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Total neoaoorta graft replacement with faucet-like coronary reconstruction technique and double valve replacement 17 years after the Norwood procedure

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Glossary of abbreviations

HLHS: hypoplastic left heart syndrome

PCWP: pulmonary capillary wedge pressure

AVV: atrioventricular valve

Central message

Maximum utilization of the anatomical features after the Norwood procedure can minimize the operative risk.

Central picture legend

Total neoaorta graft replacement with faucet-like coronary reconstruction technique.
**Case Presentation**

A 17-year-old male patient with hypoplastic left heart syndrome (HLHS) underwent the Norwood procedure (direct anastomosis, modified Blalock-Taussig shunt) and bidirectional Glenn procedure at 3-month-old, followed by the Fontan procedure (extracardiac, 16-mm expanded polytetrafluoroethylene conduit) at 2-year-old. This patient was referred to our hospital for neoaortic aneurysm (diameter of neoaorta was 60 mm [Figure 1]) and severe regurgitation of the neoaortic and atrioventricular valves. Catheterization demonstrated that Fontan pressure, pulmonary capillary wedge pressure (PCWP) and arch gradient were 8, 4 and 11 mmHg respectively, and cardiac index was 1.8 L/min/m². IRB approval was not required; patient consent was orally received for publication; there is potentially identifiable information in this article.

Cardiopulmonary bypass was established after median resternotomy (the left axillary and femoral arteries for perfusion, superior vena cava and right femoral vein for drainage). The neoaorta was clamped proximal to the connection site with the original aorta (Figure 1A), and AVV replacement was performed under electrically induced ventricular fibrillation. The AVV was severely redundant with five-segmented valves, and replaced with a 25/33-mm ON-X mechanical valve (Life Technologies). Moderate hypothermic circulatory arrest was initiated.
at 25°C, and antegrade selective cerebral perfusion was performed. Cardioplegia was delivered through the original aorta. The aortic isthmus was augmented with graft patch. Subsequently, the neoaortic root and arch were replaced with a mechanical valved composite graft (a 22-mm ATS mechanical valve [Medtronic] and a 26-mm 4-branched J-Graft [Japan Lifeline]), followed by anastomosis of brachiocephalic artery. An additional graft was bridged the original aorta and branch graft serving the brachiocephalic artery (Figure 2A). After coronary reperfusion, the left carotid and left subclavian arteries were anastomosed with a 4-branched graft (Video 1). Epicardial pacemaker leads were attached to the ventricle. The perioperative course was acceptable (bypass time: 379 minutes, antegrade cerebral perfusion time: 181 minutes, cardiac ischemic time: 170 minutes, postoperative intubation time: 43 hours, length of intensive care unit stay: 5 days, length of hospital stay: 35 days). Dopamine and dobutamine (maximum 6 mcg/kg/min for both) were used as postoperative inotropes.

One-year postoperative follow-up revealed no valve dysfunction. Catheterization revealed that Fontan pressure, PCWP and arch gradient were 12, 6 and 7 mmHg respectively, and cardiac index was 3.0 L/min/m² (Figure 2B). The patient’s condition remained favorable.

**Comment**

The early outcomes after Norwood procedure have improved. Various sequelae in the late post-Norwood period and reoperations for these sequelae have been reported.
Considering surgical indications in patients with staged repaired HLHS, it is crucial to consider their quality of life and reduce the need for future reoperations. In this case, despite the patient’s youth, we performed neoaortic root replacement rather than valve-sparing surgery, due to the need for a mechanical valve in AVV replacement, simplification of the surgical procedure and reduction the need for future reoperations.

Reoperation for patients with staged repaired HLHS is complicated and high risk. The surgical procedure should be designed to reduce the operative risk. In this case, by clamping the neoaorta proximal to the connection site with the original aorta, AVV replacement under ventricular fibrillation while maintaining coronary perfusion and controlling the neoaortic regurgitation was successful. This approach contributed to reducing the myocardial ischemia time. In addition, the faucet-like coronary reconstruction technique was expected to ensure coronary perfusion without bending. Ensuring reliable coronary perfusion is essential for a vulnerable single right ventricle. We also repaired the coarctation of aorta with elevated cardiac afterload that unfavorable effect on Fontan patient.

One-year postoperative follow-up revealed a good cardiac index with maintaining low PCWP. This report highlights that the maximum utilization of the anatomical features of patients with HLHS can minimize the operative risk. Our procedure can provide valuable insights for the management of patients with staged repaired HLHS.
References


Figure legends

Figure 1

Preoperative computed tomography image. (A) Anterior view showing marked dilation of the neoaorta. The black arrow shows connection site between original aorta and neoaorta. The dashed line shows neoaortic clamp site. (B) Left lateral view showing coarctation of aorta (black arrow). (Ao = original aorta; Neo Ao = neoaorta)

Figure 2

Postoperative image. (A) three-dimensional computed tomography image (Ao = original aorta; X = 26-mm 4-branched graft; Y = Faucet-like graft ). (B) X-ray fluoroscopy (left lateral view).

Video 1

This video demonstrates the surgical procedure of atrioventricular valve replacement, neoaortic root replacement with faucet-like coronary reconstruction technique, total arch replacement and recoarctation repair.