

## Anaesthesia recommendations for patients suffering from

### **Biliary atresia**

**Disease name:** Biliary atresia

**ICD 10:** Q44.2

**Synonyms:** Extrahepatic biliary atresia, familial extrahepatic biliary atresia, idiopathic extrahepatic biliary atresia

**Disease summary:**

Biliary atresia (BA) is a rare and fatal progressive inflammatory disease of infancy affecting the intra and extra hepatic bile ducts leading to cholestasis, fibrosis and cirrhosis. It has a varying incidence ranging from 1:10,000 to 1:20,000 live births. Persistent jaundice for more than 2 weeks in a term infant mandates evaluation for BA. Without medical intervention, BA leads to liver failure and ultimately death within the first two years of life. Kasai's portenterostomy (KPE) is the first line of treatment which aims at restoring the forward flow of bile from the liver into the intestines using a jejunal Roux-en-Y limb, which is anastomosed to the porta hepatis after resection of biliary remnants. Orthotopic Liver transplantation (OLT) is reserved for children with a failed Kasai's procedure. Kasai's portoenterostomy procedure and liver transplantation along with adjuvant medical therapy and nutritional support have improved the prognosis of infants with BA and survival up to adulthood has been documented. The main anaesthetic concerns during portoenterostomy are conduct of safe anaesthesia in an infant with mild to moderate derangement of liver function scheduled for a lengthy upper abdominal wall laparotomy. This necessitates meticulous attention to patient's intraoperative fluid status, temperature regulation, glucose metabolism and provision of adequate perioperative analgesia in the presence of a compromised and dysfunctional liver. Surgical maneuvers causing transient obstruction to inferior vena caval blood flow and hypotension need to be anticipated and managed appropriately.

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Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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Find more information on the disease, its centres of reference and patient organisations on Orphanet: [www.orpha.net](http://www.orpha.net)

## **Typical surgery**

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Liver biopsy and Intraoperative Cholangiogram, Kasai's Portoenterostomy, upper gastrointestinal endoscopy, orthotopic liver transplantation.

## **Type of anaesthesia**

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There are no current existing guidelines regarding administration of general and or regional anaesthesia in children with BA.

Anaesthetic agents with minimal hepatotoxic effects need to be administered. Changes in the body fluid compartment, low serum albumin levels can affect the drug distribution and available free fraction of drugs. Hepatic artery blood flow, presence of portosystemic shunts and decreased hepatic enzyme activity can influence the pharmacokinetics of anesthetic drugs.

Central neuraxial blocks are safe provided there is no coagulaopathy, thrombocytopenia and portal hypertension induced epidural varices.

## **Necessary additional diagnostic procedures (preoperative)**

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A baseline liver function test including serum albumin, glucose, prothrombin time and its international normalized ratio are needed to know the severity of underlying liver dysfunction.

Infants with BA associated with BASM (Biliary Atresia Splenic Malformation) can have associated anomalies of the spleen, situs inversus, absent vena cava and other cardiac anomalies. Pulmonic stenosis, Tetralogy of Fallot are also associated cardiac anomalies which need to be screened by echocardiography.

## **Particular preparation for airway management**

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Precautions in anticipation of potentially full stomach need to be taken if there is abdominal distension due to ascites or hepatosplenomegaly. Diaphragmatic splinting can decrease the functional residual capacity and cause early small airway closure leading to rapid oxygen desaturation thus the need for adequate oxygenation during induction and emergence from anaesthesia.

## **Particular preparation for transfusion or administration of blood products**

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Vitamin K administration is mandatory in infants with cholestasis. Vitamin K, a fat soluble vitamin necessary for hepatic synthesis of coagulation factors II, VII, X and IX is not absorbed in this set of patients due to impaired secretion of bile salts in the intestine. Vitamin K administration promptly corrects the coagulopathy, however Vitamin K unresponsive coagulopathy indicates severe hepatocellular failure affecting the synthetic function of liver or could indicate underlying sepsis and undernutrition. In case of documented coagulopathy, appropriate blood products need to be administered.

Although blood loss during surgery is minimal, provision of adequate units of packed red blood cells and blood components to correct coagulopathy has to be ensured before surgery.

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### **Particular preparation for anticoagulation**

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No particular recommendation for anticoagulation in patients with BA.

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### **Particular precautions for positioning, transport or mobilisation**

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No recommendations at present.

Slight head up position can be employed if there is abdominal distension to minimize respiratory compromise and avoid aspiration.

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### **Probable interaction between anaesthetic agents and patient's long term medication**

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No recommendations present.

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### **Anaesthesiologic procedure**

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Anaesthesia has to be tailored based on the patient's age, underlying liver dysfunction, cholestasis and surgical severity. Isoflurane with its ability to increase the arterial hepatic blood flow with minimal decrease in total hepatic blood flow is considered to be safe. Sevoflurane and Desflurane can also be safely used as they are devoid of any hepatotoxic effects with minimal metabolism. Atracurium and Cis atracurium have definite advantages over other neuromuscular blockers because of their unique metabolism and thus are favoured.

The hepatic synthetic and metabolic functions are relatively unimpaired till late stages of BA and thus doses of opioid analgesics remain unchanged. The hepatic artery buffer system probably responds to a decrease in portal venous blood flow by a compensatory increase in hepatic arterial blood flow in early stages of BA. Morphine in the dose of 10-40 ug/kg/hr has been used safely for infants with BA for post-operative analgesia. In children with mild to moderate Child Pugh scoring tramadol can be used but the dose needs to be halved and the dosing interval increased from 6 hours to 12 hours. Similar dose reductions are recommended for acetoaminophen if used as an adjuvant analgesic.

Metabolism of propofol is also not significantly affected in infants with BA with extrahepatic metabolism playing a more significant role in the overall elimination of propofol in this subset of population.

Metabolism of bupivacaine could be affected due to decreased clearance along with low serum alpha1 acid glycoprotein causing an increase in the unbound fraction of bupivacaine

leading to cardiovascular toxicity. A maximum dose of 0.25mg/kg/hr in infants younger than 4 months and a maximum of 0.3mg/kg/hr in infants older than 4 months has been recommended for postoperative continuous epidural infusion of bupivacaine in infants with BA.

Nitrous oxide is avoided to prevent gut distension hampering abdominal wall closure. Controlled ventilation avoiding high airway pressures with maintenance of normocarbica is essential to avoid changes in hepatic and portal blood flow.

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### **Particular or additional monitoring**

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Internal jugular venous access and or wide bore peripheral lines in the upper limb is essential for monitoring central venous pressure as well as managing major fluid shifts caused during KPE. The critical surgical event during KPE is exteriorization of liver from abdominal cavity to explore the porta hepatis which results in sudden drop in blood pressure due to kinking of inferior vena cava and subsequent obstruction of venous return. Change in the ECG amplitude (Brody's effect) or a reduction in the blood pressure/central venous pressure can be used to recognize this event. Intra-arterial catheter is recommended for continuous monitoring of blood pressure and blood gas sampling in selected cases. Hourly urine output monitoring can also help in fluid management in long cases.

Intraoperative blood sugar monitoring is mandatory. Dextrose containing fluids need to be judiciously used if hypoglycemia is documented or if the synthetic function of liver is suspected to be hampered. Core temperature needs to be monitored and measures to prevent hypothermia have to be instituted.

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### **Possible complications**

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Extensive fluid loss and surgical maneuvers leading to inferior vena caval kinking can lead to transient but significant fall in blood pressure which can be easily managed by a fluid boluses and added vasopressor therapy.

The large surface to volume ratio of infants, lack of adequate subcutaneous adipose tissue, exposure of body cavities to low environmental temperatures, infusion of cold fluids and ventilation with dry gases can increase the risk of perioperative hypothermia.

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### **Postoperative care**

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Post operatively infants with BA need High Dependency Unit care. Cardiovascular toxicity arising from continuous infusions of local anesthetic needs to be strongly suspected in infants with an epidural catheter in situ for pain management. Continuous postoperative intravenous opioid infusion for pain management necessitates monitoring of sedation score and respiratory rate to detect respiratory depression.

Episodes of ascending cholangitis can complicate the post-operative course.

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## Information about emergency-like situations / Differential diagnostics

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*caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the diseases, e.g.:*

Infants with BA who seek medical attention later in infancy can present in acute sepsis with coagulopathy which needs prompt emergent management.

BA needs to be differentiated from other infectious, congenital, metabolic and genetic causes of neonatal cholestasis causing pathological persistent conjugated hyperbiliruninemia. Timely recognition and surgical intervention is crucial event determining long term survival of infants with BA.

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## Ambulatory anaesthesia

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No current recommendations exist.

Children with BA can present for upper gastrointestinal variceal treatment in the form of sclerotherapy or variceal ligation. General anesthesia and securing the airway with tracheal intubation is recommended for all procedures except the very shortest duration surgical procedures. Gut distension during endoscopy can lead to catastrophic respiratory embarrassment and needs to be avoided.

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## Obstetrical anaesthesia

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With successful KPE and OLT, survival up to child bearing age has been documented. Pregnancy is considered high risk and although not contraindicated, several groups recommend managing pregnancy after risk assessment for hepatic failure in women with BA post KPE or OLT. Complications of portal hypertension, hypersplenism, gastrointestinal variceal bleeding, episodes of cholangitis, added cholestatic insult and extensive varices on the anterior abdominal wall can influence the course of pregnancy and attendant medical treatment. A multidisciplinary team work to manage parturient women with BA is recommended.

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**Peer revision 1**

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**Peer revision 2**

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