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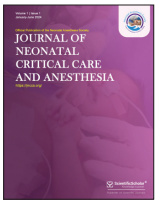

Case Report

Neonate with Single Ventricle for Emergency Colostomy – Anesthetic Considerations and Perioperative Challenges

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ABSTRACT

Single-ventricle condition involves a spectrum of congenital cardiac conditions associated with only single functioning ventricle with other ventricle usually rudimentary or non-functional. There is a mixing of systemic and pulmonary venous blood usually at the atrial level, and this leads to significant morbidity and mortality especially when they have to undergo surgery under anesthesia. We report successful management of a 3-day-old neonate, diagnosed with anorectal malformation and was presented to us for colostomy. This article describes the goals of anesthesia and risks involved in such cases with a brief discussion on various aspects of single-ventricle physiology.

Keywords: Congenital heart disease, Extubation, Neonate, Single ventricle, Tricuspid atresia

INTRODUCTION

Single-ventricle condition involves a spectrum of congenital cardiac conditions associated with only single functioning ventricle with other ventricle being usually rudimentary or non-functional. There is a mixing of systemic and pulmonary circulations at the atrial level. Mixed blood is then distributed to both systemic and pulmonary circulation from the single-functioning ventricle through an intra- or extra-cardiac shunt. Venous admixture is essential for their survival. The condition is associated with significant morbidity and mortality especially when they present for surgeries under anesthesia. We present a case of a 3-day-old baby, who presented to us in an emergency in the evening, for exploratory laparotomy. We discuss challenges that we faced in achieving the anesthetic goals that need to be met with during anesthesia induction and management as well as at the time of extubation, for optimal outcome.

The goals of anesthesia management in single-ventricle physiology include maintaining a precarious balance of pulmonary and systemic circulation with minimal interference with preload, afterload, heart rate (HR), blood pressure (BP), and myocardial contractility. In this case report, we present a neonate presented for surgical emergency found to have tricuspid atresia with single-ventricle of left ventricle (LV) type with intracardiac mixing occurring at the level of atria through ostium secundum atrial septal defect (OS ASD) and pulmonary blood flow maintained by patent ductus arteriosus (PDA). Anesthetic management involved general anesthesia (GA) with an endotracheal tube with preductal and post-ductal saturation monitoring. Maintaining good ventricular contractility, avoiding extremes of hemodynamic fluctuations, air bubble precaution, maintaining euthermia, and early extubation are a cornerstone of successful anesthetic management. Mechanical ventilation (MV) with high intrathoracic pressures will have deleterious effects on hemodynamics and pulmonary vascular resistance that can alter the distribution of cardiac output between systemic and pulmonary blood flow.

CASE REPORT

A 3-day-old male baby was brought to us in the emergency department with chief complaints of non-passage of meconium since birth, distended abdomen, respiratory distress, and bluish discoloration of lower limbs. Neonate was term born by caesarean section, in a peripheral hospital, for transverse lie, and had cried immediately after birth (APGAR 8–10). He was admitted to the Neonatal Intensive Care Unit (NICU). His birth weight was 3.1 kg. He was diagnosed with anorectal malformation and was posted for

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emergency exploratory laparotomy. On clinical examination, the neonate was active and his peripheries were cyanosed and were on oxygen therapy with a high-frequency nasal cannula (HFNC) at a rate of 5 L/min. His oxygen saturation (SpO_2) was 87% with a fraction of inspired oxygen (FiO_2) of 70%. The baby was febrile (temperature 101°F). His current weight was 2.39 kg. On auscultation, his chest was clear and S1S2 was heard along with a systolic murmur. On cardiac evaluation, his echocardiographic findings revealed complex congenital heart disease (CHD) with a single ventricle, of LV type, aneurysmal interatrial septum, and small OS ASD with the right to left shunt. His tricuspid valve was hypoplastic. The main pulmonary artery (PA) and pulmonary valve confluence were seen. Left and right PA were seen. There was a moderate PDA with a left-to-right shunt. LV systolic function was good. While he was in NICU, dobutamine infusion was started at a rate of 0.5 mL/h. Intravenous fluids therapy with D10 was on flow at a rate of 12 mL/h. Laboratory investigations showed hemoglobin of 16.8 mg%, total leucocyte count of 19.6 mm^3, serum creatinine of 2.19 mg/dL, and C-reactive protein elevated to 86.6mg/L.

In view of emergent surgery (colostomy), cardiac defect, impending sepsis, and any other complication, it was deemed a very high-risk surgery. Parents were explained in detail about the risks involved, the need for his postoperative cardiac workup, the possibility of postoperative MV, and written informed consent was taken. The neonate had already been intubated in NICU and shifted to operation theatre (OT) on T piece resuscitator with peak inspiratory pressure (PIP) of 15 cmH_2O and positive end-expiratory pressure (PEEP) of 5 cmH_2O.

The anesthesia plan included GA with regional anesthesia (RA). ASA standard monitoring with pre- and post-ductal SpO_2 monitoring was established. Vitals on arrival were non-invasive BP of 120/69 mmHg (without Dobutamine) and HR of 160/min was recorded, baseline SpO_2 of 86% (preductal) and 83% (postductal), on FiO_2 of 100%. In view of his high baseline BP, anesthesia was induced with inj. Propofol 5 mg and inj. Fentanyl 5 µg, and Inj. Atracurium 1mg, and the tube was connected to a ventilator. MV was started on pressure control mode with PIP 15 and PEEP of 5 cmH_2O. Anesthesia was maintained on oxygen (O_2), Air (50:50), and Sevoflurane 1–1.5% with a target minimal alveolar concentration (MAC) of 0.7–0.8. The caudal block was attempted but abandoned due to difficult anatomy. Intraoperatively, there was one episode of desaturation immediately after induction with a drop in preductal SpO_2 to 74%, which improved on hand ventilation and increase in FiO_2. Thereafter, the child remained clinically stable. Inj. Fentanyl 2.5 µg repeated once and inj. Paracetamol given in dose of 33 mg IV. Ventilation parameters improved after relief of abdominal obstruction and PIP was reduced to 12 cm of H_2O and FiO_2 to 30% with SpO_2 around 90%. At the end of the surgery, after ensuring adequate reversal of neuromuscular blockade with inj. Glycopyrrolate 25 µg and inj. Neostigmine 125 µg, the baby was extubated uneventfully in OT. The patient was shifted to NICU on O_2 therapy at a rate of 1L/min and preductal SpO_2 of 90–94%. In the postoperative period, the neonate again had one episode of worsening SpO_2, which was managed with HFNC at a rate of 5 L/min with FiO_2 40%. Orogastric feeds were started on 2nd postoperative day (POD) and the baby was shifted to a higher cardiac center on the seventh POD for further management of his underlying CHD.

DISCUSSION

Single-ventricle condition involves the spectrum of congenital cardiac conditions associated with only single-functioning ventricle with other ventricle usually rudimentary or non-functional.[1] Tricuspid Atresia with single ventricle of LV type with intracardiac mixing occurring at the level of atria through OS ASD and pulmonary blood flow maintained by PDA. Anesthetic management involved General anesthesia with an endotracheal tube with preductal and post-ductal saturation monitoring. Maintaining good ventricular contractility, avoiding extremes of hemodynamic fluctuations, air bubble precaution, maintaining euthermia, and early extubation are a cornerstone of successful anesthetic management. MV with high intrathoracic pressures will have deleterious effects on hemodynamics and pulmonary vascular resistance that can alter the distribution of cardiac output between systemic and pulmonary blood flow.

Regardless of the underlying defect, there are two prerequisites for patients to survive with this condition. First, there should be an unobstructed mixing of both systemic and pulmonary venous return, usually through ASD, mixed blood will flow to a single functioning ventricle. Second, the sole ventricle generates cardiac output, which is distributed to both systemic and pulmonary circulation via a patent shunt between them, which is usually a PDA or ventricular septal defect (VSD).[2] In the absence of any anatomical obstruction, systemic (Qs) and pulmonary blood flow (Qp) depend on the relative resistances of their respective beds. In view of the mixing of deoxygenated blood from the systemic venous system and oxygenated blood from the pulmonary venous system at the atrial level, they have low SpO_2 levels. Qp/Qs ratio of one indicates near adequate intra-atrial mixing, which corresponds to SpO_2 of 70–80% and partial pressure of oxygen between 40 and 50 mmHg.[3] Our neonate was having SpO_2 between 74% and 86%, which indicated near-balanced circulation. Value > 1 indicates pulmonary overcirculation with good saturation but at the cost of reduced systemic blood flow.[4] Similarly, a Qp/Qs ratio of <1 indicates high systemic blood flow presenting with increased cyanosis and signs of inadequate systemic oxygenation despite good systemic blood flow.[4] Single ventricle in such cases has both pressure and volume overload. This leads to

increased myocardial work that can precipitate heart failure and cardiogenic shock in those who present later in life.

This condition can be detected antenatally by sonography. Although usually present at birth, there may be delayed presentation after 48 h when functional closure of ductus arteriosus starts.[5] Echocardiography is diagnostic in such cases.[5] Their survival depends on the underlying defect, the presence of shunts to maintain parallel circulation, and palliation procedure is performed or not. There are case reports of unpalliated single ventricle presenting for non-cardiac surgery or obstetric procedures in later life.[6]

Anesthesia has its own implication in single-ventricular physiology (SVP) and is associated with significant morbidity and mortality. The goals of anesthetic management in single-ventricle physiology are to maintain balanced circulation with minimal interference with preload, afterload, HR, BP, and myocardial contractility. Any changes in vascular resistance of pulmonary and systemic beds will alter the Qp/Qs ratio and changes in systemic oxygenation. GA forms the mainstay of anesthetic management in these cases. Anesthetic drugs significantly affect hemodynamics due to their direct or indirect effect on the cardiovascular system. Propofol has a direct myocardial depressant effect, whereas etomidate does not have much effect on hemodynamics and can be used for induction. Ketamine may be used if LV systolic functions are good. Inhalational agents are direct myocardial depressants. Sevoflurane has minimal cardiac depressant effect of all, so is preferred with a target MAC of 0.7–0.8. Nitrous oxide is contraindicated due to its deleterious effects on pulmonary vascular resistance (PVR) and could further distend already obstructed bowel in our case. Maintaining BP, especially diastolic BP within an acceptable range, is important to maintain adequate coronary blood flow. Similarly, tachycardia will increase myocardial O_2 demand, leading to myocardial ischemic injury.

In our case, the neonate was induced with propofol in view of constantly high BP and good LV systolic function. The caudal block was attempted but could not be done due to associated spine abnormality. MV with high intrathoracic pressures will have deleterious effects on hemodynamics; targeting the lowest of PIP with optimal PEEP is suggested. During MV, we tend to keep high FiO_2 in cases with low SpO_2 but this will abolish hypoxic pulmonary vasoconstriction in them that can lead to pulmonary hyperperfusion. The ventilation strategy is to keep a high tidal volume of 8-12 ml/kg with lower respiratory rates allowing longer expiratory times and limited PEEP.[7] SpO_2 is aimed at around 80% with low FiO_2 ranging from 0.21 to 0.30 and permissive hypercapnia until 55mmHg. Early extubation is recommended, as it will allow the child to breathe spontaneously and help in maintaining a precarious balance between Qp and Qs. In our case, we extubated the neonate on the table uneventfully.

PDA is required to maintain pulmonary blood flow. Any condition leading to reduced flow from the ductus such as high PVR or hypovolemia can lead to its closure. Analgesia in such cases is of utmost importance as pain can lead to high PVR and reduced shunt flow. NSAIDs are avoided as they can lead to PDA closure. Maintaining good hydration is important to keep ductus arteriosus patent as well as to reduce the effect of high viscosity seen in these patients due to polycythemia. Restriction of longer fasting time and judicious use of intravenous fluids is advocated. However, at the same time, they are prone to pulmonary edema due to associated pulmonary abnormalities[8] and pulmonary hyperperfusion. Maintaining glucose homeostasis and euthermia is important in neonatal anesthesia. Since these babies have patent intra- and extra-cardiac shunts, strict air bubble precaution is desirous.

Long-term survival in tricuspid atresia is better around 80% at 10 years.[9] However, there are increased chances of poor neurodevelopmental outcomes in them as their brains are exposed to chronic hypoxia since birth.[10]

CONCLUSION

Single-ventricle patients can pose various anesthetic challenges. A better understanding of SVP is required to handle such cases. The main aim of anesthetic management involves GA with tracheal intubation, maintaining an adequate Qp/Qs ratio, and monitoring and maintaining pre-ductal and post-ductal saturations, which in turn depends on a good correlation between preload and afterload. An MV with high intrathoracic pressures will have deleterious effects on hemodynamics and pulmonary vascular resistance, affecting the distribution of cardiac output between systemic and pulmonary blood flow. Maintaining good ventricular contractility, avoiding extremes of hemodynamic fluctuations and early extubation form the cornerstone of anesthetic management in such cases.

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