

Hypoxia Due to Unusual Right to Left Shunt after Arterial Switch Surgery

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ABSTRACT

Hypoxia can occur after repair of transposition of great arteries. The most common cause of right to left shunt after arterial switch surgery is related to increased right ventricular pressures and persistent neonatal pulmonary arterial hypertension. We report a case of TGA repair causing right to left shunt with normal right ventricular pressures. Persistence of Eustachian valve with patent foramen ovale (PFO) is the unusual cause of hypoxia and desaturation. The patient was successfully managed by excision of Eustachian valve and closure of PFO.

Keywords: Eustachian valve, hypoxia due to right to left shunting, patent foramen ovale

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INTRODUCTION

The arterial switch operation is most commonly performed to correct transposition of great arteries. Arterial switch operation cases are increasingly more common than Senning's and Mustard procedure. After surgical repair cardiac intensivists are challenged by various peri-operative complications like coronary ischemia, hypoxia,^[1] and desaturation due to pulmonary arterial hypertension,^[2] arrhythmia and Left ventricular dysfunction, neo-pulmonary artery stenosis, aortic stenosis and neo-aortic insufficiency. Hereby we are addressing a case of hypoxia and desaturation in immediate postoperative period with normal right ventricular pressures. Echocardiographic imaging identified that persistence of Eustachian valve^[3] directly shunting through PFO causing desaturation. Patient was successfully managed by excision of Eustachian valve and closure of patent foramen ovale (PFO).

CASE HISTORY

A 2-month-old male baby of weighing 2.5 kg and height 48 cm came to our pre-operative clinic for anesthetic fitness. Child presented with history of cyanosis since birth aggravated by crying and delayed milestones in 2 months of age. His heart rate was 144/min regular, RR: 32 beats per minute and the pulse oximeter sensed the saturation of 77% on room air. Physical examination revealed central cyanosis, cold clammy skin. Bilateral breath sounds were clear. On auscultation ejection systolic murmur was heard on left parasternal area. Echocardiography report revealed D-Transposition of Great Arteries (D-TGA), atrio-ventricular concordance, and ventriculo arterial discordance. Ventricular septal defect of size 10 mm, no Atrial septal defect, no Patent ductus arteriosus, and no Left ventricle outflow obstruction. The morphology of both ventricles were good. Pre-operative investigations were

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within normal limits. Written informed consent was obtained from parents and child was kept nil oral for 4 hrs for breast milk. On arrival to the operation theatre standard monitors were placed including electrocardiogram, pulse oximetry, central venous pressure and invasive arterial blood pressure monitors. Intraoperative and immediate post-cardio pulmonary bypass course was uneventful. Surgeon had no difficulty in inserting IVC cannula. During weaning of cardiopulmonary bypass dobutamine 5 mcg/kg/min and adrenaline 0.1 mcg/kg/min administered. No epicardial echocardiogram was done on immediate postoperative period. Post bypass blood gases were normal. Patient was shifted to cardiac intensive care unit with open chest and saturation of 90% on 100% fractional inspired oxygen (FIO₂). After 5 hrs in immediate postoperative period, child started desaturating from 90% to 75% on 50% FIO₂ inspite of adequate ventilation and compliance. Since chest was open, it was planned to measure right ventricular pressures that causes shunting. Direct measurement revealed normal right ventricular and pulmonary artery pressures. After serial postoperative echocardiogram it was identified that persistence of Eustachian valve [Figure 1 and Video 1] in inferior vena cava is directly draining to right atrium with some amount of shunting taking place across Patent foramen ovale causing desaturation. Child was re-explored again, Eustachian valve was identified and excised and patent foramen ovale was closed. Saturation started improving to 75% to 95% on 50% FIO₂.

DISCUSSION AND SUMMARY

In neonates, most common cyanotic congenital heart disease is transposition of great arteries. Ventriculo

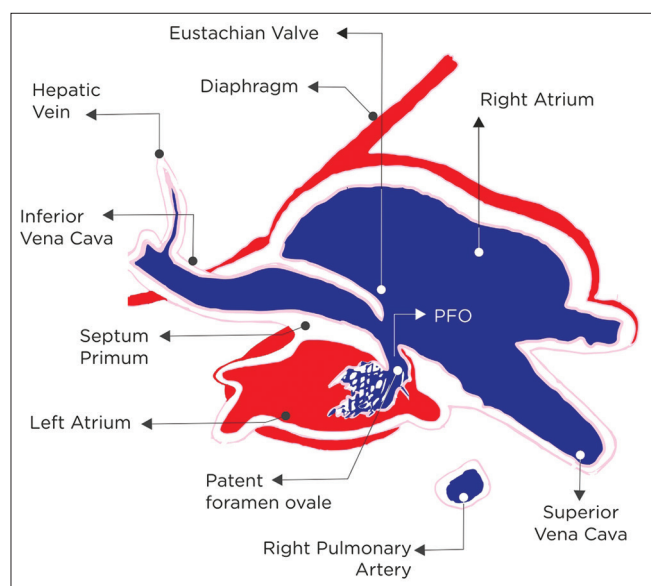


Figure 1: Persistent Eustachian valve in inferior venacava directly draining thru patent foramen ovale (diagrammatic representation of anatomical figure)

arterial discordance is the hallmark of TGA, in which aorta arises from morphological right ventricle and pulmonary artery arises from morphological left ventricle. A parallel circulation is created because of concordant atrio-ventricular and discordant ventriculo arterial connection in which recirculation of oxygenated blood occurs leading to systemic cyanosis. TGA is uniformly fatal in infant period without adequate extra or intra cardiac mixing like Arterial septal defect (ASD), PFO, Ventricular septal defect (VSD), Patent ductus arteriosus (PDA) between two parallel circulation.

Bartolomeo Eustachi, Italian anatomist first described the Eustachian valve. In the embryo, Eustachian valve^[4] is the crescent shaped membrane extending from lower margin of inferior vena cava and ostium of coronary sinus into right atrium towards fossa ovalis. It originates from right horn of sinus venosus. If involution is incomplete, it results in persistence valve of systemic venous valve/giant Eustachian valve or chairi network and divided right atrium.^[5,6,7] At birth after functional closure of foramen ovale, Eustachian valve loses its function reducing to an embryological remnant.

According to the growing evidence, a persistent Eustachian valve is a frequent finding in patients with PFO.^[8] By directing blood from inferior venacavae to inter-atrial septum, it may also prevent spontaneous closure of PFO at birth.

Adults, who presented with platypnoea-orthodeoxia syndrome,^[9] have been reported to have PFO. The platypnoea-orthodeoxia syndrome occurs due to Spinnaker effect.^[10] In upright position aortic root will be anteriorly and inferiorly displaced which leads to floppy inter atrial septum billowing toward left (spinnaker) which in turn would shift inferior vena cava toward right atrium because of increased pressure in right atrium.^[10] Usually cyanosis/desaturation with PFO occurs due to increased right atrial pressures/increased right ventricular pressures/increased pulmonary artery pressures. Almost attributable to increased right ventricular and increased pulmonary artery pressures. However, here we faced hypoxia due to normal right sided pressures. Because of normal right sided pressures there is no definitive tricuspid regurgitation. The other possible causes of hypoxia with normal right ventricular pressures include tricuspid valvular regurgitation shunting through PFO and pulmonary artery to left atrial fistula.^[11]

In our case, we faced the challenge of hypoxia with normal ventricular pressures after Arterial switch operation.

In spite of adequate ventilation and compliances, child started desaturating from 90 to 75%. There is no residual ventricular septal defect. Patent ductus arteriosus also ligated. Since child was in open chest, we measured right ventricular pressures that was normal. After our serial echoes, we found that Eustachian valve draining directly through PFO causing desaturation. Child was taken back to OT, Eustachian valve was excised, and PFO was closed. Following that saturation started improving from 75% to 95%. After 4 days of postoperative ICU stay, child was maintained in 95% saturation in room air. Child was shifted to ward with 95% saturation in room air.

Message

The presence of Eustachian valve directs the IVC blood to PFO into left atrium, and it may cause desaturation. Hence inspection of presence of Eustachian valve and excising it prevents the diversion of IVC blood.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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