

Emergency Pulmonary Artery-to-Systemic Artery Shunt to Break the Positive Feedback Loop of a Pulmonary Hypertensive Crisis after Neonatal Coarctation Repair

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A 2.5-kg neonate with coarctation of the aorta and a small left ventricle experienced a severe pulmonary hypertensive crisis. An emergency pulmonary artery-to-systemic artery shunt was placed to break the positive feedback loop caused by pulmonary hypertension and functional mitral stenosis. This shunt provided immediate relief of suprasystemic pulmonary hypertension and the resultant low cardiac output.

Key words: 1. Hypoplastic left heart syndrome
2. Pulmonary hypertension
3. Aortic coarctation
4. Shunts

Case report

Underdevelopment of the left ventricle is common in neonates with aortic coarctation [1,2]. Despite the small size of the left ventricle at the initial presentation, many patients demonstrate catch-up growth of the left ventricle after successful coarctectomy [3,4]. However, some patients suffer from low cardiac output generated by a vicious cycle in which small left ventricular volume results in diastolic failure, which aggravates pulmonary hypertension, further compressing the left ventricle early after the coarctectomy. This devastating event does not preclude the possibility of successful biventricular repair because many patients are stabilized after immediate post-operative adaptation.

We report the case of a male neonate with coar-

tation of the aorta with a small left ventricle who overcame a pulmonary hypertensive crisis with a pulmonary artery-to-systemic artery shunt after coarctectomy.

A male neonate with a birth weight of 2.05 kg at 35 weeks of gestation was diagnosed with coarctation of the aorta after birth. Echocardiography demonstrated aortic coarctation with a hypoplastic aortic arch and a small aortic valve annulus (5 mm; Z-score, -2.34). The left ventricle was small, measured as 73% of the nomogram. No ventricular septal defect was found, and the patent ductus arteriosus was 4 mm in size. A 3-mm atrial septal defect was present. Because the neonate was delivered prematurely, he was put on mechanical ventilation, and repair was delayed as long as pulmonary congestion did not develop. The baby underwent resection of aortic

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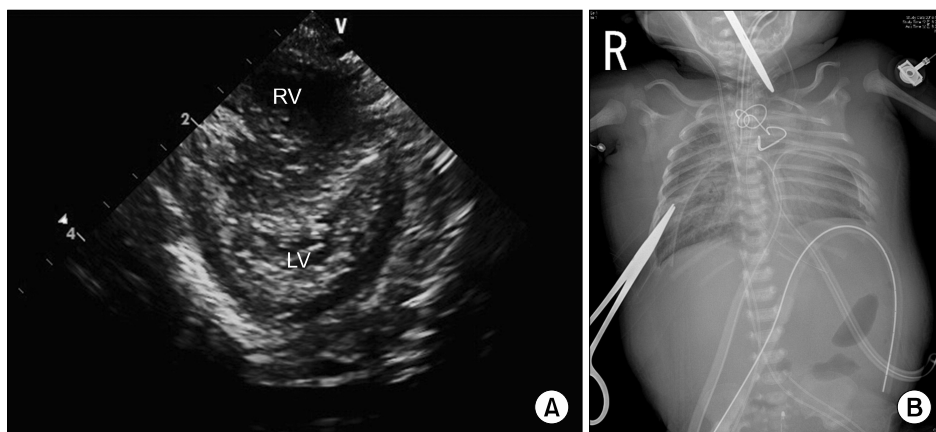


Fig. 1. (A) Postoperative echocardiogram after coarctation repair demonstrating the LV being squashed by the RV with suprasystemic pressure. (B) Chest radiography after coarctation repair demonstrating severe pulmonary congestion. LV, left ventricle; RV, right ventricle.

coarctation and extended end-to-side anastomosis under regional cerebral perfusion and moderate hypothermia at 28 days after birth when his body weight reached 2.5 kg. The neonate was doing well immediately after the repair, but soon developed progressive low cardiac output. Echocardiography showed a smaller left ventricle with interventricular septal bowing (Fig. 1A). The impinged mitral valve developed functional mitral stenosis, and the left atrium was dilated. Pulmonary venous congestion was evident on chest radiography (Fig. 1B). Suprasystemic pulmonary hypertension led to progressive dilation of the right ventricle, resulting in a more compressed left ventricle. His systemic blood pressure was approximately 50 mm Hg (systolic). Severe hypercarbia and hypoxemia caused by pulmonary venous congestion worsened despite high-frequency oscillatory ventilation with maximum settings. No improvement was shown despite conventional medical treatment using inotropes and inhaled nitric oxide. Emergency placement of a pulmonary artery-to-systemic artery shunt was performed instead of giving up on the left ventricle by performing a Norwood procedure (Fig. 2). Systemic blood pressure and oxygen saturation could not be maintained with a conventional anesthesia machine. Therefore, inhalation of nitric oxide and high-frequency oscillatory ventilation were maintained intraoperatively. The main pulmonary artery was partially clamped with a Satinsky clamp, and we waited for a few minutes to ensure that severe desaturation did not occur. Heparin (150 IU/kg) was administered intravenously. Subsequently, the innominate artery was snared both proximally and distally. The innominate artery was opened along the

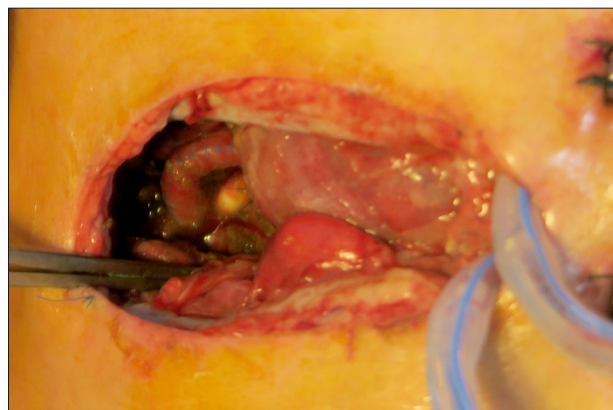


Fig. 2. Main pulmonary artery-to-innominate artery shunt with a 4-mm expanded polytetrafluoroethylene vascular graft.

previous cannulation site, and the distal end of a 4-mm expanded polytetrafluoroethylene vascular graft was anastomosed in an end-to-side manner. The main pulmonary artery was partially re-clamped, and the proximal anastomosis of the shunt was completed. An intraoperative echocardiogram demonstrated flow from the main pulmonary artery to the innominate artery through the shunt (Fig. 3A). The compressed left ventricle partially recovered its volume. The dilated right ventricle was relieved. Peripheral oxygen saturation remained at approximately 80% to 90%, and systemic blood pressure increased to 70 mm Hg (systolic). The suprasystemic right ventricular pressure was effectively relieved with the shunt. Cardiac output stabilized in 2 days after placement of the shunt. Peripheral oxygen saturation gradually increased to the mid-90% range, and serial echocardiography revealed changes in the bi-

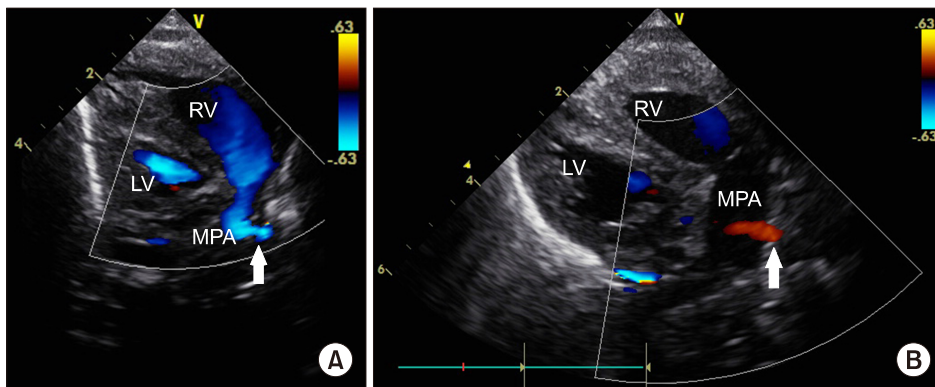


Fig. 3. (A) Right-to-left shunting through the MPA-to-innominate artery shunt (arrow). (B) Reversal of the MPA-to-innominate artery shunt direction (arrow). LV, left ventricle; RV, right ventricle; MPA, main pulmonary artery.

directional shunt, as it converted to a continuous left-to-right shunt (Fig. 3B). On day 3, the patient's diastolic blood pressure remained relatively low, mimicking the left-to-right shunting of a large patent ductus arteriosus. Therefore, the shunt was partly closed with a clip and completely closed a day later with sternum closure. The neonate showed adequate cardiac output afterwards, and pulmonary congestion improved.

Institutional review board approval was waived due to the retrospective nature of the report.

Discussion

The immediate postoperative period after coarctectomy in patients with a small left ventricle is critical. Despite good contractile function of the ventricles, patients with a small left ventricle are prone to developing functional mitral stenosis. This may lead to the aggravation of pre-existing pulmonary hypertension [3]. In addition, surgical stress from the complex operation and the vulnerability of pulmonary vascular resistance in newborns contribute to the development of suprasystemic pulmonary hypertension. The dilated right ventricle with suprasystemic pressure compresses the left ventricle, leading to the vicious cycle of a pulmonary hypertensive crisis and low cardiac output. Although this devastating phenomenon is potentially fatal, it is not appropriate to conclude that biventricular repair will necessarily be inadequate in these patients. After stabilization from the immediate postoperative changes, many patients do not experience low cardiac output and eventually show adequate catch-up growth of the left ventricle. Therefore, it is ideal to overcome the acute, unstable

period rather than to abandon the small left ventricle and proceed to the Norwood procedure. In the setting of the aforementioned positive feedback loop of a pulmonary hypertensive crisis, a pulmonary artery-to-systemic artery shunt is an effective way to alleviate compression of the left ventricle by the suprasystemic right ventricle. From a technical point of view, moreover, placement of the shunt is extremely simple and safe, even in a severely decompensated patient. The size of the shunt tube was chosen based on clinical judgment; however, a 4-mm tube was effective in relieving the suprasystemic right ventricular pressure without acute volume overload to the left ventricle when the pulmonary hypertension improved.

In conclusion, suprasystemic pulmonary hypertension after coarctectomy leads to severe low cardiac output in patients with a small left ventricle. A pulmonary artery-to-systemic artery shunt is an effective and safe method to relieve right ventricular pressure and restore cardiac output in an acute setting.

Conflict of interest

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

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