

# Hypoplastic left heart syndrome, cor triatriatum and partial anomalous pulmonary venous connection: Imaging of a very rare association

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A newborn is presented with an association of hypoplastic left heart syndrome, cor triatriatum and partial anomalous pulmonary venous connection. The diagnosis was established with echocardiography and further confirmed with computed tomography. To our knowledge the images of such an association have never been reported before.

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**H**ypoplastic left heart syndrome (HLHS) occurs in about 0.016% to 0.036% of all live births [6]. It constitutes about 1–3.8% of all congenital heart disease [6]. If left untreated, the patients do not usually survive the first week of life. Fetal interventions to prevent HLHS yielded poor results [4]. Further the offered surgeries and interventions to date are associated with high mortality, morbidity and the need for multiple interventions [1,3]. Studies are focusing on the search for genetic loci responsible for the development of HLHS [8].

We report the very unusual association of HLHS, cor triatriatum, as well as partial anomalous pulmonary venous return. According to our knowledge such a case has never been reported before.

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## Case report

A full term (38 week) infant was diagnosed antenatally by echocardiography as HLHS and was born to an 18-year-old primigravida mother by normal spontaneous vaginal delivery, which was uneventful. The baby was on room air with no respiratory distress. His SPO<sub>2</sub> was 93%, heart rate 141 min<sup>-1</sup>. He had no facial dysmorphism nor congested lungs. Abdominal examination showed

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a liver of about 3 cm below right costal margin. Auscultation showed single S2, no cardiac murmurs. His pulses were equally weak but palpable. Four limb blood pressure was measured, which showed a systolic gradient of 25 mmHg between the upper and lower limbs. The pressures were as follows: right arm 77/33 mmHg, left arm 78/30 mmHg, right leg 50/24 mmHg, left leg 54/22 mmHg.

ECG showed sinus rhythm with right ventricular hypertrophy. X-ray of the chest showed a dilated heart with increased vascularity.

Echocardiography showed situs solitus, levocardia, normal systemic veins, and the flow in superior vena cava was increased. There was no atrial isomerism. The right pulmonary veins were connected normally to left atrium (Fig. 1) but left pulmonary veins were joining a confluence and draining through a vertical vein into the innominate vein (Fig. 2). There was a small patent foramen ovale shunting left to right with a velocity of 1 m/s. The right atrium was significantly dilated with diminutive left atrium. There was a membrane in the left atrium dividing it into two parts; a small superior chamber to which two pulmonary veins were connected which was draining through a restricted hole into left atrium. The tricuspid valve was normal. The mitral valve was atretic (Fig. 3). The right ventricle was massively dilated (Figs. 3 and 4). The left ventricle was severely hypoplastic (Figs. 3 and 4). The right ventricular outflow tract, pulmonary valve, main pulmonary artery, right pulmonary artery and left pulmonary artery were normal. There was no right ventricular outflow tract obstruction. The aortic valve was se-

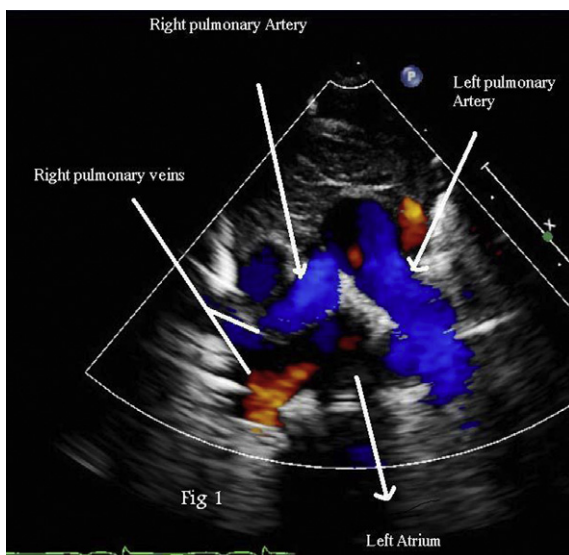


Figure 1. Crab view showing right pulmonary veins.

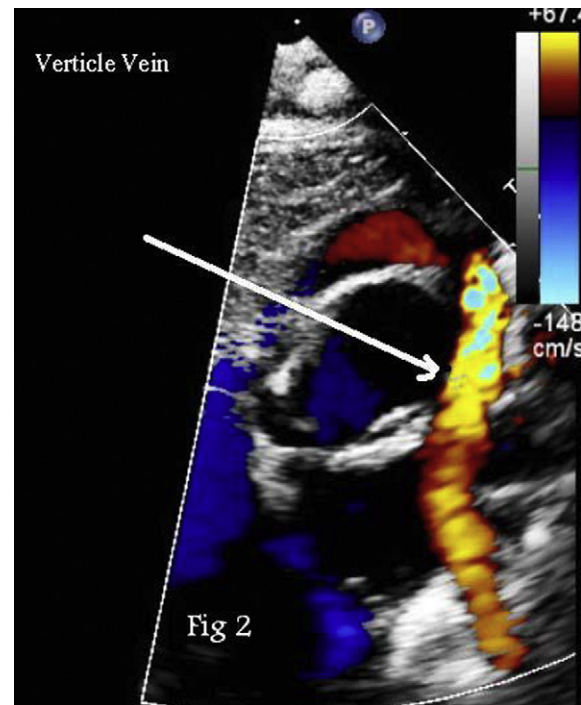


Figure 2. Crab view showing abnormal vertical vein draining to innominate vein.

verely hypoplastic and atretic (Fig. 3). The ascending aorta and transverse arch were severely hypoplastic, measuring 1.5 and 1.6 mm respectively. The descending aorta was normal with no aortic coarctation. There was a reversal flow in aorta and arch. Additionally, there was a large PDA shunting bi-directionally.

Computed tomography with angiography (CT angio) confirmed the findings of Echo (Fig. 5a and 5b).

## Management

Due to the complexity of this lesion, the parents were counseled and given the option to either proceed with surgical interference or not treat the infant. As we have no transplant option at our center, the consensus agreement among our team was not to offer the patient further management. The prostaglandin was not started. The patient expired at 30 hours of life.

## Discussion

HLHS is a well recognized entity with various grades of underdevelopment of left sided heart structures, like mitral atresia, aortic atresia, hypoplastic or interrupted aortic arch. HLHS is usually associated with normal pulmonary venous con-

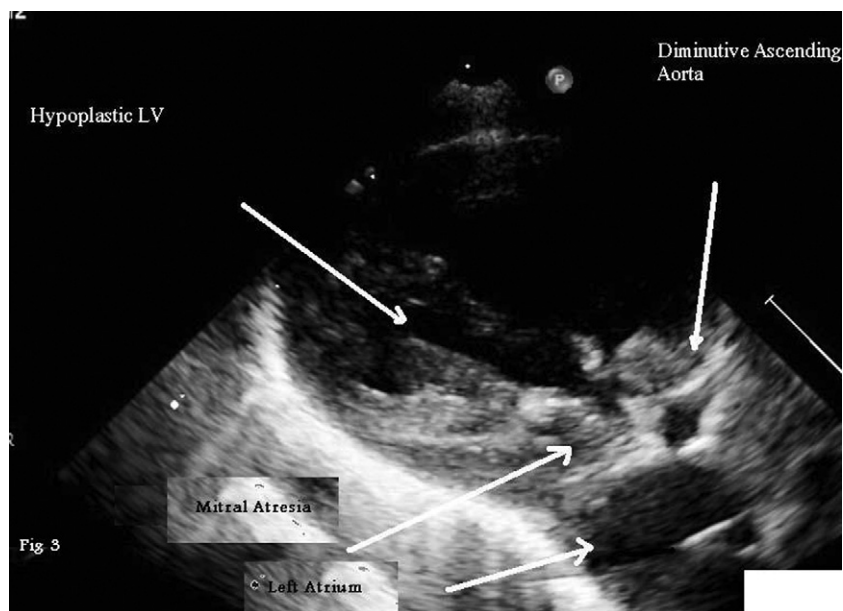


Figure 3. Parasternal long axis view showing mitral atresia, diminutive LA and hypoplastic LV.

nection, but rarely anomalous pulmonary venous connection or drainage as described in our case [10,11]. This entity also has rarely been associated with cor triatrium [2,5].

### Cardiac embryology

HLHS is a complex congenital heart lesion, the mechanism, timing and cause of which is unknown. Several mechanisms are postulated for the development of HLHS.

During the seventh and eighth weeks, the out-flow tract of the heart the conotruncus divides and forms helically arranged ascending aorta and pulmonary artery under the stimulus of blood

flow from the ventricles [7]. The systemic blood flow pressure serves as a stimulus for development of aorta, aortic annulus and arch. Any early interruption in the forward systemic flow in the presence of aortic stenosis or atresia leads to retrograde flow of low pressure blood through patent ductus arteriosus, which leads to poor growth of ascending aorta and arch and ultimately left ventricle and mitral valve as well [12].

The connection between the primitive pulmonary vein and pulmonary venous plexus occurs by the 30th day of gestation. The common pulmonary vein enlarges and incorporates in to the left atrium, and normally pulmonary veins, part

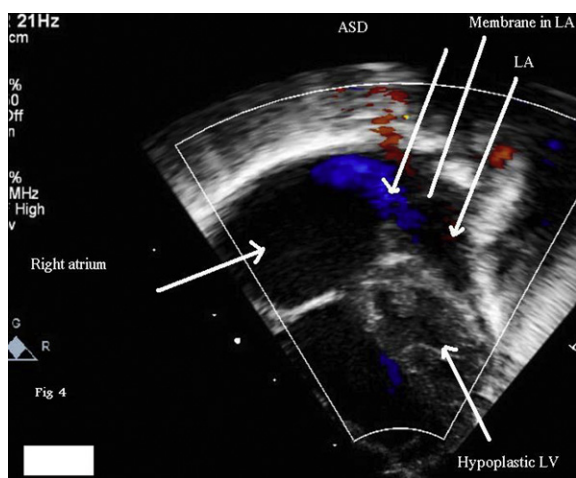


Figure 4. Apical 4 chamber view showing ASD-II shunting left to right.

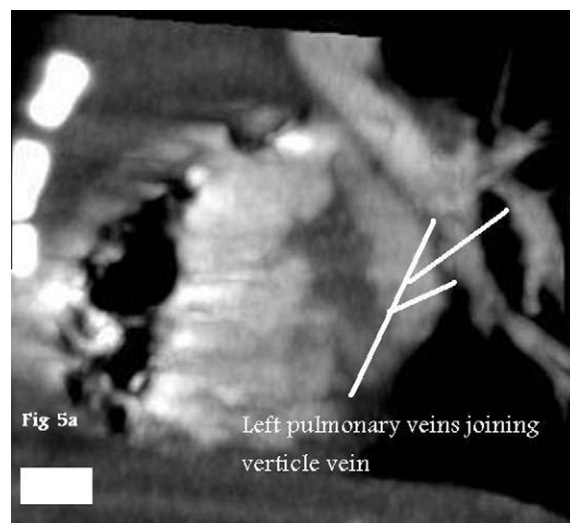


Figure 5a. CT angiogram of the heart showing vertical vein.

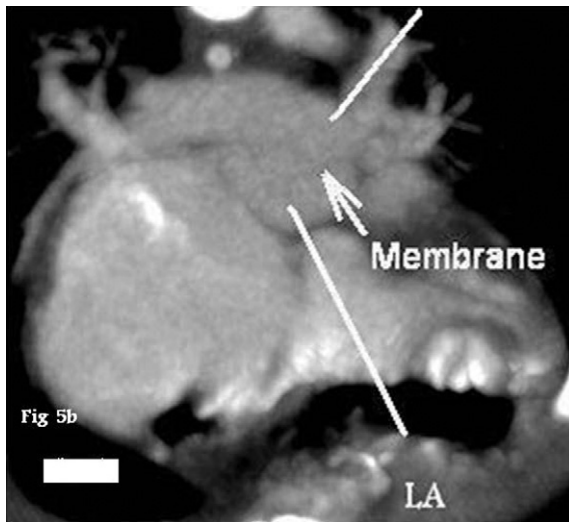


Figure 5b. CT angiogram of the heart showing membrane in left atrium.

of the splanchnic plexus gradually loses its connection with the cardinal and umbilicovitelline vein. Failure of the common pulmonary veins to connect with the pulmonary venous plexus leads to the persistence of one or more earlier venous connections to the right superior vena cava, to left vertical vein or innominate vein. Failure of incorporation of the common pulmonary vein may lead to a left atrial shelf or membrane – cor triatriatum [7,12].

The patients with HLHS and anomalous pulmonary veins may have atrial isomerism (more often right) [9]. The association of atrial isomerism with such a complex heart makes prognosis even worse [9]. Our patient did not have atrial isomerism.

In our case we found the combination of three rare diseases consisting of HLHS, partial anomalous pulmonary venous connection as well as cor triatriatum. The diagnosis was not only established by echocardiography, but also confirmed by computed tomography. The caveat of the CT image reconstruction is operator dependent, moreover, unlike echo, computed tomographic images are static. The CT resolution is not yet as good as the echocardiography, however, it can

complement the echo images for the better evaluation of the extracardiac structure which is limited by the narrow window of the echo.

In this case we chose not to offer further therapeutic management.

To our knowledge this combination has never been reported before in the literature. Additionally the images by echocardiography as well as by computed tomography have never been presented.

### Conflict of Interest

The authors have no conflicts of interest to declare.

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