

84. A Neonate of Complex Heart Anomaly With Borderline Left Ventricle Who Underwent Successful Biventricular Repair Using Multi-Staged Prudent Intervention

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Body

Background: For several decades, there has been no definitive treatment in managing borderline left ventricle. However, as we are suffering from unfavorable long-term outcome of Fontan procedure, the challenging biventricular repair became the best to pursue in any case.

Case: A 23-day-old neonate with partial atrioventricular septal defect, single atrium, severe atrioventricular valvular regurgitation and borderline left ventricle (LV) combined with left isomerism was transferred. Her inferior vena cava was interrupted, and superior vena cava and hepatic veins drained to left atrium whereas pulmonary venous connection was usual. The estimated LV volume by transthoracic echocardiography was 30–35 ml/m² at first. We planned staged operation and firstly performed mitral valve cleft repair. However, she continuously showed pulmonary edema with significant heart failure symptoms. We tried pulmonary artery banding to reduce shunt flow within single atrium. Since it worked for a while, she could be extubated, but consistently needed high-flow nasal cannula support. Also, her weight remained the same for more than a month. At her age of 4.7 months, we re-evaluated her via dual phase computed tomography that showed well-grown LV volume measuring about 90 ml/m² at end-diastole. Eventually we performed intraatrial baffling using Mustard procedure at 5 months and 4.6 kg. Although she had to undergo another procedure of pacemaker insertion due to postoperative sick sinus syndrome, she was discharged at her age of 6 months.

Discussion: The issue that how small ventricle can work as a whole ventricle supporting systemic circulation by itself is the question to solve. Encountering and managing this difficult case, we could experience such a complicated decision-making process deciding whether to use the LV as a systemic ventricle or not. Although it was a very long and tough process, we learned a lesson that putting a great effort into dealing with insufficient ventricle may lead to successful biventricular repair even in case of complex heart anomaly.

