Two Wrongs Make a Right: A Case Report on the Anesthetic Management of a Pediatric Patient with Congenitally Corrected Transposition of Great Arteries*

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ABSTRACT

Congenitally corrected transposition of the great arteries (ccTGA) or ventricular inversion, is a rare form of congenital heart disease (CHD) representing approximately 0.5% of all CHD. It is characterized by atrioventricular and ventriculoarterial discordance, in which the atria are connected to the opposite ventricle, and the ventricles are connected to the incorrect great artery. The defect is termed "corrected" because of the physiologic flow of blood through the body despite the malformation. ccTGA can be associated with other cardiac anomalies like ventricular septal defect (VSD), pulmonary outflow tract (LVOT) obstruction, tricuspid valve lesions, and coronary artery anomalies.

This paper aims to discuss the anesthetic management unique to patients with ccTGA in which the ultimate goal is to prevent hemodynamic instability that could potentially lead to cardiac failure. Here, we report the anesthetic management of a 6 year old child with ccTGA with mild tricuspid regurgitation who underwent plastic repair of cleft lip under general endotracheal tube anesthesia (inhalational). With use of balanced anesthesia to produce minimal to no cardiovascular effects, the operation concluded successfully.

Keywords: congenitally corrected transposition of great arteries, non-cardiac surgery

INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA) is a rare defect described as atrioventricular discordance with ventriculoarterial discordance (double discordance). It occurs in approximately 0.5 to 1.4% of all congenital heart diseases[1].

In this anomaly, there is ventricular inversion and malposition of great vessels (Figure 1). The morphologic right atrium empties into an anatomic left ventricle across the mitral valve, which then contracts into the pulmonary trunk. The morphologic left atrium opens into an anatomic right ventricle across the tricuspid valve, which ejects into the aorta. The aorta is usually oriented in a leftward and anterior position with respect to the pulmonary artery, thus accounting for nomenclature of L-transposition of the great arteries (L-TGA) in this lesion[2].

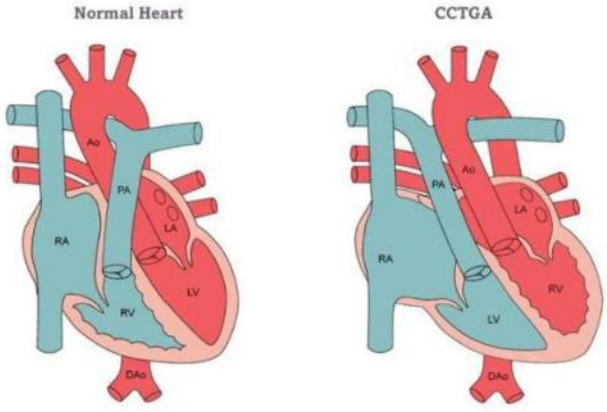

Figure 1. Congenitally Corrected Transposition of Great Arteries3

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The anatomic right ventricle functions as the systemic pump in this lesion. Cyanosis is absent because the circulations are physiologically corrected. However, associated defects are frequently present and these include pulmonary outflow tract obstruction, an interventricular communication, and tricuspid valve (left-sided) abnormalities. If without associated abnormalities, ccTGA may go unnoticed until adolescence or adulthood. This pathology may remain undetected until onset of arrhythmias or syncope is seen which may be attributed to complete heart block or the effects of concomitant defects later in life. Issues that require long term surveillance in children with congenitally corrected transposition include right ventricle (RV) performance and tricuspid valve competency[2]. The inherent vulnerability of RV to failure is due to worsening atrioventricular valve (AV) regurgitation and ischemia because there is only a single main artery supply unlike the morphological left ventricle.

This is a case report of the perioperative anesthetic management of a congenitally corrected TGA who is to undergo cleft lip repair under general anesthesia.

CASE SUMMARY

A 6-year-old female, Filipino, presented to the outpatient department for primary repair of cleft lip. No previous admissions or surgeries undergone. She has no history of seizures, asthma, nor pulmonary tuberculosis. She has no known allergies to food and drugs. Patient had complete childhood immunizations (Hepatitis B, Diphtheria, Tetanus, Polio, Hib, BCG).

Patient was born to a 30 year old GlP0 full term via normal spontaneous delivery with birth weight appropriate for gestational age. At birth, cleft lip, incomplete, right, was noted. There were no feto-maternal complications noted. The mother denies any illnesses during pregnancy. Newborn screening was unremarkable.

Patient is currently a kindergarten student, and active at school. Developmental history is at par with her age. There was no noted poor weight gain and feeding intolerance.

There was no noted familial diseases such as hypertension, diabetes, cancer, PTB. No family history of mental retardation, congenital anomalies nor growth problems.

On physical examination, blood pressure (BP) was 90/60 mmHg, heart rate (HR) was 89 beats per minute, respiratory rate (RR) was 19 breaths per minute, temperature was 36.5 deg C, and body weight was 21.3 kg. Cardiac examination showed adynamic precordium, with normal heart rate, S2>S1, and a grade 1 systolic murmur heard at the 5th left interspace midclavicular line. No peripheral cyanosis or clubbing of the fingers noted. Lung, abdominal and neurological examination was unremarkable.

Patient was then referred to Pediatric Cardiology service for which cardiopulmonary work-up was done. Chest radiograph was unremarkable. Electrocardiogram showed sinus rhythm with a normal PR interval and left axis deviation, while her 2D echo revealed ventricular inversion with associated mild tricuspid regurgitation. Results of other laboratory examinations, such as CBC with platelet count, prothrombin and partial thromboplastin time, electrolytes and kidney function tests, were within normal limits. Hence, patient was classified as American Society of Anesthesiologists (ASA) Class 2 for Ventricular Inversion. The anesthetic plan (GETA), risks, and possible complications were explained, understood and accepted by the patient. Informed consent for anesthesia was obtained from the mother. Patient was maintained on NPO as follows: Intravenous line (gauge 22) established prior to OR and venoclysis done with 5% dextrose in 0.3% sodium chloride at 62-63 cc/hr.

Patient was received in the operating room and standard monitors were attached. Vitals signs were: BP 100/70 mmHg, HR 120 bpm, RR 28 cpm and temperature of 36.5 degrees celsius. Preoxygenation with 100% oxygen via a tight fitting mask for 5 minutes was done.

Induction was started with IV midazolam 2mg (0.1mg/kg) and ketamine 35mg (1.5mg/kg), sevoflurane 3% vol, neuromuscular blockade was

achieved with intubating dose of atracurium 10mg (0.5 mg/kg) IV. Two pumps of lidocaine spray 1.0% (1mg/kg) was also given topically. The patient was then intubated under direct laryngoscopy using MAC3 with endotracheal tube (ETT) 5.0mm ID, secured at level 15cm, and cuff inflated with 1mL of air, taped and secured. Endotracheal placement was confirmed using 5 point auscultation, and capnography. Vital signs were as follows: BP 100/70 mmHg, HR 111bpm, RR 22 cpm, temperature of 36.5 C, end tidal carbon dioxide (EtCO2) of 33. Preventive analgesia with Paracetamol 300 mg (15mg/kg) IV was given while patient was being prepped prior to surgery.

Mechanical ventilation was set at volume assist control. Respiratory frequency was adjusted to achieve a tidal volume (VT) of 140 (7cc/kg) and an intraoperative EtCO2 35–45 mmHg. Sevoflurane was maintained at 2.5% volume in 100% oxygen. Vital signs remained stable all throughout the procedure (BP 90-102/60-68 mmHg, HR 102-106 bpm, SpO2 99-100%), and the surgery lasted for 45 minutes. There was minimal blood loss of about 30 mL, with total of 200 mL crystalloid given and urine output of 30 mL. The patient was extubated awake and follows commands. Suctioning was done with minimal secretions. Vital signs were: BP 100/60 mmHg, HR 110 bpm, RR 23 cpm and temperature of 36.4 C. Patient was then transferred to the post-anesthesia care unit (PACU). For post-operative pain, paracetamol 300mg (15mg/kg) TIV was continued every 8 hours and started with ketorolac 10 mg every 8 hours for 24 hours. She was then shifted to oral paracetamol 300mg every 8 hours for the next 3 days. Her postoperative course was uneventful and was discharged after 1 day. On follow up, 1 week post-operative, the patient had no complaints.

DISCUSSION

Any cardiac patient that is to undergo surgery is always a challenge to the anesthesiologist. These patients usually undergo a relatively long period of preoperative preparation. The most important factor in minimizing the problems of anesthesia itself is the skill and knowledge with which the anesthetic agents are chosen and administered. It consists of careful selection of preanesthetic medication, flawless technique, and use of minimal quantities of anesthetic agents and adjuncts.

The cardiac abnormalities present in the patient must first be taken into consideration.

The main complication of ccTGA affecting patients' function and life expectancy is systemic (morphologic right) ventricular dysfunction resulting from pressure and volume overload.4 The pressure overload occurs as the RV pumps blood against the high systemic pressures, while volume overload can be caused by tricuspid regurgitation and relative myocardial hypoperfusion (supplied with a single (right coronary) artery). The morphologically right ventricle is unable to sustain the demands of a systemic ventricle due to absent helical myocytic arrangement, typically seen in a normal left ventricle, which is important in twisting or torsion during systole5. Hence, any suppression of contractility may induce RV failure. In addition to these, there can also be conduction abnormalities. Every year, around 2% of patients with ccTGA acquire complete AV block and was said to be caused by the displacement of AV node and abnormal course of conduction tissue[6].

In our patient who has ventricular inversion with mild TR, the perioperative hemodynamic goal thus, is to maintain forward flow in the regurgitant valve, prevent arrhythmias and right ventricular dysfunction. To maintain forward systemic flow, the following must be achieved: 1) avoid bradycardia to decrease regurgitation, 2) avoid decrease in preload since low preload results in inadequate cardiac output, 3) contractility should be maintained or increased to decrease the morphologic RV volume. These were achieved by balanced anesthesia with sevoflurane, ketamine (1mg/kg) and atracurium (0.5mg/kg).

A popular drug used in induction is propofol. However for this case, since propofol is a cardio-depressant drug, ketamine was used instead. The advantage of ketamine over propofol is that it has cardio-stimulant properties which make it a useful drug for patients with impaired cardiac function7. It stimulates the cardiovascular system resulting in an increase in heart rate, blood pressure and increase in cardiac

output, mediated principally through the sympathetic nervous system. It has minimal effects on central respiratory drive and produces airway relaxation by acting on bronchial smooth muscles, thus resulting to increased pulmonary blood flow. The increase in pulmonary blood flow then leads to improved oxygenation[8]. All of which are beneficial to our patient.

Volatile anesthetics also have an impact on the cardiovascular system through its effects on the myocardium and on the systemic vascular resistance. Studies suggest that cardiac output during sevoflurane anesthesia, at clinically relevant concentrations, is not substantially decreased, but rather preserved. According to the authors, there is also no evidence for a relevant coronary steal phenomenon in patients given sevoflurane. Sevoflurane is said to act as a weak coronary vasodilator, with coronary blood flow maintained[9].

Despite the reported hemodynamic changes due to release of histamine after administering atracurium, several studies have used atracurium in patients with congenital heart disease [10,11]. Its use is still being recommended given that its dose is decreased[12].

Intraoperative hemodynamics is also an important issue the anesthesiologist must deal with. Hypotension is common post-induction and during maintenance of anesthesia. Hypotension can decrease coronary artery perfusion. In patients having noncardiac surgery, the occurrence of perioperative hypotension was associated with cardiovascular events regardless of the degree of coronary artery disease[13]. Having said, hypotension should be avoided. There are several factors contributing to intraoperative hypotension and preoperative dehydration is one of them. Preload and afterload can be preserved by simple adequate hydration and liquid replacement. Dopamine or vasoconstrictors are helpful in cases of volume depletion and shock[12]. In our case, these were not necessary since there was no volume deterioration. Moreover, since there was adequate diastolic function, liberal IV fluid administration can be done.

Effective pain control must be also achieved to reduce stress, adverse hemodynamics and hypercoagulable states. In our case, aside from preventive analgesia, a topical anesthesia was given prior incision.

CONCLUSION

Any cardiac patient to undergo non-cardiac surgery poses great challenges to the anesthesiologists. The key to success is a thorough evaluation and planning -- from preparation of patient preoperatively, to the use of balanced anesthesia with ketamine, sevoflurane and atracurium in maintenance of hemodynamic stability intraoperatively and finally to the use of multimodal analgesia postoperatively.

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APPENDIX

2D ECHOCARDIOGRAPHY

FINDINGS:

Normal abdominal situs

Apex at the left side of the chest

Atrioventricular discordance and ventriculoarterial discordance

Intact interatrial and interventricular septae

Apically displaced and septophilic atrioventricular valve is attached to the coarsely trabeculated left sided ventricle

Septophobic atrioventricular valve is attached to the finely trabeculated right sided ventricle

Non thickened and well coaptating atrioventricular and semilunar valves

Normal chamber sizes

Good right and left ventricular function

Tricuspid annular plane systolic excursion of 1.73cm

Tricuspid regurgitation, mild TR jet of 12mmHg

Normal color flow doppler across mitral and semilunar valves

Normal sized coronary arteries arising from two different ostia

Normal estimated pulmonary artery pressure by PAT, kindly correlate clinically

Left sided aortic arch

No coarctation of aorta

No patent ductus arteriosus

FINAL INTERPRETATION: Ventricular inversion