Discussion
The present patient is one of the lowest-birth-weight neonates to undergo an arterial switch operation. Recent reports show that the results of arterial switch operations in patients with intramural coronary artery, as well as those of open surgery in very low birth weight babies, are improving.1-3 Therefore, in the present case, an arterial switch operation was favored over an atrial switch operation or a delayed 2-stage arterial switch strategy. In patients with intramural coronary arteries, several techniques of arterial switch operation have been advocated, including the Aubert procedure, the aortocoronary flap plus pericardial hood technique, and the modification of the standard 2-button technique.1,4,5 However, we were reluctant to use any of these techniques. In the Aubert procedure, the placement of the sutures on the neopulmonary arterial intima in close proximity to the intramural artery may cause coronary artery distortion. In the aortocoronary flap plus pericardial hood technique or the modified 2-button technique, the undersized coronary artery may easily twist. Thus, we adopted the new technique that we described; it is technically easy and unlikely to induce coronary insufficiency, even in a very small neonate. At the expense of mild neopulmonary valve regurgitation, surgery-induced coronary insufficiency was eliminated by a large neoaorta–coronary artery tunnel, the absence of coronary artery rotation, and a suture line placed on the valve tissue near the intramural coronary artery.

References

Norwood modification in a patient with hypoplastic left heart and a right aortic arch

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We report a modification of the Norwood procedure in a neonate with a right aortic arch, an aberrant left subclavian artery, and a left ductus arteriosus in a variant of hypoplastic left heart syndrome.

Clinical Summary
A male infant (3.4 kg) was born with unbalanced atrophicventricular septal defect with a hypoplastic left-sided atrophicventricular valve, left ventricle, and aortic valve situs solitus, total anomalous pulmonary venous connection to the right atrium, right aortic arch, aberrant left subclavian artery, left ductus arteriosus, and left descending aorta. The ascending aorta of the right aortic arch was 2 to 3 mm in diameter, and no left aortic arch was present. He underwent a Norwood procedure with a Sano shunt 2 days after birth.

In the operating room, a 3.5-mm polytetrafluoroethylene shunt (Gore-Tex shunt; W. L. Gore & associates, Inc, Flagstaff, Ariz) was sutured to the larger left carotid artery and cannulated. The right atrium was cannulated and the patient was cooled to 18°C. The right aortic arch crossed posterior to the trachea. The left common carotid artery started from the arch rightward of the trachea and passed anterior to the trachea up to the normal left carotid position. Reconstructing the arch posterior to the trachea was not possible owing to compression from the trachea. Therefore, an innovative approach was undertaken to reconstruct the aorta (Figure 1). The arch was divided just distal to the right subclavian artery on the right side of the trachea. Arch reconstruction included a Damus–Kaye–Stansel anastomosis and use of the left common carotid artery as the transverse arch. The left common carotid artery was opened up longitudinally as it crossed over the trachea. This incision was carried proximally to the sinotubular junction of the hypoplastic aorta. After ductal tissue excision, the back wall of the proximal descending aorta was sutured to the opened left common carotid artery. The proximal descending thoracic aorta was spatulated anteriorly and a homograft patch was sutured to augment the left common carotid artery, completing the arch and augmenting the ascending aorta. Arch reconstruction was...
performed under low-flow cerebral perfusion and the atrial septicotomy was done with a brief period of circulatory arrest. While the patient was being rewarmed, the proximal Sano anastomosis was performed. The patient was weaned from cardiopulmonary bypass, and chest closure was delayed.

Follow-up serial echocardiograms revealed unobstructed neo-aorta with no significant gradient across the arch reconstruction. Cardiac catheterization at 3 months of age demonstrated excellent arch repair with a peak systolic gradient of 5 mm Hg into the proximal descending aorta beyond the region of the aortic repair (Figure 2).

Discussion
Coarctation of the aorta with a right aortic arch in a variant of hypoplastic left heart syndrome is extremely uncommon and poses challenging diagnostic and therapeutic dilemmas. Right aortic arch is estimated to be present in about 0.1% of the normal population. The majority of right aortic arches will descend on the right side of the spine with either mirror-image brachiocephalic vessel branching or an aberrant left subclavian artery that arises from the proximal descending aorta behind the esophagus. Some right aortic arches will cross the midline behind the esophagus, descending on the left side of the spine, a variant known as the right circumflex retroesophageal aortic arch.

The most important question raised by this case is how best to surgically approach this lesion. McElhinney and associates reported on 6 patients who underwent surgery for an obstructed right cervical arch; access for repair was through a standard right posterolateral thoracotomy in 3 patients and through a median sternotomy in 3. Patch augmentation aortoplasty was used in 2 patients, a tube graft from the ascending to descending aorta in 2, end-to-side anastomosis of the descending aorta to the proximal arch in 1, and direct anastomosis to reconstruct an atretic left-sided component of a double arch in 1. This report describes an innovative technique of reconstructing the arch that allows for continued growth and avoids compression behind the trachea.

The management of patients with coarctation and unusual arch anatomy needs to be individualized. The arch laterality, brachiocephalic branching pattern, and the presence of a vascular ring need to be taken into consideration when deciding on the surgical approach to these patients. We provide a technique of reconstructing a hypoplastic right aortic arch that should prove valuable to the congenital cardiac surgeon.

References