A new surgical challenge: Reconstruction of aortic arch aneurysm late after the Norwood procedure in an adult with Fontan circulation

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With increasing survival into adulthood after Norwood-type reconstruction, there is a growing population of patients with aneurysmal degeneration of the reconstructed ascending aorta and aortic arch.1 A redo aortic arch reconstruction poses significant risk in patients with Fontan circulation.

CLINICAL SUMMARY
A 27-year-old patient, with original diagnosis of hypoplastic left heart syndrome variant and tricuspid atresia with malposed great vessels, underwent Norwood and subsequent Fontan operation in in the neonatal period, and presented with an asymptomatic aortic arch aneurysm, dilated neoaortic root, severe neoaortic valve regurgitation, and mildly reduced ventricular function. Chest computed tomography imaging showed an aneurysmal ascending aorta measuring 63 × 58 × 57-mm abutting the sternum (Figure 1). The surgical challenges include high-risk reentry and arch aneurysm dissection while avoiding left recurrent nerve and left pulmonary artery injury, myocardial and cerebral protection, and recreation of the aortic arch without kinking. The patient’s informed consent was obtained. Valve-sparing root replacement was not feasible in this case because of severe aortic insufficiency in the setting of a heavily dysplastic neoaortic valve. At our institution, there is a low threshold to offer the Bentall procedure. Rather than risking an unsuccessful valve repair, the reliability of valve replacement minimizes repeat sternotomies in Fontan patients, of whom many eventually require a heart transplant, for which there is compounding risk of allosensitization and adhesions associated with multiple operations.

SURGICAL TECHNIQUES
Axillary artery cannulation using a Dacron side graft was performed before redo sternotomy, reducing the risk of sternal reentry, and providing selective cerebral perfusion during arch reconstruction. The chest was entered unevenly, and cardiopulmonary bypass was initiated with axillary arterial and bicaval cannulation. The next challenge was navigating the large aneurysmal aortic arch while dissecting between the aortic arch and the left pulmonary artery. Placement of the crossclamp was managed after careful dissection, and antegrade cardioplegia was delivered to achieve cardiac arrest. The ascending aorta was quickly opened near the Damus–Kaye–Stansel anastomosis and additional direct coronary cardioplegia was given. The aneurysmal portion was resected while maintaining continuity between the descending aorta and arch vessels on the posterior...
aspect. When the patient was cooled to 20 °C for deep hypothermic circulatory arrest. A 26-mm Dacron graft was beveled and anastomosed to the descending aorta and the aortic arch with the patient under full circulatory arrest. The remaining aortic arch reconstruction was performed with the patient under selective cerebral perfusion. To address the acute angle formed by the Dacron graft at the transition from the ascending aorta to the aortic arch, the proximal end of the graft was oversewn in anticipation for the end-to-side anastomosis to the distal Bentall graft. The native pulmonary valve (ie, the neoaortic valve) was excised and a 29-mm Bentall composite mechanical valve was implanted. The distal end of the Bentall graft was then anastomosed to the inferior aspect of the proximal aortic arch graft in an end-to-side fashion. Finally, a rectangular opening was created on the anterior aspect of the ascending aortic graft and the native aorta was reanastomosed to the graft (Figure 2). Cardiopulmonary bypass was terminated without difficulty. The postoperative course was complicated by an acute right middle cerebral artery infarct, acute respiratory distress syndrome, and left vocal cord paralysis. The patient was discharged to stroke rehabilitation on postoperative day 22. At 3 months postrepair, his neurological deficits and vocal cord function significantly improved, and he has returned to work.

DISCUSSION
With more single-ventricle palliation patients surviving into adulthood, neoaortic dilatation after Fontan reconstruction is becoming increasingly recognized. In one study it was reported that 98% of post-Fontan patients develop an aortic root z-score of >2 at a median follow-up of 9.2 years. The long-term concerns of Norwood reconstruction are dilatation of the pulmonary homograft from high-pressure circulation, and cystic medial degeneration of the aortic arch in the absence of connective tissue disease as a result of myxoid matrix proliferation. Seven cases of neoaortic aneurysms after Fontan reconstructions have been previously reported, with ages at the time of repair ranging from 4.5 to 24 years and aneurysm size ranging from 5.4 to 17 cm. Repair techniques involved valve-sparing root replacement, ascending aortic replacement with or without external polytetrafluoroethylene strip reinforcement for neoaortic root downsizing, bio- or mechanical Bentall, and 1 case of aortic arch aneurysm repair using a 2-graft technique with end-to-end anastomosis.

To our knowledge, our 27-year-old male patient is the oldest of reported cases and our report is the first to describe FIGURE 1. Preoperative cardiac computed tomography showing 63 × 58 × 57 mm ascending aortic and aortic arch aneurysm in a 27-year-old patient with Fontan circulation shown in (A) coronal and (B) axial views.

FIGURE 2. Surgical illustration of complete ascending and arch aneurysm repair in Fontan circulation, involving end-to-side aortic graft anastomosis and mechanical Bentall.
end-to-side graft anastomosis to facilitate transition between the arch and ascending neoaorta. This technique navigates the curvature of the aortic arch while minimizing graft kinking associated with single-graft arch reconstruction or inadequate bevelling of an end-to-end graft anastomosis (Figure 3).

The case also highlights the need for lifetime imaging surveillance in late survivors of Norwood reconstructions, as well as the need for expert consensus on timing of imaging surveillance and reintervention thresholds. The indications for surgical intervention include symptomatic presentation, significant neoaortic regurgitation, compromise of the Fontan circulation because of pulmonary artery obstruction secondary to extrinsic aneurysmal compression, and progression of neoaorta dilatation. In asymptomatic and hemodynamically stable patients, reoperation is commonly deferred until implantation of an adult-size graft is feasible. When surgery is decided, careful preoperative planning is paramount for reoperations after Fontan completion because of dense adhesions and tenuous Fontan physiology.

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