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Anaesthetic Management during Transposition of Great Arteries (TGA) Correction: Points to be Focussed

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Authors' contributions

This work was carried out in collaboration between all authors. Author SG designed the study, performed the statistical analysis, wrote the protocol and the first draft of the manuscript. Authors AS and RM managed the analyses of the study. Author ND managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Transposition of great arteries (TGA) is combination of concordant atrioventricular and discordant ventriculo-arterial connections and is divided into dextro-looped (d-TGA) and levo-looped (l-TGA) based on whether the atria and ventricles are concordant or discordant, respectively. Arterial switch operation is the procedure of choice but other surgical surgical options exists. In our case, a 4 month old boy presented with cyanosis, delayed milestone, diagnosed d-TGA with ASD came for surgical repair. Arterial switch repair was planned but due to decreased right ventricular pressure and raised pulmonary arterial pressure, mustard-senning procedure was performed. Hypoxia, systemic hypoxemia, metabolic acidosis, hypercarbia, sympathetic nervous stimulation due to light plane of anaesthesia can trigger a rapid rise in PVR, and even a pulmonary hypertensive crisis. Our aim was to prevent alteration in SVR and PVR in order to prevent desaturation especially at time of induction and off CPB bypass.

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1. INTRODUCTION

Transposition of great arteries (TGA) is the group of cyanotic congenital heart lesions in which the pulmonary artery originates completely or partly from the morphological left ventricle (LV) and the aorta originates completely or partly from the morphological right ventricle (RV). It is relatively frequent congenital cardiac defect accounting for more than 5% of all cardiac malformations with a prevalence of 0.2 per 1,000 live births and male preponderance [1,2]. TGA is divided into dextro-looped (d-TGA) and levo-looped (l-TGA) based on whether the atria and ventricles are concordant or discordant, respectively. Complete TGA, d-TGA, TGA are often used synonymously. Traditionally d-TGA is divided further into 3 subtypes: TGA with intact ventricular septum (TGA-IVS), TGA with ventricular septal defect (TGA-VSD), and TGA with VSD and left ventricular outflow tract obstruction (TGA-VSD-LVOTO) with incidence of 70%, 25%, and 5-10%, respectively [3,4].

The arterial switch operation (ASO), currently the procedure of choice has recently improved the 20 year survival rate to nearly 90% [4]. Perioperative anaesthetic considerations include comprehensive preoperative evaluation, management of hypoxaemia, regulation of systemic vascular resistance, pulmonary vascular resistance, post bypass bleeding tendencies and ventricular dysfunction.

2. CASE REPORT

A 4 month old boy presented with history of cyanosis since birth aggravated by crying, delayed milestone, diagnosed d-TGA with ASD soon after birth with recuurent attacks of chest infection came for surgical correction. His heart rate was 144 beats/minute regular, respiratory rate 32 breaths/minute, and had room air oxygen saturation of 77%. Physical examination revealed central cyanosis, cold clammy skin, bilateral breath sounds were clear. On auscultation systolic ejection murmur was heard in pulmonic area. Electrocardiogram showed right axis deviation, Echocardiography report revealed d-TGA, enlarged right atrium and ventricle, intact interventricular septum, OS-ASD (6.5 mm) bidirectional shunt, left ventricle ejection fraction 60%, Ventriculoatrial (VA) concordance and Ventriculoarterial (Va) discordance. Preoperative laboratory investigations revealed Hb 14.4%, Plt counts 3.95 lakhs/ml, K^+ 3.57 mMol/L. The prothrombin time was 14.1 seconds and INR was 1.3.

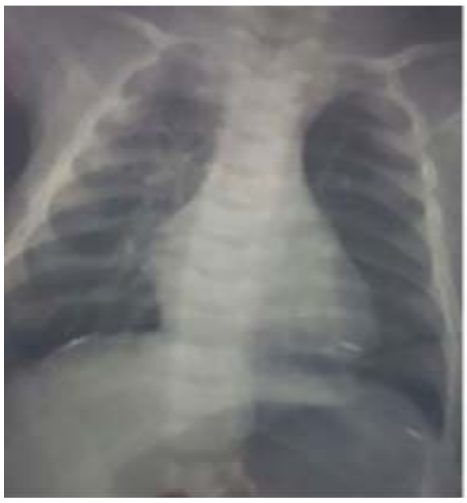

Fig. 1. X-ray chest PA view

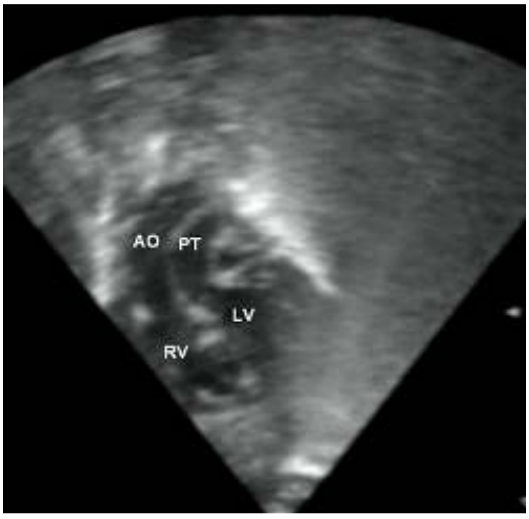

Fig. 2. Subcostal view showing right ventricle giving rise the aorta and left ventricle pulmonary trunk, proximal portion of arteries running parallel

Written informed consent was taken from parents and child was kept nil per oral for 4hrs. On arrival to Operation Theatre standard monitors were placed which includes Electrocardiography, non-invasive blood pressure and pulse oxymeter. Intravenous line secured using 24G canula. Inj. fentanyl 5 microgram/kg and inj. midazolam 0.02

mg/kg was given and inhalational induction was done with sevoflurane in 100% oxygen. Tracheal intubation was facilitated with single dose of rocuronium 5 mg. Antibiotic prophylaxis was done with ceftriaxone. Following tracheal intubation, femoral arterial cannula, and a triple lumen central venous catheter were placed. The cannula was connected to a pressure transducer for continuous monitoring of the vital signs. Capnography, temperature, urine out-put and airway volume and pressure monitoring was also done.

The baseline arterial pressure was 68/42 mm of Hg and CVP was 15 cm of H_2O. Maintenance anesthesia consisted of sevoflurane and fentanyl infusion adjusted to maintain the mean arterial pressure at 30-50 mmHg. Vecuronium was used as neuromuscular blocking agent.

Prior to insertion of arterial and venous cannulas for bypass, heparin 20 mg was administered through the central catheter. The activated coagulation time (ACT), used for monitoring heparin activity was 540 seconds. Plan of surgery was to perform arterial switch operation (ASO) but due to low left ventricular pressure which was insufficient to sustain systemic circulation, atrial switch was preferred over arterial switch and ASD was repaired. During cardiopulmonary bypass, anaesthesia was maintained with fentanyl, midazolam, vecuronium. ACT and blood gases were monitored every 20-30 mins to ensure ACT> 480 secs and prevention of hypoxia. During cardiopulmonary bypass the hematocrit was maintained between 20-25%, and lactate levels were closely monitored to assess the adequacy of oxygen delivery. Following surgical repair and prior to separation from bypass, inotropic agents were started and the heart rhythm was evaluated.

Weaning from bypass was facilitated with inotropic support which included milrinone and epinephrine. Milrinone was used to control pulmonary and systemic vascular resistance.

Once the surgical repair and patient hemodynamics were optimal, inj. protamine was administered through peripheral line after giving test dose. Blood products were transfused and trenexamic acid 10 mg/kg was administered to minimise post operative coagulopathy. After 5 mins ACT was repeated and was found 92 sec.

Patient was shifted to ICU in warm blanket on inotropic support and extubated next day.

3. DISCUSSION

Transposition of the great arteries (TGA) is the most common cyanotic congenital heart lesion that presents in neonates. The hallmark of transposition of the great arteries is ventriculoarterial discordance, in which the aorta arises from the morphologic right ventricle and the pulmonary artery arises from the morphologic left ventricle.

This combination of concordant atrioventricular and discordant ventriculo-arterial connections creates parallel circulation systems in which recirculation of oxygenated blood occurs within the pulmonary circuit via the left ventricle and pulmonary trunk while deoxygenated systemic blood recirculates to the body via the right ventricle and aorta that result in systemic cyanosis. Without adequate intra- or extracardiac mixing, between the 2 separate circulations, TGA is uniformly fatal in the infant period, with 30% mortality in the first week of life and 50% within the first month [2]. Atrial septum defect (ASD), patent foramnen ovale (PFO), Ventricular septal defect (VSD), or a patent ductus arteriosus (PDA) may provide mixing between the parallel circuits.

In ASO, the aorta and pulmonary trunks are sectioned and their distal extremities are transposed and anastomosed; coronary arteries are then translocated to the neo-aorta. ASO for TGA-IVS should ideally be performed in neonatal period to prevent involution of left ventricle. Delay in surgical procedure in children with TGA-IVS might lead to failure of low pressure LV to support systemic pressures after the ASO [5,6]. This condition was encountered in our case as the child was 4 months of age with systemic right ventricle so the arterial switch was not feasible. An alternative approaches are required in that case. A repair at atrial level, either by a Mustard or a Senning procedure, is particularly suitable for hearts with an intact ventricular septum. Our patient echocardiography showed intact IVS with OS-ASD (6.5mm) bidirectional shunt which was suitable for Mustard or Senning procedure.
Our case presented with d-TGA-IVS with ASD after neonatal period posing great risk of right ventricular failure and low left ventricular function. Our utmost aim was to maintain cardiac output by maintaining heart rate, contractility, preload and afterload. Systemic arterial desaturation could occur by decrease in cardiac output.

Low PVR in comparison to SVR ensures effective blood flow to lungs and thus intercirculatory mixing. Alterations in Systemic Vascular Resistance (SVR) to Pulmonary Vascular Resistance (PVR) were minimised in order to prevent desaturation of patient especially at the time of induction, by using induction technique causing minimal myocardial depression.

Preserving right ventricular function was also our one of the major concern during pre CardioPulmonary bypass period for which adequate preload and afterload was maintained using vasodilators and monitoring right atrial filling pressure regularly.

Hypoxia, systemic hypoxemia, metabolic acidosis, hypercarbia and sympathetic nervous system stimulation due to light plane of anaesthesia and pain can trigger a rapid rise in PVR, and even a pulmonary hypertensive crisis. A pulmonary hypertensive crisis is characterized by a rapid increase in PVR to the point that the Pulmonary arterial pressure (PAP) exceeds systemic blood pressure, RV ejection fraction acutely drops leading to a decrease in pulmonary blood flow, decreased cardiac output and biventricular failure [7,8]. Strategies to maintain PVR included the use of opioid and sevoflurane in induction, low tidal volumes (6 mL/kg predicted body weight), a high respiratory rate (25/min) and optimal levels of positive end-expiratory pressure (3 cmH_2O), adequate depth of anaesthesia and use of milrinone.

Perioperative use of milrinone is useful in patient with both pulmonary hypertension and RV dysfunction. It increases cardiac output by augmenting contractility while decreasing PVR and SVR. However, with milrinone, reduction in SVR and systemic hypotension remains the rate-limiting factor.

Another concern was prevention of neurological risk associated with circulatory arrest which was addressed by maintaining adequate perfusion pressure, glucose, arterial blood gases. Urine output was maintained 1 to 2 mL/kg/hour. Glucose, potassium, and calcium were measured throughout the bypass period. Glucose was measured to prevent inadvertent effect of hypoglycemia and hyperglycemia. Both situations could provoke neurologic injury and worsen outcome. The potassium in cardioplegia could raise serum potassium levels above normal and affect myocardial conduction and contractility. The infant myocardium is particularly sensitive to ionized calcium levels. Low ionized calcium levels could impair contractility and separation from cardiopulmonary bypass (CPB).

Post bypass coagulopathy could be a particular problem in children with cyanotic cardiac defects that are multifactorial in nature. Antifibrinolytics and ultrafiltration are useful in reducing post bypass bleeding [9,10].

The main complications of this technique are sinus node dysfunction, baffle stenosis, obstruction to either pulmonary or systemic venous return, supraventricular tachyarrythmias, residual interatrial shunt, right ventricular dysfunction and pulmonary vascular obstructive disease.

4. CONCLUSION

Alternative procedures are required if child is reported late for complex corrective operation for TGA requiring long and complex anaesthesia techniques. Alteration in SVR and PVR are to be prevented especially at time of induction and off CPB bypass and cardiac output should be maintained using multiple anaesthetic techniques including the use of opioid and sevoflurane in induction, low tidal volumes with a high respiratory rate and optimal levels of positive end-expiratory pressure, adequate depth of anaesthesia and use of milrinone.

ETHICAL APPROVAL

Institutional ethics committee approval was obtained in accordance with the “Declaration of Helsinki” laid down in 1964.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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