

Staged interventional solution for a diagnostic dilemma caused by hypoplastic left ventricle with severe aortic arch hypoplasia

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ABSTRACT

Duct dependent aortic arch obstruction with borderline left ventricular hypoplasia presents a diagnostic dilemma. If the left ventricle (LV) is adequate to sustain systemic cardiac output without a patent duct, arch obstruction is relieved surgically and duct is divided. Inadequate LVs do not tolerate duct division, and these patients need more complex Norwood type surgeries. However, catheter-based interventions for arch obstruction can retain ductal patency. The progressive changes in anatomy and physiology of a neonate who presented with hypoplastic left heart structures and patent duct with advancing age and modified by serial catheter-based interventions are presented to highlight the use of interventions in this therapeutic dilemma.

Keywords: Biventricular repair, catheter interventions, coarctation stenting, hypoplastic left heart, transverse arch hypoplasia

INTRODUCTION

Hypoplastic left ventricle (LV) with outflow obstruction and duct-dependent systemic circulation creates a dilemma in decision-making regarding univentricular versus biventricular repair.^[1,2] Prediction of postnatal ability of LV to sustain a systemic cardiac output without a ductus is based on the severity of left ventricular hypoplasia and multiplicity of levels of obstruction.^[2] The echocardiographic indices proposed to predict the outcome after surgical repair have been observed to improve progressively on follow-up when the neonate is medically managed for weeks, thereby allowing biventricular repair in some infants who were initially deemed unsuitable for sustaining a two-ventricle circulation.^[3] This questions the validity of these echocardiographic predictors.^[4] Physiological principles that improve the growth of a hypoplastic LV include reducing afterload by the relief of obstruction and improving preload by spontaneous or surgical reduction

of atrial septal communication.^[5] Ductal division during arch repair is safe only when the adequacy of systemic cardiac output is assured.^[6] While surgical arch repair cannot retain patency of the duct, it can be retained by catheter-based interventions on the hypoplastic aortic arch by the use of balloons and uncovered stents. A child with duct-dependent severe aortic arch obstruction who presented in extremis was managed in catheterization laboratory by serial interventions that progressively normalized the circulation. The changes in the anatomy and hemodynamics with advancing age and further modified by the catheter interventions are discussed.

CASE REPORT

Following an antenatal diagnosis of hypoplastic aortic arch at 5 months of gestation, severe maternal pregnancy-induced hypertension forced a premature

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induction of delivery at 8 months of gestation. Postnatal echocardiogram of the neonate weighing 1.05 kg showed a hypoplastic apex-forming LV, apical muscular ventricular septal defect with bidirectional flows, hypoplastic transverse arch, right to left ductal flows into the descending aorta, and a small patent oval foramen with vigorous left to right flows [Figure 1]. High risks involved in apical muscular ventricular septal defect closure combined with arch repair in extreme prematurity forced a compassionate care and discharge from neonatal unit 1 month later on diuretics. As the distal transverse arch and isthmus measured 1 mm in diameter in this patient weighing 1 kg, angioplasty or stenting was not considered.

After being lost from follow-up due to a pessimistic outlook, the child was hospitalized at 2 years of age in cardiogenic shock and severe respiratory distress with a weight of 7 kg and significantly delayed gross-motor development, corresponding to 6 months. After resuscitation, blood pressure difference between the upper and lower limbs was 20 mmHg. The dilated LV showed severe systolic dysfunction with a very large 43 mm × 23 mm thrombus filling the chamber [Figure 2]. The apical muscular ventricular septal defect noted in neonatal imaging had spontaneously closed. The reduction in size of the atrial septal communication to 2 mm and increase of interatrial gradient to 11 mmHg increased left ventricular preload improving its size [Figure 3]. The transverse arch was hypoplastic with a moderate sized duct supplying the descending aorta led to differential cyanosis and oxygen saturations of 70% in legs. In view of severe pulmonary hypertension with right to left ductal flows and uncertain pulmonary vascular resistance, a decision was taken to relieve the arch obstruction without closing the pop-off through the duct.

The pulmonary artery pressures were marginally higher than descending aortic pressures, and the systolic arch gradient was 40 mmHg [Table 1]. The

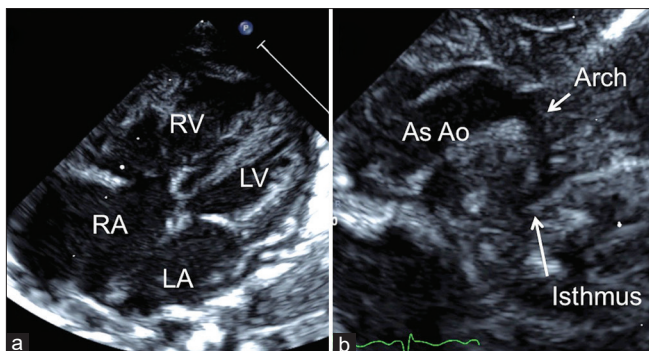


Figure 1: Apical four-chamber echocardiographic view (a) shows hypoplastic left ventricle, dilated right ventricle, left atrium decompressed by foramen ovale to right atrium. The transverse aortic arch and isthmic narrowing is seen in the suprasternal long-axis view (b)

distal transverse arch between the left carotid artery and left subclavian artery was 12 mm in length and 3.5 mm in diameter (Z-score was - 4.7) was stented with 8 × 12 Formula 418 stent (Cook medical, Bloomington, IN, USA). The isthmus measuring 2.5 mm (Z-score was - 4.1) and coarctation segment measuring 1.5 mm was stented with a 7 × 16 Formula 418 stent. The ductal patency was maintained through the struts of the stent. Optimal oral anticoagulation with heart failure medications in the next 6 months normalized the LV function, dissolution of LV thrombus and dramatic improvement in the motor developmental milestones and disappearance of differential cyanosis. A repeat catheterization showed a 12% step up at ductal level. The duct was crossed through the sidestruts of the lower stent and closed with a low-profile 5-4 Amplatzer duct occluder II (Abbott medical, Plymouth, MN, USA) resulting in fall of pulmonary artery pressures to 40% of systemic pressures [Figure 4]. The two stents were dilated to 10 mm during this procedure. At 1-year follow-up, the LV function normalized with no arch gradients and normal pulmonary artery pressures. Carvedilol and enalapril were continued to maintain LV function [Figure 5].

DISCUSSION

Hypoplastic LV refers to a wide spectrum, ranging from mildly hypoplastic LV as sometimes seen in 45% of neonates with aortic coarctation to extremely severe form as seen in hypoplastic left heart syndrome.^[1]

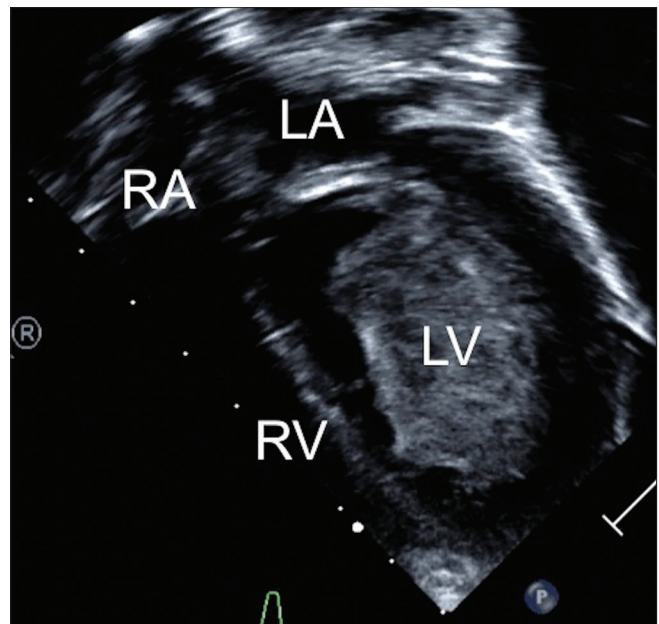


Figure 2: A large left ventricular thrombus occupying three-fourth of the cavity is seen on apical four chamber view. The foramen ovale between the right atrium and left atrium is spontaneously closed. Right ventricle is comparatively smaller than the dysfunctional left ventricle

Table 1: Hemodynamics and Oximetry

Site	First procedure at 2 years of age			Second procedure after 6 months		
	SO ₂ (%)	Before stenting (mmHg)	After stenting (mmHg)	SO ₂	Before PDA device (mmHg)	After PDA device (mmHg)
SVC	69.4			70.1		
IVC	40.9			61.9		
RA		Mean 6	Mean 2		Mean 1	Mean 1
RV		Systolic 90			Systolic 70	Systolic 50
PA	60.4	90/40/55		80.1	70/42/56	50/20/30
AA	92.6	120/60/90	95/45/78	96.2	105/42/70	120/60/86
DA	66.2	80/45/55	90/45/75	96.2	105/42/70	120/60/86

RA: Right atrium, RV: Right ventricle, PDA: Patent ductus arteriosus; SVC: Superior vena cava; PA: Pulmonary artery; AA: Ascending aorta; DA: Descending aorta; IVC: Inferior vena cava

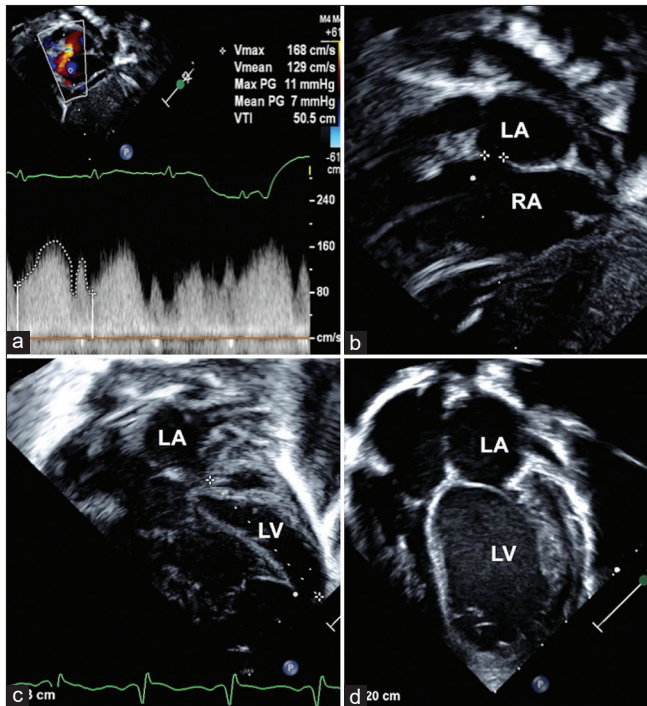


Figure 3: The Doppler gradient (a) between the left atrium and right atrium increased to 11 mmHg due to reduction in size of the atrial septal communication (b) to 2 mm on sibxiphoid view. The increased preload improved the long axis of left ventricle from 2.6 cm in neonatal period (c) to 6.2 cm at 2 years (d) on apical view

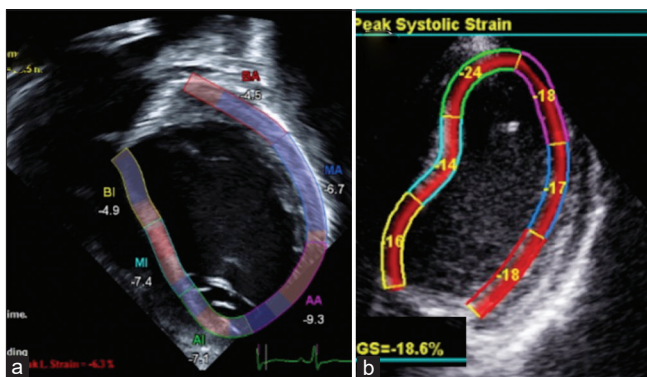


Figure 5: The severely dysfunctional left ventricle with global longitudinal strain of -6.3% and ejection fraction of 15% before the stenting (a) improved to values of -18.6% and 65% after the interventions (b)

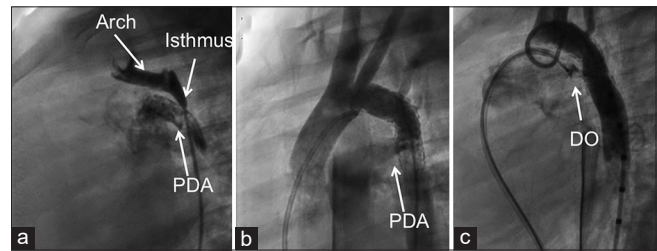


Figure 4: Severe transverse arch and isthmic narrowing seen on lateral view aortogram (a) is relieved after stent angioplasty of aorta (b) and large patent duct closed (c) on a staged sitting with a duct occluder device

Decision-making is simpler when the hypoplastic LV is at either extremes.^[2] Severe forms require Norwood palliation or cardiac transplantation; mildest forms get complete biventricular repair. Echocardiographic scores based on left heart dimensions and body surface area, proposed to define mortality risk after biventricular repair are questioned as they progressively improve with growth of the neonate.^[2,3] When afterload reduces with relief of outflow tract obstruction and preload improves with the closure of oval foramen, LV dimensions progressively show improvement.^[4,5] Biventricular repair in duct-dependent neonates with hypoplastic, but morphologically normal LV is feasible with the restoration of normal loading conditions and relief of LV obstruction.^[6] Staged surgical approaches with initial univentricular palliation in the neonatal period with later conversion to biventricular repair after one to three stages have demonstrated potential for the growth of left heart structures.^[6]

Conservative approach due to extreme prematurity and body-weight in our patient forced a study of the evolving natural history in this situation. Spontaneous closure of oval foramen improved LV preload and dimensions, which remained severely dysfunctional due to persistent arch obstruction leading to cavitory thrombus formation. Despite the LV growth, a unique challenge of persistent right-to-left ductal flows and unclear pulmonary vascular resistance prohibited surgical arch repair, as duct division was obligatory. Catheter approaches that are not usually considered in young patients with severely narrowed aortic arch may

retain the ductal patency in spite of complete relief of gradients. After initial coarctation stenting and relief of left heart obstruction, pulmonary vascular resistance dropped significantly changing the ductal flows and permitting complete transcatheter duct closure at a later date when hemodynamics improved.

Transverse arch and isthmic coarctations are known to be associated with repeated reinterventions in 11%–19% on follow-up.^[7] Despite capability of the two stents for postdilatation to a final diameter of 14–16 mm, the need for repeated catheterization and dilatations in this patient remain as points of concern. The current postdilatatable stents can be dilated to twice their nominal diameters, thereby allowing somatic growth of young patients.^[8]

CONCLUSION

Definitive biventricular repair is a therapeutic dilemma in hypoplastic LV associated with duct dependent aortic arch obstruction. Conventional echocardiographic indices sometimes fail to predict adequacy of LV accurately and may progressively improve with age, thereby changing the surgical approach over time on follow-up. While surgical strategies for arch obstruction obligatorily divide duct in biventricular approaches, catheter-based strategies are more flexible and can retain ductal patency. Such catheter interventions may be of value in selected neonates and infants with duct-dependent arch obstruction.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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