Hepatic Portoenterostomy:
An Assessment of Its Value in the Treatment of Biliary Atresia

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A series of 12 infants undergoing hepatic portoenterostomy for
correctable-type biliary atresia is reviewed. There has been no
evidence of a sustained postoperative increase in bile excretion or
improvement in biliary cirrhosis in any patient. Survival statistics
for the group as a whole are poor with a mean total survival time of 11 months and a mean total survival time of 15.5 months.
These survival times are worse than that previously reported for infants with untreated biliary atresia. Our disappointing results with hepatic portoenterostomy raise doubts concerning its value
in the treatment of biliary atresia.

Adequate surgical drainage of the biliary tree in infants with biliary atresia remains an unsolved problem for the majority of such infants have an incorrectable type lesion, i.e., no extrahepatic bile duct is available for
anastomosis to the intestine. The history of attempts to surgically circumvent this problem has been most depressing. Claims of success with such procedures as the use of artificial bile ducts, and the drainage of thoracic duct lymph, and hepatic lymphatic drainage to the G.I. tract have all eventually proven to be false.

Biliary atresia, for some unexplained reason, appears to be more common in Japan than elsewhere. Interest in amelioration of the disease by surgical means has therefore flourished in that country. In 1957 Kasai first reported the use of hepatic portoenterostomy as a means of dealing with incorrectable types of biliary atresia, and subsequently claimed a cure rate of approximately 10%. A more recent report, combining several large Japanese series, claims an approximately 40% cure rate for infants

with biliary atresia using Kasai's basic technique together with sundry modifications.

The optimism engendered by the reports of Kasai and others in Japan led us 5 years ago to begin performing hepatic portojejunostomies on all infants with biliary atresia in whom an extrahepatic bile duct was not available for anastomosis to the intestinal tract. The diagnosis of an incorrectable-type lesion was made at surgery after operative cholangiograms and careful dissection of the porta hepatitis. All infants undergoing hepatic portoenterostomy have been under continual close postoperative surveillance.

Clinical Material

Between June 1969 and December 1974, we have seen 12 infants with incorrectable-type biliary atresia at Oklahoma Children's Memorial Hospital. Prior to surgical exploration, all underwent extensive evaluation to rule out other possible causes of persistent neonatal jaundice. The evaluation of persistent neonatal jaundice at our institution includes serum Lipoprotein-X determinations before and after a short course of cholestyramine. The first 10 patients in our series were also evaluated with double I-Rose Bengal excretion tests as reported by Poley, et al. Percutaneous needle biopsy of the liver was a routine part of the diagnostic work up in all patients studied.

All infants were explored as soon as possible after making the diagnosis of biliary atresia. Exploration was done through a long, right subcostal incision which ex-
tends across both rectus muscles. The peritoneal cavity is explored and wedge biopsies of the liver obtained. We then proceed exactly as outlined by Kasai in his original reports. We have deemed it inadvisable to use any of the technical modifications which were reported by Kasai and his co-workers after the onset of our study. Attempts are first made to locate the extrahepatic biliary tree or a portion thereof through which to perform a cholangiogram. If an extrahepatic bile duct or gallbladder is identified, it is cannulated with a fine polyethylene catheter and flushed with sterile saline. An operative cholangiogram is then performed by the gentle injection of 4 to 5 cc of 50% Hypaque through the catheter. If an extrahepatic biliary tree or portion thereof is not identified with the above maneuvers, the gallbladder and cystic duct are dissected free and dissection of the porta hepatis carried out. If an extrahepatic biliary tree is still not identified, a transverse incision is made on the anterior surface of the hepatoduodenal ligament and the mass of tissue lying anterior to the hepatic artery and portal vein dissected up to the hilum of the liver. This mass of prevascular tissue is dissected as close as possible to the liver substance and the specimen removed for microscopic examination. A Roux-en-y type portoenterostomy is carried out between the small remnant of connective tissue left at the liver hilum and the proximal jejunum. The Roux-en-y anastomosis is constructed so as to form a 25 to 30 cm jejunal spur. This segment of jejunum is brought up to the stump of the tissue dissected off the vessels of the porta hepatitis through the transverse mesocolon. The area of anastomosis is drained through a separate lateral stab wound.

All infants undergoing hepatic portoenterostomy have received close continuous postoperative surveillance. All survivors are readmitted every 3 months and evaluated with liver function studies and percutaneous needle biopsy of the liver. For the past two years, we have also included serum Lipoprotein-X determinations and I-Rose Bengal excretion studies as a routine part of this evaluation.

Autopsies were carried out in 4 of the 9 deaths. A needle biopsy of the liver had been performed within 3 months of death in all infants in whom an autopsy was not performed.

Results

We have had one postoperative death in our series of 12 patients for an operative mortality of 8.3% (Table 1). This death was due to respiratory complications which developed on the day of surgery. The patient (L.W.) was 13 months of age and in end-stage liver failure at the time of surgery.

There have been three significant postoperative complications for a complication rate of 25% (Table 2). Patient K.W. (Table 1) developed massive upper gastrointestinal bleeding secondary to multiple gastric stress ulcers in the first week postop. She required vagotomy and pyloroplasty for control of the hemorrhage. Patient P.B. required exploratory laparotomy 8 days postop for intestinal obstruction secondary to adhesions. At the time of exploration, a right subphrenic abscess was discovered and drained. Patient M.H. suffered from moderately severe upper G.I. bleeding in the first week postop. He required blood transfusions, but the bleeding ceased spontaneously without surgical intervention. Upper G.I. series failed to reveal the source of the bleeding.

Liver biopsies taken at the time of operation have shown evidence of biliary cirrhosis in all patients, despite the fact that the majority were under 3 months of age. The degree of cirrhosis has correlated roughly with the age at operation. Infants operated on at 3 months of age or younger have shown primarily severe cholestasis and early fibrosis while several of the older patients have had full blown biliary cirrhosis at the time of surgery. Subsequent evaluation of hepatic morphology by both 3 monthly needle biopsy and autopsy examination has revealed a steady progression of the cirrhosis in all 12 infants. The microscopic findings have been those of increasing fibrosis, cholestasis and hepatocyte loss.

### Table 1. Survival Statistics in Infants Undergoing Hepatic Portoenterostomy

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at Operation (Mo.)</th>
<th>Postop Survival Time (Mo.)</th>
<th>Total Survival Time (Mo.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>JW</td>
<td>5</td>
<td>52*</td>
<td>57*</td>
</tr>
<tr>
<td>LW</td>
<td>13</td>
<td>0</td>
<td>13</td>
</tr>
<tr>
<td>JF</td>
<td>3</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>JV</td>
<td>3</td>
<td>10</td>
<td>13</td>
</tr>
<tr>
<td>GM</td>
<td>3</td>
<td>10</td>
<td>13</td>
</tr>
<tr>
<td>KW</td>
<td>2.5</td>
<td>3.5</td>
<td>6</td>
</tr>
<tr>
<td>DG</td>
<td>2.5</td>
<td>11</td>
<td>13.5</td>
</tr>
<tr>
<td>AS</td>
<td>2</td>
<td>15</td>
<td>17</td>
</tr>
<tr>
<td>MH</td>
<td>8</td>
<td>4.5</td>
<td>12.5</td>
</tr>
<tr>
<td>AF</td>
<td>2.5</td>
<td>14**</td>
<td>16.5**</td>
</tr>
<tr>
<td>CB</td>
<td>4</td>
<td>3.5</td>
<td>7.5</td>
</tr>
<tr>
<td>BB</td>
<td>2.5</td>
<td>7**</td>
<td>9.5**</td>
</tr>
<tr>
<td>MEAN</td>
<td>4.3</td>
<td>11.0</td>
<td>15.3</td>
</tr>
</tbody>
</table>

*Orthotopic liver transplant at 52 mos post hepatic portoenterostomy
**Alive, terminal liver failure

### Table 2. Postop Complications in Infants Undergoing Hepatic Portoenterostomy

<table>
<thead>
<tr>
<th>Complication</th>
<th>Count</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Upper G.I. Bleeding</td>
<td>12</td>
<td>16.7%</td>
</tr>
<tr>
<td>Requiring Transfusion</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Intestinal Obstruction</td>
<td>12</td>
<td>8.3%</td>
</tr>
<tr>
<td>Secondary to Adhesions</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total Post Op Complication</td>
<td>25%</td>
<td></td>
</tr>
</tbody>
</table>


Survival data is outlined in Table 1. The average age at operation was 4.3 months. This is skewed by patient L.W. who was 13 months of age at the time of hepatic portoenterostomy. The mean postoperative survival time is 11 months. Patient J.W., the first patient in our series, underwent an orthotopic liver transplant 52 months post hepatic portoenterostomy for end-stage liver failure. Her postoperative survival time has been calculated to the time of liver transplant.

The mean total survival time for the 12 patients is 15.3 months (Table 1). Three patients are still alive. Patient J.W., who underwent a liver transplant, is still alive and doing well 14 months post transplant. Her total survival time has been calculated to the time of transplant. Two other patients, A.F. and B.B. are still alive 14 and 7 months post hepatic portoenterostomy. Both patients are in end-stage liver failure with markedly abnormal liver function studies and liver biopsies showing evidence of rapidly progressing biliary cirrhosis. Both suffer from severe ascites which is difficult to control with diuretic therapy. Both are now considered candidates for liver transplant.

Comprehensive evaluation of our postoperative survivors, which is carried out every 3 months, has been remarkably similar in all patients. Although serum bilirubin levels will sometimes show intermittent improvement, we have not seen continued improved bile excretion or a return of bilirubin levels to normal in any patient. Hepatic excretory function as measured by LP-X determinations and 

![Graph showing survival comparison in infants with untreated biliary atresia and those with biliary atresia posthepatic portoenterostomy.](image_url)

Fig. 1—Survival comparison in infants with untreated biliary atresia and those with biliary atresia posthepatic portoenterostomy.

- Infants with untreated biliary atresia
- Infants with biliary atresia, post hepatic portoenterostomy (present study)
Discussion

Bill has recently reviewed a combined series of infants in Japan undergoing hepatic portoenterostomy for incorrectable biliary atresia between 1968 and 1971. This combined series shows a 43% cure rate for infants undergoing hepatic portoenterostomy before 3 months of age, and 7% cure rate for infants operated on after 3 months of age. Eight or 67% of the infants in our series were 3 months of age or younger at the time of surgery. We have seen no cures with hepatic portoenterostomy. In fact, the mean postop survival time is only 11 months despite patient J.W., who lived for 52 months before liver transplant. The mean total survival time is only 15.3 months, which is actually less than the average total survival time reported previously by Hays and Snyder for infants with untreated biliary atresia. Hays and Snyder had two patients who survived longer than 53 months without treatment, so one must be wary of supposed cures reported as early as one year postop. If one plots the survival statistic for our series of 12 infants who have undergone hepatic portoenterostomy, against the series of untreated infants reported by Hays and Snyder (Figure 1), one notes that there is no significant improvement in survival with hepatic portoenterostomy. One must conclude that hepatic portoenterostomy provides no therapeutic advantage in the treatment of biliary atresia.

Since the Japanese combined series totals 215 patients, one can question whether our series is large enough to be significant. Statistical evaluation of our data shows that if the expected cure rate for the procedure is 10%, the probability of our having no cures in 12 patients solely by chance is 28.2%. If the expected cure rate is approximately 40%, as reported in the latest Japanese series, the probability of seeing no cures in 12 patients drops to 0.21%. One must conclude that the probability of our having no cures, purely by chance, is quite remote.

One may logically ask if we are continuing to perform hepatic portoenterostomies on infants with incorrectable types of biliary atresia. We are continuing to perform the procedure for, at present, we have nothing other than liver transplant to offer these unfortunate infants. Transplant surgeons, in general, do not believe that a previous hepatic portoenterostomy interferes with the performance of a liver transplant. They, in fact, use the Roux-en-Y limb of jejunum to drain the biliary tree of the transplanted liver. This is considered an advantage for it means one less anastomosis to perform at the time of transplant and also one less anastomosis to cause problems in the post transplant period. In addition, statistical evaluation reveals that if one accepts the 10% cure rate originally reported by Kasai, there is a 28.2% probability that we have seen no cures with hepatic portoenterostomy because our series is not yet large enough.

References


We have had experience with only three patients. Two are still alive, one for 18 months and one for four months. The first initially had a repair of a jejunal atresia and an accompanying gastrostomy. A portocholecystostomy was done one month later with resulting clearling of the jaundice. One year later the liver was biopsied at the time of closure of the gastrostomy. Residual cirrhosis was prominent but the fibrosing process did not appear to be active. The baby has continued to do well. The liver is still palpable. We admit the baby may eventually show liver failure, but we certainly are better off than if we had not tried.

In the last patient we performed the Kasai procedure, a double Roux-en-Y with a jejunojunostomy stoma. Bile drainage through the stoma has been so persistent that the mother has said, "Please close it because the bile is running all over the baby's clothes, and I cannot keep her clean." Kasai has recommended that the jejunojunostomy be made as a safety valve to guard against retrograde cholangitis from any intercurrent gastrointestinal upset. He recommended that this be left open for six months to one year. The baby's liver enzymes have returned to normal and bilirubin from 16 mg% to normal.

In the tissue removed from these two cases microscopic ducts, or at
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least epithelial lined tracts, were found. 50 μ in diameter in one and up to 100 μ in diameter in the other. No ducts were found in the baby who put out no bile.

Our results so far are encouraging, and we feel the procedure is worth the effort as the best currently available.

DR. JUDSON G. RANDOLPH (Washington, D.C.): This is a very discouraging problem, and I think the group at Oklahoma is to be congratulated on this frank courageous presentation.

I'd like to present to you data from the work that's going on in our clinical research center under the direction of my colleague, Dr. Peter Altman. We have studied 16 patients with biliary atresia in the last 22 months. One of the differences in our experience from that reported by Dr. Williams and Dr. Campbell and their co-workers may be based on our decision not to operate on any children that are over three months of age.

(Slide) The dissection must be carried well into the hilum. A frozen section of the tissue to be used for anastomosis is essential to assure the presence of duct structures. We chip away at the liver until we find a series of tiny ducts, then the stoma loop is put around this area, as Kasai had described.

(Slide) Here's one of our patients whose preoperative liver scan had no isotope coming from the liver. Yet, here in the postoperative period some months later, normal activity flow into the intestine can be seen.

(Slide) This shows one of our patients who is completely free of jaundice. Naturally, I picked one of our best babies to show you.

(Slide) Here are the data from this group of patients. Sixteen patients, in seven of whom there was no drainage. In this group are two patients over three months of age. Here there were nine who had biliary drainage, and these patients have been followed from 6 to 22 months. One died early in the postoperative period. These other eight survivors have liver scars, showing normal amounts of radioactive material excreted, and none of them are clinically jaundiced. Their bilirubin range is between 3 and 1. So, 50% of the patients are draining apparently normal amounts of bile.

But note that five of the eight survivors have continued cirrhosis, as indicated on their subsequent biopsy studies; some open, some by needle. Only one had no cirrhosis, although two are not yet biopsied, they are so early in their postoperative course.

This would indicate that in spite of biliary drainage by the Kasai procedure, we're still seeing on-going fibrosis, as if there is more to the etiology, or the pathophysiology, of biliary atresia than just absence of ducts. But we are encouraged, and will continue to study and operate on those patients that are under three months of age.

DR. DAVID P. CAMPBELL (Closing discussion): I would like to thank our discussants. I agree with both Dr. Shaffner and Dr. Randolph that these patients are, in general, a very discouraging group. We have, however, not given up the procedure despite our rather disappointing results.

One must be hesitant in claiming satisfactory operative results in these infants prior to two to three years postop, because, as per the last slide shown by Dr. Williams, they may live for four years or more without any treatment. I would also like to emphasize the fact that there may be great fluctuations in liver function studies, including serum bilirubin, in the postoperative period. Statements regarding immediate improvement in liver function postoperatively must therefore be evaluated with caution. A rather prolonged period of postoperative evaluation is necessary, for despite intermittent decreases in serum bilirubin levels and what seems to be evidence of improved hepatic excretion of bilirubin, there is a continual discouraging increase in the fibrotic process within the liver.

I think that Dr. Randolph hit the nail on the head when he said there is much more to the pathophysiology of biliary atresia than obstructed bile ducts. We presently believe, as do other workers in the field, that biliary atresia and neonatal hepatitis are different expressions of the same disease, i.e., an inflammatory process of unknown etiology involving the biliary tree and periductal hepatic tissue. This inflammatory process may clear spontaneously and be labeled neonatal hepatitis or, in a small percentage of patients, progress to fibrosis of the biliary tree and surrounding tissue and present as biliary atresia.