Connection of superior vena cava to left atrium in a newborn followed with unexplained mild cyanosis

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SOUHRN

Abnormální vyústění horní duté žíly do levé síně srdeční představuje vzácnou vrozenou malformaci srdce, která vede v důsledku pravolevého zkratu k rozvoji cyanózy. I když se v takových případech obvykle jedná o izolované propojení horní duté žíly s levou síní, může souviset i s postižením síňového septa typu sinus venosus a s dalšími vrozenými srdečními vadami.
V této kazuistice popisujeme strategii léčby 43denního novorozence, vyšetřeného pro mírnou cyanózu přetrvávající od narození, u něhož byla nakonec stanovena diagnóza perzistující levostranné horní duté žíly, která odvádí krev do levé síně.

Keywords:
Abnormal systemic venous connection
Cyanosis
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ABSTRACT

Abnormal drainage of the superior vena cava into the left atrium is a rare congenital cardiac malformation. It leads to cyanosis due to right-to-left shunt. Although the condition is usually an isolated form of superior vena cava to left atrial connection, it may also be associated with sinus venosus type atrial septal defects and other congenital cardiac lesions.
In this report we present the management strategy of a 43-day-old baby who was investigated for mild cyanosis since birth and was diagnosed with superior vena cava draining into the left atrium.

Introduction

Anomalous systemic venous connection to the left atrium (LA) is an uncommon congenital cardiac pathology leading to right-to-left shunt. The most prevalent variant is persistent left superior vena cava (SVC) which usually occurs approximately in 5% in the population with congenital heart defects.[1] An anomalous right SVC connecting to the LA may also be seen in less than 10% of the cases.[2] Drainage of the SVC commonly manifests with other congenital heart defects such as tetralogy of Fallot, atrial septal defect, ventricular septal defect, single atrium, and various forms of single ventricle pathologies. The patients with isolated SVC-LA connection often present with unexplained cyanosis. Surgical correction is mandatory, otherwise complications related with chronic hypoxemia, paradoxical embolism, and brain abscess may develop. Kirsch et al. were the first to report SVC-LA connection in the literature.[2]

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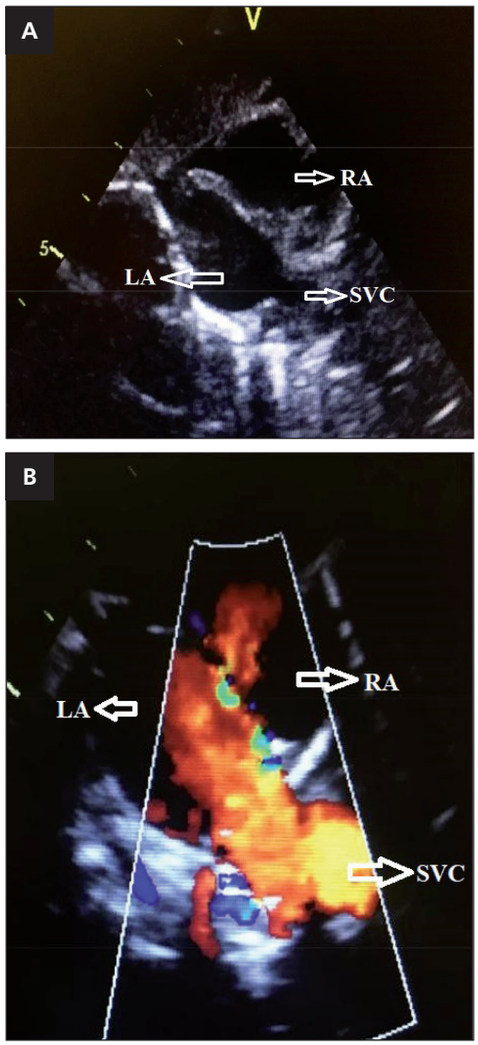
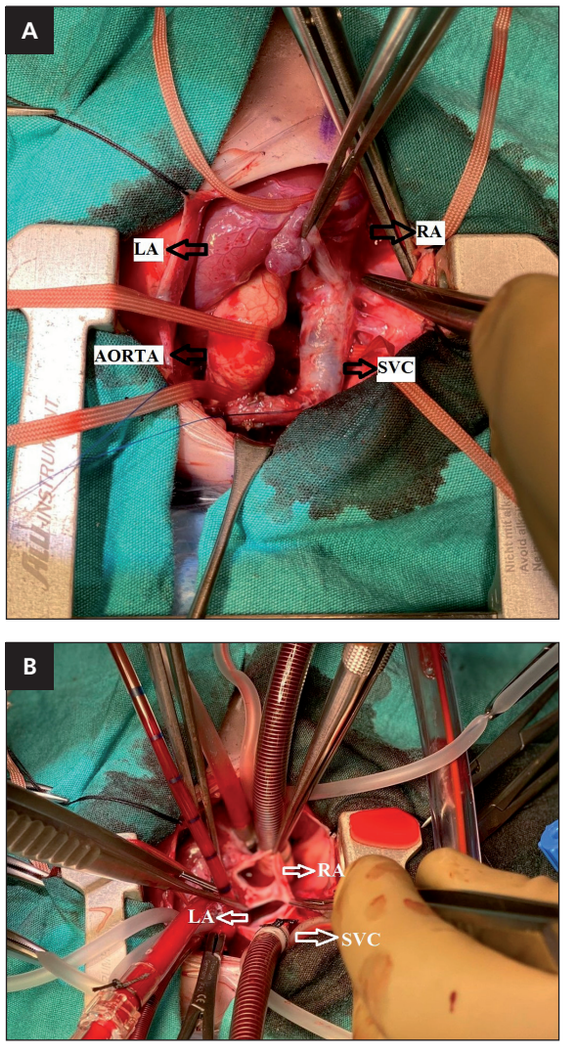


Figs 1 – (A, B) Echocardiographic view of the superior vena cava – left atrium connection. LA – left atrium; RA – right atrium; SVC – superior vena cava.



Figs 2 – (A) Perioperative view of the superior vena cava – left atrium connection. (B) Additionally we see the right and left atrium. LA – left atrium; RA – right atrium; SVC – superior vena cava.

In this case report we present our management strategy in a newborn who had mild cyanosis since birth and was diagnosed with right SVC-LA connection.

Case report

A male newborn, weighing 3320 g and measuring 48 cm, with five minutes Apgar score of 7/10 was delivered via cesarean section at 34+2 weeks. The baby was admitted to the neonatal intensive care unit (NICU) 6 hours after birth due to desaturation. Physical examination at NICU admission revealed oxygen saturation between 90–93%, otherwise completely normal. A slight increase up to 96% was observed in saturation with nasal oxygen support. Chest X-ray was not remarkable. A transthoracic echocardiography examination revealed an insignificant patent ductus arteriosus and a very small sinus venosus type arterial septal defect (ASD) (**Fig. 1**). The baby was thoroughly investigated and considered to have central hypoventilation syndrome due to shallow breathing and further desaturation especially during sleep. He was also neurologically and metabolically tested; however, anything significant could not be found. The polysomnography on the 19th day of NICU admission indicated severe sleep apnea. Non-invasive respiratory support of nasal BIPAP was started with high level of oxygen. However, arterial blood gas analysis indicated decreased partial oxygen pressure and saturation despite 100% oxygen inhalation, suggestive of a congenital cardiac defect. After prolonged NICU stay complicated with desaturation attacks and alimentation problems, a contrast echocardiography on the 40th day of NICU admission using agitated saline injection into a venous line from an antecubital vein was decided. Agitated saline appeared inside the LA and left ventricle, but not inside the right side of the heart, suggestive of a right SVC-LA drainage. Pathology was confirmed with contrasted computerized tomography of the chest (**Figs 2A, 2B**).

Despite the pros and cons of operating on a delicate neonate at a very young age, since he experienced deep

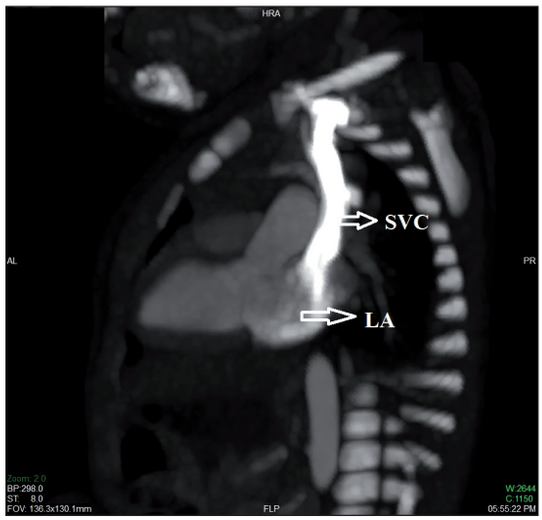


Fig. 3 – Contrasted computerized tomography indicating superior vena cava – left atrium connection. LA – left atrium; SVC – superior vena cava.

saturation decrease attacks especially during sleep, an early surgery was decided by the multidisciplinary team comprised of neonatologist, pediatric cardiologist, and cardiovascular surgeons. We operated on the baby on the 43rd day of life after explaining in details the risks and benefits and the consent of the family. Operation was performed through median sternotomy, with cardiopulmonary bypass, and cardiac arrest. The SVC draining into the LA was clearly identified with right atriotomy through a small superior sinus venosus type ASD (**Fig. 3**). The defect was enlarged incising the septum towards the foramen ovale and re-closed with an autologous fresh pericardial patch draining the SVC into the right atrium. Operation was finalized successfully and patient could be weaned off mechanical ventilator after 12 hours after the operation. Postoperative course was uneventful and desaturation was not observed. Patient was discharged home after 8 days from the NICU with nasal BIPAP respiratory support to be used especially during sleep.

So far, no problems have been detected since the discharge of the patient during more than 18 months follow-up. He has been active with normal growth and parents reported they no longer used respiratory support.

Discussion

Systemic venous return anomalies are rare disorders and among the reasons for desaturation in the newborns. According to the reviews of Mayo Clinic, SVC draining into the LA was detected in approximately 0.5% of patients with congenital cardiac cases and only 19 patients in their cohort had right SVC draining into the LA.[2] The hemodynamic effects of anomalous systemic venous drainage mimics lesions with right to left shunt. In addition, since the SVC carries quite a significant amount of blood into the heart during the newborn period, signs and symptoms of left-sided volume overload may occur. Brain abscess and paradoxical embolization are possible life-threatening conditions, especially in chronically hypoxic patients.[3]

Drainage anomalies of the SVC commonly manifest together with atrial and ventricular septal defects, coarctation of the aorta, pulmonary arteriovenous fistulae, patent ductus arteriosus, and abnormalities of the inferior vena cava. The patients with isolated SVC-LA related condition often present to the Department of Pediatric Cardiology with unexplained cyanosis, since congenital heart diseases are among the most common causes of desaturation.[3] Van Praagh et al.[4] reported multiple cases of sinus venosus defects from echocardiographic and postmortem anatomic findings. They reported co-occurrence of SVC type sinus venosus defects and connection between right SVC and LA.[4] A SVC type sinus venosus defect results from a deficiency in the wall shared by the right SVC and the right upper pulmonary veins. The formation of SVC to LA anomaly occurs due to an amalgamation of predominant blood flow between those structures and atresia that affects the right atrial orifice of the SVC.[4] Current understanding suggests that SVC to LA is also part of the spectrum of superior sinus venosus defect and the embryonic abnormality result from the commitment of SVC to LA while the sinus venosus defect is closed in fetal life.[5] In our case, we detected a small sinus venosus type ASD in addition to mainly directed SVC flow into the LA.

Connection of SVC to the LA as an isolated or co-existent congenital heart disease has been reported in all age groups.[4,6–8] Hammouada et al.[6] reported right SVC-LA drainage in a 17-day-old neonate presenting with cyanosis, and Rakha et al.[7] detected right SVC-LA drainage in an 11-month-old girl who had cyanosis while crying since birth. Similarly, Moradian et al.[8] detected abnormal right SVC-LA connection in an 8-year-old girl who presented with unexplained cyanosis.[8] Bagget et al.2 reported a 34-year-old hypoxic female patient in the peripartum period and had a transition between right SVC-LA. As can be seen in the literature, diagnosis of SVC-LA connection in the early neonatal period may not be easy. Since the pathology is associated with persistent central cyanosis a wide range of disorders including metabolic, neurologic, infectious, hematologic, and pulmonary problems other than cardiac pathologies should be reminded.[9] On the other hand it should be kept in mind that congenital cardiac disorders are the most common causes of persistent arterial desaturation in neonates. In our particular patient, since we failed to explain the reason of cyanosis with initial echocardiographies which only indicated a small sinus venosus type ASD, we searched for metabolic, neurologic, hematologic and pulmonary disorders. Polysomnography indicated sleep apnea syndrome; however, cyanosis persisted despite appropriate treatment.

In grown up patients, SVC-LA drainage may be detected during the investigation of clubbing of digits and shortness of breath, paraxial embolism, and stroke in adolescence and early adulthood.[4] Some other findings in echocardiography are also helpful in identifying venous anomalies such as unexplained dilatation of the left chambers of the heart; however, careful imaging studies are required for a definitive diagnosis. The diagnosis was generally made with saline contrast echocardiography which is an inexpensive technique[10] and with addi-

tional radiologic measures such as thoracic computerized tomography angiography or magnetic resonance angiography which were preferred in various cases reported in the literature. In our case, we diagnosed the pathology with contrast echocardiography and confirmed with contrasted thoracic computerized tomography. Similar to the literature, our case was diagnosed with the regular diagnostic techniques while investigating the reason of cyanosis very early in the neonatal period.

Regardless of clinical presentation, surgical correction is indicated once the diagnosis of systemic venous connection to the LA is determined.[11] In our case, we performed surgical correction when he was 40 days old as soon as he was diagnosed with the condition despite the pros and cons on operating on a fragile neonate. The corrective procedure was performed by enlarging the atrial septal defect and re-routing the SVC and RA. This report highlights the importance of investigating SVC-LA connection by contrast echocardiography in patients with or without sinus venosus type atrial septal defects and the presence of central apnea. Central cyanosis and sleep apnea immediately improved after surgical correction of right SVC-LA connection.

Conflict of interest
None.

Funding
None.

Ethical statement
The study was conducted in accordance with the principles of the Declaration of Helsinki.

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