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Behcet's disease

Biliary atresia

orphanaesthesia

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

SUPPLEMENT NR. 10 | 2019

OrphanAnesthesia –
ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patienten mit seltenen Erkrankungen. Damit will Orphan Anesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesisten damit keine Erfahrungen gesammelt haben, so dass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem ein Anästhesist sowie ein weiterer Krankheitsexperte (z.B. Pädiater oder Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, so dass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

OrphanAnesthesia –
a common project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommenations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

A survey of until now in A&I published guidelines can be found on:

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Anaesthesia recommendations for

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 extrahepatic 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 portoenterostomy (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 manoeuvres causing transient obstruction to inferior vena caval blood flow and hypotension need to be anticipated and managed appropriately.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

Find more information on the disease, its centres of reference and patient organisations on Orpha.net: www.orpha.net

▶ **Citation:** Ganigara A: Biliary atresia. Anästh Intensivmed 2019;60:S440–S447.
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Typical surgery

Liver biopsy and intraoperative cholangiogram, Kasai's portoenterostomy, upper gastrointestinal endoscopy, orthotopic liver transplantation.

Type of anaesthesia

There are no current guidelines regarding the 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 or low serum albumin levels can affect the drug distribution and the available free fraction of drugs. Hepatic artery blood flow, presence of portosystemic shunts and decreased hepatic enzyme activity can influence the pharmacokinetics of anaesthetic drugs.

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

Necessary additional pre-operative testing (beside standard care)

A baseline liver function test including serum albumin, glucose, prothrombin time and its international normalised 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 and Tetralogy of Fallot are also associated cardiac anomalies which need to be screened by echocardiography.

Particular preparation for airway management

Precautions in anticipation of a 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

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 the liver or could indicate underlying sepsis and undernutrition. In case of documented coagulopathy, appropriate blood products need to be administered.

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

Particular preparation for anticoagulation

No particular recommendation for anticoagulation in patients with BA.

Particular precautions for positioning, transportation and mobilisation

No recommendations at present.

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

Interactions of chronic disease and anaesthesia medications

No recommendations present.

Anaesthetic procedure

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 cisatracurium 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 µg/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 acetaminophen if used as an adjuvant analgesic.

The 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.

The metabolism of bupivacaine could be affected due to decreased clearance along with a low level of 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 post-operative continuous epidural infusion of bupivacaine in infants with BA.

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

Particular or additional monitoring

Internal jugular venous access and/or wide bore peripheral lines in the upper limb is essential for monitoring the central venous pressure as well as managing major fluid shifts caused during KPE. The critical surgical event during KPE is exteriorization of the liver from the abdominal cavity to explore the porta hepatis, which results in a 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. An 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 hypoglycaemia is documented or if the synthetic function of liver is suspected to be hampered. The core temperature needs to be monitored and measures to prevent hypothermia have to be instituted.

Possible complications

Extensive fluid loss and surgical manoeuvres leading to inferior vena caval kinking can lead to a 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.

Post-operative care

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

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

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Disease-related acute problems and effect on anaesthesia and recovery

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 hyperbilirubinaemia. Timely recognition and surgical intervention is crucial for determining long-term survival of infants with BA.

Ambulatory anaesthesia

No current recommendations exist.

Children with BA can present for upper gastrointestinal variceal treatment in the form of sclerotherapy or variceal ligation. General anaesthesia 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.

Obstetrical anaesthesia

With successful KPE and OLT, survival up to child-bearing age has been documented. Pregnancy is considered a 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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