

Severe Persistent Cyanosis in a Newborn Due to Prominent Eustachian Valve

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ABSTRACT

The valves of right horn of systemic venous sinus are prominent structures within the right atrium during early embryonic period. Involution of these structures may not be complete, resulting in a spectrum of anatomical presentations such as cyanosis. A full-term male neonate referred to our hospital for precise evaluation of severe cyanosis on the first day of life. Echocardiographic examination revealed right-to-left interatrial shunting through patent foramen ovale due to prominent eustachian valve with normal estimated right heart pressures from peak tricuspid regurgitation velocity. He was maintained with supplemental oxygen and a PDE-5 inhibitor (sildenafil) and subsequently improvement in oxygen saturation was achieved. The patient was discharged after 2 weeks of treatment. In conclusion; the embryologic remnants of the sinus venosus rarely may lead to right-to-left shunting resulting in severe cyanosis. Pulmonary vasodilators such as sildenafil may improve oxygen saturation in these patients even in case of normal right heart pressures.

Keywords: Cyanosis, Chiari network, Eustachian valve, Newborn, Sildenafil

Introduction

The valves of right horn of systemic venous sinus are prominent structures within the right atrium during early embryonic period. With the normal development of the heart, these structures involute between the 9th and 15th week of gestation and cranially involutes into terminal crest and caudally develops rudimentary eustachian valve and thebesian valve (1). 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 (1). 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. Cyanosis in neonates has been reported with either forms. The eustachian valve may lead to right-to-left interatrial shunting in case of atrial septal defect or patent foramen ovale even in the presence of normal right heart pressures (2-4).

Herein, we report a neonate presenting with severe cyanosis shortly after birth due to prominent eustachian valve in the right atrium that improved after use of sildenafil.

Case presentation

A male infant was born at 37 weeks of gestation to a 28 years old woman by caesarean section because of cord entanglement with a birth weight of 3500 g. Cyanosis was detected shortly after birth and referred to our hospital for precise evaluation. On admission to the hospital pulse oximetry showed 64% oxygen saturation without

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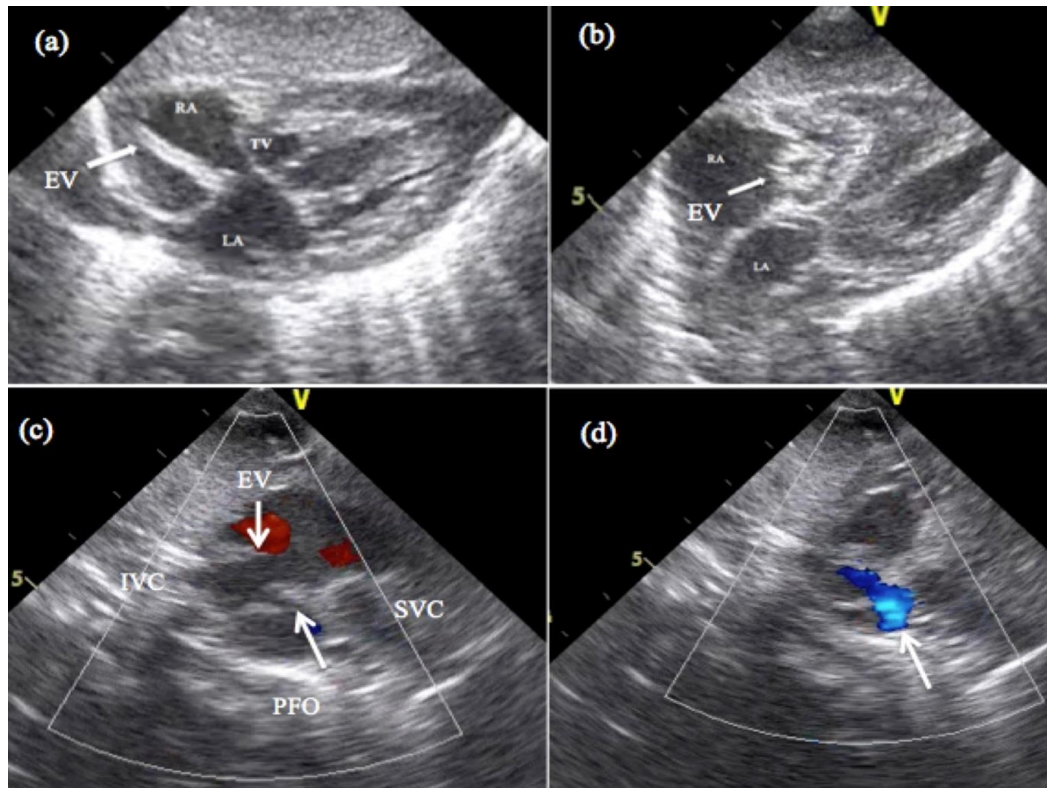


Figure 1. Transthoracic echocardiographic findings of the patient; (a,b) prominent Eustachian valve from the subcostal view, (c,d) subcostal bicaval view demonstrating eustachian valve and right-to-left shunting through foramen ovale (arrow). (EV; Eustachian valve, RA; right atrium, LA; left atrium, TV; tricuspid valve, IVC; inferior vena cava, SVC; superior vena cava, PFO; patent foramen ovale)

any difference between upper and lower extremities. His physical examination revealed a heart rate of 140 per min, and respiratory rate of 50 per min without any signs of respiratory distress. A grade 2/6 systolic murmur best heard at the left sternal border and normal femoral pulses were noted. Complete blood count and biochemical parameters were in normal limits. An arterial blood gas showed normal results except hypoxia. Chest x-ray was normal. Transthoracic echocardiography demonstrated mild tricuspid regurgitation with a peak velocity of 2.8m/s, prominent eustachian valve in the right atrium directing blood from inferior vena cava to left atrium through patent foramen ovale resulting in right-to-left shunting (Figure 1). First he was maintained with low supplemental oxygen (1-2 l/min) and his oxygen saturation increased to 72%. On the second day of life sildenafil (1mg/kg/day) was started to contribute postnatal decrease of pulmonary vascular resistance and thus to decrease the interatrial right-to-left shunting. The oxygen saturation gradually increased to a level of 92% and he was discharged from hospital at 2 weeks old with sildenafil treatment. Subsequently, the

cyanosis gradually disappeared and colour Doppler echocardiography revealed no interatrial shunt flow at 2 months old and then sildenafil was discontinued.

Informed consent was obtained from the parents of the patient to produce this manuscript.

Discussion

During fetal life, eustachian valve contributes to direction of oxygen-rich blood from the inferior vena cava towards the foramen ovale into the systemic circulation and away from the tricuspid valve (1). After birth with closure of foramen ovale its function ends. It may disappear or persist as a thin, filamentous structure. Prominent eustachian valve is essentially a benign tissue in the right atrium, but also its presence as a redundant and prominent valve had been reported to be related with atrial pre-excitation, infective endocarditis, thrombosis, paradoxical embolism (1). As in our case prominent eustachian valve can lead to right-to-left shunting in case of atrial septal defect or patent foramen ovale resulting in cyanosis even in the presence of normal right heart pressures.

The management of persistent valve of systemic venous sinus 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 persistent valve of systemic venous sinus with significant right ventricular inflow obstruction. 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 (1). In the literature the some of the described patients needed surgical excision of the redundant tissue and closure of interatrial communication resulting in complete disappearance of the cyanosis (4). Since right-to-left interatrial shunting was through the patent foramen ovale in our patient, and expectation of improvement in oxygen saturation with postnatal physiological decline in pulmonary vascular resistance, surgical intervention was not considered in our patient. In addition to presence of the redundant eustachian valve, fetal distress due to cord entanglement may be involved in the mechanism responsible for the patient's cyanosis, by adversely affecting the process of circulatory adaptation after birth.

After realizing that absence of ductal dependent congenital heart disease, we treated the patient with supplemental oxygen to reverse hypoxemia resulting in minimal response. However several studies demonstrated that even brief exposures to hyperoxia diminish pulmonary vascular response to endogenous and exogenous NO (5). Sildenafil is a PDE-5 inhibitor producing pulmonary vasodilation by increasing cGMP levels and cGMP response to endogenous or exogenous NO that had been shown to be effective in persistent pulmonary hypertension in newborns (6). Treatment of pulmonary artery smooth muscle cells with sildenafil partially restored the cGMP response to exogenous NO, further highlighting that inhibition of PDE5 activity can counterregulate abnormal vascular cGMP responses after hyperoxia exposure (5). Although the estimated right heart pressures were in normal limits we started sildenafil to facilitate the decline of pulmonary vascular resistance and this

might reverse the effects of hyperoxia. After then we observed progressive improvement of oxygen saturation reaching to levels of 80-85% in 3 days of sildenafil treatment.

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. In some cases, with decreasing pulmonary vascular resistance, during physiologic process or with pulmonary vasodilators such as sildenafil, the oxygen saturation may improve as we observed in our case.

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Conflicts of interests

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