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A NEW OPERATION FOR NONCORRECTABLE BILIARY ATRESIA

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ABSTRACT

An improved operative technique to transect the fibrous cord by dividing the ligamentum venosum (Arantius' canal) is described for noncorrectable biliary atresia. The Arantius' canal is situated cranial and posterior side to the bifurcation of the umbilical portion and the portal branch of the Quinous' segment 3. The portal vein is fully mobile and the porta hepatis can be widely exposed by dividing the Arantius' canal. The fibrous cord of the porta hepatis can easily be dissected posteriorly and laterally where there is an extensive number of bile ducts. Eight patients with biliary atresia underwent this procedure. Jaundice resolved completely (serum total bilirubin concentration: ≤1 mg/dl) in 7 patients within 40 days. Postoperative cholangitis did not occur. By dividing the Arantius' canal, the portal vein comes free from the portal fissure to make the hepatic hilum wider, and surgeons are able to work within a larger porta hepatic space without causing portal vein compression. Free drainage of the bile from the porta hepatis may prevent postoperative cholangitis and promote resolution of jaundice.

Key Words: biliary atresia, Kasai's operation, hepatic portoenterostomy, Arantius' canal ductus venosus

INTRODUCTION

Biliary atresia was first identified as a specific disease entity in 1892.[1] In 1916, Holmes[2] introduced the concept of correctable and noncorrectable biliary atresia. The first successful surgery for correctable biliary atresia was reported by Ladd in 1928.[3] For noncorrectable biliary atresia, several unsuccessful surgical trials were reported in the 1950s and early 1960s. Kasai et al.[4] first reported hepatic portoenterostomy for noncorrectable biliary atresia in 1959. Recently, survival in children with biliary atresia has markedly improved with the development of Kasai's hepatic portoenterostomy.[5] However, there are many problems associated with hepatic portoenterostomy, such as postoperative ascending cholangitis, recurrent jaundice, and portal hypertension. In this manuscript, an improved operative technique designed to prevent these complications is described for noncorrectable biliary atresia.

CONVENTIONAL HEPATIC PORTOENTEROSTOMY

In patients with biliary atresia, there is a cone of fibrous tissue which is usually attached to the hepatic parenchyma in the middle portion of the portal fissure. It is believed that the portal fibrous tissue, which is thought to be the remnant of the extrahepatic bile ducts, contains many

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small bile ducts communicating with major intrahepatic ducts. Kasai believed that bile flows through these small bile ducts. Careful dissection of the portal fibrous tissue is critical to the success of this operation because it preserves drainage of bile from the intrahepatic bile ducts to the intestinal tract.[6,7,8] In our experience, however, only 11 (25.0%) of 44 patients survived without jaundice (serum total bilirubin concentration: ≤1 mg/dl).

HEPATIC PORTOENTEROSTOMY WITH EXTENDED EXPLORATION OF THE PORTAL AREA

The portal fibrous cord in patients with biliary atresia is usually attached to the middle portion of the portal fissure (Fig. 1). This area, however, is not the site of the portal tract in the normal liver. In a normal liver, the right and left hepatic ducts are situated in the ventral and cranial side to the right portal vein where it divides into the anterior and posterior segmental branches and cranial side to the transitional portion of the left portal vein (Fig. 2). Macroscopic and microscopic studies of the portal tract in liver specimens obtained at autopsy from patients with noncorrectable biliary atresia have demonstrated that the anatomic relationship of the major bile ducts to the portal veins and hepatic arteries is similar to that in the normal liver.[9] In addition, the major intrahepatic bile ducts were absent with varying distance from the portal fissure. Therefore, exploration of the portal tract should be carried out ventral and cranial side to the right portal vein where it divides into the anterior and posterior segmental branches, and cranial side to the transitional portion of the left portal vein, close to the obstructed ends of the major intrahepatic bile ducts (Fig. 3). A Roux-en-Y hepatic portoenterostomy is performed on the exposed portions of the right and left portal tracts (Fig. 4). A 3-cm incision is made on the antimesenteric border of the jejunum. The jejunum is anastomosed to the porta hepatic space. The lateral corners of the jejunal opening are brought across the blood vessels and sutured to the hepatic parenchyma.

Between 1980 and 1995, hepatic portoenterostomies with extended exploration of the portal area were performed on 66 patients with noncorrectable biliary atresia. Jaundice, which resolved in 73.5 ± 41.4 days, cleared completely in 41 patients (62%). These results were superior to those achieved with conventional hepatic portoenterostomy. However, postoperative cholangitis was observed in 32 patients (48.5%). Additional operations for recurrent jaundice were required in 15 patients (22.7%) due to occlusion of the explored portion of the portal area between the liver and the portal vein.

HEPATIC PORTOENTEROSTOMY WITH DIVISION OF THE ARANTIUS' CANAL

One reason why the explored area is easily occluded is that the space is so narrow that it is difficult to open the jejunal lumen. In addition, exploration of the portal tract is difficult laterally and cranio-posterior side to the left portal vein, because the portal vein is fixed at the hepatic porta. A solution to these problems is to detach the portal vein from the hepatic porta. The portal vein is fixed by several branches to the caudate lobe and the ligamentum venosum (Arantius' canal). The portal vein can be freed from the hepatic porta by resecting of these vessels.

Such resection begins by separating the gallbladder from the liver bed, after which the fibrous cord is dissected upward, denuding the portal vein and the hepatic artery. After isolation of the portal vein from the hepatic parenchyma, several fine venous branches to the left and right caudate lobe are ligated and divided. The ligamentum venosum, branching from the left

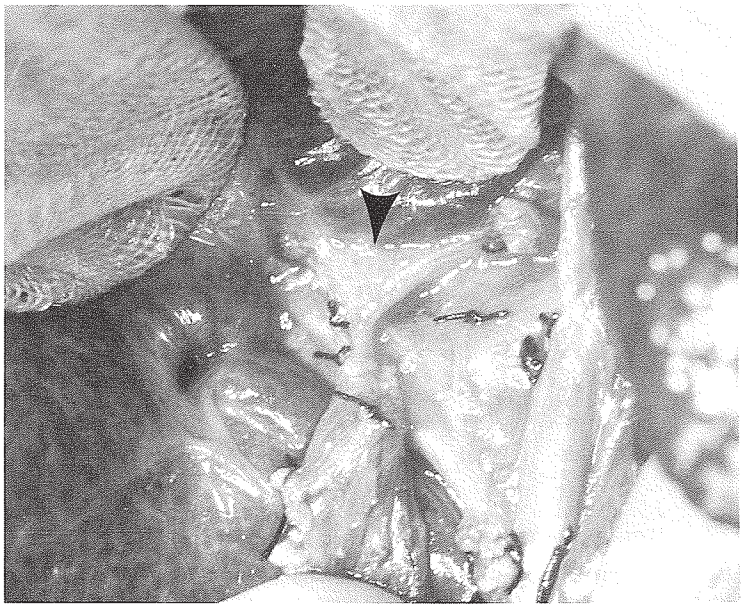

Fig. 1: Attachment of the portal fibrous cord. A cone of fibrous tissue is attached to the hepatic parenchyma in the middle portion of the portal fissure at the porta hepatis (arrow).

Fig. 2: Anatomy of the porta hepatis. The hepatic ducts (arrow) are seen ventral and cranial side to the portal vein. HA: hepatic artery, PV: portal vein

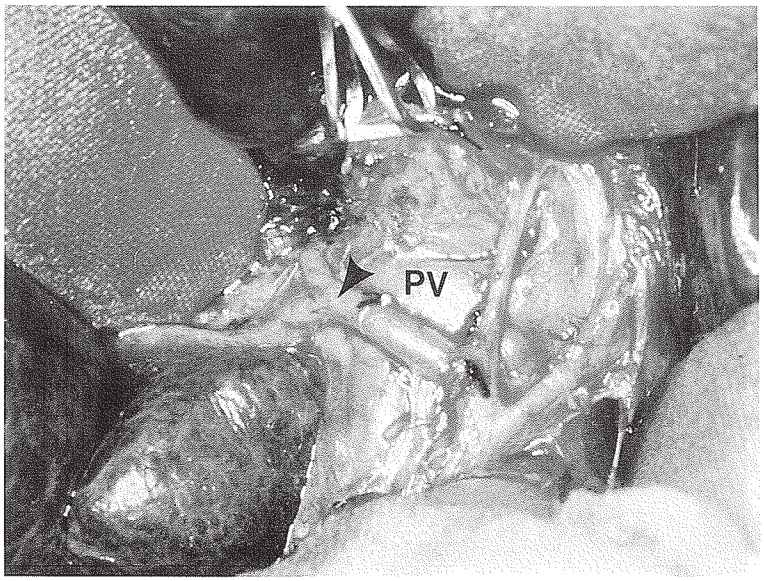

Fig. 3: Extended exploration of the hepatic porta. Exploration of the portal area is carried out beyond the point where the right hepatic artery (arrow) divides into the anterior and posterior segmental branches. PV: portal vein

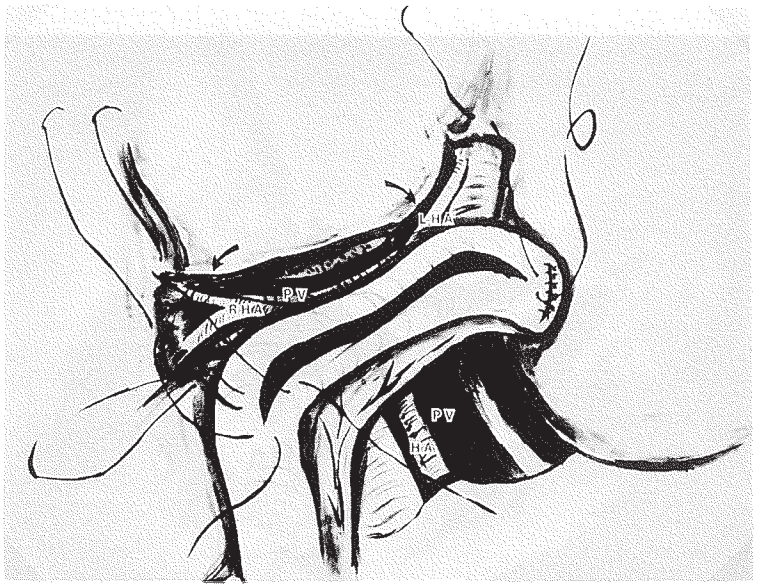

Fig. 4: Hepatic portoenterostomy with extended exploration. The jejunum is anastomosed to the caudate lobe posteriorly, and to the quadrate lobe anteriorly. On the right lateral border, the suture line skips over the portal vein and the hepatic artery.

portal vein at the bifurcation of the dorsal-lateral segment and the umbilical portion, is then ligated and divided (Fig. 5). The portal vein is then fully mobile, and the hepatic porta can be widely exposed (Fig. 6). The posterior aspect of the bile duct remnant is freed completely. The fibrous cord can be dissected posteriorly and laterally. The portal fibrous cord of the hepatic porta is transected using sharp scissors, and a small part of the hepatic hilar tissue is left attached to prevent injury to the liver parenchyma. The bile duct remnants are transected at the level of the posterior surface of the portal vein. The lateral margins of the fibrous cord are transected where there are many bile ducts. If the middle hepatic artery interferes with transection of the fibrous cord or the subsequent anastomosis to the jejunum, it can be ligated and divided. Bleeding points after transection should be identified with magnification and controlled using fine needle electrocautery after irrigation with warm saline. Bile can be seen flowing from the transected lateral surface of the bile duct remnant.

Thereafter a Roux-en-Y portoenterostomy is performed. The Y loop of this construction is approximately 50 cm long, and the limb is not externalized. The closed end of the Roux-en-Y limb is brought up to the porta hepatis through the transverse mesocolon. A 3-cm incision is made on the antimesenteric border of the jejunum. The jejunum is anastomosed to the caudate lobe posteriorly, and the quadrate lobe anteriorly. On the right lateral border of the anastomosis, the suture line skips over the right portal vein, running parallel to the anterior aspect of the right hepatic artery. The anastomosis of the left lateral border is made behind the left portal vein. This provides a large work area within the porta hepatic space without causing portal vein compression. Therefore a portoenterostomy is easy to perform.

Between February 1996 and January 1998, 8 patients with biliary atresia underwent this procedure. Jaundice resolved completely in 7 patients (87.5%) within 40 days (Figs. 7, 8). One patient underwent a living donor liver transplantation because of persistent jaundice. Neither postoperative cholangitis nor recurrent jaundice occurred in any of the patients.

DISCUSSION

Most modifications of the Kasai procedure, such as external jejunostomy,[10,11,12] intussuscepted ileocecal conduit,[13] and jejunal valve conduits,[14,15] have been developed to prevent ascending infection by intestinal bacteria. The technique of hepatic portal dissection itself has received little attention.[8,9,16] Although many factors determine the ultimate outcome of treatments for biliary atresia, adequate biliary drainage following transection of the fibrous cord, and protection of the hepatic hilum from portal vein compression, are the two most important factors in the prevention of ascending cholangitis. In hepatic portoenterostomy with division of the Arantius' canal for noncorrectable biliary atresia, transection of the fibrous cord is easily accomplished, and portal vein compression at the hepatic hilum is prevented. The portal vein moves freely, and the porta hepatis is wider after division of the Arantius' canal. We have encountered no problems with ligation and division of this ligament because it is an obliterated vestige of the ductus venosus.[17] Free drainage of the bile from the porta hepatis may prevent postoperative cholangitis and promote resolution of jaundice.

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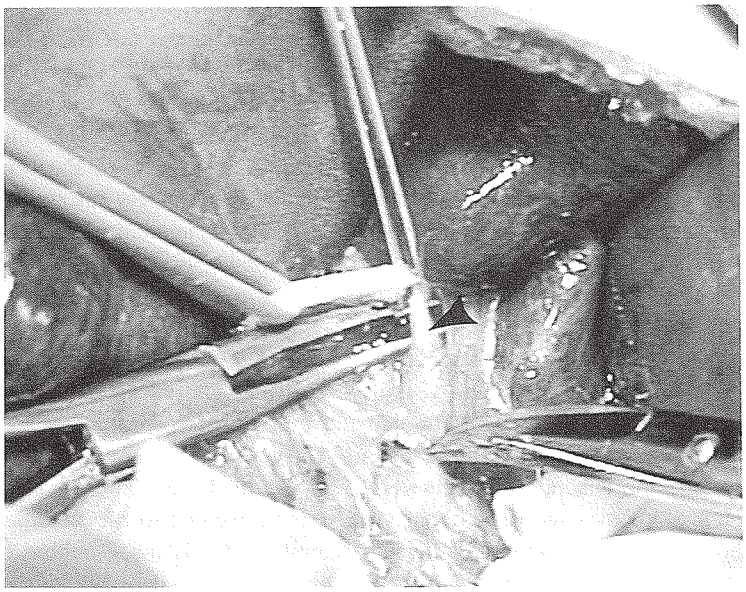

Fig. 5: Separation of the Arantius' canal: The Arantius' canal (arrow) is cranial and posterior side to the bifurcation of the umbilical portion and the portal branch of Couinaud's segment III.

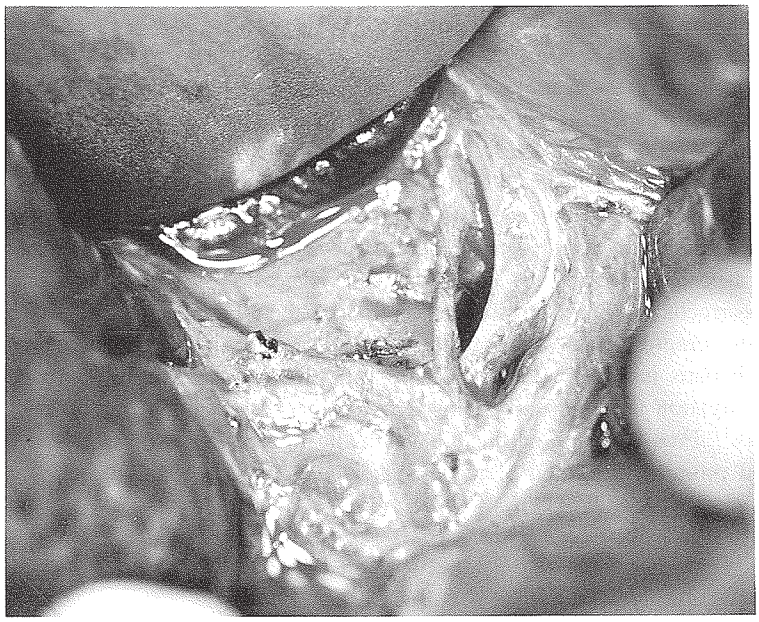

Fig. 6: The porta hepatis after division of the Arantius' canal. The porta hepatis is widely exposed.

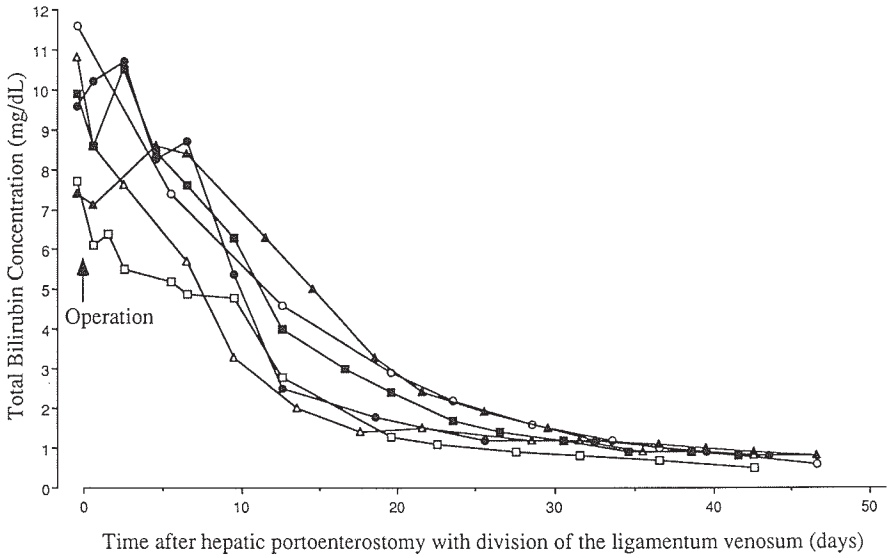

Fig. 7: Total bilirubin concentration after hepatic portoenterostomy with division of the Arantius' canal. Total bilirubin concentration decreased than 1 mg/dL within 40 days after surgery.

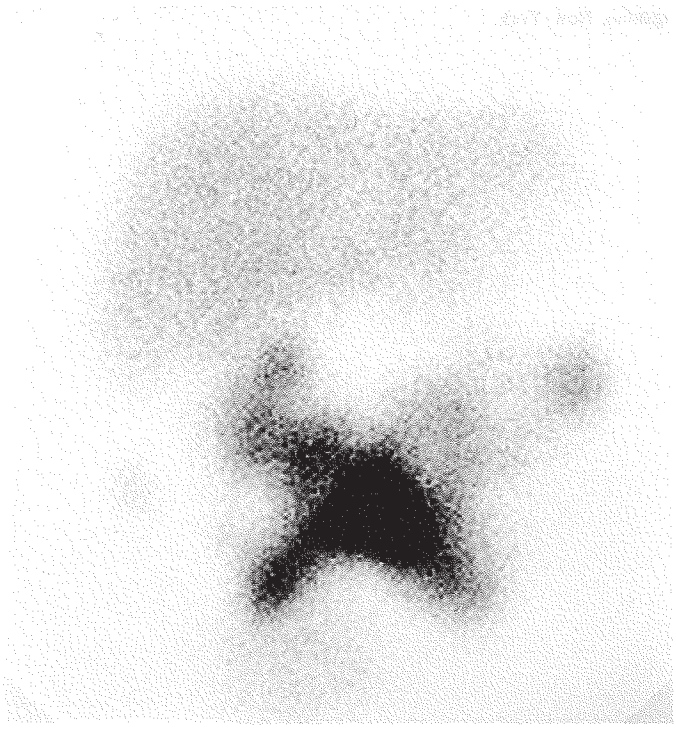

Fig. 8: Postoperative scintigram (1 hr after injection). Smooth bile drainage is shown by Tc^{99} scintigraphy.

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