CASE REPORT

Anaesthetic considerations in a rare presentation of transposition of great arteries in adulthood with an unusual association of severe valvular aortic stenosis

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Abstract

Transposition of the Great Arteries (TGA) with Intact Ventricular Septum (IVS) is incompatible with life due to systemic circulation of deoxygenated blood. It usually presents in the neonatal period with cyanosis and an arterial switch operation is done within 3 weeks of life. We present a case of TGA with IVS and severe valvular Aortic Stenosis (AS) presenting for the first time in adulthood. The occurrence of valvular AS with dextro-TGA is a rarity and the anaesthetic considerations in such a case have not been reported. The patient survived till adulthood due to the presence of a large atrial septal defect. Worsening of AS leading to heart failure led to the delayed presentation. Understanding the circulatory pathway and pathophysiology of congenital heart disease before and after surgical correction is essential in managing heart failure peri-operatively. Discussing the anaesthetic considerations in such a case is paramount due to multiple unusual circulatory abnormalities.

Keywords: d-TGA, Aortic stenosis, milrinone, atrial switch operation.

Introduction

Transposition of the Great Arteries (TGA) is the most common cyanotic heart disease presenting in neonates [1]. With a male preponderance, the incidence of TGA is 31 per 1,00,000 live births [2]. Dextro-TGA (d-TGA) can present with intact ventricular septum (IVS) in 70% causing cyanosis in the neonatal period or a ventricular septal defect in 25% of patients allowing the mixing of oxygenated blood to be circulated systemically [2]. Our patient presented in adulthood despite IVS with a large Atrial Septal Defect (ASD) and inter-mixed oxygenated blood. Pulmonary stenosis can occur in 5-10% causing Left Ventricular Outflow Tract Obstruction (LVOTO). However, Right Ventricular Outflow Tract Obstruction (RVOTO) is rare and can be in the form of coarctation of the aorta or interrupted aortic arch [2-3].

Valvular Aortic Stenosis (AS) with d-TGA has not been reported. Arterial Switch Operation (ASO) is done in infancy or can be delayed with adequate left ventricular training to correct d-TGA.The delayed presentation and presence of RVOTO in our case precluded this definitive surgery posing multiple challenges.

Case Report

A 22-year-old malnourished male had dyspnea on exertion, orthopnoea and paroxysmal nocturnal dyspnea for 3 months. He had occasional giddiness, chest pain and numbness in the lower extremity. Birth and developmental history were significant for frequent chest infections without any hospitalization. He was stunted, poorly built with peripheral cyanosis and had grade 3 clubbing.

Peripheral pulses were weak, and irregular at 68/min, left upper limb BP measured 110/70 mmHg, with a room air saturation of 89-91%. Grade 2 ejection systolic murmur was heard over the aortic area. Other systemic and airway examination findings were normal. The chest radiograph showed severe cardiomegaly with a right ventricular-type apex. Significant right axis deviation with T inversions in leads II, III, aVF and V1-V6 was seen in the Electrocardiogram (ECG). Blood investigation was significant for polycythemia with haemoglobin of 22 g/dl and elevated liver enzymes. Cardiac Computed Tomography (CT) (Figure 1) and pre-operative echocardiography (Figure 2) revealed d-TGA with Right Ventricular Hypertrophy (RVH) and mild tricuspid regurgitation (TR). A 24 mm ostium secundum ASD with a left-to-right shunt and thickened, sclerosed and calcified aortic valve with severe AS [peak / mean gradient = 85/51 mmHg] was noted. Right coronary artery was arising from the posterior cusp at the 6 o'clock position. In cardiac catheterization, the right and left ventricular (RV and LV) pressures were noted to be 162/8 (17) mmHg and 47/6 (16) mmHg respectively. Pre-operatively, the patient was started on oral furosemide 40 mg and metoprolol 25 mg once daily. On the day of surgery, informed consent and fasting protocols were followed.

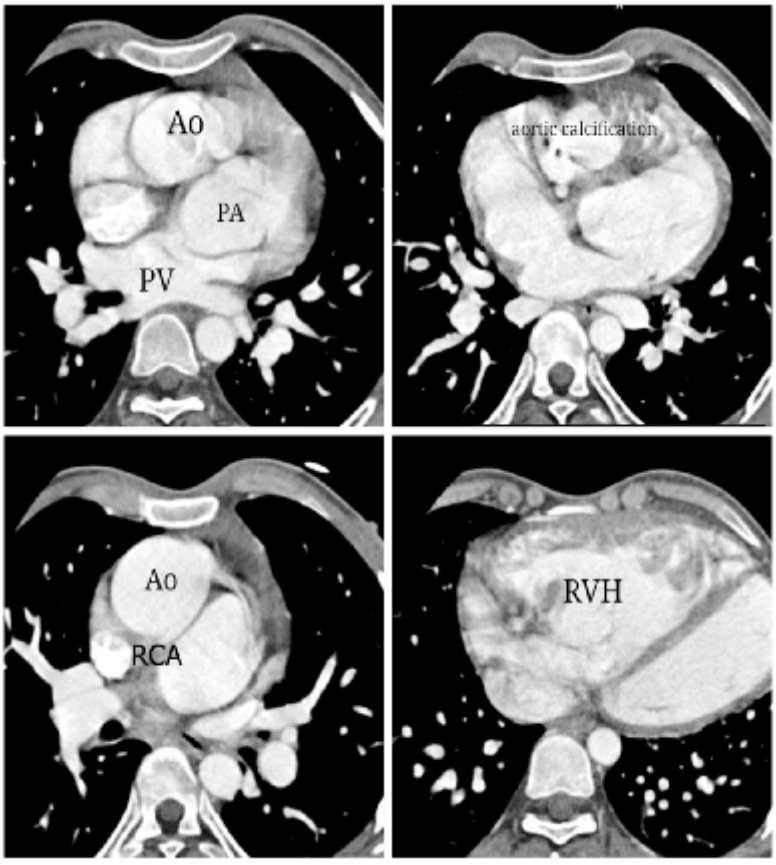

Figure 1: Preoperative cardiac CT showing Aorta (AO) anterior and to the right of Pulmonary Artery (PA), calcified aortic valve, Right Coronary Artery (RCA) arising from the non-coronary cusp at 6'o clock position and Right Ventricular Hypertrophy (RVH)

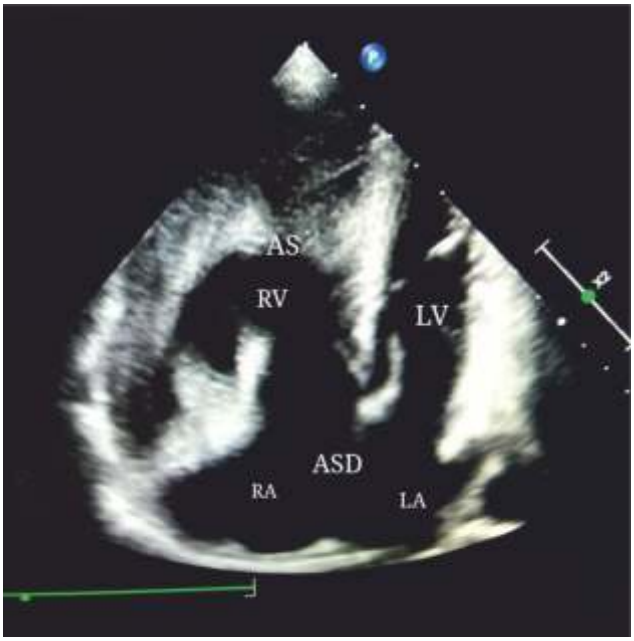

Figure 2: Pre-operative echo showing a large ASD, thickened and calcified aortic valve. The interventricular septum is shifted towards the left ventricle resulting in a banana-shaped LV. Right ventricle appears dilated.

All standard monitors, invasive blood pressure, and cerebral and renal oximetry monitoring were done. Pre-oxygenation improved the baseline saturation to 96-98%. Under local anaesthesia and conscious sedation with midazolam 0.02 mg/kg and fentanyl 2 mcg/kg, the radial artery, Internal Jugular Vein (IJV) and femoral vein were cannulated and venous let down of 350 ml blood was replaced with an equal volume of colloid reducing haemoglobin to 17 g/dl. Induction was with etomidate 0.2 mg/kg and rocuronium 1.2 mg/kg aided the intubation. A nasal temperature probe and transesophageal echocardiography probes were inserted post-induction.

Maintenance was with sevoflurane, fentanyl and vecuronium infusions. Injection dexamethasone 8 mg and injection tranexamic acid 500 mg were given after induction. After adequate heparinisation, at an activated clotting time of 772 seconds, Aorto-Bicavalcardiopulmonary Bypass (CPB) with retrograde cardioplegia and Deep Hypothermic Cardiac Arrest (DHCA) was initiated. An additional dose of thiopentone was given on the pump during low-flow CPB. Hematocrit was maintained between 20-27%. An ambient temperature of 16^0 C and a core body temperature of 22^0 C were maintained.

A modified Senning procedure was done creating a systemic venous-tricuspid-LV chamber and pulmonary venous-mitral-RV chamber. The aortic valve was replaced with a St Jude Medical 21 mm metallic valve. The total CPB time was 5 hours 16 minutes and the aortic cross-clamp time was 3 hours 56 minutes. To facilitate weaning from CPB, adrenaline and noradrenaline infusions were titrated to bring up systolic BP and heart rate.

Milrinone and nitroglycerine were titrated to reduce pulmonary and systemic vascular resistances to accommodate return from CPB. Ventricular pacing leads were also inserted. After the reversal of heparin with an appropriate dose of protamine, the patient was weaned from CPB. Heterologous blood products in the form of single donor platelet, fresh frozen plasma, cryoprecipitate and packed red cells were administered as required.

Post-operatively, the patient was ventilated overnight with inotropes and dexmedetomidine infusions. He was extubated on first post-operative day and pacing leads were removed. Post-operative blood work-up was normal. He was started on tablet warfarin 5 mg and tablet aspirin 75 mg to maintain the patency of the mechanical aortic valve and discharged on the 7^{th} postoperative day.

Discussion

The confluence of valvular AS and d-TGA is a rarity [4]. In addition to the challenges in surgical correction due to delayed presentation, the presence of valvular abnormality in d-TGA made the peri-operative management of heart failure particularly complicated.

The anaesthetic challenges included: management of complications of cyanotic heart disease and polycythemia added upon with adverse effects of DHCA and CPB; peri-operative management of heart failure in a complex circulatory abnormality; and understanding the post-operative circulatory pathway and effects of cardiotomy to wean off from CPB successfully.

Delayed ASO have been described beyond the neonatal period as a single-stage or two-stage operation after training the LV which involves pulmonary artery banding to raise the LV after-load and increase the LV muscle mass. Right ventricular failure with Pulmonary Hypertension (PH) allows the performance of delayed ASO without LV preparation [5]. However, in our patient, PH was due to ASD and right heart failure was due to worsening AS causing RVOTO. The resulting RV dilatation shifted the IVS towards the LV resulting in a D-shaped or banana-shaped LV (Figure 2). While this helped the RV to adapt to the increasing afterload, it deterred LV training, as it would cause further RV dilatation. Hence an atrial switch operation was done with pulmonary artery banding and AV replacement. This created cavo-mitral and pulmonary-tricuspid chambers. The vena cava drained into LV which was pumped into pulmonary arteries. The pulmonary veins drained in RV which remained as the systemic ventricle (Figure 3). Hypercarbia and acidosis should be avoided to prevent increase in right ventricular pressure [6].

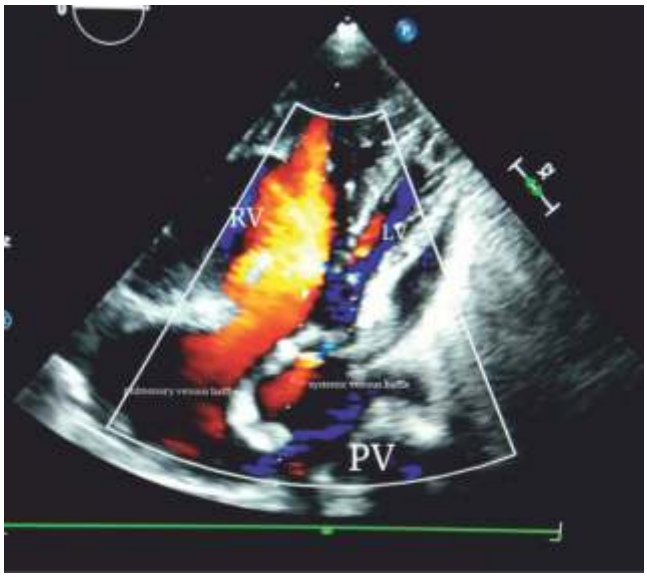

Figure 3: Post-operative echo showing pulmonary venous baffle draining into the Right Ventricle (RV) and systemic venous baffle draining into the left ventricle. The right ventricle is the systemic ventricle pumping blood into the aorta.

Table 1: Pre-operative and post-operative investigations

Investigations	Pre-operative	Post-operative
Hb/TLC/Plt	22/11,700/1,44,000	10.7/13,400/1,27,000
BUN/Serum Creatinine	10/0.8	23/1.1
Serum bilirubin	1.1	2.9
AST/ALT	73/78	183/71
Total protein/albumin	7.6/4.7	5.1/3.2
PT/INR	16.6/1.29	
Na+/ K+	147/3.01	157/4

Hb/TLC/Plt – Hemoglobin/Total Leucocyte Count/Platelets, BUN – Blood Urea Nitrogen, AST/ALT – Aspartate Transaminase/Alanine Transaminase, PT/INR – Prothrombin Time/International Normalised Ratio, Na+/K+ - Sodium/Potassium

DHCA is required in the correction of congenital heart disease. CPB in such cases usually requires a bi-caval venous port and an aortic port. The IJV cannula tip was interrupted by the SVC port and hence was clamped. The femoral vein cannulation helped in venous let-down pre-operatively, maintaining infusions intra-operatively and removed after the surgery. Haemostatic abnormalities associated with DHCA and prolonged CPB were added upon with that of polycythemia due to cyanotic heart disease. Polycythemia can result in depletion of iron reserves due to decompensated erythrocytosis and hyper-viscosity can present as headache, dizziness, visual disturbance and paraesthesias. It can also cause thrombocytopenia, platelet dysfunction, disseminated intravascular coagulation, impaired liver function, decreased production of coagulation factors, vitamin K deficiency and primary fibrinolysis [7]. Our patient had paraesthesia, indicating hyper-viscosity symptoms and hence it was prudent to do a phlebotomy and begin the surgery at a normal haematocrit to avoid further thrombo-embolism. Additionally, intravenous tranexamic acid, a potent antifibrinolytic agent was given before initiation of CPB.

Transfusion with autologous blood from phlebotomy is limited by deficiency of coagulation factors, platelet dysfunction and increased fibrinolysis. The adverse neurological outcomes of DHCA are related to its duration [5-6]. Cerebral protection measures such as an additional dose of thiopentone in CPB, ice packs around the head, surface and core cooling measures were taken during CPB. Near-infrared spectroscopy estimates tissue oxygenation even in non-pulsatile states such as CPB. We used cerebral and renal oximetry to monitor organ perfusion throughout the procedure. Maintaining a mean arterial pressure of 40 mmHg, ensuring adequate urine output and closely monitoring serum lactate levels indicated

adequate organ perfusion during CPB. Prudent phlebotomy and judicious transfusion of heterologous blood products and monitoring of ACT and haematocrit regularly prevented coagulopathy post-operatively.

Milrinone is useful in patients with PH and RV dysfunction. It improves myocardial contractility and reduces Pulmonary Vascular Resistance (PVR). It also reduces Systemic Vascular Resistance (SVR). It is recommended pre-operatively for d-TGA patients with RV failure [7]. However, maintenance of after-load for the systemic Ventricle (RV) was essential to maintain myocardial perfusion in the presence of severe AS pre-operatively in our case. Hence it was not started pre-operatively but was given after the surgical repair to reduce PVR and continued in the post-operative period in a controlled fashion along with inotropic drugs. Hemodynamic considerations in patients with AS include maintaining systolic pressure with preloading to ensure myocardial oxygenation. A target HR of 60-80/min was achieved by pre-operative beta blockers to allow adequate diastolic time and maintain sinus rhythm.

A balance between PVR and SVR was targeted to maintain the baseline arterial saturation and prevent further hypoxemia. Hence etomidate was used as the induction agent of choice. An increase in PVR was avoided intra-operatively by maintaining adequate oxygenation, normocarbia, depth of anaesthesia and preventing acidosis. Intra-atrial reentrant tachycardia and atrial flutter commonly occur after atrial switch operation. Cardiac resynchronisation therapy can improve symptoms of arrhythmias. Pacemakers are considered for symptomatic bradycardia or atrioventricular block [8]. Ventricular pacing leads were inserted as a precaution in our patient. The impact of the ASO on early mortality is excellent. Late complications include systemic RV failure, baffle obstruction and/or leak [10]. Warfarin titrated to prothrombin time-international normalised ratio of at least 2.5 helps to maintain the patency of the mechanical aortic valve.

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