KASAI PROCEDURE

Surgical Specialty:	Pediatric Surgery
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Background:

- General Considerations
  - The Kasai procedure is a palliative surgery performed to restore biliary flow in patients with biliary atresia or severe cholestasis. Biliary atresia is an obliterative cholangiopathy that, without treatment, leads to fibrosis of the biliary tree, cholestasis, and liver failure.[1,2]
  - The Kasai procedure is a portoenterostomy. The procedure always starts with an intraoperative cholangiogram. The absence of extrahepatic bile duct opacification confirms the diagnosis. Then, dissection of the extrahepatic biliary tree to the liver capsule is performed to expose the intrahepatic bile ducts and dissect the porta hepatis. A jejunal Roux-en-Y is created and anastomosed to the exposed porta hepatis to allow free-flowing bile drainage from the liver.[1,2] The gallbladder (if present) is removed.
  - Kasai procedures are commonly performed as an open procedure but can be done laparoscopically.
  - In patients without atresia involving the gallbladder, cystic duct, and common bile duct, an alternative surgery, hepatic portocholecystostomy, can be performed. This procedure preserves the gallbladder, sphincter of Oddi, and ampulla of Vater, which is postulated to decrease the risk of ascending cholangitis postoperatively.[3]
  - The Kasai procedure is best performed within the first 60 days of life due to the rapid progression of the obliterative cholangiopathy, leading to intrahepatic biliary fibrosis and cirrhosis of the liver.[1,2] The earlier the Kasai procedure is performed, the greater its likelihood of success.
  - However, many children who undergo a Kasai procedure eventually need orthotopic liver transplantation, even if drainage is successful due to ongoing and progressive portal hypertension or cirrhosis.[1,2]
- Patient Considerations
  - Patients are at risk of malabsorption of fat-soluble vitamins, and therefore, the continuation of vitamin ADEK supplementation is essential to prevent osteomalacia and coagulopathy.[1,2]
  - Biliary atresia can be associated with other congenital abnormalities, most commonly splenic anomalies.[1,2] Biliary Atresia Splenic Malformation (BASM) syndrome can

consist of splenic abnormalities (asplenia, polysplenia) with other features such as a preduodenal hepatic vein, absent inferior vena cava, malrotation, cardiac anomalies, and abdominal situs inversus.[1,2]
    - It is rare for patients to have portal hypertension at the time of the Kasai procedure. In patients with advanced disease or rapid disease progression, cirrhosis may develop, resulting in portal hypertension and ascites.

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - Labs should include the following:
        - Coagulation panel
            - Standard coagulation tests (PT, PTT, fibrinogen)
            - Consider Thromboelastography (TEG) or Rotational Thromboelastometry (ROTEM)
        - Complete blood count
        - Albumin level
        - Electrolytes
        - Consider an echo to exclude cardiac malformations in cases with malformative or syndromic associations (e.g., polysplenia/ asplenia, BASM)
    - Physical Exam
        - A focused physical exam should be completed. Specifically, these infants generally present with jaundice, acholic stools, manifestations of failure to thrive, and nutritional deficiencies.
        - Ascites and abdominal distention may result in shallow respirations and tachypnea.
- Specific or Unique Room Set-up Requirements
    - The airway should be secured with an endotracheal tube (ETT)
        - The patient may have limited pulmonary reserve due to ascites causing interference with diaphragmatic excursions and basilar atelectasis. This pathophysiology can result in limited oxygen reserve during induction and intubation.
    - Placement of standard American Society of Anesthesiologists monitors, including accurate temperature monitoring.
        - If the patient has sepsis secondary to cholangitis or sequala of advanced hepatic disease, there is a low threshold to place an intra-arterial catheter.
    - Although relatively uncommon, there is a potential for sudden and rapid blood loss, so a large bore or second intravenous (IV) line should be placed for rapid resuscitation.
    - A dextrose-containing maintenance fluid should be run throughout the case. Although isotonic intravenous solutions without lactate (such as Plasmalyte or Normosol) are usually favored in patients with poor hepatic reserve, most infants undergoing the Kasai procedure will still have adequate hepatic synthetic function to tolerate Lactated Ringer's.
    - If significant ascites fluid is removed, albumin should be given to replace losses. In patients with impaired hepatic synthetic impairment and hypoalbuminemia, albumin is often also used in the maintenance and loss replacement fluid regimen.[1,2]
    - Foley catheter placement depends on surgeon preference and the anticipated case duration.
    - Blood Availability

        - Blood loss is usually minimal but can be sudden and rapid in some situations.
        - Red blood cells and fresh frozen plasma should be crossmatched and available.
    - Antibiotics should be given prior to incision. Most commonly, these include cefoxitin or gentamycin[3].
- General
    - An upper abdominal procedure may result in postoperative diaphragmatic insufficiency.
    - The placement of retractors can interfere with ventilation, and pressure control ventilation may result in inconsistent ventilation. A volume-guaranteed ventilation mode with positive-end expiratory pressure (PEEP), preferably with a decelerating flow pattern (such as PRVC), is favored.
- Induction
    - An IV induction or mask induction is appropriate.
- Positioning

  Surgery is performed supine. Careful management of ETT to avoid kinking and line management should be performed before draping.
- Maintenance
    - Total intravenous anesthesia (TIVA), inhalational anesthesia, or a balanced technique are appropriate.
    - Neuromuscular blockade can be maintained throughout the case utilizing neuromuscular blockade monitoring.
    - Avoid nitrous oxide due to distention of the bowel.[1,2]
    - If no coagulopathy is present, some centers have advocated for neuraxial anesthesia to improve postoperative pain control.[2,4] The epidural can also be used intraoperatively to augment general anesthesia.
    - The incision is typically subcostal. Therefore, alternative regional anesthesia techniques could include abdominal plane blocks[5], erector spinae plane blocks, or paravertebral blocks.
    - If no regional block is placed, multi-modal analgesia with acetaminophen and opioids is appropriate.
    - Hepatic function is usually preserved, therefore dose adjustment of medications is unnecessary[1]. If the patient is premature, changes in metabolism and clearance should be considered when determining an appropriate dose.
- Hemodynamic/Physiologic goals
    - Although hemodynamic stability is typical in these cases, more severe cirrhosis, elevated portal pressure, cholangitis, or sepsis can result in hypotension and instability in some patients.[1,2]
- Surgical Considerations
    - Retractors and abdominal packing can elevate the diaphragm and cause the ETT to migrate endobronchially. It can also lead to increased airway pressures during the case due to increased impedance to diaphragmatic descent.
    - The surgeon may need to reflect the liver during the open procedure to improve exposure. This can lead to kinking of the inferior vena cava, decreased venous return, and hypotension.[1,2]
- Emergence/Disposition
    - Plan for extubation at the end of the case, assuming minimal blood and crystalloid transfusion during the case.
    - ICU is used for postoperative monitoring, depending on the monitoring capabilities on the floor at one's institution.

- Post-op Care
    - The surgery is successful if jaundice improves and biliary flow is restored.
    - Antibiotics are given for approximately five days post-operatively to prevent ascending cholangitis.[2]

Case-Specific Complications/Pitfalls

- Immediate surgical complications such as a biliary leak, ascending cholangitis, and ascites can occur.[1,2]
- Long-term complications include ongoing hepatic fibrosis and portal hypertension.[1]

References

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