Persistent pulmonary hypertension in a neonate with transposition of the great arteries: Preoperative ECMO, Timing of Surgical Repair, and Anesthetic Management

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INTRODUCTION

Persistent pulmonary hypertension (PPHN) occurs in approximately 1% to 3% of neonates with transposition of the great arteries with intact ventricular septum (TGA/IVS). PPHN in this patient population causes severe right to left shunting at the ductus arteriosus and poor interatrial mixing leading to hypoxemia and ventricular dysfunction. In fact, PPHN with TGA/IVS often necessitates the use of extracorporeal membrane oxygenation (ECMO) when other supportive measures have failed.

In cases that result in ECMO cannulation, the appropriate timing of separation from ECMO and surgical correction is controversial. The following describes the anesthetic and surgical considerations involved in a case of a neonate born with TGA/IVS who developed severe PPHN, needing extracorporeal support, and the timing of her surgical repair.

CASE PRESENTATION

A 39 week gestational age female born with TGA/IVS developed PPHN secondary to meconium aspiration syndrome. On DOL 2, despite a balloon atrial septostomy and maximal medical therapy including inhaled nitric oxide (NO), the patient was hypotensive, acidicotic, and had severe reversed differential desaturation (preaductal SaO₂ of 33% and postduductal oxygen SaO₂ of 74%). The patient was placed on venaoceral ECMO support.

During a temporary period of decreased flow on ECMO, an ECHO obtained on DOL 6 demonstrated hyperdynamic LV function, normal RV function, and left to right shunting across both the ductus arteriosus and the atrial septal defect on no inotropic support.

On DOL 7, the infant underwent an arterial switch operation (ASO), repair of the atrial septal defect, and decannulation from ECMO. Immediately following the ASO, the neonate was successfully weaned from cardiopulmonary bypass on NO 20 ppm, epinephrine 0.08 mcg/kg/min, milrinone 0.5 mcg/kg/min, and calcium chloride 10 mg/kg/hr.

The infant was weaned from NO on POD 3, extubated on POD 4, and discharged home on POD 13. At 4 months of age, she continues to be growing and developing well.

DISCUSSION

There are few case reports describing the combination of TGA/IVS and PPHN. The primary anesthetic and surgical challenges included:

1) Timing of repair

The literature indicates that perioperative NO and/or ECMO support is often required in this patient population. It has been speculated that preoperative ECMO causes LV “deconditioning”. Based on this, some studies recommend ECMO be limited to 3 days and an attempt to wean from ECMO be made to allow “reconditioning” of the myocardium prior to surgical repair. However, in our case, preoperative ECHO demonstrated hyperdynamic LV function and L to R shunting at the ductus arteriosus suggesting that the patient would successfully wean from cardiopulmonary support following her ASO.

2) Intraoperative evaluation of LV function

Transesophageal ECHO allowed dynamic assessment of ventricular function intraoperatively which aided in decision making regarding safe transition from ECMO to CPB as well as separation from CPB.

3) Teamwork

All care team members met at the patient’s bedside preoperatively to discuss key portions of the case including roles during transport, equipment positioning in the OR and other surgical/anesthetic concerns. Clear communication was of great importance to ensure safe and successful care of our patient.

CONCLUSION

Intraoperative weaning from ECMO is possible at the time of ASO in this patient population provided early institution of ECMO, good preoperative biventricular function, and empiric administration of NO post-separation from cardiopulmonary bypass.

REFERENCES