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Case Report

Persistence of Eustachian Valve-Rare Cause of Intra Cardiac Shunting Leading to Neonatal Cyanosis

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Abstract

Eustachian valve (EV) is an embryological remnant of the inferior vena cava (IVC) which helps in diverting oxygenated blood from IVC towards foramen ovale during fetal life. It usually regresses after birth however its persistence can lead to complications in the neonatal period. In this case report, we present a newborn with persistence of EV presenting with central cyanosis. Echo cardio graphic examination revealed right-to-left atrial level shunting through patent foramen ovale due to prominent/persistence of EV. The patient was managed conservatively and discharged in stable condition with regular cardiology follow up in clinics

Keywords: *Eustachian Valve; Newborn; Cyanosis; Echocardiogram; Infant*

Introduction

Eustachian valve (EV) is located in the superior portion of the inferior vena cava (IVC) and protrudes into the right atrial cavity. It is considered to be a functional valve in the fetus that helps direct oxygenated blood from the IVC toward the foramen ovale, thereby bypassing the pulmonary circulation [1]. In the early embryonic period, the valve of right horn of systemic venous sinus is prominent within the right atrium. With the normal development of the heart, this structures involutes into rudimentary Eustachian valve between the 9th and 15th week of gestation [2,3]. This Involution may not be complete, resulting in a spectrum of anatomical presentations; (i) Persistent valve of systemic sinus venosus, as a prominent or giant Eustachian valve, (ii) Chiari network representing a relatively more incomplete involution of the embryonic structure presenting in 2 - 4% of population and (iii) divided right atrium or cor triatriatum Dexter with no or minimal involution of valve tissue [2]. There are no strict criteria to distinguish between these anatomical forms, however the redundant tissue extending from Eustachian valve with no attachment to interatrial septum is defined as prominent or giant Eustachian valve. It may present with clinically apparent central cyanosis in neonates with either forms. The Eustachian valve may lead to right to-left atrial level shunting in case of atrial septal defect or patent foramen ovale even in the presence of normal right heart pressures [4-6]. To the best of our knowledge, this is the first clinical case report from Pakistan of a neonate presenting with central cyanosis at birth due to prominent Eustachian valve as specialized cardiology services are not available in all tertiary centers.

Case Report

A baby boy was born via Caesarean section at 38 weeks of gestation to a primigravida mother who received regular antenatal care at a local hospital with a birth weight of 2700 grams. Parents noticed blue discoloration of the skin which increased during crying. Suspecting cyanotic congenital heart disease baby was referred to our center for cardiac evaluation. On admission, his pulse oximetry at room air showed 70% oxygen saturation without any difference between upper and lower extremities. His physical examination revealed a heart rate of 140 per minute, and respiratory rate of 50 per minute without any obvious signs of respiratory distress. The heart sounds were normal with no significant murmur. Capillary refill time was < 3 seconds with regular and normal volume femoral pulses. Complete blood count, C reactive Protein and biochemical parameters were within normal limits. Hyperoxia test was performed which was suggestive of Intra cardiac right to left shunting raising the suspicion of congenital cyanotic heart disease. Chest x-ray was normal. A transthoracic echocardiogram showed elevated RV/PA pressures, more than half of systemic pressures (based on ventricular septal position), suggestive of moderately raised pulmonary pressures along with a persistent Eustachian valve in the right atrium directing blood from inferior vena cava to left atrium through patent foramen ovale resulting in intra-cardiac right to left shunting (Figure 1). It was confirmed by bubble contrast study which also revealed bubbles, directed to left atrium via Patent foramina ovale (PFO) due to persistence of eustachian valve. As moderate pulmonary hypertension cannot explain nearly complete right to left shunting across PFO, so our primary diagnosis was Persistence of Eustachian with pulmonary hypertension being the contributing factor. During the three days of NICU stay, the baby remained stable on breast feeding with no signs of distress. His oxygen saturations were maintained in acceptable range between 75 to 80% on room air so planned to discharge home with regular follow up in cardiology clinic for possible intervention if needed.

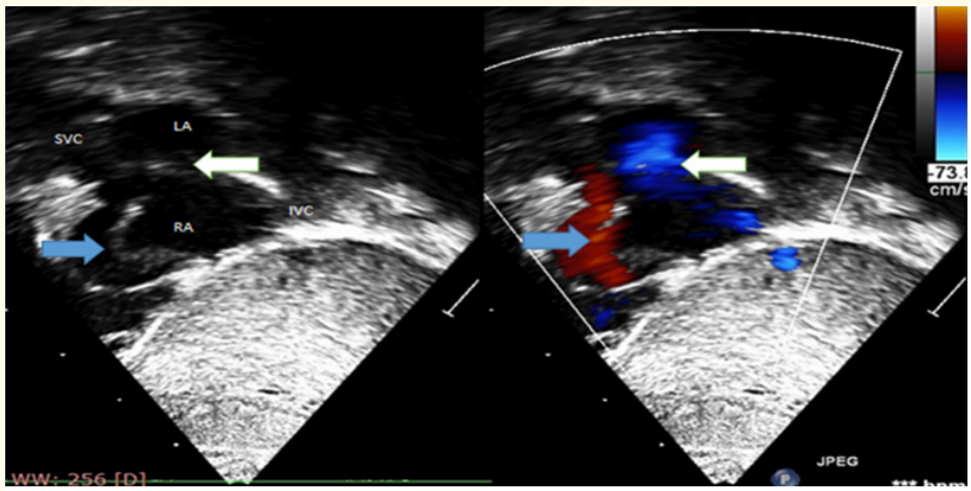

Figure 1: *Subcostal echocardiogram: Bi-caval view showing persistent Eustachian valve (blue arrow), blood coming from lower body (IVC) is shunted through PFO (white arrow) into LA due to persistent Eustachian valve.*

At the time of follow up after 8 weeks, the baby was thriving well on mother feed with no signs of respiratory distress or obvious cyanosis. A follow up echocardiography showed an expected natural reduction in Right ventricular (RV) and Pulmonary artery (PA) pressures. There was a bidirectional shunting at PFO with persistence of EV. The oxygen saturation was 92% on room air, explaining the EV being the major cause of desaturation in this baby. Informed consent was obtained from the parents of the patient to produce this manuscript.

Discussion

We report a case of persistent Eustachian valve leading to transient cyanosis in the neonatal period. Persistent EV can lead to central cyanosis, cardiac arrhythmias, infective endocarditis, thrombosis and paradoxical embolism [2]. Celia., *et al*. reported a case of premature neonate with EV complicated with infective endocarditis [8]. Its management depends on the degree of right ventricular inflow obstruction and cyanosis resulting from right to left interatrial shunting. Surgical resection is the treatment of choice for EV with significant RV inflow obstruction [7]. In case of cyanosis without any obstruction to right ventricular inflow, physiologic decrease in pulmonary vascular resistance in postnatal period may lessen the degree of right-to-left interatrial shunting and improve the oxygen saturation [2]. In literature, some of the described patients needed surgical excision of the redundant tissue and closure of inter-atrial communication resulting in complete disappearance of the cyanosis [6].

In our case right-to-left interatrial shunting was through the patent foramen ovale with no RV inflow obstruction, improvement in oxygen saturation with postnatal physiological decline in pulmonary vascular resistance was expected so surgical intervention was not considered. After proving the absence of duct dependent congenital heart disease, we treated the patient conservatively with supplemental oxygen to reverse hypoxemia with the strategy of watchful waiting, anticipating that the PFO will gradually reduce in size or close physiologically and pulmonary vascular resistance is going to drop progressively. In a case series conducted by Ashraf., *et al*. reported four cases with almost similar presentation and being diagnosed with persistent EV that were managed conservatively, requiring no surgical intervention [9]. Cyanosis in newborns is an urgent condition that requires quick assessment and treatment. The embryologic remnants of the sinus venosus such as prominent Eustachian valve rarely may lead to right to-left shunting resulting in severe cyanosis. Persistent EV is usually a benign finding, but complications may arise that necessitate surgery, and in rare cases can be fatal. This condition was the etiology of cyanosis and O_2 desaturation in our newborn patient. Clinicians should also be aware of the variable presentations and natural history of EV to determine the optimal management for such patients.

Conclusion

Central cyanosis in neonate is a common presentation which is associated with varied etiology. Persistent EV is a rare cause of neonatal central cyanosis which usually resolves spontaneously, but complications may arise that can be fatal requiring surgical intervention. Surgical resection is usually required if patient is symptomatic and associated with RV inflow obstruction. For optimal management of the patients, this is a good example to Pediatric physicians to have a high index of suspicion of intra-cardiac shunting while evaluating a neonate with cyanosis especially in the absence of associated respiratory distress.

Conflict of Interest

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

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