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Biventricular Repair after Bilateral Pulmonary Artery Banding as a Rescue Procedure for a Neonate with Hypoplastic Left Heart Complex

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Hypoplastic left heart complex (HLHC) consists of less severe underdevelopment of the left ventricle without intrinsic left valvular stenosis, i.e., a subset of hypoplastic left heart syndrome (HLHS). HLHC patients may be able to undergo biventricular repair, while HLHS requires single ventricle palliation (or transplant). However, there is no consensus regarding the likelihood of favorable outcomes in neonates with HLHC selected to undergo this surgical approach. This case report describes a neonate with HLHC, co-arctation of the aorta (CoA), and patent ductus arteriosus (PDA) who was initially palliated using bilateral pulmonary artery banding due to unstable ductus-dependent circulation. A postoperative echocardiogram showed newly appearing CoA and progressively narrowing PDA, which resulted in the need for biventricular repair 21 days following the palliation surgery. The patient was discharged on postoperative day 13 without complications and is doing clinically well seven months after surgery.

Key words: 1. Congenital heart disease
2. Hypoplastic Left Heart Syndrome
3. Pulmonary artery
4. Biventricular repair

CASE REPORT

A male neonate weighing 3.63 kg was delivered at Asan Medical Center after 39 weeks of uneventful gestation. He was prenatally diagnosed with hypoplastic left heart syndrome (HLHS) and/or coarctation of the aorta (CoA) by a fetal echocardiogram. Upon delivery, his vital signs were stable with oxygen saturation of 90% in the right arm and 88% in the right leg. Initial postnatal echocardiogram revealed mitral stenosis, aortic stenosis, bicuspid aortic valve (AV), patent ductus arteriosus (PDA) with a bidirectional shunt, intact atrial septum, and near normal left ventricle (LV) function with an ejection fraction (EF) of 68%. The mitral valve (MV) and the AV annulus diameters were 7.1 mm (Z-score=−3.47) and 4.2 mm (Z-score=−4.86), respectively. Prostaglandin E was infused immediately (0.008 mcg/kg/min). However, on day 3 of hospitalization, the patient's vital signs became unstable, including severe fluctuation of blood pressure, increasing blood lactate level, diminishing urine output, and differential cyanosis up to 30% when blood pressure was elevated. Echocardiographic and computed tomography (CT) findings at that time showed right to left dominant PDA flow, retrograde

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Table 1. Echocardiogram and volume measured CT findings

Variable	After birth	After pulmonary artery banding	After biventricular repair	
Age (day)	3	20	31	136
Echocardiogram				
Aortic valve annulus (mm)	4.2	5.0	5.9	7.8
Z-score	−4.855	−3.501	−1.846	−1.165
MV annulus (mm)	7.1	6.8	8.6	9.5
Z-score	−3.469	−3.996	−2.412	−3.009
Fractional shortening (%)	19.45	50.50	30.65	38.92
EF (%)	36.32	84.4	61.27	72.26
PDA size (mm)	6.4	3.3	(−)	(−)
Flow	Right to left dominant	Bidirectional	(−)	(−)
Volume measured CT				
MV area (cm^2)	NC	0.4	NC	0.8
Tricuspid valve area (cm^2)	NC	2.3	NC	2.5
Left ventricle EF (%)	32.5	63.8	NC	66.0
EDV (mL/m^2)	35	29	NC	56
ESV (mL/m^2)	24	10	NC	19
Right ventricle EF (%)	63.2	63.2	NC	65
EDV (mL/m^2)	46	71	NC	75
ESV (mL/m^2)	17	25	NC	26
PDA size (mm)	8	3	(−)	(−)
Ascending aorta (mm)	5	8	NC	10
Proximal aortic arch (mm)	3	4.6	NC	8
Distal aortic arch (mm)	3.7	5.6	NC	8
Aortic isthmus (mm)	4.6	5.5	NC	8

CT, computed tomography; MV, mitral valve; EF, ejection fraction; PDA, patent ductus arteriosus; NC, not checked; EDV, end diastolic volume; ESV, end systolic volume.

aortic arch flow, and severely decreased left ventricular function with an EF of 36% (Table 1). Volume-measured heart CT produced the following measurements: subaortic area 4 mm; ascending aorta 5 mm; proximal aortic arch 3 mm; distal aortic arch 3.7 mm; and aortic isthmus 4.6 mm. The LV EF was 32.5%, the LV end-diastolic volume (EDV) was 35 mL/m^2, and the LV end-systolic volume (ESV) was 24 mL/m^2 (Table 1). The patient also had a hypoplastic aortic arch and CoA was not definite (Fig. 1). The patient underwent emergent bilateral pulmonary artery banding using a 3.5-mm Gore-Tex graft as a rescue procedure. Immediately after the surgery, the mean left atrial pressure (LAP) decreased from 33 to 24 mmHg and the systolic blood pressure increased from 64 to 76 mmHg. Six hours after the surgery, the LAP had decreased to 13 mmHg and became almost normal 12 hours later (Fig. 2) owing to reduction of pulmonary blood flow and LV volume unloading.

On postnatal day 15, follow-up echocardiogram due to gradual development of differential cyanosis revealed diminished PDA size from 8 to 3 mm in addition to distinct CoA (Table 1). The LV EF was 84.4%, and the MV annulus and AV annulus diameters were 6.8 mm (Z-score=−4.00) and 5.0 mm (Z-score=−3.50), respectively. To determine the LV volume and function, heart CT was performed on postnatal day 20, which showed that the LV EDV changed from 35 to 29 mL/m^2 and the LV ESV from 24 to 10 mL/m^2, with increased LV wall thickness and improved wall motion (Table 1, Fig. 3). On postnatal day 21, the patient underwent biventricular (BV) repair including pulmonary artery debanding, extensive aortic arch augmentation with main pulmonary artery (MPA) patch and end-to-end anastomosis, MPA and right pulmonary artery angioplasty using glutaraldehyde-fixed pericardium, and creation of an atrial septal defect (ASD, 3.5 mm). ASD creation was performed in order to better support the systemic flow while preventing the left heart from ex-

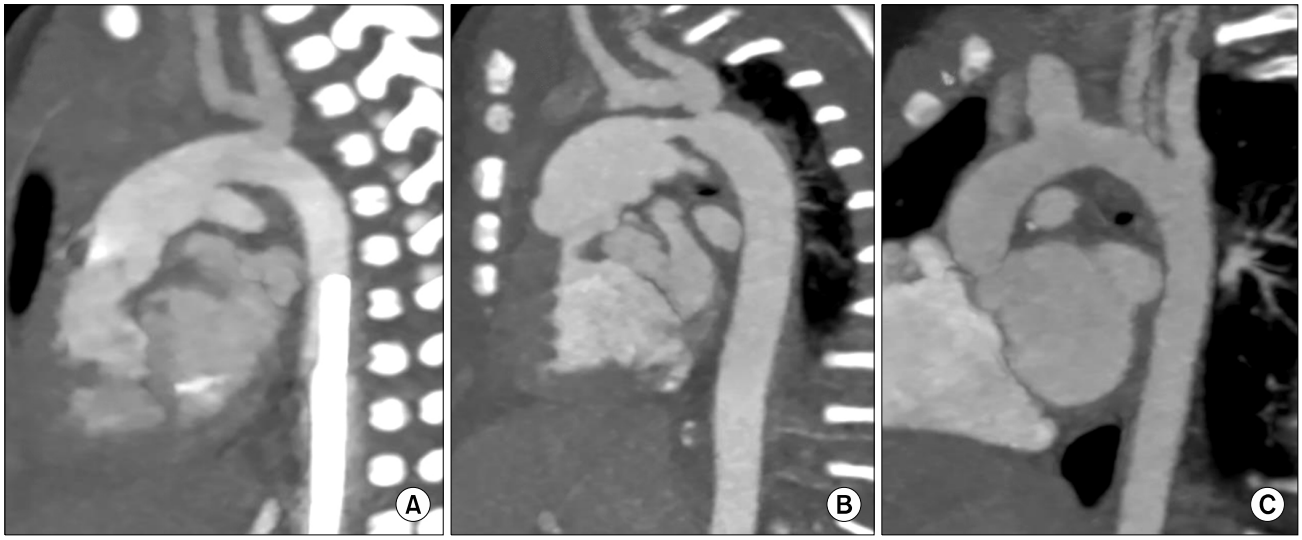

Fig. 1. Serial follow-up of the sagittal view of the aorta. (A) On two days after the birth. (B) On day 20 of his life after pulmonary artery banding surgery. (C) On day 136 of his life after the biventricular repair surgery.

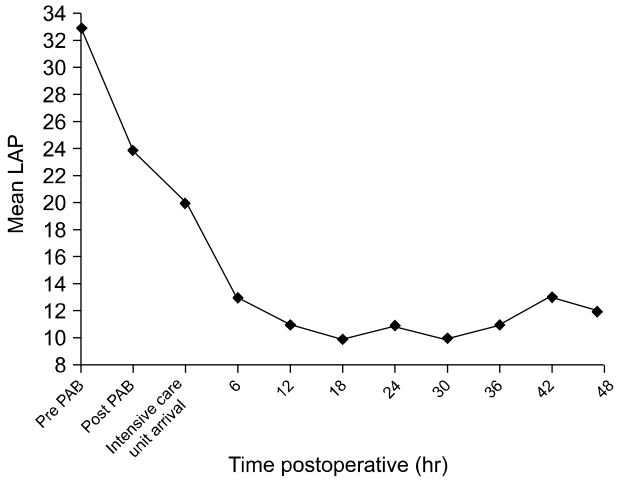

Fig. 2. Trend of LAP until 48 hours after PAB. LAP, left atrial pressure; PAB, pulmonary artery banding.

cessive overload and failure [1]. All procedures were performed under beating heart, and selective cerebral perfusion was instituted by prosthesis-aided cannulation of the right brachiocephalic trunk with moderate hypothermia at 28°C to 30°C. The cardiopulmonary bypass time and selective cerebro-myocardial perfusion time were 141 and 36 minutes, respectively. To avoid compression by the sternum, the patient was transferred to the intensive care unit with an open sternum and delayed sternal closure was performed the next day. During this period, his vital signs and LAP were within a normal range. The patient was extubated on postoperative day 5 and was discharged on postoperative day 13 without complications. A postoperative follow-up echocardiogram demonstrated normal ventricular function, no mitral or aortic stenosis, no residual stenosis at the arch repair site, and iatrogenic ASD (3 mm). The diameters of the MV and AV annulus had increased to 8.6 mm (Z-score=−2.41) and 5.9 mm (Z-score=−1.85), respectively (Table 1). The patient was clinically well seven months following his surgery and with similar echocardiographic and CT findings (Table 1, Fig. 1).

DISCUSSION

HLHS comprises congenital structural cardiac malformations characterized by variable underdevelopment of the structures in the left heart-aorta complex [2]. Those at the favorable end of this group have been defined as having hypoplastic left-heart complex (HLHC), which presents with aortic and mitral valve hypoplasia without intrinsic valve stenosis or atresia, hypoplasia of the LV, hypoplasia of the left ventricular outflow tract, and hypoplasia of the ascending aorta and of the aortic arch and/or CoA [1]. Surgical decisions are not difficult when patients present at the extreme end of the spectrum, which implies univentricular (UV) or BV repair.

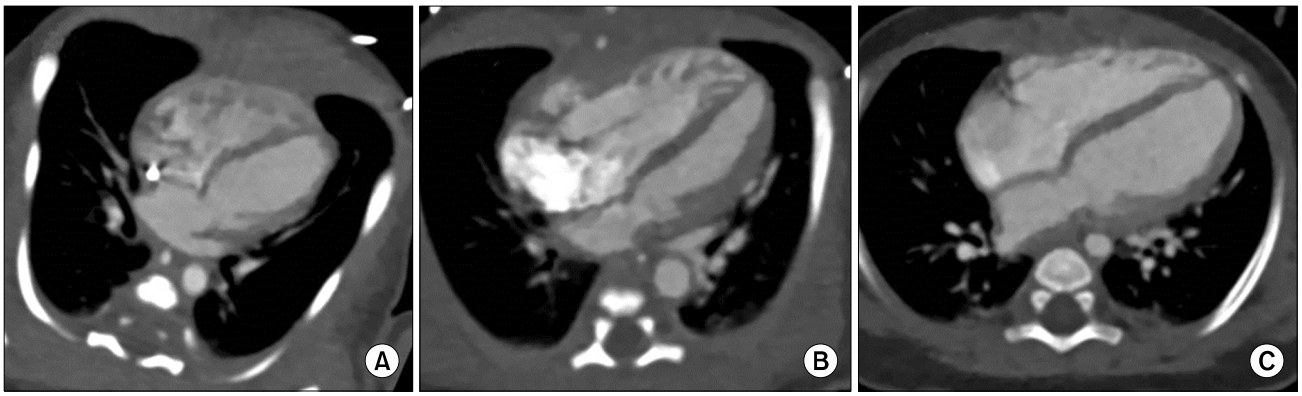

Fig. 3. Serial follow-up of the axial view of the heart. (A) On two days after the birth, diffuse thinning of LV wall is found. (B) On day 20 of his life after pulmonary artery banding surgery, increased LV wall thickness and enlarged right cardiac chambers are seen. (C) On day 136 of his life after the biventricular repair surgery, increased LV size and interval decrease in right cardiac chambers are found. LV, left ventricular.

However, it is sometimes difficult to decide which surgical correction to perform in patients with HLHC, due to the absence of absolute criteria and because inadequate surgical management for these patients can lead to terrible consequences [3].

Since Rhodes et al. [4] first reported the parameters associated with predicted survival following BV repair in patients with critical aortic stenosis, such as a cut-off value for the indexed mitral valve area of 4.75 cm^2/m^2, a left ventricular length to heart long-axis ratio of 0.8, or an indexed aortic root size of 3.5 cm/m^2, there has been limited success in determining the criteria for which patients have sufficient left heart structure and function to support the systemic circulation. This is largely because most previous reports dealt with critical aortic stenosis alone, and applying a cut-off value targeted to simple left-heart hypoplasia was not valid in patients with HLHC [5,6]. Indeed, the decision to pursue a UV or BV repair is generally made by experienced pediatric cardiologists and cardiac surgeons based on echocardiographic images, volume-measured heart CT, and/or information obtained from cardiac catheterization of the left heart.

In the clinical field, it is sometimes too difficult to decide which type of surgical correction to perform in neonates with HLHC, especially in those whose initial vital signs are unstable, as in our patient. The consequences of an incorrect decision, such as conversion to UV circulation after a failed BV repair, are often deleterious, whereas the reverse sequence of these procedures seems to have a favorable and successful outcome [4]. Therefore, in cases of HLHC, an initial palliation surgery including bilateral pulmonary artery banding, augmentation of the right ventricular systemic flow, and maintaining the ductus patency by stenting or prostaglandins could be an alternative option without the need for an early decision and major surgery [6]. Although one-stage BV repair has the advantage of establishing normal physiology at an earlier point in time, Yerebakan et al. [7] reported that initial palliative surgery for neonates with HLHC allows time for the growth of the LV, thus avoiding major surgery and the associated prolonged myocardial ischemia and bypass during the neonatal period. The UV and BV options could be retained as alternatives to be performed at around four to eight months of age with satisfactory long-term outcomes [7]. Although the period from palliative surgery to BV repair was relatively short in our patient, the diminished PDA size and simultaneous development of CoA and improved LV contractility that occurred in the meantime enabled us to perform BV repair (Table 1).

In conclusion, we have successfully achieved BV repair after first-stage palliation for a neonate with HLHC, a subset of HLHS. In selected patients with HLHC, bilateral pulmonary artery banding is a suitable alternative surgical option that could delay early decision-making on more invasive and risky procedures. It serves as an initial rescue procedure in patients who are clinically unstable. Although there is no absolute cri-

teria for which type of surgical repair to perform for neonates with HLHC, meticulous assessment of the left heart-aorta complex and timely surgical treatment that considers the patient's vital signs may be required for optimal treatment.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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