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Case report: duct dependency in hypoplastic left heart complex can be reversible without surgery

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Brief Report

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Abstract

We describe an infant with duct-dependent hypoplastic left heart complex with moderate hypoplasia of the left ventricle and aortic arch who was not operated due to resource limitations. The left-sided structures grew remarkably due to favourable loading condition changes of the left ventricle, allowing weaning from prostaglandin at the age of 3 months and discharging the patient without intervention.

Case report

The patient was a full-term infant with a birth weight of 2.8 kg antenatally diagnosed with hypoplasia of the left ventricle and coarctation. Initial echocardiography showed moderate hypoplasia of the left ventricle. The mitral valve was a parachute like and the annulus measured 6.4 mm (z score: −3.9). The aortic valve was trileaflet, and the annulus measured 4.4 mm (z score: −3.9) with no stenosis. The left ventricle inflow length was 22 mm and the heart long axis length measured 30 mm. No endocardial fibroelastosis was noted. The patient's Rhodes and Congenital Heart Surgeons' Society scores are displayed in Table 1.

Significant aortic arch and isthmus hypoplasia were observed; the transverse arch measured 2.8 mm (z score: −6.3), as shown in Fig 1a–c and Table 1. The maximum gradient across the aortic arch was 12 mmHg in the presence of a large patent ductus arteriosus. A moderate secundum atrial septal defect measuring 7 mm with left-to-right shunting (peak/mean gradient, 0.8/0.4 mmHg) was observed. The ventricular septum was intact. There was moderate tricuspid regurgitation. The estimated right ventricular systolic pressure was 65 mmHg plus right atrial pressure. The patient has a good left ventricular function. A left superior vena cava draining to the coronary sinus and normal pulmonary venous drainage was observed (Supplementary video S1). The findings fit the definition of a hypoplastic left heart complex.

The patient started on prostaglandin E1 infusion at birth based on the antenatal diagnosis, which was later discontinued to evaluate duct dependency. However, the patient developed tachypnoea, poor perfusion, and weak pulses once the ductus arteriosus was almost closed. The systolic functions of the right and left ventricles and the degree of tricuspid regurgitation significantly worsened, and pulmonary hypertension was noted.

The patient was again placed on prostaglandin treatment with the impression that she had a duct-dependent coarctation and her left ventricle could not support the systemic circulation. Due to the unavailability of paediatric cardiac surgery services in our centre, the patient remained on a long waiting list and was ineligible for other local cardiac centres. Serial echocardiography showed a progressive reduction of atrial communication size, leading to a higher gradient across the mitral valve and atrial communication (Table 1). The patient has severe pulmonary hypertension for the first 2 months. However, pulmonary vascular resistance started to decline slowly, allowing more left-to-right shunting across the patent ductus arteriosus, thus pushing more flow across the left side.

When the patient reached the age of 3 months, we decided to stop the prostaglandin treatment since the left ventricle and aortic arch had increased in size. The patient tolerated this well, and the ductus arteriosus became smaller. She continued to have a mild but tolerable degree of mitral stenosis and arch hypoplasia (Fig 1d–f). She was discharged from the hospital and has regularly been followed up. She was last seen at the age of 4 years, and her mitral and aortic valves continued to show good growth with mild mitral stenosis and minimal gradient (16 mmHg) across the aortic arch (Table 1 and Supplementary video S2).

Discussion

Hypoplastic left heart complex is a milder form of hypoplastic left heart syndrome with coarctation or hypoplastic aortic arch with the presence of hypoplasia of left heart structures including small

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Table 1. Echocardiographic measurements of left-sided heart structures over time

Age	BSA*	MV mm (z Score**)	AV mm (z Score)	T arch mm (z Score)	LVDD	LV inflow Ln mm	Heart long axis Ln mm	MV inflow (peak/mean) mmHg	PDA size (mm)	PDA shunt	ASD size (mm)	ASD gradient (mmHg)	Degree of TR	TR gradient (mmHg)	Rhodes score	CHSS score
1 day	0.18	6.4 (−3.9)	4.4 (−3.9)	2.8 (−6.3)	11 (−4.7)	22	30	6.7/2.4	5.5	Bidirectional	7	0.8/0.39	Moderate	65	−2.49***	−27.48
12 days	0.19	6.7 (−3.5)	4.5 (−3.2)	3.5 (−4.8)	12 (−3.9)	28	34	5/1.2	closed	Closed	7.4	5.1/2.2	Severe	96	−2.12***	−41.7
27 days	0.2	6.8 (−4.0)	5.3 (−2.4)	4.2 (−3.9)	13 (−3.4)	30	35	_	3.8	Bidirectional	4.2	3.8/2.1	Very severe	90	−1.63***	−34.45
54 days	0.21	8.3 (−2.4)	5.7 (−1.8)	4.7 (−3.3)	21 (+1.3)	30	34	24/13	4.5	L-R 16 mmHg	3	24/9.7	Moderate	79	−1.13***	−41.7
3 months	0.24	9.7 (−1.7)	7.2 (−0.1)	5.3 (−2.9)	22 (+1.1)	31	32	27/10	4.2	L-R 30 mmHg	2.2	19.7/8.2	Moderate	73	−0.26	−50.57
4 months	0.24	10 (−1.6)	8.1 (+1.1)	5.4 (−2.8)	23 (+1.5)	35	35	10/5	3.4	L-R 126 mmHg	2	6/2.3	Mild	66	NA	NA
4 years	0.61	16 (−1.4)	13 (+0.8)	8.4 (−3.0)	37 (+1.3)	46	48	12/5	1	L-R	0	NA	Trivial	24	NA	NA

ASD = atrial septal defect; AV, aortic valve; BSA = body surface area; CHSS = Congenital Heart Surgeons' Society; Ln = length; L-R = left to right; LVDD = left ventricular diastolic dimension; MV = mitral valve; PDA = patent ductus arteriosus; T arch = transverse aortic arch; TR = tricuspid regurgitation.

*BSA is measured using DyBois formula.

**z score was based on the Detroit data.

***Score prefers univentricular repair.

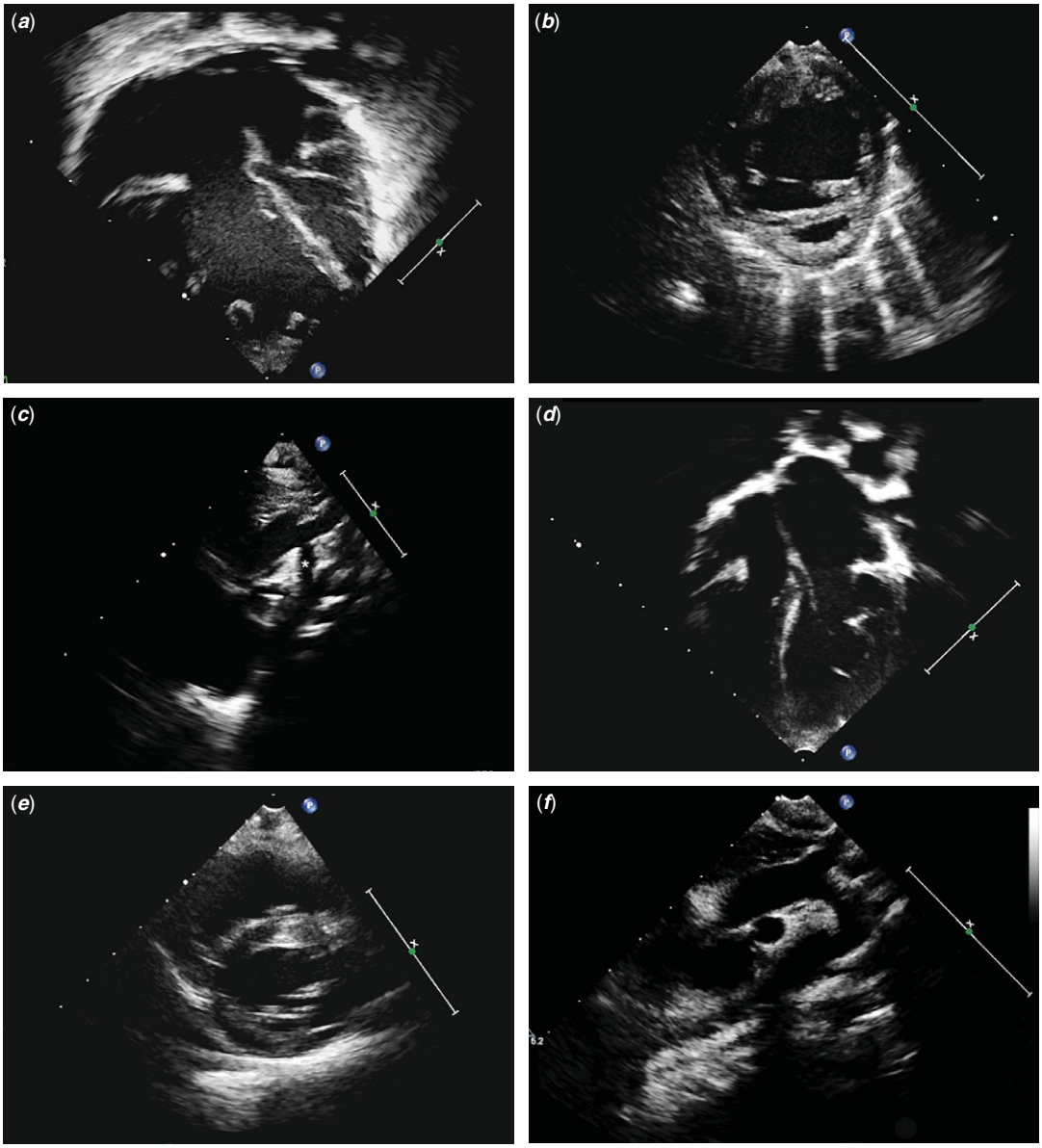


Figure 1. Two-dimensional echocardiographic images at birth and on the last follow-up. (***a–c***) Echocardiography at birth showing the small mitral valve, hypoplastic left ventricle (LV), dilated right ventricle (RV), and hypoplastic distal aortic arch (*). (***d–f***) Echocardiography on the last follow-up showing good growth of LV and normalisation of the size of the RV and aortic arch.

aortic and mitral valves without intrinsic aortic or mitral stenosis.[1] The pathophysiology of this disease is characterised by a holosystolic antegrade flow in the ascending aorta and duct dependency.[1,2] Neonatal surgical repair with aortic arch reconstruction, ductal ligation, and removal of ductal tissue is considered the mainstay approach in patients with this condition.[1,3,4] The objective of surgery is to normalise the loading conditions of the left ventricle by reducing afterload and/or increasing the preload to allow left ventricular growth.[4] The closure of a concomitant atrial septal defect remains controversial. Early closure might lead to very high left atrial pressure with a stormy post-operative course and left ventricular decompensation. Alternatively, early closure of the atrial septal communication will augment the preload of the left side, allowing further growth.[2,3] The decision to go for surgical repair is straightforward in almost all patients due to duct dependency. The early elimination of aortic arch obstruction supposedly facilitates left ventricular growth.[3,4]

In our patient, arch obstruction was alleviated by prostaglandin E1 therapy (afterload reduction), and the natural reduction of the size of the atrial septal defect allowed gradual increases in the left ventricular preload leading to spontaneous growth and adaptation of left-sided chambers. The left atrial pressure was quite high when the ductus arteriosus was still open due to the small mitral valve and additional volume loading. The ductus arteriosus eventually became restrictive following a normal drop in pulmonary pressure. The outcome was surprisingly adequate left ventricular growth with a minimal gradient across the aortic arch and no pulmonary hypertension.

This case suggests that in selected patients with duct-dependent hypoplastic left heart complex with aortic arch hypoplasia, left ventricular hypoplasia in the presence of an atrial septal defect waiting for a few weeks might allow significant growth of the mitral and aortic valves, enabling the medical team to stop prostaglandin

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therapy and re-evaluate the need for surgical therapy. Such a conservative approach can be utilised in selected conditions when delaying surgical therapy is needed. However, the clinician should be aware of the adverse outcomes of prolonged prostaglandin E1 therapy and avoid delaying surgery in patients who are undoubtedly destined for a univentricular repair as such delay will worsen the long-term outcome.[5,6]

Conclusion

Left ventricular rehabilitation in patients with duct-dependent hypoplastic left heart complex is feasible under appropriate haemodynamic conditions, and these patients may or may not require interventions for left ventricular rehabilitation to be achieved in the first few weeks of life.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951120003935.

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