

KASAI PROCEDURE- HEPATOPORTOENTEROSTOMY

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Question

A 1-month-old infant with jaundice and pale stools has been diagnosed with biliary atresia and is scheduled for a portoenterostomy. Which of the following is the **STRONGEST** prognostic indicator for successful portoenterostomy?

- A. Age less than 60 days at the time of surgery
- B. Development of postoperative cholangitis
- C. Elevated gamma-glutamyl transpeptidase (GGT) after surgery
- D. Portal pressure greater than 10 mmHg

Key Points

- Biliary atresia (BA) is a rare but severe liver disease in newborns marked by progressive obstruction of bile ducts preventing bile from exiting the liver into the intestine.
- The Kasai procedure, or hepatoportoenterostomy (HPE), is a surgical intervention aimed at restoring bile flow in infants with BA. The Kasai procedure improves survival in infants with BA, although predicting long-term outcomes remains difficult.¹
- Anesthetic management includes a focused preoperative evaluation specifically focused on comorbidities related to liver dysfunction (e.g., coagulopathy, electrolyte imbalances, cardiac anomalies), laboratory studies (e.g., CBC, coagulation profile, and electrolytes), and intraoperative management involving precise monitoring (e.g., temperature, glucose, and hemodynamics) and preparation for potential blood and fluid resuscitation.

Biliary Atresia

- BA is an obliterative cholangiopathy that can lead to biliary fibrosis, liver inflammation, cirrhosis, and, ultimately, liver failure.
- The earliest sign of BA is jaundice with persistence beyond the first two weeks of life. Patients also experience pale or clay-colored stools due to the absence of bile in the intestines.
- Key prognostic factors include the age at which HPE is performed, the absence of cholangitis, and post-surgical serum bilirubin levels showing early decline with normalization by 12 weeks.²
- Early intervention tends to yield the best results; studies show that infants undergoing HPE before 60 days old have better survival rates with their native liver.³
- Elevated serum bilirubin (>2 mg/dL) and low albumin (<3.5 g/dL) at three months post-surgery were independent predictors of the need for transplantation.⁴

Surgical Procedure

- The Kasai procedure begins with a cholangiogram to identify any bile ducts. Diseased bile ducts and fibrosed tissue are removed at the portal area. This is followed by the creation of a Roux-en-Y anastomosis by which a loop of intestine is connected directly to the liver's hilum (Figure 1).
- The anastomosis allows bile to flow into the small intestine, leading to a reduction in

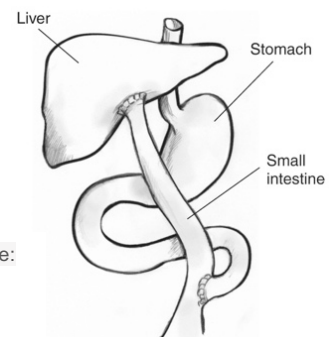


Figure 1: Kasai procedure. Source: National Institute of Diabetes and Digestive and Kidney Diseases, National Institutes of Health.

jaundice over subsequent weeks. However, while this procedure can significantly improve bile drainage, many patients eventually develop progressive liver disease and may eventually require transplantation. Regrettably, the disease persists as chronic cholangiopathy, characterized by persistent ductular reaction and escalating portal fibrosis, eventually leading to cirrhosis and liver failure.¹

Anesthetic Management⁵

Anesthetic Considerations	Management Recommendations
Preoperative Assessment	Detailed history and physical exam, evaluation of comorbidities. CBC, coagulation panel, electrolytes, possibly an echocardiogram to exclude cardiac malformations especially in patients with Biliary Atresia Splenic Malformation.
Setup and Monitoring	General anesthesia with an endotracheal tube. Standard ASA monitors; +/- arterial line. Large bore IV for potential rapid blood loss. Central venous access should be considered for patients with difficult or inadequate IV access, and when prolonged parental nutrition is needed. Temperature and glucose monitoring.
Anesthetic Induction and Maintenance	Ensure adequate oxygenation. Anticipate oxygen desaturation during induction due to limited pulmonary reserve and reduced functional residual capacity. Prepare for full-stomach precautions if abdominal distension is present. Maintenance with TIVA, inhalational, or balanced anesthesia is appropriate, with neuromuscular blockade as needed. Dose adjustment of medications is rarely needed as hepatic function is maintained.
Hemodynamic and Physiologic Goals	Liver retraction may cause inferior vena cava kinking, reducing venous return and causing hypotension. Albumin replacement may be required if significant ascitic fluid is removed. Packed RBCs and FFP should be available.
Postoperative Care	Plan for extubation if intraoperative stability is achieved. ICU monitoring may be necessary. Consider placing an epidural catheter or abdominal plane blocks for postoperative pain management if there is no evidence of coagulopathy.

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Answer: A. The most significant predictor of successful Kasai procedure in biliary atresia is the patient's age, which ideally should be less than 60 days at the time of surgery (A). Early intervention is paramount, as delays significantly elevate the risk of irreversible liver damage and diminish the chances of successful bile drainage. While complications such as postoperative cholangitis (B) are prevalent, they do not determine the success of the procedure. Elevated gamma-glutamyl transpeptidase (C) levels after surgery are associated with poor outcomes and the potential necessity of liver transplantation. Lastly, portal pressure exceeding 10mmHg (D) signifies advanced disease and adversely impacts the prognosis.