Anaesthetic management of a child with Caroli's disease presenting for incidental surgery: A case report

Sir,

Caroli's disease (CD) is a rare disease involving mainly the liver. It affects 1 in 100,000 live births, with higher frequency rate in females than males.[1,2] We are reporting a male patient with CD who was posted for closed reduction of fracture femur followed by hip spica application.

A 3-year-old male presented with pain and swelling of the left thigh following a fall. He was posted for closed manipulation followed by hip spica under general anaesthesia. He was a known case of CD with enlargement of liver and kidney. He was on treatment with atenolol, amlodipine and ursodeoxycholic acid. He had history of uneventful adenotonsillectomy at the age of two.

On examination, the child was active but uncooperative. He had a distended abdomen and was in mild respiratory distress. On systemic examination, the chest was clear, heart sounds were normal and no murmurs. The abdomen was moderately enlarged probably due to enlarged kidneys. Laboratory investigations were within normal limits.

In the operation theatre, his heart rate was 140/min. Non-invasive blood pressure was found to be high, 166/90 mmHg. An intravenous cannula number 22 was introduced after inhalational induction with sevoflurane; intravenous fentanyl 2 mcg/kg was given. After deepening the anaesthesia with sevoflurane, size 2 ProSeal Laryngeal Mask Airway (PLMA) was introduced. Anaesthesia was maintained with infusion of remifentanil 100 μg/hour and sevoflurane in oxygen - nitrous oxide mixture. Airway pressures were slightly elevated at 15 mmHg. Systolic blood pressure was maintained between 90 and 110 mmHg throughout the surgery. At the end of the surgery, sevoflurane and remifentanil were discontinued and PLMA was removed. Post-operative pain was controlled with rectal paracetamol 500 mg.

CD is a rare inherited disorder. The main features are multifocal, segmental enlargement of large intrahepatic bile ducts.[3,4] This can be associated with varying severity of the renal cystic disease. Two variants are described. They are CD and Caroli's syndrome. The less common CD is characterised by ectasia of bile ducts without other abnormalities of the liver.

In Caroli's syndrome, dilatation of the bile duct is associated with hepatic fibrosis.[4] This disorder can occur as an isolated finding, or it can be associated with congenital hepatic fibrosis.[1] Most cases are transmitted as autosomal recessive disorders. Prognosis is unpredictable and depends on associated complications and the rapidity of the progress of the disease. Long-term survival is difficult unless the child undergoes corrective surgeries. The pathogenesis is incompletely understood. Once the diagnosis is established, treatment is largely supportive.[5]

Children may be posted for investigations such as magnetic resonance imaging, endoscopic retrograde cholangiopancreatography or liver biopsy. As age advances, they may require a biliary drain placement, nephrectomy or liver transplant.[6] They may also need a central line for long-term intravenous access or dialysis.

Preoperative investigations to exclude the presence of an altered electrolyte and acid–base imbalance should be performed. Anaesthetic considerations include the presence of systemic hypertension as well as the severity of renal and hepatic involvement. Patients with poor hepatic function may present with coagulopathy, low serum albumin, impaired metabolism of drugs, hypoglycaemia, ascites, etc. The renal dysfunction can lead to volume overload, decreased excretion of certain drugs and changes in total body water. Splenomegaly, secondary to portal hypertension can cause pancytopenia and thrombocytopenia. Distended abdomen can lead to a reduction in functional residual capacity and impaired respiratory function.

During induction of anaesthesia, maintaining 15°–20° head-up position helps to minimise the risk of diaphragmatic splinting and aspiration. Fluctuations in blood pressure should be avoided. It is prudent to avoid drugs that alter the potassium levels like suxamethonium. During maintenance, a narcotic like remifentanil will help to control the fluctuation in blood pressure. Since it is metabolised by tissue esterases, it is considered to be safe. We did not use muscle relaxants since it was a relatively short procedure, although cisatracurium and atracurium are considered safe to use. Paracetamol can be used in small doses.

In conclusion, CD is a rare genetic disorder. Anaesthetic considerations are related to compromised hepatic and renal function. Hypertensive patients on multiple drugs may pose problems perioperatively. Careful titration of drugs should be done in view of the altered drug metabolism. Meticulous planning and execution of perioperative care will result in safe outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

Murugesh Sukumar, Rajagopal R Nair, Nikhil Kumar Gosalia

Department of Anaesthesia and ICU, Khoula Hospital, Ministry of Health, Muscat, Oman

Address for correspondence:
Dr. Murugesh Sukumar,
Department of Anaesthesia and ICU, Khoula Hospital,
Ministry of Health, Mina Al Fahal, Muscat, Oman.
E-mail: msukumar66@gmail.com

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