Cardiol Young 2005; 15: 523–524

ISSN 1047-9511

Brief Report

Hypoplastic left heart syndrome in mirror-imaged arrangement

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Abstract To date, almost all patients reported with hypoplasia of the left heart have had usual atrial arrangement, with a small proportion known to have left or right isomerism. As far as I am aware, however, only one patient has previously been described with mirror-imaged arrangement of the heart and organs. In this report, I describe the second case.

Keywords: Congenital heart disease; dextrocardia; atrial arrangement

THE HYPOPLASTIC LEFT HEART SYNDROME includes a spectrum of congenitally malformed hearts, their common denominator being underdevelopment variously of the left atrium, mitral valve, left ventricle, and aortic valve. Most patients have atresia of the aortic valve. The pathology was first described by Lev,[1] who described "hypoplasia of the aortic tract complexes". It was Noonan and Nadas, in 1958, who coined the term "hypoplastic left heart syndrome".[2] Thus far, nearly all patients described with this entity have left-sided hearts, and most have usual arrangement of the atrial chambers and other organs. A small proportion is known to have right or left isomerism.[3] To the best of my knowledge, the first description of a patient showing mirror-imaged arrangement was given in 2004.[4] In this report, I describe another patient in whom the heart was right-sided in the setting of mirror-imaged atrial arrangement.

Case report

The female patient was born weighing 3,840 grams, being 53 centimetres long. Delivery was normal, and the newborn was asymptomatic. There had been no prenatal fetal studies. After 20 hours of life, the patient became irritated, tachypnoeic, and cyanotic. When seen, the blood pressure was 75 over 52 millimetres of mercury. The saturation of oxygen was 89 percent. On auscultation, the second heart sound was single, but two murmurs were audible. A continuous murmur, of grade 3 out of 6, was best heard along the upper sternal border, and a systolic murmur, judged to be grade 2 out of 6, was present at the low sternal border. The peripheral pulses were weak.

The chest radiograph (Fig. 1) revealed an enlarged right-sided heart and discrete pulmonary plethora. The liver was left-sided. It was not possible to determine the arrangement of the bronchial tree. The electrocardiogram revealed sinus tachycardia, with right ventricular enlargement. The P wave was negative in lead D1, and positive in lead avF.

The transthoracic echocardiogram demonstrated mirror imagery of the atrial chambers and the thoraco-abdominal organs, along with hypoplasia of the left heart (Fig. 2a). Doppler interrogation showed blood passing from the small right-sided morphologically left atrium to an enlarged left-sided morphologically right atrium, passing through a restrictive defect in the oval fossa. The mitral valve was atretic (Fig. 2b). The aortic valve was also atretic, and the blood from the right ventricle passed to the brachiocephalic and coronary arteries having traversed a large persistently patent arterial duct. The pulmonary arterial pressure was 80 millimetres of mercury, as estimated by tricuspid insufficiency. Abdominal ultrasound confirmed that the liver was on the left, and a solitary spleen of normal size was found on the right. A haemodynamic study revealed

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Accepted for publication 12 April 2005

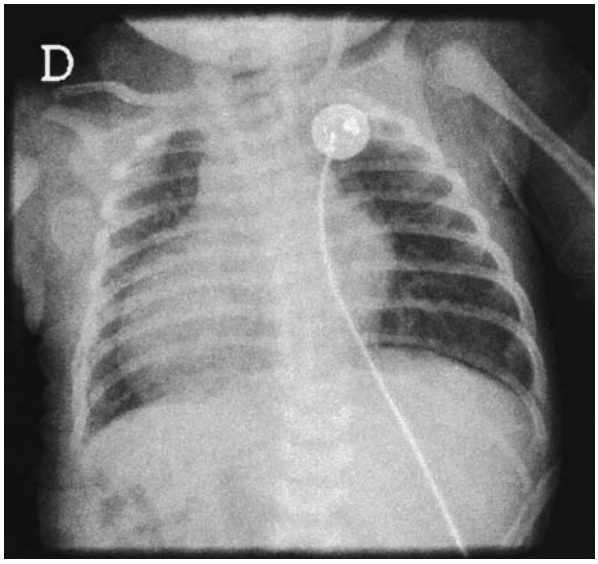

Figure 1.
The chest radiograph, seen in frontal projection, shows a right-sided and enlarged heart. The liver is on the left.

that the hepatic segment of the inferior caval vein was interrupted, the blood returning through a left-sided azygos vein to the superior caval vein, which drained all the superior and inferior caval venous systems to the roof of the left-sided morphologically right atrium. All the pulmonary veins drained to the right-sided morphologically left atrium. We proposed a hybrid intervention, with banding of the pulmonary trunk and stenting of the arterial duct, but the patient died before the procedure could be attempted. Permission was not granted for post-mortem examination.

Discussion

Most patients exhibiting hypoplasia of the left heart have usual arrangement of the atrial chambers and organs,[3] and almost all hearts in this setting are left-sided. As far as I am aware, only one patient has previously been described with mirror-imaged arrangement.[4] This patient underwent the first stage of the Norwood sequence, and the authors stress that no technical difficulties were encountered during the operation on account of the mirror imagery.[4] My report, therefore, is the second to describe a patient with a right-sided heart in the setting of mirror-imaged atrial arrangement. In the light of the discovery of an interrupted inferior caval vein, we obviously considered the possibility of left isomerism, but no further evidence was found to support this diagnosis.

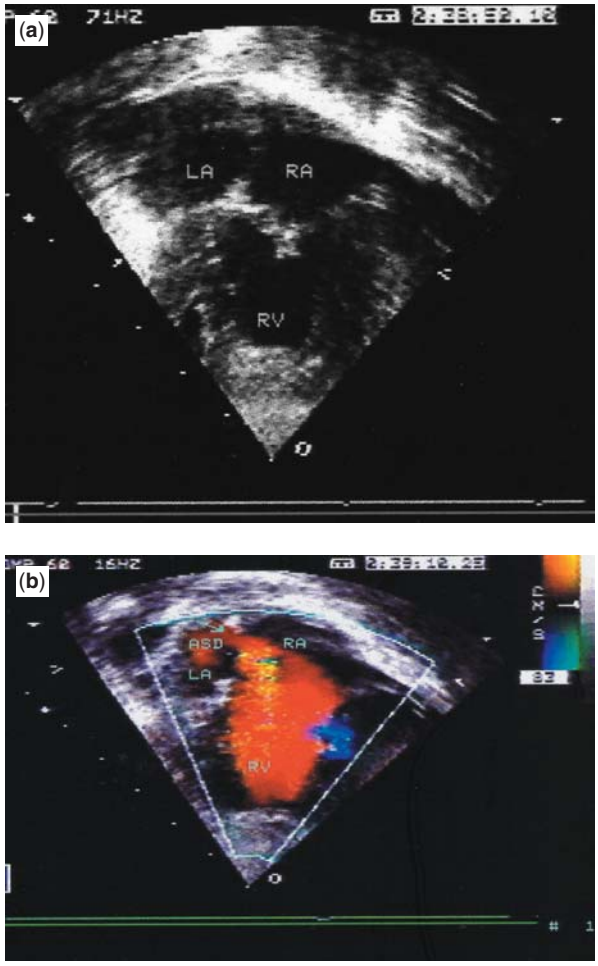

Figure 2.
*The cross-sectional echocardiogram, in the four-chamber plane (**a**), shows the heart on the right side. The right ventricle and atrium are left-sided, with the hypoplastic left ventricle and the small left atrium on the right side. Doppler interrogation (**b**) shows the flow from the left to the right atrium through a restrictive atrial septal defect, and then to the right ventricle. The mitral valve is atretic. RA: right atrium; RV: right ventricle; LA: left atrium; ASD: atrial septal defect.*

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https://doi.org/10.1017/S1047951105001423 Published online by Cambridge University Press