Cardiology in the Young

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Anomalous systemic venous connection to left atrium in a heart with usual atrial arrangement

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Brief Report

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Abstract

Anomalous systemic venous connection to left atrium is rare anomaly. Previously published cases described this anatomy in patients with left isomerism. Depending on the size of the atrial septal defect, patients usually present with varying degrees of cyanosis and right heart hypoplasia. Here, we report a case of anomalous systemic venous connection to left atrium in a newborn with the usual atrial arrangement.

Case report

The baby was born to a 36-year-old healthy mother. Fetal echocardiogram was done in the context of a previous child with atrioventricular septal defect. Fetal echocardiography showed dilated left ventricle and bowing of the interatrial septum to the right instead of left and mild

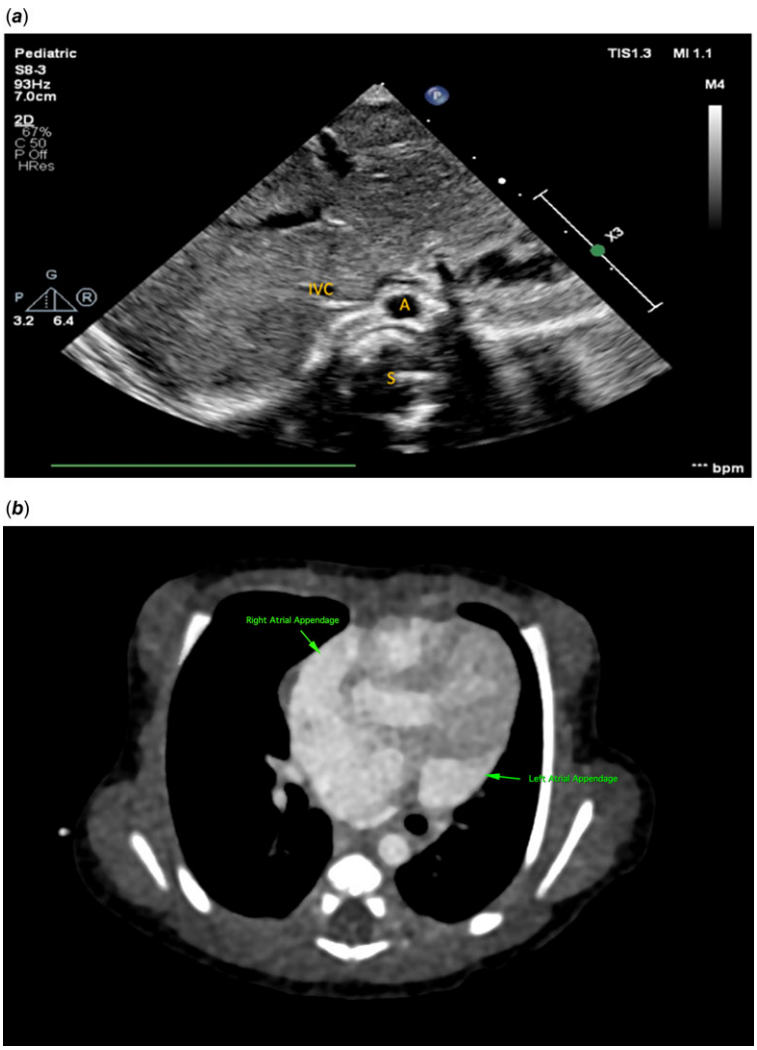

Figure 1. (***a***) Transthoracic subcostal echocardiography showing normal situs solitus. A: aorta, S: spine, IVC: inferior vena cava. (***b***) Chest computerized tomography in cross section showing the right and left atrial appendanges. (***c***) Chest computerized tomography in coronal section showing the bronchial arrangment. (***d***) Transthoracic echocardiogram showing bicaval and hepatic vein connections to left atrium. SVC: superior vena cava, LA: left atrium, RA: right atrium, HV: hepatic veins, IVC: inferior vena cava.

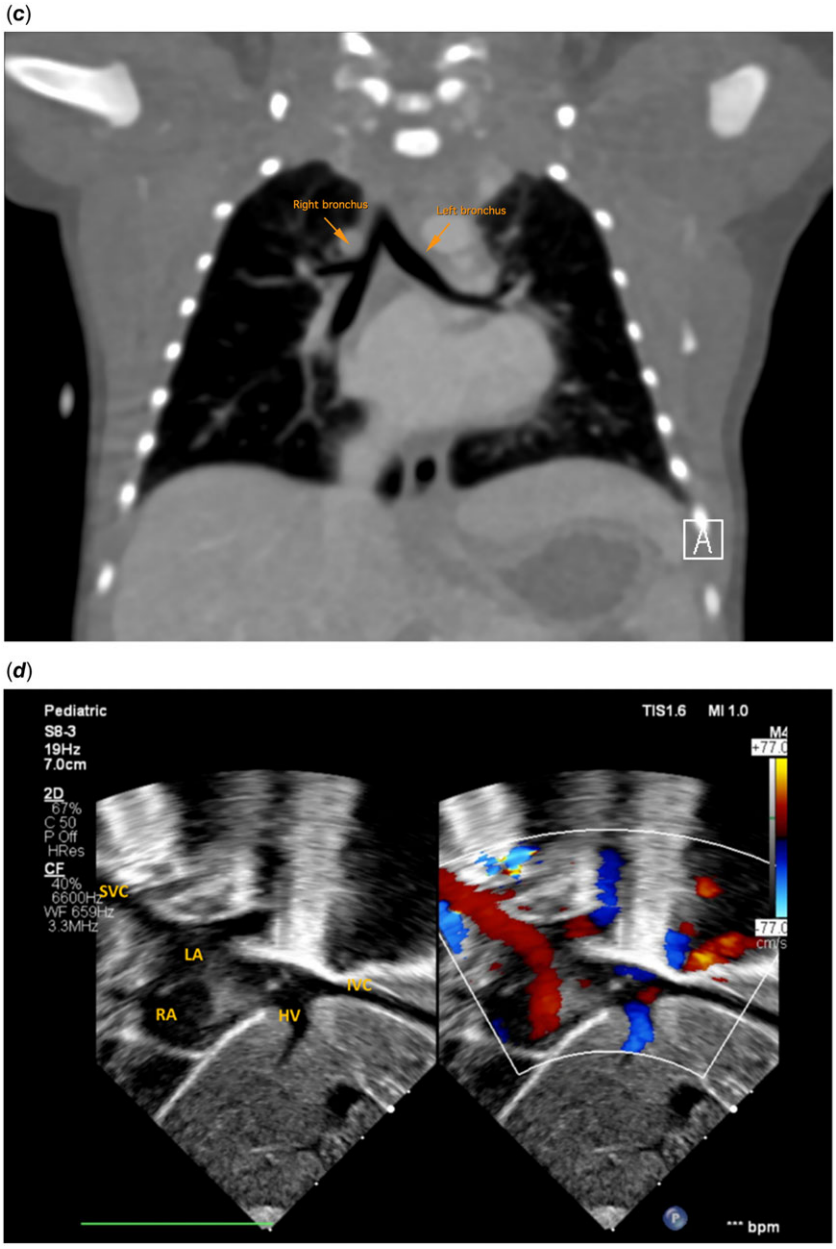

Figure 1. (Continued).

hypoplasia of right ventricle. The baby was born at 40 weeks of gestation with a birth weight of 2.6 kg. He was noted to have cyanosis with a saturation of 88 % in room air. ECG showed low right atrial rhythm at 157 bpm. Echocardiogram (Fig 1a and 1d) showed situs solitus heart with anomalous connection of superior, inferior caval veins along with hepatic veins to left atrium. The left ventricle was dilated (Left ventricle end diastolic dimension Z score 2.8), and there was mild right heart hypoplasia (tricuspid valve Z score – 2.1).

The external morphology of the atrial appendages and the arrangement of bronchial morphology on computerised tomography scan were indicative of normal atrial arrangement (Fig 1b and 1c).

Due to a restricted atrial septal defect, the patient underwent balloon atrial septostomy. Angiography in the inferior vena cava confirmed the diagnosis (Fig 2a and 2b). The septostomy was done using 13.5 mm Z-5™ atrioseptostomy balloon (NuMed) through 6F femoral venous sheath. Using 0.014" balance middleweight coronary wire (Abbot), the atrial septum was crossed from left atrium to right atrium then a reverse septostomy was done with a pullback from right to left atrium. The septostomy was done to ensure adequate systemic blood flow to the lungs until the planned definitive surgery is performed at 6 months of age. During follow-up at 4 months of life, patient saturation was 88% and was well and growing.

Discussion

Anomalous systemic venous connection to left atrium is rare anomaly rarely reported in fetal and postnatal life.[1] Most cases

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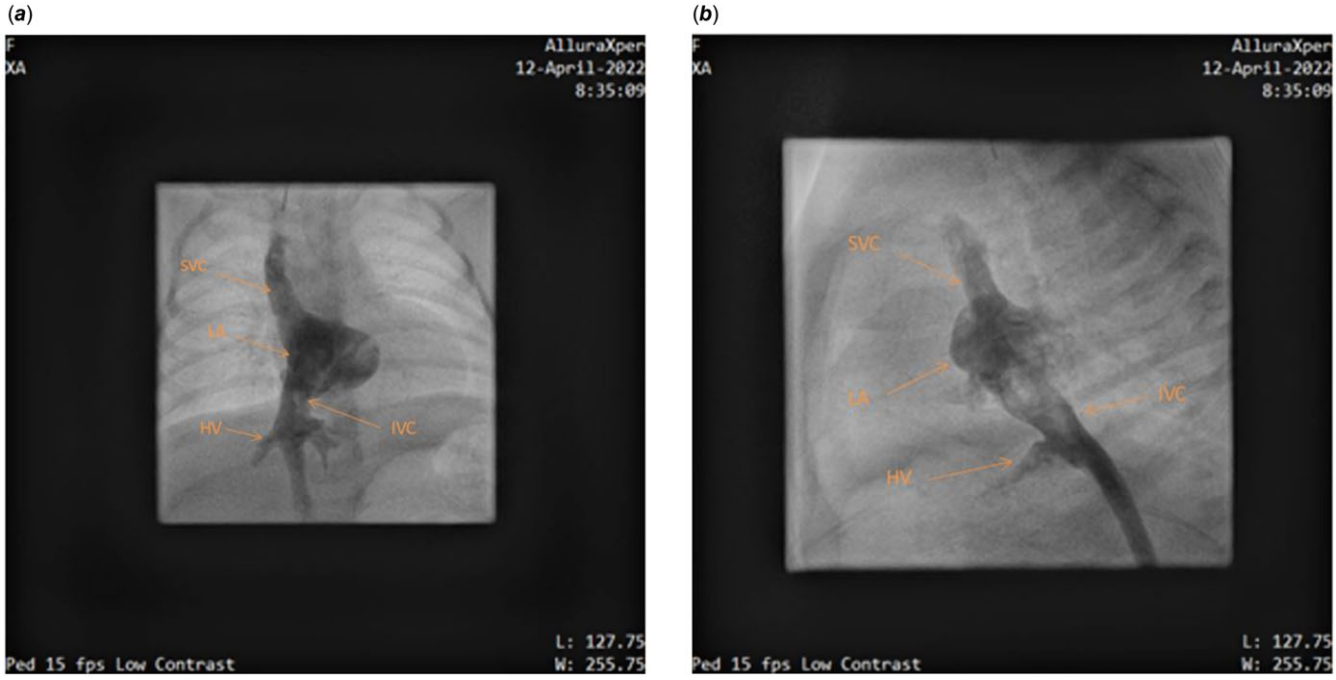

Figure 2. (***a***) Angiography in frontal plane showing Bicaval and hepatic veins connection to left atrium. (***b***) Angiographic lateral plane showing Bicaval and hepatic veins connection to the posterior atrium.

were described in hearts with left isomerism and a few in hearts with the usual atrial arrangement.[2-4] Late diagnosis in adulthood was reported in this rare form of systemic venous connection.[5] Certain clues point to the diagnosis during fetal and postnatal life. First during fetal life, ventricular symmetry with dilated left ventricle and right ventricle hypoplasia along with associated obligatory left to right shunt across the atrial septum should serve as a clue toward this diagnosis. In postnatal life, mild cyanosis without respiratory distress is expected. The degree of cyanosis is determined by the amount of the shunt at the atrial, ventricular, or arterial level. The presence of left to right shunt across the atrial septum in the setting of cyanosis should serve as a clue toward anomalous systemic venous connections to left atrium

During embryological development, incorporation of systemic veins into the right atrium is determined by the venous valves.[6] Therefore, the embryological origin of this anomalous connection is likely due to failure of regression of right valve of right systemic venous sinus or because the systemic venous sinus is being incorporated into left atrium.[2] Although the majority of cases of anomalous systemic veins were described in the setting of left isomerism, our case demonstrates such anomaly occurs in the heart with usual atrial arrangement arguing against using anomalous systemic vein as a marker of left isomerism.

The initial management is dictated by the adequacy of the shunt across the atrial septum to ensure adequate left to right shunt. Definitive surgical therapy will require baffling of the systemic veins to right atrium or a cavopulmonary connection if the right ventricle is hypoplastic.

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Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation (Ministry of health, Oman) and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by research ethics board of Royal hospital, Oman.

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