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Anaesthetic considerations in a neonate with unrepai red D-transposition of great arteries with non-syndromic holoprosencephaly for emergency exploratory laprotomy: A case report

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Abstract

D-Transposition of great arteries (D-TGA) is a cyanotic congenital heart disease (CHD) characterized by parallel circulation. Non syndromic holoprosencephaly is an abnormality of brain development resulting from failure or incomplete division of embryonic forebrain into distinct cerebral hemisphere. Major challenges in patients of D-TGA are hemodynamic instability, cyanotic spell, coagulopathy, acid base and electrolyte imbalance. We report successful management of one day old neonate posted for emergency resection anastomosis for ileal atresia with unrepai red d-TGA and non-syndromic holoprosencephaly.

Keywords: Cyanotic congenital heart disease, uncorrected D-Transposition of great arteries, non-cardiac surgery, holoprosencephaly

Introduction

Transposition of great arteries (TGA) is a congenital malformation characterised by atrioventricular concordance and ventriculoarterial discordance. The incidence is 1 in 3500-5000 live births ^[1]. Cardiac malformation which are associated with TGA are ventricular septal defect (VSD), atrial septal defect (ASD) & patent ductus arteriosus (PDA) which allows intermixing of arterial and venous blood hence chances of post natal survival ^[1]. Neonates and infants with congenital heart disease are at two fold increased risk from non cardiac surgery ^[2]. Pre-operative diagnosis is done by 2- D echocardiography, which gives anaesthetist the idea about direction of shunt in VSD, ASD, PDA and amount of pulmonary artery hypertension (PAH) ^[3].

Non syndromic holoprosencephaly is an abnormality of brain development that also affects the head and face. The incidence of holoprosencephaly is 1 in 13000 births. Normally brain divides into two hemispheres in early development. Holoprosencephaly occurs when brain fails to divide properly in to right and left hemispheres. Patients with non syndromic holoprosencephaly have microcephaly, although they can have hydrocephalus that causes macrocephalus. Patients also present with features like cleft lip and cleft palate, single maxillary central incisor, flat nasal bridge. HP is characterized by hypoplasia of prosencephalon, facial anomalies (hypotelorism, flat nose, and/or small prolabium), abnormality of autonomic nervous system functions (hypernatremia and/or poikilothermia), and clonic convulsion ^[4].

Main anaesthetic concerns in this patient for an anaesthetist is paediatric age group, direction of shunt, maintain a balance between pulmonary vascular resistance and systematic vascular resistance to avoid intra-operative reversal of shunt, to prevent hypoxia, maintain haemodynamic parameters, prevent inadvertent paradoxical air embolism. Presence of holoprosencephaly with hydrocephalus and cleft palate poses challenges for mask holding and intubation for patient. Temperature regulation in HP patients also possess a challenge. We came across an neonate posted for emergency resection anastomosis for ileal atresia with unrepai red TGA and non-syndromic holoprosencephaly.

Case report

A 1 day old term neonate born of second degree consanguinous marriage with unrepai red TGA and non syndromic holoprosencephaly which was diagnosed antenatally, presented to

us with complaints of abdominal distension and no passage of meconium diagnosed on ultrasound abdomen, posted for emergency exploratory laparotomy with resection of ileal segment and ileo-ileal anastomosis. On pre-op examination patient had peripheral cyanosis, pulse rate was 154/min, saturation in all four limbs was between 62-65% on air, with oxygen saturation in all 4 limbs was 90-92 percent. Head circumference of that neonate was 45 cm and abdominal girth was 32 cm. Patients complete blood count and renal function test were within normal limits. Patients 2-D Echo findings showed AV concordance, VA discordance, large sub aortic VSD, aorta arising from RV, origin of pulmonary artery cannot be visualised, large PDA with severe tricuspid regurgitation with severe pulmonary artery hypertension. Patients Xray showed egg shaped heart.

Patient was kept nil by mouth on the day of surgery. Patient was taken on the OT table, all ASA monitors attached, 24 G cannula secured, patients heart rate was 156/ min regular, SPO2 on room air on pre ductal probe was 67% and postductal probe was 71%. Patient was pre-oxygenated with 100% oxygen for 3 minutes. After oxygenation SPO2 increased to 92%, pre medication was done with Inj. Atropine 0.01mg/kg. Patient was induced with Inj. Ketamine 2mg/kg i.v. and Inj. Propofol 1mg/kg. After checking for ventilation patient was paralysed with Inj. Atracurium 0.5 mg/kg, for ease of intubation a soft bolster was kept below patients shoulders, patient was intubated with 3.5mm uncuffed endotracheal tube. Anaesthesia was maintained on 50% oxygen and 50% air and Sevoflurane. After induction of anaesthesia to provide pain relief single shot caudal was given to the patient. Intra-operative fall in blood pressure was managed by boluses of phenylephrine 0.1mg/kg to avoid reversal of shunt. At the end of surgery patient was reversed with inj atropine 0.01 mg/kg and inj. Neostigmine .05mg/kg, and extubated. Patient was observed in the recovery area for 30 minutes and shifted to NICU for further management.

Discussion

Transposition of great arteries is an anomaly where aorta arises from right ventricle and pulmonary artery arises from left ventricle, hence parallel circulation^[2,3]. This anomaly is incompatible with life unless there's some shunt which allows adequate mixing of blood. Neonates and infants are at increased risk of mortality with CHD undergoing non cardiac surgery^[4]. Patients with holoprosencephaly have poorly developed central nervous system. In addition there are periods of temperature instability, apnea^[5]. Difficult intubation. Even though intubation was difficult in this patient, it was managed carefully. Patient had hydrocephalus with increased head circumference posing a challenge for intubation. Anaesthetic considerations in neonate with intestinal obstruction is to maintain adequate hydration, prevent hypothermia, hypotension, avoid metabolic acidosis and minimisation of intracardiac shunting. Avoid decrease in systemic vascular resistance and increase in pulmonary

vascular resistance.

Pre-operative evaluation of patients with CHD undergoing non cardiac surgery should use a multidisciplinary approach that includes the participation of anaesthesiologist, cardiologists, intensivists, and surgeon. But as this case was an emergency it was not possible for all to participate. Pre-medication with hypnotics must be undertaken very cautiously because hypoventilation and hypercapnia may produce deleterious increase in pulmonary vascular resistance.

Standard conventional non invasive monitoring including pulse oximetry, electrocardiogram, non-invasive blood pressure, capnography and temperature are used in all patients. There are no evidence based recommendations to guide the anaesthetic management of patients with CHD undergoing non-cardiac surgery^[6]. This case was one of the unique and challenging cases we have encountered. Due to airway and metabolic problems, it is important to utilise capnography and pulse oximetry and to measure arterial blood gas tensions as indicated by the clinical situation^[7]. The anaesthesiologist must be prepared to deal with extremes of hyper and hypothermia when administering anaesthesia to these patients of holoprosencephaly^[8]. Our case was unique as it presented with CHD, holoprosencephaly and intestinal obstruction which posed us with a challenge of anaesthetic management and intubation difficulty.

Preoperative SpO₂, chest radiography, electrocardiography, and echocardiography are must before anaesthesia^[6]. Avoiding prolonged preoperative fasting and maintaining adequate hydration are important to prevent hyper viscosity^[2]. Large bore IV cannula must be secured, and all lines must be flushed to avoid air bubbles as these patients are at high risk of paradoxical embolism. We premedicated the patient with inj atropine to avoid bradycardia and reversal of shunt, for induction we used inj ketamine along with inj propofol to maintain systemic vascular resistance and pulmonary vascular resistance. For maintenance instead of using 100% oxygen we used 50% oxygen and 50% air to avoid retrolental fibroplasia. Avoid nitrous oxide as it may increase chances of increase in size of air bubble which may enter through the surgical site. Pain management in this patient was done by caudal anaesthesia. Maintenance of intravascular volume, avoidance of precursors to acidosis and minimisation of intra cardiac shunting⁹. Intraoperative hypotension can be managed with adequately replacing intravascular volume and direct acting alpha agonist like phenylephrine. Intraoperative episodes of hypotension were managed by boluses. Patient was extubated and shifted to paediatric ICU.

This case represents that with proper knowledge of pathophysiology of the condition and anaesthetic management we can prevent possible complications and successfully anaesthetise a complex case of CHD for non cardiac surgery.



Image 1: showing boot shaped heart with dilated bowel loops



Image 2: showing patient positioned for caudal block

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