

26-Year-Old Survivor of Kasai Procedure With Native Liver

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Abstract

A 26-year old woman with history of congenital biliary atresia and Kasai procedure as a newborn presented with recurrent nausea and abdominal pain. Physical examination showed anicteric sclera and tender right upper quadrant. Elevated liver function tests suggested obstruction and hepatitis. Percutaneous transhepatic cholangiogram showed generalized ductal dilation, innumerable intrahepatic bile duct stones, and multiple filling defects. No anastomotic stenosis was noted. The patient was treated for acute symptoms and referred to a liver transplant center. Long-term survivors of the Kasai procedure with a native liver are rare, and this represents the third known U.S. case in the literature.

Introduction

Congenital biliary atresia is the most important cause of neonatal cholestasis.¹ The Kasai procedure was introduced in 1959 as a neonatal hepatic portoenterostomy to re-establish bile flow.² The long-term survival with a native liver and the natural history prior to ultimate liver transplantation are not well known, with only a few reported cases in the U.S. literature.³

Case Report

A 26-year-old woman with congenital biliary atresia presented initially with neonatal jaundice and acholic stool. She underwent the Kasai procedure within 3 weeks of birth and had a normal childhood. She was hospitalized 2 years ago with nausea and right upper quadrant pain. Magnetic resonance cholangiopancreatography (MRCP) at that time revealed diffuse intrahepatic biliary dilation with narrowing at the anastomotic site. Her symptoms abated without intervention.

She recently presented with the same symptoms of increased severity. Physical examination revealed anicteric sclera, and a soft, non-distended abdomen tender to palpation in the right upper quadrant, with no rebound or guarding. Lab studies revealed total serum bilirubin of 1.1 mg/dL, conjugated bilirubin of 0.8 mg/dL, alkaline phosphatase of 901 U/L, serum aspartate aminotransferase of 142 U/L, alanine aminotransferase of 169 U/L, and elevated serum lipase of 192 U/L.

MRCP revealed worsening stenosis at the hepaticojejunostomy site and increasing intrahepatic biliary dilatations and strictures (Figure 1). No hepatic lesions were seen. Push enteroscopy showed no abnormalities. The Kasai reconstruction anatomy was normal, but the adult colonoscope and injected contrast could not reach the suspected stricture in the Roux-en-Y limb. Subsequent percutaneous transhepatic cholangiogram (PTC) showed generalized ductal dilation, innumerable intrahepatic bile duct stones, and multiple filling defects in the most central distended duct communicating with the enteric loop, but no evidence of anastomotic stenosis (Figure 2). A core liver biopsy showed stage 2 moderate fibrosis.

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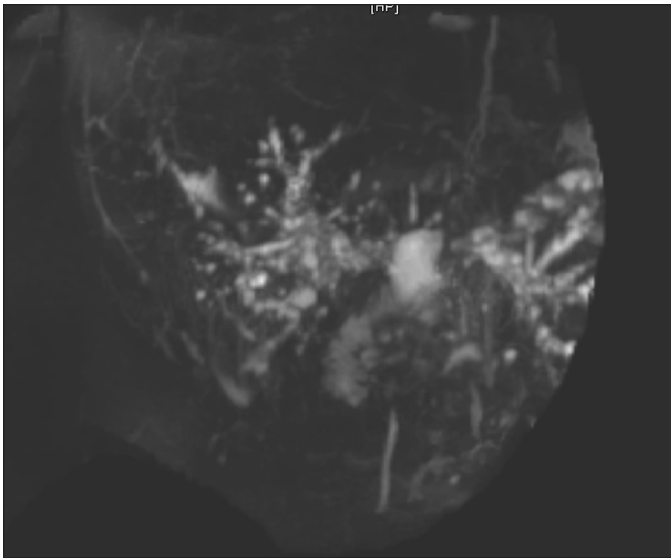


Figure 1. MRCP showing increased intrahepatic biliary dilatations and strictures.

The patient was treated symptomatically with intravenous fluids and pain medications. An external drain was not placed, as PTC revealed preferential flow into the small bowel. The patient improved clinically and was ultimately discharged on ursodiol. She was referred to a transplant center for evaluation for a liver transplant.

Discussion

Congenital biliary atresia is an inflammatory obstruction of extra- and intrahepatic bile ducts.^{1,4} It occurs in 1/8,000 to 1/18,000 live births. Neonates commonly present within the first few months of life with jaundice, acholic stools, and hepatomegaly. Surgical correction can restore proper bile flow with an 80% success rate, but 80% of these patients will ultimately need a liver transplant. Half of these patients receiving the transplant will do so before 2 years of age.¹

The Kasai procedure was introduced in Japan to re-establish bile flow from the liver to the intestine.⁵ Jejunal anastomosis is created with a Roux-en-Y loop to the portahepatis. The procedure postpones liver transplant by at least 10 years. Worldwide, there are less than 200 long-term survivors in the literature, and the 20-year survival rate with the native liver ranges from 21–44%.^{4,6} While the Kasai procedure addresses the extrahepatic duct atresia, intrahepatic lesions worsen over time. Sixty percent of long-term patients eventually develop a severe complication, 100% with cholangitis, 80% with portal hypertension, 45% with gastrointestinal bleeds, and 1.3% with hepatocellular car-

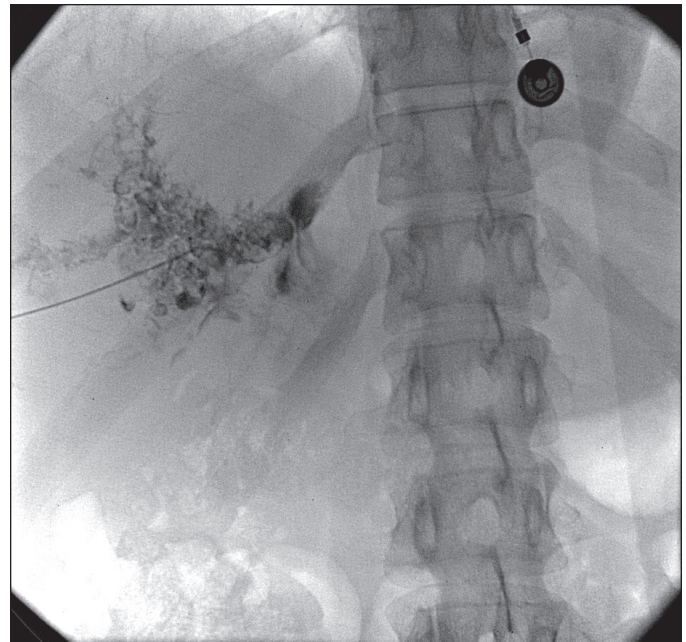


Figure 2. PTC showing ductal dilation, innumerable intrahepatic bile duct stones, and multiple filling defects.

cinoma.^{5,7} Intrahepatic cholelithiasis is seen in one-third of cholangitis cases.⁶

The procedure was not widely accepted in the U.S. until the early 1970s.³ Previously, the neonates were considered inoperable, and the use of artificial duct prosthesis often failed. The other 2 long-term U.S. survivors underwent the procedure in the 1960s, within the first 8 weeks of life, and first reported symptoms before age 20 years. Our patient's status is remarkably good more than 25 years after her initial surgery. The literature estimates about 160 patients alive at >20 years without a liver transplant, the majority of whom in Japan, where the procedure was adopted early.⁵ It is therefore important to report the long-term outcomes of the procedure in a non-Asian population, as underlying genetics and environmental factors can differ regionally.

One patient's Kasai surgery was performed before age 70 days, which likely doubled her chance of survival and lowered her complication rate.⁶ It is important for gastroenterologists to be aware of the long-term complications of Kasai patients, as they often transition to adulthood. Our patient may be able to live with her native liver for a few years longer, but liver transplantation is unfortunately inevitable. The ideal time for liver transplantation is still not known, but should be done earlier in patients with recurrent cholangitis and portal hypertension.

Disclosures

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Informed consent was obtained for this case report.

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