Beating-Heart Surgery for Hypoplastic Left Heart Syndrome With Coronary Artery Fistulas
Shunsuke Matsushima, MD, Yoshihiro Oshima, MD, PhD, Ayako Maruo, MD, PhD, Tomomi Hasegawa, MD, PhD, Hironori Matsuhashi, MD, PhD, Rei Noda, MD, and Ryuma Iwaki, MD
Department of Cardiovascular Surgery, Kobe Children’s Hospital, Kobe, Japan

The presence of ventriculocoronary connections in patients with hypoplastic left heart syndrome and mitral stenosis-aortic atresia is a suggested risk factor for myocardial ischemia after surgical palliation. We describe a neonate with this anatomic variant of hypoplastic left heart syndrome who underwent a successful beating-heart Norwood operation, with continuous coronary perfusion. The ventricular condition could be visually confirmed during the procedure, and a postoperative echocardiogram showed preserved ventricular function. This technique is considered an effective option for minimizing myocardial damage in this patient subtype.

Ventriculocoronal connections (VCCs), also called ventriculocoronal fistulas or sinusoids, are frequently seen in patients with the mitral stenosis-aortic atresia (MS-AA) subtype of hypoplastic left heart syndrome (HLHS) [1, 2]. The presence of VCCs is a risk factor for early failure after stage I palliation [1]. However, the exact mechanism for the increased mortality in this subtype remains to be elucidated. Here, we describe a neonate with HLHS and extensive VCCs who underwent a successful, modified, beating-heart Norwood operation, with continuous coronary perfusion.

A 2.93-kg male neonate was born at 39 weeks gestation, with a prenatal diagnosis of HLHS and MS-AA, requiring prostaglandin E1 infusion. Transthoracic echocardiography confirmed the diagnosis and the presence of extensive VCCs (Fig 1). Color Doppler flow mapping showed prominent systolic antegrade flow through the VCCs and diastolic retrograde flow through the 4.5-mm diameter ascending aorta. Gentle balloon atrial septostomy was performed and subsequent bilateral pulmonary artery banding was performed on postnatal day 3. In the immediate postoperative period transthoracic echocardiography showed transient ventricular dysfunction, owing to the decreased left ventricular preload, that was resolved by volume loading.

Fig 1. Preoperative, color Doppler flow mapping echocardiograms. (A) Parasternal short-axis view showing extensive ventriculocoronary connections (white arrows). (B) Parasternal long-axis view showing prominent antegrade systolic flow in the ascending aorta (yellow arrow). (aAo = ascending aorta; LA = left atrium; LV = left ventricle; RV = right ventricle.)
During the reconstruction of the aortic arch, right ventricle ischemic changes were observed. Then, an 18-gauge cardioplegia spike was placed in the proximal ascending aorta and connected to the side arm of the arterial circuit (Fig 2). Soon after the start of proximal coronary perfusion complete recovery of right ventricular contractions was confirmed and maintained during atrial septectomy, arch reconstruction completion, and right ventricle to pulmonary artery (RV-PA) shunt placement, using a 6-mm GoreTex tube. Finally, the patient was weaned from the CPB without any complications.

The patient’s postoperative course was uneventful. Repeated echocardiography revealed preserved right ventricular function and an unchanged pattern of coronary perfusion. A bidirectional Glenn shunt was performed when the patient was 5-months old. During the Glenn anastomosis, ventilation was continued, and the left pulmonary blood flow was maintained through the RV-PA shunt to preserve left atrial oxygen saturation. After establishing the Glenn shunt, the RV-PA conduit was subtotally excised. At the 6-month follow-up visit, the patient’s condition was stable, with preserved ventricular function, and he was waiting for a Fontan procedure.

**Comment**

The presence of VCCs in patients with HLHS and MS-AA has been implicated as a risk factor for increased hospital mortality after surgical palliation. Vida and colleagues [1] found a 50% failure rate for stage I palliation in patients with this anatomic variant and Sathanandam and colleagues [2] suggested that large VCCs were associated with a poor prognosis. Although the exact mechanism for the increased mortality in this disease subtype remains unclear, some authors have proposed that the VCCs might interfere with adequate myocardial protection during heart decompression on CPB, resulting in ischemia-induced ventricular dysfunction [1, 3]. This condition can be referred to as left ventricle-dependent coronary circulation, as a similar condition involving the right side of the heart in patients with pulmonary atresia and intact ventricular septum is called right ventricle-dependent coronary circulation. We hypothesized that the left ventricular systolic pressure might play a vital role in creating coronary perfusion pressure, and that left ventricular collapse could be prevented by proximal coronary perfusion in patients with extensive VCCs. In the present case, therefore, we employed the modified Norwood operation on a beating heart with continuous coronary perfusion and ventricular contraction.

Kishimoto and colleagues [4] first reported the beating-heart Norwood operation, in which continuous coronary perfusion was established using flow from the brachiocephalic artery with an aortic cross-clamp at the level of the proximal arch [4]. Since then, several modifications have been described [5, 6], including the use of continuous coronary flow using a cardioplegia spike or an olive-tip cannula secured in the proximal ascending aorta [6]; the authors reported that this modification was a safe alternative for use with any size of aorta. For smaller ascending aortic diameters, an olive-tip cannula connected to the arterial circuit can be placed into the lumen of the opened aorta and secured with a vessel loop tourniquet. In addition to their technique we employed mild hypothermic CPB, with lower body perfusion [7], to further maintain the ventricular contractions.

Despite the use of this beating-heart strategy, a short period of atrial decompression during atrial septectomy is unavoidable. In addition, the efficacy of this technique for patients with the severest forms of VCCs, such as coronary artery interruption, remains unclear. However, the myocardial condition can be visually checked during the procedure, which is another advantage of this beating-heart technique.

In summary, the beating-heart Norwood operation, with continuous coronary perfusion, is an effective option for providing myocardial protection in HLHS patients with MS-AA and large VCCs.

**References**


