

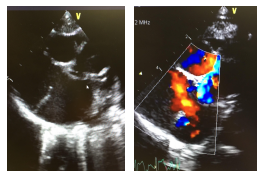
Liver Transplantation in an Infant with a Large Secundum ASD

Yehuda Salamon MD, Dritan Prifti MD, Surendrasingh Chhabada MD, John Seif MD

Department of Pediatric Anesthesiology, Cleveland Clinic Foundation, Cleveland, Ohio

Introduction and Significance

Our patient is an 8 month old 4.3 kg female with biliary atresia status post Kasai at three months of age, and a large secundum atrial septal defect (ASD) presenting for liver transplant. Prior studies discussing pediatric liver transplants in patients with congenital heart disease recommend cardiac surgery before liver transplant, but this patient presented for urgent transplantation due to her liver failure and so she could not tolerate open heart surgery with cardiac bypass. We describe the preoperative evaluation and perioperative monitoring and anesthetic management of this complex patient with congenital heart disease and liver failure.



Case

Past Medical History

- Biliary atresia s/p Kasai at 13 weeks of age, s/p failure of her Kasai
- Fat soluble vitamin deficiency
- Severe malnutrition requiring a nasogastric tube, Situs ambiguus
- Large ASD with right ventricular dilatation
- VSD with spontaneous closure
- Interrupted inferior vena cava (IVC) with azygous continuation

Case (continued)

Presentation

She presented with jaundice, bilateral scleral icterus, a distended but nontender abdomen with abdominal ascites and hepatosplenomegaly.

- Because of her interrupted IVC and the size of the ASD, she was not a candidate for percutaneous ASD closure.
- In a multidisciplinary meeting, it was decided that the patient should have liver transplantation before undergoing ASD closure.
- Because of possible esophageal varices due to portal hypertension, TEE could not be used during the surgery. Instead, precordial doppler was used to monitor for the presence of any air in the heart.

Intraoperative Course

To maintain PVR and SVR:

- FiO₂ remained slightly higher than room air
- Ventilatory parameters were vigilantly adjusted
- Fluids were administered to prevent any hypotension (decreased SVR) or hypoperfusion
- Started on a norepinephrine drip at 0.03 mcg/kg/hr in the pre-anhepatic phase, and over the course of the case received 50 ml/kg of albumin 5%
- Frequent ABGs were obtained, and TEG was performed for close monitoring and to make adjustments in the anesthetic management.

Postoperative Course

The transplant was successful and liver function normalized within a few days. The patient failed extubation two times in the postoperative period, which was found by otolaryngology to be due to *the abutment of a prominent pulmonary artery on the left main bronchus*. Multidisciplinary discussion in the postoperative period recommended closing the ASD, as the enlargement may have been due to pulmonary overcirculation, and would be the only way to facilitate the removal of the tracheostomy. ASD repair was performed without significant improvement in respiratory function.

Conclusion

Although liver transplants have been performed successfully with limited morbidity in those with an ASD, the safe anesthetic that was utilized has not been described. Close attention was paid to the central venous pressure, as well as electrolytes and acid-base physiology to minimize any changes in SVR and PVR.

References

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