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Case Series

Balancing Parallel Circulations: Anaesthetic Management of Non-Cardiac Surgery in Neonates with Unpalliated Hypoplastic Left Heart Syndrome—A Case series.

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ABSTRACT

Hypoplastic left heart syndrome (HLHS) is a complex congenital heart disease characterized by marked hypoplasia of left ventricle and ascending aorta leading to inadequate systemic perfusion following post-natal closure of patent ductus arteriosus. We describe the perioperative management of two neonates, each weighing 2.2 kg, one underwent thoracotomy for tracheoesophageal fistula repair, while the other underwent laparotomy for duodenal atresia. Both neonates received prostaglandin E1 to maintain ductal patency and Dobutamine for myocardial support. The primary goal in the management of patient with HLHS, is balancing the pulmonary and systemic circulation with maximal systemic oxygen delievery at Qp/Qs=1:1. Hypercarbia in combination with lowest possible FiO2 is used to maintain pulmonary vascular resistance and Inotropic support is also required to maintain adequate cardiac output. Both procedures were completed with stable intraoperative haemodynamics, and the neonates were transferred to intensive care with ongoing prostaglandin E1 infusion and inotropic support. The anaesthetic management of neonates with HLHS posted for non-cardiac surgery is challenging and include maintenance of ductal patency ,balance between systemic and pulmonary vascular resistance to optimize systemic oxygen delievery and perfusion pressure.

Keywords*: Hypoplastic left heart syndrome; neonate; non-cardiac surgery; prostaglandin E1; single-ventricle physiology; anaesthetic management.*

INTRODUCTION

Hypoplastic left heart syndrome (HLHS) is a complex congenital heart disease characterized by hypoplasia of left sided heart structures leading to inadequate systemic perfusion following post-natal closure of patent ductus arteriosus. [1] Neonates born with HLHS are dependent on a patent ductus arteriosus and an interatrial communication for survival until surgical intervention. [2] We discuss the anaesthetic management of neonates with HLHS posted for non-cardiac surgery.

CASE 1

We discuss a case of Day 1 term male neonate weighing 2.2 kg with trachea-oesophageal fistula (TEF) and hypoplastic left heart, born via normal vaginal delievery and was posted for right posterolateral thoracotomy with TEF repair. The baby was antenatally diagnosed with hypoplastic left heart with Ventricular septal defect and dilated Right Ventricle. Immediately after birth, patient was intubated for apnea not responding to tactile stimulus and kept on Time cycled Pressure Limited (TCPL) mode with 21% fiO2 maintaining SpO2-85-90%. PGE1 infusion was started at 50 ng/kg/min to maintain ductal patency and infusion Dobutamine at 10mcg/kg/min to maintain adequate cardiac output. [1,2,10] Patient was extubated on Day 2 of life and kept on NIMV with pre-operative blood gas showing pH-7.47/pCO2-38.4/pO2-42.2 and HCO3-18.4. After taking written informed parental consent, patient was wheeled to OR and standard monitors including ECG, NIBP, Pre-ductal and post-ductal pulse oximetry, temperature and capnography were attached. With 24 G iv cannula in situ, patient was induced with incremental doses of fentanyl upto 10 mcg/kg and injection cis-atracurium 0.4 mg was given and videolaryngoscopy was done with C-MAC Miller Videolaryngoscope Blade Size 0 and endotracheal tube 3.5 mm uncuffed inserted. Anaesthesia was maintained with 50%oxygen and nitrous mixture with intermittent doses of fentanyl 10 mcg repeated every 1 hour with PGE1 and Dobutamine infusion ongoing. Under Ultrasound guidance right internal jugular vein

(IJV) was secured with 3 french 6 cm central venous catheter and left radial artery was cannulated for intraoperative hemodynamic monitoring and arterial blood gas analysis. A 2% dextrose-containing balanced crystalloid solution was administered at 10 mL/kg/h. Intraoperative Hemodynamics was stable with IBP-62/40mmHg and PR-142/min with blood gas showing pH-7.323/pCo2-46/pO2-42/HCO_3-18 mmol/Litre on 25% fio2.Post operatively the patient was shifted back to NICU with 3.5 mm uncuffed endotracheal tube in situ on TCPL mode of ventilator with Dobutamine and PGE1 infusion ongoing. Post operative blood gas showed pH-7.22/pCO2-62/pO2-55/ and HCO_3-18.4mmol/Litre. Postoperatively, the neonate remained haemodynamically stable and was successfully extubated on postoperative day 4. Further cardiac palliation was planned following multidisciplinary evaluation.

CASE 2:

We report a case of Day 3 term male neonate weighing 2.2 kg who was diagnosed with duodenal atresia with hypoplastic left heart and was posted for exploratory laparotomy. The patient was intubated and mechanically ventilated in neonatal intensive care unit in view of respiratory distress with subcostal retractions (Downe's score >6) and kept on Pressure controlled (PC) mode of ventilation with 40%fio2/Pi-16/PEEP-5 maintaining target SpO2-88%. As antenatal scan suggestive of hypoplastic left heart syndrome, post-natal 2D echo was done which confirmed severely hypoplastic left ventricle with hypoplasia of the mitral and aortic valves. The right ventricle was dilated and constituted the dominant systemic ventricle. An atrial septal defect was present, allowing unrestricted left-to-right atrial-level shunting. A patent ductus arteriosus (PDA) measuring 1.8 mm was noted, providing duct-dependent systemic blood flow. In view of duct-dependent systemic circulation, prostaglandin E1 infusion was initiated @150ng/kg/min to maintain ductal patency and infusion Dobutamine @10mcg/kg/min was started in view of poor cardiac contractility. Patient was planned to be taken up exploratory Laparotomy. Pre operative venous blood gas was done with pH-7.34/pCO2-47/pO2-30/lactate-3/HCO3-26.2 with Haemoglobin of 16.2gm%. After taking written informed parental consent; patient was shifted to Operating room (OR) intubated with infusion of PGE1 on-going @150ng/kg/min and dobutamine @10mcg/kg/min. In the OR, all the standard monitors including ECG, NIBP, preductal and post ductal pulse oximetry, capnography and temperature were attached. Baseline BP was 82/44 mmHg and PR-154/min Anaesthesia was induced with graded doses of intravenous fentanyl 2mcg/kg every 5 minutes and upto 5 mcg/kg followed by cis Atracurium 0.4 mg and mechanical ventilation started with PCV mode with 50% fio2/Pi-15/PEEP-5 with target SpO2-88-90% and target end-tidal CO_2 of 45-50 mmHg. Ultrasound guided caudal block was given with 1.25 mL/Kg of 0.25% Bupivacaine. Following anaesthesia induction, Ultrasound guided left brachiocephalic vein cannulated and 3 french 6 cm central line secured .Right radial artery was cannulated under Ultrasound guidance and connected to arterial transducer for intraoperative hemodynamic monitoring and blood gas analysis.Anaesthesia was maintained with 50%oxygen and nitrous oxide and intermittent doses of fentanyl 5mcg repeated every 30 minutes for the surgical duration of 4 hours.2% Dextrose in Ringer Lactate(DRL) was given intraoperatively with maintenance rate of 10 mL/kg/hr. Intraoperative hemodynamics were stable with BP-72/36mm Hg and PR-136/min on Dobutamine at 10 mcg/kg/min with PGE 1 infusion ongoing. Mild hypothermia was maintained with temperature of 36.2ºC to maintain Pulmonary vascular resistance. Intraoperative arterial blood gas revealed pH-7.233/pCO2-54/pO2-100/HCO3-23.5 and lactates of 3.0 with SpO2 90% signifying adequate systemic oxygen delievery and cardiac output. Post operatively patient was shifted to NICU intubated for further post-operative care on Pressure controlled mode of mechanical ventilation with 40% fio2, On PGE1 and Dobutamine infusion maintaining SpO2-88%. Post operative Arterial blood gas revealed pH-7.278/pCo2-48/pO2-140/HCO3-17.7. The neonate was extubated on postoperative day 5 and subsequently supported with non-invasive ventilation. Further cardiac palliation was planned for following multidisciplinary evaluation.

DISCUSSION:

In hypoplastic left heart syndrome, there is complete mixing of pulmonary venous and systemic venous blood in a single ventricle, which is connected in parallel to both pulmonary and systemic circulation and the systemic circulation depends on PDA. [3,7,8] The Left ventricle in an infant with HLHS does not serve any function. In neonates with HLHS and a patent ductus arteriosus, blood is ejected from the heart through the pulmonary valve and into the main pulmonary artery to provide a source of blood for the pulmonary circulation. After the main pulmonary artery, blood flows into the right and left branch pulmonary arteries. From the left pulmonary artery, a portion of the blood flows through the ductus arteriosus. Blood flow through the ductus arteriosus supplies the aortic arch, ascending aorta and coronary arteries in a retrograde fashion and the descending aorta in an antegrade direction, thereby supplying blood to the systemic circulation. [10] Survival depends on balance between systemic (Qs) and pulmonary blood flow (Qp), because both circulation are supplied from the right ventricle in parallel. The major determinant of distribution is the relative systemic and pulmonary vascular resistance. [3,8,9] The primary goal in the management of patient with HLHS, is optimization of systemic oxygen delievery and perfusion pressure to prevent end organ ischemia and injury. This goal is achieved by balancing the pulmonary and systemic circulation with maximal systemic oxygen delievery at Qp/Qs=1:1. [9] **Shunt flow ratio**

$$QP/Qs = \frac{(SAOO2 - SMVO2)}{(SPVO2 - SPAO2)}$$

An increase in pulmonary blood flow will decrease systemic cardiac output and leads to systemic hypoperfusion, metabolic acidosis in the presence of high arterial oxygen saturation· [3,8,9] Thus in SV physiology patient, PVR manipulation is essential and is accomplished most reliably with ventilatory intervention. [4,7-9] Hypercarbia in combination with lowest possible fiO2 is used to increase Pulmonary vascular resistance (PVR) and to decrease Pulmonary blood flow. Inotropic support is also required to maintain adequate cardiac output· [1,2] Increase in heart rate(>140/min) can cause myocardial ischemia in patients with low aortic diastolic pressures. A target PaO2-40-45mm Hg and SaO2-70-80% are associated with adequate systemic oxygen delievery. [3,9,10] Thus optimal anaesthetic management of neonate with HLHS entails a thorough understanding of spectrum of anatomical and physiological findings that can comprise this syndrome. Management of the effect of anaesthetics and intraoperative events on pulmonary blood flow is important because rapid flux in PVR during post-natal and perioperative period is the major destabilising factor in these neonates. [2,7,8]

CONCLUSION:

Neonates with hypoplastic left heart physiology undergoing major non-cardiac surgery present a significant anaesthetic challenge because systemic perfusion is critically dependent on ductal patency and a delicate balance between pulmonary and systemic blood flow. [2,3,8,9] Successful perioperative management requires an individualized approach aimed at maintaining ductal patency, adequate myocardial contractility and systemic perfusion while avoiding excessive pulmonary blood flow. Judicious oxygen administration, controlled ventilation, appropriate inotropic support, invasive haemodynamic monitoring and uninterrupted prostaglandin E1 infusion are important components of management. These cases demonstrate that, with a clear understanding of single-ventricle physiology, meticulous monitoring and close multidisciplinary coordination, urgent non-cardiac surgery can be successfully undertaken in these high-risk neonates. [1,2,5,6]

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