105 d	Died		
Group III							
1	HPC	39	HPE	66 d	Good	Good	5 yr, 6 mo
2	HPC	51	HPE	79 d	Good	Good	2 yr
3	HPC	126	HPE	157 d	Good	Mild	1 yr, 6 mo
Group IV							
1	HPE	54	HPE	5 yr, 9 mo	Mild	Good	2 yr, 1 mo
2	HPC	152	ChD	9 yr	Good	Mild	2 yr, 3 mo
3	CyD	45	HPE	7 yr	Poor	Poor	6 mo
4	CyD	48	HPE	11 mo	Good	Good	12 yr
5	HPE	69	HPE	1 yr, 3 mo	Mild	Poor	1 yr
6	HPC	57	ChJ	5 yr, 2 mo	Good	Good	1 yr
Group V							
1	HPC	28	HPE	126 d	Died		
2	HPE	78	HPE	151 d	Died		
3	HPE	57	HPE	92 d	Poor	Poor	1 yr
4	HPC	15	HPC	60 d	Good	Good	2 yr

Good, bilirubin < 17 μmol/L; mild, $17 <$ bilirubin < 50 μmol/L; poor, bilirubin > 50 μmol/L.

Abbreviations: HPE, hepatoportoenterostomy; HPC, hepatoportocholecystostomy; CyD, cystoduodenostomy; ChD, cholecystoduodenostomy; ChJ, cholecystojejunostomy.

just over 2 years. The last one is considered a failure at 6 months. In group V, only one child is cured 2 years after a second HPC, the first procedure done elsewhere having set the gallbladder in an uncorrect position. The three other cases were not improved and two of them died.

DISCUSSION

Some recent publications, mostly from Japanese literature[1-3] have underlined the need for reoperation in BA in specific cases. According to Kasai, most patients with once active bile drainage will recover bile flow after reoperation.[1] Postoperative cholangitis is as a rule considered the source of fibrosis causing secondary obstruction at the anastomotic site. In the case of severe infection with positive blood or liver samples culture, reoperation should of course be postponed until all inflammatory signs have subsided while biliary retention persists. In the case of HPC where ascending infection does not occur, fibrosis may likewise develop and obstruct the anastomosis. As it appears from patients studied in group IV, late recurrence of jaundice that occurred after 5 years for four children, may also be in relation to secondary development of scar tissue at the site of the anastomosis. A well-known complication leading to reoperation is postoperative intraperitoneal leakage of bile, as already reported by Altman[4] and by the authors.[5] The cause is probably accumulation of blood and mucosities inside the minute and disused cystic duct. Kasai[2] recommends that HPC should not be constructed in cases where the cystic duct appears to be too small on operative cholangiogram. Another late obstructive complication in one case of HPC has been cholelithiasis. In another case, lithiasis was suspected but could not be confirmed since hemobilia following a transhepatic cholangiogram led to an emergency cholecystojejunostomy with impossibility in locating the presumed gallbladder stone. Last, indication for reoperation would be the case operated elsewhere by an inexperienced surgeon. One of our patients had been formerly submitted to HPC; at reoperation, the gallbladder was found to be positioned in the wrong site, near the porta hepatis. Some children with incomplete bile flow recovery in our series of BA treated by HPE will show on abdominal ultrasonogram the presence of small intrahepatic cysts communicating with the irregular bile duct system (Figs 1 and 2), as shown on transhe-

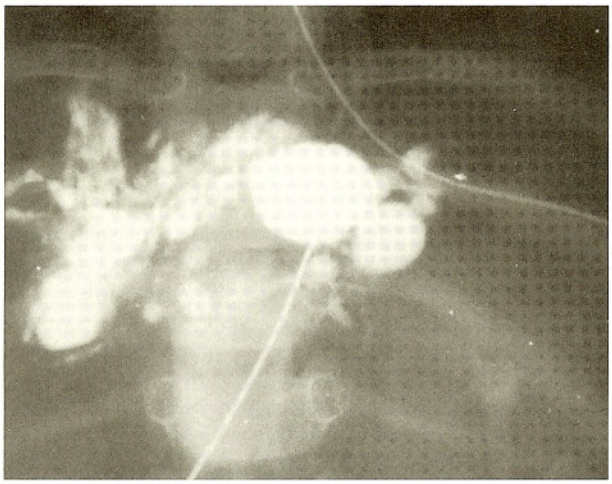

Fig 1. Girl treated by HPE at age 78 days. Persistent jaundice (group II). Reoperation at age 276 days with moderate recovery of bile flow. Transhepatic cholangiogram 2 months later, when jaundice recurred, showing numerous intrahepatic cystic structures; bile drainage from these cysts through a percutaneous catheter.

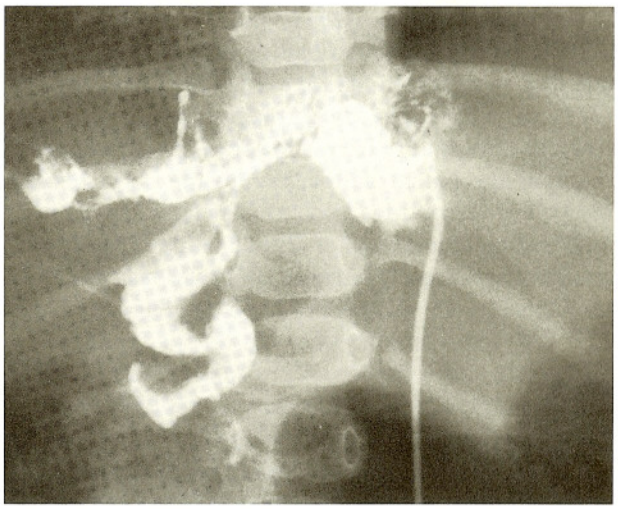

Fig 2. Same child as in Fig 1, 2 months later, with very moderate bile retention. Transhepatic cholangiogram showing good flow of dye through the anastomosis.

patic cholangiogram. In no case, however, was another anastomosis possible with these cystic cavities deeply situated within the liver parenchyma.

In 18 cases, technique for reoperation in our series was construction of an HPE after removal of fibrous tissue at the porta hepatis, using the same jejunal limb in the 13 cases of previous HPE, or cystoportoenterostomy when a small cyst had been discovered during the first operation; or replacing the gallbladder with a Roux-en-Y anastomosis in five other cases. In three cases, a different kind of anastomosis was constructed, joining the gallbladder to the jejunum or the duodenum in two cases, and transposing the gallbladder to the correct position in the last one. Suruga et al[3] have advocated curettage of the porta hepatis via the jejunal loop for children who had secondary cessation of bile flow, with the possibility of a good result, providing curettage was done within 30 days after recurrence of bile retention. If the dissection during the first operation has been correctly carried out, with removal of the fibrous remmants of the biliary ducts close to the posterior aspect of the division of the portal vein, the revision of the anastomosis should only consist in removing scar tissue without further penetration of liver parenchyma at the porta hepatis, which could only provoke bleeding and more fibrosis.

The outcome after reoperation is unpredictable. In our series, complete bile flow recovery was not observed in cases where the first operation had not brought at one time complete disappearance of jaundice. Out of the 21 children with BA reoperated, seven have died, three after partial recovery of bile flow, three have mild biliary retention, and eight are considered cured with normal serum bilirubin levels at last examination. There are survivors from each group, free from jaundice, except from group II, where jaundice had never cleared completely before the second operation. No statistical analysis can be done from this study, since the variety of conditions of failure after the first operation are numerous, and each group too small. Kasai states that 17 of his 32 patients reoperated had excellent bile drainage after reoperation,[2] but that on the other hand, only 11 demonstrated disappearance of jaundice. Suruga considers curettage as an effective mode of treatment, since four out of 17 cases have been cured from jaundice by those means.[3]

There were three postoperative deaths in our series. Two children from group V submitted to re-HPE died respectively, 1 and 5 months later from liver failure; another one from group II, submitted 3 months after HPC to HPE, died in the immediate postoperative period from sepsis. Other series report immediate postoperative deaths.[4] Although none of these patients should be considered for reoperation because they have ascitis and severe portal hypertension, most of the patients reoperated are not in a very good nutritional state, and decompensation of the liver function postoperatively must be feared.

CONCLUSION

During a 16-year period, 21 cases of biliary atresia, among a total series of 353 children treated by the authors since 1968, were submitted to surgical revision of the anastomosis. They were divided into five groups corresponding to the different circumstances leading to

a second operation. Good results could only be achieved in patients who had demonstrated complete bile flow recovery at one time after the first operation (2/2) or when a hepatoportoenterostomy was constructed after failure from bile peritonitis after anastomosis of the patent gallbladder to the porta hepatis (2/3). Late second-look operations could also be indicated after recurrence of bile retention, with good results when a precise cause of cholestasis could be found, such as cholelithiasis (2/6). However, the chances of cure at second operation are few, and the hazards of postoperative complications in these children should lead to careful selection of potential candidates, especially if a liver transplant is foreseen for the child in the near future.[6]

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