



Perioperative Management of a Child with Hypoplastic Left Heart Syndrome Undergoing Cryptorchidism Surgery

Shaban Memeti^{1*} , Saimir Kuci², Alfred Ibrahim², Marsela Goga², Altin Veshti³, Sokol Buba⁴, Arben Baboci³

¹Department of Paediatrics Surgery, Faculty of Medicine, Ss Cyril and Methodius University of Skopje, Skopje, Republic of Macedonia; ²Department of Cardiac Anesthesia, Mother Teresa University Hospital Center, Tirana, Albania; ³Department of Cardiac Surgery, Mother Teresa University Hospital Center, Tirana, Albania; ⁴Department of Paediatrics Surgery, Mother Teresa University Hospital Center, Tirana, Albania

Abstract

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Correspondence: Shaban Memeti, Department of Paediatrics Surgery, Faculty of Medicine, Ss Cyril and Methodius University of Skopje, Skopje, Republic of Macedonia. E-mail: shab28me@yahoo.com
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BACKGROUND: Hypoplastic left heart syndrome (HLHS) is a complex congenital heart condition which includes abnormal development of the left-sided cardiac structures leading to inadequate systemic perfusion following postnatal closure of the patent ductus arteriosus (PDA). Surgical palliation may be accomplished through a 3 staged process known as the hybrid approach. Shortly after birth, the first procedure includes bilateral pulmonary artery banding to restrict pulmonary blood flow and placement of a stent into PDA to allow for systemic blood flow without the ongoing need for prostaglandin therapy to maintain ductal patency. Patients with HLHS may need to undergo other non-cardiac surgical procedures during the 1st years of life posing a real challenge to the anaesthesiologist, surgeon, and the entire medical team.

CASE PRESENTATION: We present the case of a 18-months old, 9 kg infant who presented for cryptorchidism surgery after performing the first surgical stage for the repair of HLHS. Cryptorchidism or undescended testis is one of the most common pediatric disorders of the male endocrine glands and the most common genital disorder identified at birth. The main reasons for treatment of cryptorchidism include increased risks of impairment of fertility potential, testicular malignancy, torsion, and/or associated inguinal hernia. The intraoperative implications of the hybrid anatomy are discussed, options for anesthetic care presented, and previous reports of anesthetic care for such patients reviewed.

CONCLUSION: Children with palliated HLHS have anesthetic considerations that must be followed to reduce perioperative morbidity and mortality in this high-risk pathology.

Introduction

Hypoplastic left heart syndrome (HLHS) is a complex congenital heart condition which includes abnormal development of the left-sided cardiac structures leading to inadequate systemic perfusion following postnatal closure of the patent ductus arteriosus (PDA) [1], [2].

Various surgical strategies have been employed for the palliation of HLHS in the immediate newborn period including the Norwood procedure and more recently, the hybrid procedure. The hybrid procedure includes bilateral pulmonary artery (PA) banding through a median sternotomy and placement of a stent into PDA [3], [4].

Patients with HLHS may need to undergo other non-cardiac surgical procedures during the 1st years of life posing a real challenge to the anesthesiologist, surgeon, and the entire medical team. We present the case of a 18-months-old baby with HLHS undergoing orchidopexy surgery after Norwood procedure at

2 months old. Orchidopexy in the first 18 months of life is recommended to preserve available fertility potential. In the majority of cases the total number of germ cells is within the normal range in cryptorchid testes during the first 6 months of life, but about 25% of the cryptorchid boys are born with a reduced number of germ cells.

Cryptorchidism or undescended testis (UDT) is one of the most common pediatric disorders of the male endocrine glands and the most common genital disorder identified at birth. The main reasons for treatment of cryptorchidism include increased risks of impairment of fertility potential, testicular malignancy, torsion, and/or associated inguinal hernia [5].

Case Report

We present the case of a 18-month-old child, weighing 9.0 kg, diagnosed at birth with HLHS. At 2 months old the child underwent a successful Norwood

procedure. The base of the PA is attached to the aorta and a shunt is inserted between the aorta and the PA leading blood to the lungs, bypassing the left side of the heart.

Objective examination shows an afebrile child with axillary temperature of 37°C, no abnormal facie, with peripheral cyanosis, no peripheral edema. He was not pale and not jaundiced. The chest wall was symmetrical with equal chest expansion; he had a respiratory rate of 36 breaths/min, vesicular breath sounds with fine basal crepitations. The heart rate was 140 beats/min and regular. The first and second heart sounds were heard, they were of normal intensity but with a pansystolic murmur grade 4/6 maximal at the left lower sternal edge. The abdomen revealed a subtle hepatomegaly (1 cm below the costal margin), no ascites. An undescended testicle was not felt in the right scrotum at all.

The packed cell volume was 43%, white cell count of 5800 with neutrophils of 30%, lymphocytes of 62%, monocyte of 6%, and eosinophils of 2% all within normal range. Preoperative testing showed a normal (Complete cell blood count) as well as normal PT, PTT, and INR. Blood sugar, serum electrolytes and creatinine were within normal, except for a mildly elevated urea of 70 mg/dL.

His SpO₂ was 90% with 0.5 L of O₂ through a nasal cannula. There were no other relevant signs in the physical examination. An EKG showed no arrhythmias and an echocardiographic evaluation showed adequate single ventricle function and permeability of the Norwood shunt. The chest X-ray showed no abnormal infiltrates.

At the time of the assessment, he has only a peripheral 24G IV catheter for the administration of maintenance fluids.

We used general anesthesia with laryngeal mask airway (LMA) combined with local anesthesia. Standard monitors were placed including invasive blood pressure monitoring through a cannula in the right radial artery. Before surgery he received 1 ml/kg/min of 5% albumin, 0.05 mg/kg midazolam. The patient was pre-oxygenated, and general anesthesia was induced with fentanyl in small doses sevoflurane and propofol. A LMA was placed; anesthesia was maintained with 1 MAC (minimum alveolar concentration) of sevoflurane, oxygen, and air and IV propofol. The patient remained hemodynamically stable during the case (blood pressure and heart rate were maintained within 20% of pre-operative baseline values).

Orchidopexy was performed. During the procedure, a small incision (cut) in the groin was made and was located the testicle. The testicle was examined to make sure it was healthy. The hernia sac was repaired. A second incision in the scrotum was made to make a "pocket" under the scrotal skin and place the testicle

into the scrotum. Once the procedure was complete, we closed both incisions with dissolvable sutures.

At the end of the case, the LMA was removed without complication. The patient was transferred to the ICU, under 2 l of O₂ through nasal cannula, without the need for additional opioids. The child had no episodes of emesis, and pain was well controlled. His hemodynamics remained stable postoperatively.

Discussion

Cryptorchidism or UDT is one of the most common pediatric disorders of the male endocrine glands and the most common genital disorder identified at birth. The main reasons for the treatment of cryptorchidism include increased risks of impairment of fertility potential, testicular malignancy, torsion, and/or associated inguinal hernia. Cryptorchidism has evolved significantly over the past half century, with respect to both diagnosis and treatment. The current standard of therapy in the United States is orchidopexy (also referred to as orchiopexy in the literature), or surgical repositioning of the testis within the scrotal sac, while hormonal therapy has fewer advocates. Successful scrotal relocation of the testis, however, may reduce but does not prevent these potential long-term sequelae in susceptible individuals [5].

Congenital heart disease (CHD) adds a significant risk of mortality in children requiring noncardiac surgery, especially in patients with single ventricle physiology who cannot have a "definite" surgical repair [6].

HLHS is a complex congenital heart condition which includes abnormal development of the left-sided cardiac structures. The hybrid procedure includes bilateral PA banding through a median sternotomy and placement of a stent into PDA [4], [7].

Surgical palliation may be accomplished through a 3 staged process.

1. Norwood procedure. This surgery is usually done within the first 2 weeks of your child's life
2. Bidirectional Glenn procedure. This procedure is generally the second surgery. It's done when your child is between 3 and 6 months of age.
3. Fontan procedure. This surgery is usually done when your child is between 18 months and 4 years of age
4. Cardiac transplant is the best treatment, in those parts of the world where the expertise and necessary equipment are available with survival following surgery at 5 years and 10 years being 65% and 55%, respectively [3].

The primary goal in the management of patients with single ventricle physiology is optimizing systemic

oxygen delivery and perfusion pressure. This is achieved by balancing systemic and pulmonary circulations (Q_p : Q_s). It is important to avoid hyperventilation and hyperoxia because it would reduce pulmonary vascular resistance (PVR) and cause pulmonary “overcirculation;” hypoxia and hypercarbia will elevate PVR and cause pulmonary “undercirculation.” Elevation of systemic vascular resistance (SVR) could generate lactic acidosis and low cardiac output (high afterload). LMA showed to be effective in avoiding hyperventilation and hyperoxia.

Hypothermia should also be avoided because it could elevate SVR and also elevate VO_2 [8], [9].

Anesthetic maintenance is usually achieved with volatile anesthetics and opioids. There are no specific recommendations or contraindications for the use of regional anesthesia techniques. The arterial oxygen saturation usually increases once the patient is anesthetized due to reduced peripheral O_2 extraction and improved cardiac output (reduced afterload) [7].

The induction and maintenance of anesthesia are achieved with a combination of opioids, volatile, and intravenous agents. During both the induction and maintenance of anesthesia, the anesthetic agents chosen should be modified to limit their deleterious effects on hemodynamic parameters [8], [9].

In our patient, anesthesia was induced and maintained with a combination of a volatile agent (sevoflurane or isoflurane) and fentanyl. Although synthetic opioids such as fentanyl offer the advantage of hemodynamic stability, the use of high doses will often require postoperative mechanical ventilation [5], [6]. Given that our patient already required chronic mechanical ventilation, we chose a high dose opioid technique with fentanyl to effectively blunt the surgical stress response and maintain hemodynamic stability. Alternatively, if the goal had been early LMA removal, the dose of fentanyl could be decreased to 3–5 $\mu\text{g}/\text{kg}$ and higher doses of isoflurane could be used for maintenance anesthesia. In addition to our patient’s comorbid CHD, the surgical procedure may also impact intraoperative hemodynamic and respiratory function. Concerns exist as to whether or not, laparoscopic surgery is safe in patients with CHD, especially those with HLHS, given the increase in intra-abdominal pressure (IAP) with insufflation and its effects on cardiac function. In the absence of associated CHD, an IAP of 12 mm Hg, but not 6 mm Hg, has been shown to decrease cardiac index assessed by TEE [6]. IAP above 6–8 mm Hg may decrease preload, increase afterload, and increase PVR which may be poorly tolerated in patients with HLHS. Echo- Doppler monitoring has shown decreases in aortic blood flow, stroke volume, and cardiac index in children at an IAP as low as 10 mm Hg [6]. In addition, increases in PaCO_2 induced by laparoscopy should be expected and minute ventilation increased, as an increase in PaCO_2 levels may have deleterious effects on myocardial function and pulmonary vascular resistance. A significant increase in

the arterial pressure of CO_2 and end-tidal CO_2 gradient after abdominal insufflation has shown end-tidal CO_2 to be an insensitive monitor in healthy infants [10].

Adequate post-operative pain control in children has historically been difficult to achieve and continues to present a challenge to the anaesthesiologist and intensive care unit physician. The challenge of adequately treating post-operative pain is particularly relevant after surgery for complex CHD such as HLHS as pain treatment must be carefully weighed against the risks associated with opioids. Our experience has suggested that nurse-controlled anesthesia with fentanyl achieves effective analgesia with limited adverse effects as demonstrated by low pain scores, successful early LMA removal, infrequent need to change to an alternate opioid, and a low incidence of adverse effects [9]. Fentanyl is commonly preferred in this setting due to its rapid onset, easy titratability, lack of active metabolites, and limited effect on myocardial and hemodynamic performance. Dexmedetomidine may be added to the regimen to provide sedation and potentiate opioid-induced analgesia thereby decreasing opioid and potentially opioid-related adverse effects [11].

Conclusion

Children with palliated HLHS have anesthetic considerations that must be followed in order to reduce perioperative morbidity and mortality in this high-risk pathology. LMA combined with local anesthesia was effective to maintain optimal cardiac function of this patient with HLHS.

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