CASE REPORTS

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Partial abnormal drainage of superior and inferior caval veins into the left atrium: two case reports

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Abstract

Abnormal connection of the right superior caval vein to the left atrium is an uncommon systemic vein drainage anomaly, with only a few cases reported among congenital heart disease (CHD), around 20 cases published in the medical literature. The inferior vena cava connection with the left atrium, also very rare, can appear directly or in heterotaxy. Clinical suspicion arises due to the presence of cyanosis in the absence of other specific clinical signs (without other associated CHD). We present the cases of two children with abnormal superior and inferior systemic venous return. The first case is an abnormal connection of right superior vena cava to the left atrium associated with persistent left superior vena cava draining into the right atrium through the coronary sinus. The second case is an interruption of the inferior vena cava with hemiazygos continuation, drained into the left superior vena cava, which drained into the left atrium. The diagnosis was imagistic – echocardiography and angiography. Surgical treatment solutions vary from one case to another, usually following anatomic correction. Hypoxia accompanied by cyanosis must bring into question the pathology of systemic venous drainage anomaly, after other common causes have been excluded. Surgery is indicated in all cases due to the risk associated with the presence of right-to-left shunt.

Keywords: abnormal, systemic venous return, left superior vena cava.

Introduction

Systemic venous return anomalies occupy a special place in the congenital heart disease (CHD), because they are heterogeneous and very rare, with a prevalence of approximately 0.5% of all congenital heart diseases [1]. From all these 0.5% of all congenital heart diseases, the most frequent is the persistence of the left superior caval vein with drainage either into the coronary sinus or directly into the left atrium (the second being a form of CHD associated with central cyanosis due to the right-to-left shunt). Abnormal connection of the right superior caval vein to the left atrium is an uncommon systemic vein drainage anomaly, with only a few cases (around 20 cases) reported among congenital heart disease [2–8]. The inferior vena cava connection with the left atrium, also very rare, can appear directly or in heterotaxy [9]. They may be associated with other cardiac abnormalities and rarely can be met isolated [2].

The clinical appearance is variable because, depending on associated anomalies, may coexist with normal or corrected physiology or with important right-to-left shunt, and significant cyanosis. Diagnosis can be made by echocardiography or other imaging methods: cardiac catheterization, contrast-enhanced computed tomography (CT) or magnetic resonance imaging (MRI) angiography.

Aim

We present two cases: one with right superior vena cava (RSVC) drainage in the left atrium, associated with left superior vena cava (LSVC) persistence, which drains in the coronary sinus and then in the right atrium. The second case presents inferior vena cava (IVC) flow drainage to the left atrium, through hemiazygos continuation, and persistence LSVC. The importance of these cases rely on identifying them as a cause of central cyanosis, especially in the absence of other cardiac anomalies, and avoid in this way appearance of right-to-left shunt consequences (septic or trombotic embolization, desaturation and dyspnea).

Case presentations

Case No. 1

A 30-month-old female child with situs solitus viscero-atrialis, right vena cava connected to the left atrium, persistent LSVC connected to the right atrium through a dilated coronary sinus, normal connection of the pulmonary veins to the left atrium, persistent foramen ovale, mild enlargement of the left ventricle, and normal the subsequential heart anatomy, presented with a medical history of chronic systemic desaturation (90–92%). The patient was asymptomatic and had a normal somatic development. There was no family history of congenital heart disease. Her biological parameters (weight 11.6 kg, height 93 cm) and vital signs (blood pressure 90/52 mmHg, heart rate 122 beats/min.) were normal, without murmur on cardiac examination. The laboratory findings and electrocardiography were normal. On chest radiography, there was an enlarged cardiac silhouette.

To confirm the echocardiographic diagnosis (Figures 1 and 2) and assess the hemodynamic significance, both right and left cardiac catheterizations were performed. The right cardiac catheterization was done after heparinization (100 IU/kg) by two vascular access sites: right internal jugular vein (4 Fr) and the right femoral vein (4 Fr). We measured pressures in the pulmonary and systemic circulation, with consensual measurement of saturation, Qp:Qs = 0.8:1. A normal pressure regime was found in the pulmonary circulation.

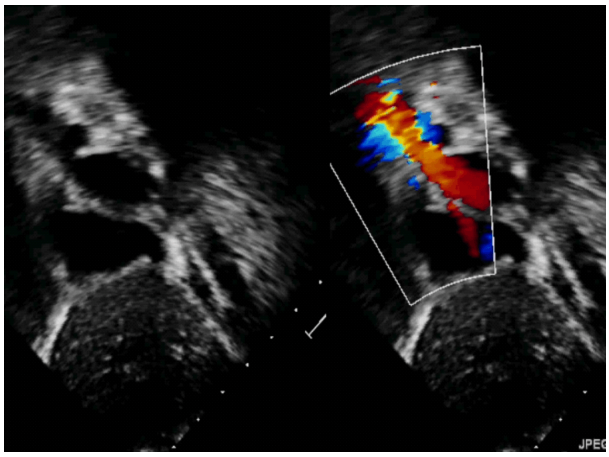

Figure 1 – ***Right superior vena cava connected to the left atrium, in subcostal bicaval image.***

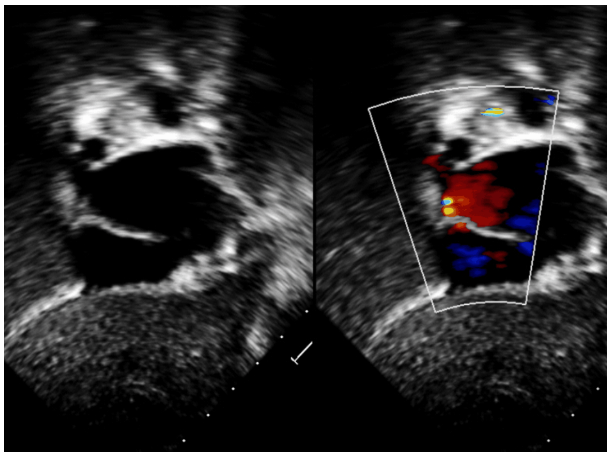

Figure 2 – ***Dilated coronary sinus – suggestive, in this case, for persistence of left superior vena cava draining in coronary sinus, in subcostal two chambers image.***

The RSVC angiography showed a normal dimension right superior vena cava draining into the left atrium. Communication with the right atrium through the permeable foramen ovale. The selective LSVC angiography was possible through the dilated coronary sinus and showed no communication with the right caval vein. The pulmonary artery angiography showed a normal pulmonary vascular bed, a normal pulmonary venous return, and excluded arterio-venous pulmonary fistula. No evidence of right heart opacified structures during the pulmonary artery angiography (Figures 3 and 4).

Although the patient is asymptomatic and the age is still relatively small, surgical correction is indicated because the risks of brain abscess and paradoxical embolization are high.

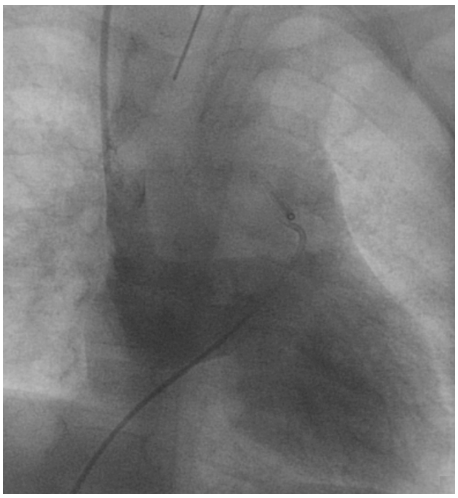

Figure 3 – ***Right superior vena cava (RSVC) angiography (right jugular vein 4 Fr access) visualizing RSVC connected to the left atrium, which is connected to the left ventricle.***

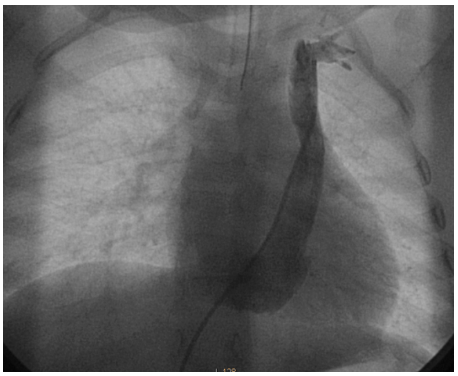

Figure 4 – ***Persistence of left superior vena cava draining in the right atrium, through the dilated coronary sinus.***

Case No. 2

A 40-day-old male infant with situs solitus visceralis, left atrial isomerism, interruption of the inferior vena cava with hemiazygos continuation to the left superior vena cava connected to the left sided atrium, normal drainage of the pulmonary artery (to the left sided atrium), large atrial septum defect, double outlet left ventricle, large perimembranous ventricular septal defect, normally related great arteries presented with cyanosis, and mild respiratory distress. The child had a prenatal diagnosis of double outlet left ventricle (DORV) and was born by Caesarean section, weighing 2.5 kg. No family history of congenital heart disease was reported. Somatic parameters (weight 4050 g, height 56 cm) and vital signs (blood pressure 80/43 mmHg, heart rate 170 beats/min.) were normal, saturation 88–90%, with a 3/6 systolic murmur mid-sternal on cardiac examination. The laboratory findings were normal. The postnatal echocardiographic diagnosis was confirmed on angiography.

The inferior vena cava angiography showed interruption of the IVC with hemiazygos continuation draining in persistent LSVC, which ended in the left sided atrium. Opacified right cavities were evident through the large atrial and ventricular septal defects (Figures 5 and 6).

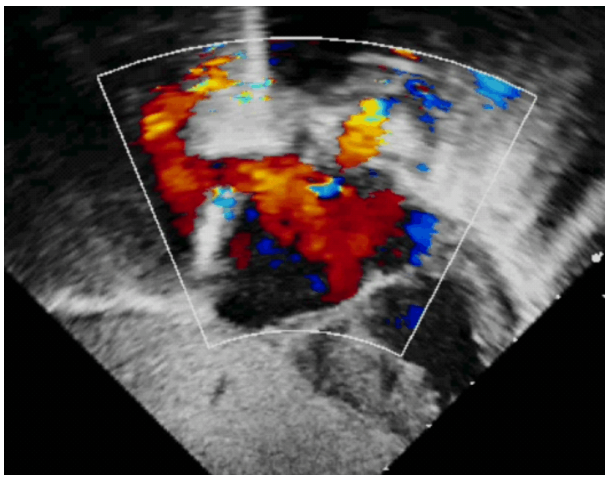

Figure 5 – ***Persistent left superior vena cava receiving the hemiazygos vein and draining in the left atrium.***

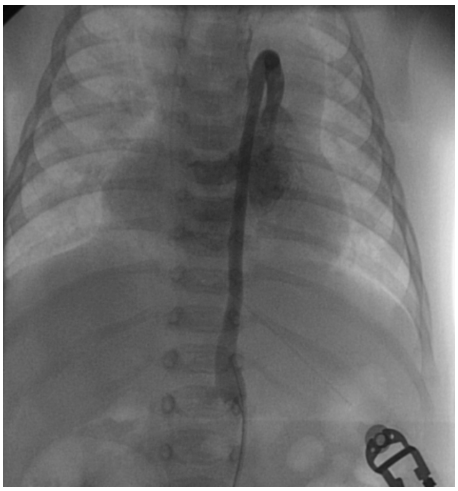

Figure 6 – ***Hemiazygos continuation of the IVC, draining into the left superior vena cava, which drains directly in the left atrium.***

This child underwent a single stage of surgical repair at our institution, by redirecting the flow of the left superior vena cava to the right atrium using a heterologous pericardium patch, atrial septation, and tunneling of the ventricular outflow with heterologous pericardium patch closure. The surgery was complicated by the occurrence of junctional tachycardia, which was treated with amiodarone. Subsequently, pacing was necessary for bradycardia occurrence.

Discussion

Abnormal connection of the right superior caval vein to the left atrium is an uncommon systemic vein drainage anomaly, with only few cases reported among congenital heart disease (CHD). In 1975, de Leval *et al.* reported it, around 0.5% in all CHD, including the persistence of the LSVC [1]. Venous return anomalies in the superior caval system include: persistent left superior vena cava (LSVC) (with drainage in the coronary sinus or left atrium), the anomaly of the right superior vena cava drainage (RSVC) in the left atrium, or their combination (RSVC to the left atrium and LSVC to the right atrium through the coronary sinus). Anomalies of the inferior caval system usually are met in heterotaxy, but direct connection of the IVC to the left atrium is also described. In heterotaxy syndrome, the persistence of the left superior vena cava is found in 30–50% of the cases. The persistent LSVC rarely drains directly to the left atrium, situation that is followed by the necessity to separate the two venous returns, systemic and pulmonary by an interatrial baffle [9, 10].

In terms of embryological development, this anomaly is not yet fully understood [11–14]. There are some hypotheses for the RSVC connected to the left atrium. One possibility about malposition of the right horn of the sinus venosus in a left cephalic position, regarding the right superior vena cava anomaly in the left atrium has been described [15]. Another hypothesis was developed by Van Praagh *et al.* and supports the superior vena cava (SVC) orifice atresia associated with a sinus venosus atrial defect [16].

In the absence of other significant cardiac abnormalities, patients are asymptomatic [17] and the diagnosis can be made even in adulthood [18]. For instance, in our first reported case we found the Qp:Qs ratio equal to 0.8:1, without a real clinical significance, expressing only the systemic desaturation.

Clinical suspicion arises due to the presence of cyanosis in the absence of other specific clinical signs (without other associated CHD). Hypoxia causes are numerous. In a cyanotic patient, abnormal systemic venous drainage should be considered after frequent causes were excluded (primary lung disease, pulmonary embolism, cardiac malformation with right-left shunt, arteriovenous malformations, hemoglobinopathies or methemoglobinemia) [2]. Dyspnea, cardiomegaly, cerebral embolism or brain abscess may also be present in this spectrum of disease [14].

Cardiac abnormalities are commonly associated with this pathology including atrial septal defect, ventricular septal defect, single atrium, single ventricle, Eisenmenger's syndrome, tetralogy of Fallot, transposition of the great vessels. Frequently associated extracardiac anomalies are coarctation of the aorta, pulmonary arterial-venous fistula, abnormal ductus arteriosus permeable and inferior vena cava [2], and abnormal pulmonary vein drainage [14, 19].

Diagnosis involves all imaging techniques, from the simplest [20], echocardiography and contrast echocardiography with injection of the left arm [21] in persistent LSVC or to the right arm [22] with RSVC draining in the left atrium, cardiac MRI [2] or contrast-enhanced CT [23] to the most invasive cardiac catheterization, as a confirmatory test [2]. Many times, echocardiography is limited by the poor acoustic window or by operator variability.

The clinical significance of these anomalies relies on the presence of the right-to-left shunt. The right-to-left shunt increase the risk for cerebral embolism or brain abscess. Out of the cases reported in the literature, 20% were complicated by brain abscess [2]. Therefore, surgical correction is indicated even in asymptomatic cases. Surgical technique is adapted to each case. For the RSVC connected to the left atrium several techniques have been described. Clamping RSVC and its connection to the right atrium is the most common. Bidirectional Glenn may be recommended when there is right ventricular hypoplasia [14]. For the IVC interruption with hemiazygos continuation in

persistent LSVC draining in the left atrium, the surgery is more complex. Usually, this anomaly is associated with many other cardiac defects. The used techniques followed the principle of systemic and pulmonary venous return separation, with atrium septation, if it is necessary, and intra-atrial baffle redirection of the systemic flow. The left atrium appendage can be used for the baffle procedure [9].

Conclusions

Hypoxia accompanied by cyanosis must bring into question the pathology of systemic venous drainage anomaly, after other common causes have been excluded. Echocardiography and contrast echocardiography from both arms and leg should be performed when there is a diagnostic doubt. Cardiac catheterization may help to define the precise anatomy. Surgery is indicated in all cases due to the risk associated with the presence of right-to-left shunt.

Conflict of interests

The authors declare that they have no conflict of interests.

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