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

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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 months of age, followed by the Fontan procedure (extracardiac, 16-mm expanded polytetrafluoroethylene conduit) at 2 years of age. This patient was referred to our hospital for a neoaortic aneurysm (diameter of the 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 mm Hg, respectively, and cardiac index was 1.8 L/min/m². Institutional review board 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 1, A) 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 mm Hg, respectively, and cardiac index was 1.8 L/min/m². Institutional review board approval was not required; patient consent was orally received for publication; there is potentially identifiable information in this article.

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respectively, and cardiac index was 3.0 L/min/m² (Figure 2, B). 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 neo-aortic 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 to reduce cardiac afterload which adversely affects Fontan patients. One-year postoperative follow-up revealed a good cardiac index with maintaining low PCWP.

This report highlights that the maximum use of anatomic features of patients with HLHS can minimize the operative risk. Our procedure can provide valuable insights for the management of patients with staged-repair HLHS.

Conflict of Interest Statement
The authors reported no conflicts of interest.

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