

# **Title: Anesthetic Management of Kasai Hepatopuertoenterostomy for Extrahepatic Biliary Atresia in a Neonate with Unpalliated Complex Congenital Heart Disease**

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## **ABSTRACT BODY:**

**Introduction:** Extrahepatic biliary atresia (EBA) (incidence 1/ 15,000) has a 10-25% association with heterotaxy syndrome (HS). Predominant malformations occur in cardiovascular, respiratory, gastrointestinal systems. [1, 2] Early surgical correction before 8-weeks-old is crucial to successful restoration of biliary drainage. [3] We describe the anesthetic management of a 29-day-old with EBA and unpalliated complex congenital heart disease (CHD) with HS (dextrocardia, complete AV canal, polysplenia, malrotation) requiring Kasai hepatopuertoenterostomy (KHPE), resection choledochal cyst, and modified Ladd's.

**Case report:** The 3660 gm, full-term, 29-day-old female patient was not in respiratory distress after birth. Her original procedure two weeks previously included an exploratory laparotomy/intraoperative cholangiogram/broviac/liver biopsy. Combined with the subsequent procedure, this constituted a staged KHPE for biliary atresia. In the intervening time, the patient underwent liver/spleen scan, and MRI/MRA Brain/Abdomen for which she received midazolam 0.5 mg/kg, for better definition anatomy. Manifestations of HS in this patient: preduodenal portal vein, situs ambiguous/intestinal malrotation, polysplenia, multiple liver lobes, small pancreas, absent intrahepatic inferior vena cava (IVC) with azygous continuation IVC. Echocardiogram: single atrium, single atrioventricular valve, large right ventricle, small left ventricle, double outlet right ventricle, transposition great arteries; subpulmonic stenosis with pressure gradient 40 mm Hg, dextrocardia with no cyanotic episodes with crying or activity or problems feeding. On physical exam, BP 83/59 mm Hg, HR 161 bpm, RR 40, no facial dysmorphic features, grade III/VI SEM LSB, palpable femoral pulses. CXR: full endocardial cushion defect. EKG: sinus tachycardia 140 bpm. Laboratory findings: normal electrolytes, hemoglobin 13.5 gm/dl, hematocrit 38.7%, platelets 314,000. Room air oxygen saturation: 82-93 percent.

We minimized alterations in pulmonary blood flow, decreases in afterload and intracardiac shunting by (1) inducing with atropine 0.1 mg, fentanyl 25 mcg/kg, pancuronium 0.1 mg/kg, (2) maintaining with minimal inspired oxygen concentration of 50% O<sub>2</sub>/air, 0.2-0.4% sevoflurane after uneventful tracheal intubation, and titrating additional fentanyl, pancuronium, (3) achieving normocarbida and utilizing pressure-controlled mechanical ventilation (Ohmeda) I/E 1:2, PP 16, PEEP 4, RR 35, FIO<sub>2</sub> 50%, and (4) maintaining normovolemia with fluid replacement D10LR and PRBC for ongoing blood loss. Anesthesia time 6 hours 5 minutes. Neuromuscular blockade was not antagonized, patient transported intubated on Ambu 100% O<sub>2</sub> to NICU. Postoperative course was uneventful.

**Discussion:** Despite a 47% incidence of adverse events during non-cardiac surgery in patients with CHD [5], we safely managed a neonate with unpalliated complex CHD/HS for KHPE. The lower pressure gradient mitigated the risk of uncompensated CHF and inotropic support was not required. We achieved circulatory stability with no dysrhythmias whereby atropine and pancuronium countered cardiodepressant effects of opioid-induced bradycardia. Potential hepatic dysfunction required attention to drug metabolism.

## **References:**

1. J Pediatr Surg 1991;26(5):524-527.
2. J Pediatr Surg 1999;34(11):1687-90.
3. Ped Anesth 2000;10:581-89.
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5. Anesthesiology 1991;75:A1056.