Chapter

27 Hepatic Portoenterostomy; Kasai Procedure

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A nine-month-old male recently adopted from abroad with a past medical history significant for biliary atresia, arrives for preoperative evaluation for the Kasai procedure.

He was recently admitted for hematemesis requiring transfusion, followed by esophagogastroduodenoscopy with sclerotherapy and variceal band ligation. His oxygen saturation is 88% on room air. Physical examination is consistent with nail bed clubbing, scleral icterus, and a distended abdomen from ascites. He has scratch marks on his skin, some with scabbing and some appearing infected.

The preoperative clinic asks for guidance in performing a preoperative evaluation.

What Is Biliary Atresia?

Biliary atresia, with an incidence between 1 in 8,000 and 1 in 18,000 live births, is the most common cause of neonatal cholestasis and the most frequent indication for liver transplantation in children. Biliary atresia is a progressive fibro-inflammatory cholangiopathy leading to a complete obliteration of the extra-hepatic bile ducts. There is a slight female predominance with a female to male ratio of 1.7:1.

What Is the Natural History of Biliary Atresia?

Biliary atresia presents as neonatal jaundice. Neonatal jaundice beyond the first two weeks of life should raise suspicion for biliary atresia as morbidity and mortality depend on the timing of surgical intervention. Biliary cirrhosis, portal hypertension, and end-stage liver disease will occur in about 50% of children by two years of age with no surgical intervention. Clinical findings may include a conjugated hyperbilirubinemia, elevated gamma-glutamyl transferase, and elevated serum liver transaminases.

Neonatal jaundice progresses to hepatic fibrosis at four to six weeks of age. From two to six months, chronic hepatic fibrosis/cirrhosis results in the changes noted later in the chapter. If untreated, end-stage liver disease requiring liver transplantation occurs.

Associated congenital anomalies include polysplenia, situs inversus, absent vena cava, malrotation, and cardiac anomalies.

What Are the Associated Pathophysiologic Issues Related to Chronic Biliary Atresia?

- Biliary obstruction leads to hepatic fibrosis and eventually cirrhosis.
- Hepatic failure (usually after a few months of life) results in:
  - Anemia secondary to liver disease
  - Ascites (hypoalbuminemia)
  - Coagulopathy: intrahepatic factor production and inability to absorb vitamin K due to lack of bile salts
  - Hypoxemia: reduced functional residual capacity secondary to large volume ascites
  - Jaundice: acholic stool and dark urine
  - Malnutrition: malabsorption
  - Portal hypertension and esophageal varices
  - Secondary splenomegaly
  - Recurrent cholangitis/sepsis

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What Is a Kasai Procedure (Hepatic Portoenterostomy)?

The Kasai procedure, or a hepatic portoenterostomy, involves:

(1) Mobilization of the liver to identify the porta hepatis
(2) Excision of all extrahepatic fibrous biliary remnants at the point where the portal vein enters the hepatic parenchyma.
(3) Creation of a Roux-en-Y loop of the proximal jejunum.
(4) Anastamosis of the Roux-en-Y loop of the jejunum to the porta hepatis.

The result is a conduit for biliary drainage to exit the liver.

What Is the Timing of and Prognosis for the Kasai Procedure?

The timing of the surgical intervention affects the morbidity and mortality of children with biliary atresia. Early intervention (8–12 weeks) affords the best prognosis.

Approximately one-third of children who have undergone a Kasai procedure will require no further surgical intervention whereas the other two-thirds of children will require a liver transplantation with development of portal hypertension, ascites, malnutrition, poor growth, and recurrent cholangitis.

What Is the Cause of Hematemesis in This Case?

Portal hypertension is an elevated portal venous pressure above 10–20 mmHg causing increased resistance to portal venous inflow. The increased resistance occurs not only because of hepatic fibrosis but also because of nitric oxide deficient vasoconstriction. Esophageal variceal bleeding is the most common presentation of portal hypertension with increasing blood flow from inadequate forward portal venous inflow, causing variceal expansion and rupture.

Why Is the Patient Hypoxemic?

Hypoxemia with impaired respiratory function occurs secondary to mechanical impairment from ascites and hepatosplenomegaly with reduced functional residual capacity. Pulmonary edema and pleural effusions, often reaccumulate despite drainage, due to hypoalbuminemia and reduced oncotic pressure. These changes combine to reduce functional residual capacity and increased work of breathing.

What Is Hepatopulmonary Syndrome (HPS)?

Hepatopulmonary syndrome (HPS) may also cause hypoxemia with right to left shunting secondary to intrapulmonary arteriovenous shunting and intrapulmonary vasodilation. HPS is diagnosed clinically with a PaO_2 <70 mmHg or an alveolar-arterial gradient greater than 20 mmHg on room air.

Technetium-99 radiolabeled microalbumin may also be used to diagnose HPS; normally, 95% of the microalbumin is taken up in the lungs, whereas in HPS secondary to intrapulmonary right to left shunting, the microalbumin is taken up in the systemic capillary beds. Additionally, a bubble study with a transthoracic echocardiogram may also be used to assess right-to-left shunting in the pulmonary vasculature bed with appearance of bubbles in the left atrium. Supplemental oxygen is used as supportive therapy. Hepatopulmonary syndrome is reversible following a liver transplantation.

What Is Hepatorenal Syndrome (HRS)?

Hepatorenal syndrome may be acute (type 1) or chronic (type 2). HRS is characterized by renal insufficiency in the setting of portal hypertension, presumably caused by prerenal azotemia or acute tubular necrosis. Children with metabolic disorders are prone to renal insufficiency and may be on renal replacement therapy preoperatively. Type I is characterized by rapid progression of renal failure with 100% rise in creatinine in 2 weeks, whereas type 2 progresses over several weeks to months. Liver transplantation is the definitive therapy for HRS.

What Other Major Organ Systems Are Affected by End-Stage Liver Disease?

End-stage liver disease is a multisystem disorder.

Circulatory. The cardiovascular system undergoes significant changes with end-stage liver disease. End-stage liver disease is associated with a hyperdynamic circulation with an increased cardiac output

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and systemic vasodilation. The pathophysiology of this condition has been attributed to sympathetic nervous system hyperactivity, inadequate clearance of vasoactive substances, arteriovenous (A-V) shunting, and tissue hypoxemia. The hyperdynamic circulation decreases sensitivity to catecholamines and vasoconstrictors, exaggerating the vasodilatory effects of most anesthetics.

Pulmonary Vasculature. Portopulmonary hypertension (PPHN) is defined as a mean pulmonary artery pressure greater than 25 mmHg with a normal pulmonary capillary wedge pressure in the presence of portal hypertension.

PPHN may present with a new-onset murmur with a loud P2 sound, dyspnea, and syncope. PPHN may be diagnosed by echocardiography or catheterization. Patients with a mean pulmonary artery pressure (mPAP) <35 mmHg do not have increased mortality during liver transplantation whereas those with a mPAP between 35 and 45 mmHg have a mortality of approximately 50%, and those with a mPAP > 50 mmHg have a 100% mortality.

Gastrointestinal. Gastrointestinal complications of portal hypertension include ascites and gastrointestinal bleeding from esophageal and gastric varices.

Coagulation. Hepatic synthetic dysfunction reduces plasma concentrations of albumin, plasma cholinesterases, and coagulation proteins. Hypoalbuminemia reduces serum oncotic pressure predisposing to intravascular hypovolemia, ascites, pulmonary edema, and pleural effusions. Decreased plasma cholinesterase concentrations only mildly prolong neuromuscular blockade after succinylcholine administration since this enzyme is produced in excess even in children with end-stage liver disease.

What Is Hepatic Encephalopathy?

Hepatic encephalopathy is graded by the severity of the altered mental status as defined by West Haven (Table 27.1). The etiology of hepatic encephalopathy is multifactorial, including electrolyte disturbances such as hyponatremia, hypoglycemia, hyperammonia, and coagulopathy. Low blood urea nitrogen with elevated ammonia is present in children with end-stage liver disease. Furthermore, gastrointestinal hemorrhage may produce hyperammonia from bacterial deamination of intestinal blood.

Table 27.1 West Haven criteria for hepatic encephalopathy

Grade	Clinical presentation
0	Minimal: no detectable changes in personality or behavior, no asterixis
1	Lack of awareness, shortened attention span, sleep disturbance, altered mood, and slowing of the ability to perform mental tasks; asterixis may be present
2	Lethargy or apathy, disorientation to time, amnesia of recent events, impaired computations, inappropriate behavior, and slurred speech; asterixis present
3	Somnolence, confusion, disorientation to place, bizarre behavior, clonus, nystagmus, and positive Babinski sign; asterixis absent
4	Coma, lack of verbal, eye, and oral response

Ammonia crosses the blood brain barrier, resulting in cerebral edema. Ammonia levels greater than 200 μg/mL have been associated with cerebral herniation and death. Lactulose is prescribed to produce an osmotic diuresis and acidify the gut lumen to trap ammonia and minimize absorption, yet has not been shown to improve survival.

Children with grade 3–5 hepatic encephalopathy require endotracheal intubation to protect airway reflexes and therapeutic measures to reduce increased intracranial pressure, including elevation of the head of the bed, sedation, acute hyperventilation, osmolar diuretics, and hypertonic saline.

What Are the Anesthetic Implications of the Kasai Procedure?

Preoperative Considerations

Blood products should be prepared prior to surgery (especially in the setting of past transfusion and potential for antibody presence).

Blood should be irradiated as these patients should always be considered for future transplantation potential. Blood prior to transfusion undergoes brief irradiation to reduce donor lymphocytes. The aim is to prevent graft vs. host disease in a newly immune suppressed patient.

The patient has severe itch due to build up of bile salts; secondary infection should be treated.

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Laboratory examinations should include: CBC, coagulation studies, electrolyte, glucose, albumin, blood urea nitrogen (BUN)/Cr, and liver function tests.

A chest X-ray to evaluate for fluid overload and an echocardiogram to evaluate for PPHN are also important preoperatively especially given this patient's late presentation.

Intraoperative Considerations

- In the absence of large ascites, inhalation induction is appropriate.
- Large IV access should be obtained, specifically in the upper extremities.
- Central line (internal jugular vein or subclavian) if inadequate access, pressor support or TPN considered.
- Blood should be in the operating room.
- Arterial access should be obtained for hemodynamic monitoring and frequent laboratory examination.
- Temperature maintenance should be carefully monitored, especially in the setting of coagulopathy.
- Frequent monitoring of electrolytes, glucose, and lactate is needed.
- Transfusion of blood products as required for bleeding (especially in small neonates undergoing procedure).
- Fluid homeostasis is vital with third spacing and evaporative losses during long intra-abdominal procedure.
- Vasopressors should be immediately available.

Postoperative Considerations

Depending on the preoperative severity of illness and the intra-operative course these children may be suitable for extubation at the end of the procedure.

Analgesia is critical with the large intra-abdominal incision. In the absence of coagulopathy, thoracic epidural (direct vs. caudally threaded epidural is appropriate). The epidural may be used to reduce intra- and postoperative opioids and associated complications.

Suggested Reading

Bromley P, Bennett J. Anesthesia for children with liver disease. *Cont Educ Anaesth*. 2014:14(5):207–12.

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