Case report - Congenital
The bilateral pulmonary artery banding for hypoplastic left heart syndrome with a diminutive ascending aorta
Takahiro Tomoyasu, Kagami Miyaji*, Takashi Miyamoto, Nobuyuki Inoue
Department of Cardiovascular Surgery, Kitasato University School of Medicine, Sagamihara, Japan
Received 31 August 2008; received in revised form 4 December 2008; accepted 8 December 2008

Abstract
A one-day-old neonate who was diagnosed with hypoplastic left heart syndrome (HLHS), aortic atresia, with a diminutive ascending aorta, and mitral atresia, was referred to us for cardiogenic shock because of excessive pulmonary blood flow. The patient underwent bilateral pulmonary artery banding (bPAB). After bPAB, the patient’s hemodynamics were still unstable because of coronary malperfusion, to proceed to undergo Norwood procedure at the age of 3 days. In this case, the stenosis of the ascending aorta, just proximal to the innominate artery caused coronary ischemia. The precise evaluation of the ascending aorta is necessary to perform the bPAB for HLHS with diminutive ascending aorta. If there is a sign of stenosis of the ascending aorta, the Norwood procedure should be performed as the first stage palliation, even for high-risk HLHS patients.

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Keywords: Aortic arch; Hypoplastic left heart syndrome; Norwood; Ischemia; Pulmonary arteries

1. Introduction
Surgical results for hypoplastic left heart syndrome (HLHS) are improving with resulting higher survival rates [1–3]. Nowadays, new indications for pulmonary artery banding have been considered in HLHS, either as a rescue procedure in critical neonates or as an elective preparation for the subsequent surgical stage. Because a surgical palliation using a cardiopulmonary bypass was considered to be invasive, the bilateral pulmonary artery banding (bPAB) was introduced to high-risk HLHS patients. Here, we report a case of an unsuccessful bPAB, followed by the Norwood procedure for HLHS with a diminutive ascending aorta.

A 2.9 kg female newborn had severe respiratory distress that developed shortly after birth. Two-dimensional echocardiography revealed HLHS with aortic atresia, with a diminutive ascending aorta, mitral atresia, and atrial septal defect. The diameter of the patent ductus arteriosus (PDA) was 6 mm, and the right ventricular function was normal. Prostaglandin E1 was administered. On day 1 (actually 12 h after birth), her hemodynamics became unstable because of excessive pulmonary blood flow, and her arterial oxygen saturation increased up to 97%. Thus, she was intubated and managed with mechanical ventilation. Because the Norwood procedure was considered to be too risky, bilateral pulmonary artery banding (bPAB) was performed on the same day.

*Corresponding author. Department of Thoracic and Cardiovascular Surgery, Kitasato University School of Medicine, 1-15-1 Kitasato, Sagamihara, Kanagawa 228-8555, Japan.
E-mail address: kagami111@aol.com (K. Miyaji).
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2. Bilateral pulmonary artery banding
A median-sternotomy was performed. The diameter of ascending aorta was 2 mm. Bilateral pulmonary artery bandings were dissected and encircled. The bPAB was performed to create a circumference on the right 10 mm and on the left 9 mm by polytetrafluoroethylene (PTFE) banding tape. The right banding tape was placed just proximal to the right upper branch, away from the ascending aorta. Arterial oxygen saturation decreased from 95% to 80%, and the systolic systemic blood pressure increased from 45 mmHg to 60 mmHg.

Three hours after surgery, her hemodynamics became unstable again. When the systemic blood pressure became <60 mmHg, the ST segment on the ECG depressed and soon after the blood pressure went down with bradycardia. To maintain the blood pressure, an epinephrine transfusion was needed. The echocardiogram showed a poor single ventricular function (ejection fraction 38%) and stenosis of the ascending aorta (1.5 mm in diameter), just proximal to the innominate artery (Fig. 1). The patient’s hemodynamics was still unstable, because of coronary malperfusion, to proceed to undergo the Norwood procedure on day 3.

3. The Norwood procedure
A re-median sternotomy was performed. The banding tapes did not migrate, and the right banding tape and right pulmonary artery did not affect the ascending aorta. There was a stenosis just proximal to the innominate artery in the ascending aorta. A 3.5 mm-PTFE tube graft was anastomosed to the innominate artery as an arterial line. The cardiopulmonary bypass was started with bicaval venous
drainage. The patient was cooled down to 25 °C. The atrial septal defect was enlarged, and the PDA was ligated and divided during cooling. The main pulmonary artery was transected just proximal to the branch, and the PTFE patch was anastomosed. Then a 5 mm-PTFE tube graft was secured to the distal orifice of the pulmonary artery. The ascending aorta was transected at the stenotic site, the diameter of this portion was approximately 1.5 mm, and a cardioplegic solution was given using a 4-Fr tube. After cardiac arrest, the ascending aorta was incised vertically down to the sinus level and anastomosed to the main pulmonary artery in a side-to-side fashion to maintain sufficient coronary blood flow. Under the regional cerebral perfusion, the descending aorta was clamped. The ductal tissue was removed, and the ridge on coarctation was resected. The lesser curvature of the aortic arch was incised and the main pulmonary artery anastomosed directly to the aortic arch (Fig. 2). The right ventricular pulmonary artery conduit (RV-PA conduit) was contracted through the right side of the neo-aorta using a 5-mm ePTFE graft, which was anastomosed to the patch-closed pulmonary orifice (Fig. 2). Because of the emergent Norwood procedure under cardiogenic shock, a delayed sternal closure was performed nine days after surgery. The patient was discharged without any major complications. She underwent bidirectional Glenn shunt at three months old and Fontan procedure at 18 months old without any complications.

4. Comment

The short-term and long-term results for patients with HLHS have continued to improve due to modification of surgical and medical management [1, 2]. bPAB is performed widely as the first procedure for HLHS [4–6]. Recently the hybrid procedures of bilateral pulmonary artery banding and ductal stenting are common [4–7]. After ductal stenting, coronary ischemia, also called retrograde coarctation, was reported [5].

In the present report, bPAB was selected as the first stage palliation, followed by prostaglandin E1 administration, to maintain PDA flow instead of ductal stenting. We experienced coronary ischemia because of critical stenosis of the small ascending aorta, which had not been evaluated precisely before bPAB. After birth, the deterioration of the patient’s hemodynamics was very fast in this case, which suggested that the coronary malperfusion caused by the stenosis of ascending aorta may have affected her hemodynamics.

Preoperative accurate assessment of the retrograde, transverse aortic flow, using echocardiography is important for patients with diminutive ascending aorta. However, in our case, the stenosis of ascending aorta was not diagnosed, before bPAB. The patient’s clinical course was extraordinary, because 12 h after birth, her hemodynamics became unstable. In such cases, more precise assessment of the retrograde aortic flow, using echocardiography should be necessary. In order to rule out this critical stenosis, the multi-slice CT-scan following the 3-D images of aortic arch may be useful. If there is a sign of stenosis of the ascending aorta, the Norwood procedure should be performed as the first stage palliation, even for high-risk HLHS patients.

References

