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Brief Report

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Successful biventricular repair following strategic treatment focused on left ventricular growth in a patient with hypoplastic left heart complex and unbalanced right dominant atrioventricular septal defect: a case report

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Abstract

A patient with borderline left ventricle successfully underwent biventricular repair following a staged surgical approach to promote left ventricular growth. Despite initial concerns about left ventricle size, apex formation and adequate size of atrioventricular valve indicated potential for future growth. The patient demonstrated significant left ventricular growth, resulting in stable biventricular circulation and a favourable outcome over a three-year postoperative follow-up period.

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Introduction

Considering the long-term complications and outcomes for patients with univentricular circulation, the benefit of achieving biventricular circulation has been re-emphasised in recent years.[1] Previous studies have proposed criteria to define a left ventricle as adequate for biventricular repair, such as left ventricular volume, aortic valve diameter, and mitral valve diameter,[2] but there is still no established consensus on these parameters. Here, we report a case of borderline left ventricle with successful biventricular repair after strategic staged treatment focusing on left ventricular volume loading to promote growth.

Case presentation

Prenatal echocardiography revealed an unbalanced right dominant complete atrioventricular septal defect, hypoplastic left ventricle, mild aortic stenosis, hypoplastic aortic arch, coarctation of the aorta, and moderate atrioventricular valve regurgitation. Notably, prenatal echocardiography showed a right-to-left shunt at the foramen ovale, with moderate regurgitation from the left-sided atrioventricular valve passed through the primary foramen and reached the right atrium. Antegrade flow was maintained across the arch and isthmus. The patient was born at 39 weeks' gestation with a birth weight of 2746 g. The patient did not exhibit any systemic diseases. Initial postnatal echocardiography (Fig 1) showed an aortic valve diameter of 5 mm (z-score[3] −2.1), ascending aortic diameter of 5.1 mm (z-score −3.4), aortic isthmus of 1.5 mm (z-score −5.0), left ventricular end-diastolic diameter of 7.8 mm (z-score −6.8) with left ventricular apex formation, and left-sided atrioventricular valve interpapillary muscle distance of 8.1 mm (z-score for mitral valve diameter -1.7) as a left ventricular inflow dimension.[4] The cor triatriatum was also identified, and the abnormal septum interrupted the pulmonary venous flow to the left ventricle and redirected flow to the right atrium through the secondary foramen ovale. There was no evidence of left ventricle endocardial fibroelastosis and the left ventricle ejection fraction was 78%, indicating good systolic function. Moderate regurgitation of atrioventricular valve was observed with dysplastic leaflets. Blood flow across the ascending aorta was antegrade, and flow across the ductus arteriosus was initially bidirectional but became predominantly left-to-right. Despite the markedly small left ventricular volume based on left ventricular end-diastolic diameter, the adequate size of the aortic and left-sided atrioventricular valves and left ventricular apex formation suggested the potential for left ventricle growth.

At 4 days of age, the patient underwent aortic arch repair and pulmonary artery banding. Atrioventricular valvuloplasty (bivalvation) was performed at 28 days of age. Cor triatriatum repair and patent foramen ovale closure were also performed to increase left ventricular volume loading. Although moderate atrioventricular valve regurgitation persisted postoperatively, significant left ventricular growth was achieved. The left ventricular end-diastolic diameter

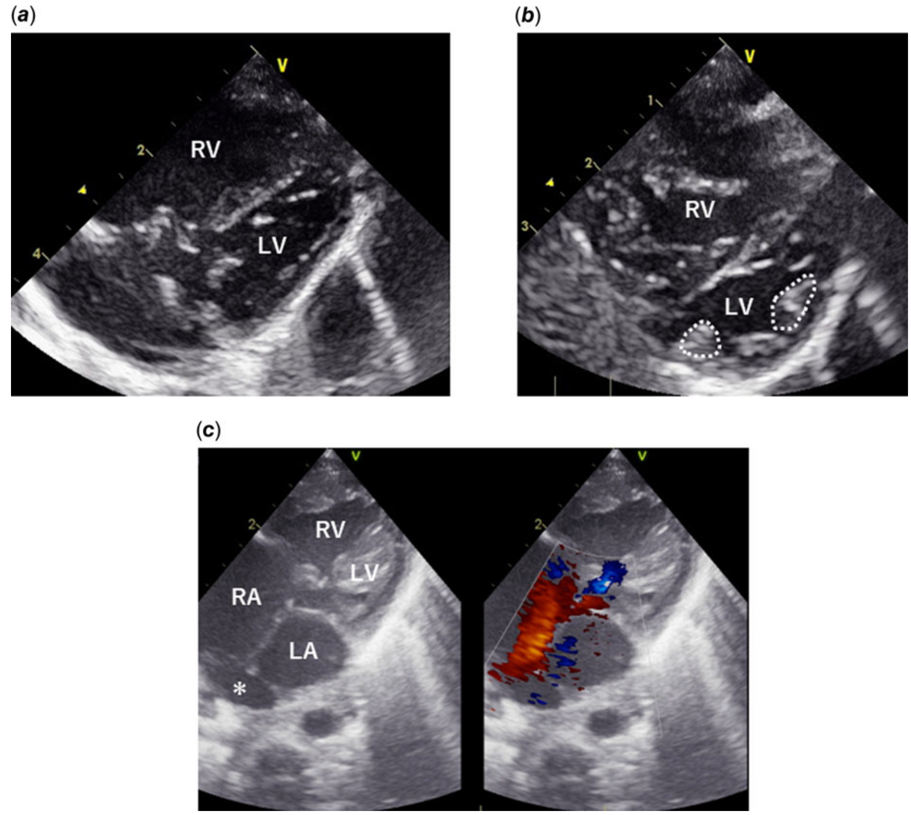

Figure 1. Postnatal echocardiography. (***a***) The four-chamber view demonstrates a small left ventricle with apex formation. (***b***) The short-axis view reveals a interpapillary muscle distance (dotted white line) of 8.1 mm, corresponding to a Z-score of -1.7 for the mitral valve diameter. (***c***) The colour doppler four-chamber view shows pulmonary venous blood flow returning to the accessory chamber (*) and flowing back into the right atrium through a patent foramen ovale.

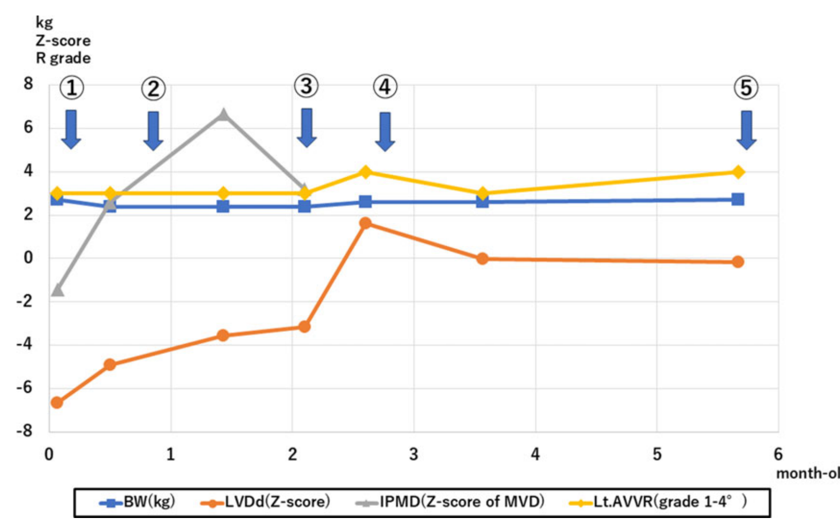

Figure 2. Graphical overview of the clinical course. The graph shows the patient's body weight, left ventricular end-diastolic diameter (Z-score), interpapillary muscle distance (Z-score of mitral valve diameter), and left atrioventricular valve regurgitation (grade 1-4) over time from birth to 6 months. The main surgical interventions are: (①) aortic coarctation repair and pulmonary artery banding, (②) cor triatriatum repair and common atrioventricular valve plasty, (③) complete atrioventricular septal defect repair, (④) right and left atrioventricular valve plasty, and (⑤) replacement of left atrioventricular valve.

z-score and the left-sided atrioventricular valve interpapillary muscle distance became −3.4 and 3.1, respectively (Fig 2). Pre-atrioventricular septal defect repair catheterisation data showed a left ventricular end-diastolic volume index of 35.3 mL/m^2, and a left ventricular end-diastolic pressure of 7 mmHg, which further supported for decision to proceed. Atrioventricular septal defect repair was completed at 2 months of age. Severe regurgitation of both the left and right atrioventricular valves developed post-operatively, necessitating replacement of the left-sided atrioventricular valve with a mechanical valve (ATS-AP 360 Mitral 16 mm) by semi-circumferential translocation at 5 months of age.

The patient was discharged 111 days after left-sided atrioventricular valve replacement.

Three years after valve replacement the patient remains well. Cardiac catheterisation performed one year after valve replacement revealed a central venous pressure of 7 mmHg, pulmonary artery pressure of 26 mmHg, left ventricular end-diastolic pressure of 12 mmHg, cardiac index of 3.4 L/min/m^2, and left ventricular end-diastolic volume index of 75.8 mL/m^2. These results indicated mild diastolic dysfunction of left ventricle with mild pulmonary hypertension but demonstrated well-preserved biventricular circulation with an adequate cardiac index and low central venous pressure. The patient did not require readmission for heart failure during follow-up. The patient's physical growth had declined to a minimum of -5.0 standard deviations during the course of the disease but has recovered since the valve regurgitation was

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controlled with the replacement after the atrioventricular septal defect repair and is now up to 0.5 standard deviations.

Discussion

Previous reports have established criteria for determining the feasibility of biventricular repair in cases of borderline left ventricle, such as left ventricular end-diastolic diameter, mitral valve diameter, and aortic valve diameter.[2,5–7] In this case, the left ventricular volume (Z-score of left ventricular end-diastolic diameter -6.8) suggested that biventricular repair might be inadequate. However, the presence of left ventricular apex formation on echocardiography at birth, as well as the aortic valve diameter (Z-score -2.12) and interpapillary muscle distance of left-sided atrioventricular valve (Z-score of mitral valve diameter -1.7), met the criteria for biventricular repair.

Cheng et al. reported that when the left ventricle shows apex formation, left ventricular growth can be expected even when the initial volume is small if sufficient volume loading is applied.[8] In this case, the assessment of left ventricular volume at birth was inaccurate due to the presence of the coarctation of the aorta and Cor triatriatum. Taking these factors into account, a staged surgical approach to increase the volume loading of the left ventricle resulted in significant growth of left ventricle.

On the other hand, volume load to left ventricle may worsen the atrioventricular valve function. In this case, the management of postoperative atrioventricular valve dysfunction proved difficult and ultimately necessitated prosthetic valve replacement. Although prosthetic valve was successfully implanted with semi-circumferential translocation, closed follow-up is required for potential complications such as reduction of left atrial cavity, progression of pulmonary venous obstruction, and subaortic stenosis. Furthermore, ingenuity in redo valve replacement is mandatory.

This report suggests the growth potential of left ventricle should be considered comprehensively and staged approach to promote left ventricular growth may achieve left ventricle adequate for biventricular repair. As in this case, it is important to accurately assess all left ventricular components and loading status, rather than focusing on a single component.

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Competing interests. None.

Ethical Standards. The authors declare that all procedures contributing to this work complied with the ethical standards of the relevant national guidelines for human experimentation (Ethical Guidelines for Medical and Biological Research Involving Human Subjects, Japan) and the Helsinki Declaration of 1975 as revised in 2008, and were approved by the institutional committees (Institutional Review Board of Osaka City General Hospital, number 1902139.).

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