Arterial switch in a 1146-gram neonate with transposition of the great arteries and an intramural coronary artery

Yukihiro Kaneko, PhD,^a Keiji Tsuchiya, MD,^c Yusuke Yamamoto, MD,^a Hitoshi Yoda, MD,^b Wakako Yamamoto, MD,^b and Jotaro Kobayashi, PhD,^a Tokyo, Japan

A male monozygotic twin weighing 1146 g delivered at 31 weeks' gestation was cyanotic. An echocardiogram disclosed a simple complete transposition of the great arteries. The right and left coronary arterial orifices were located in sinus 2 (right posterior facing sinus).¹ The left coronary artery was hypoplastic, had an intramural course between the great arteries, and gave rise to the left anterior descending artery and the circumflex artery. The aortic root was located anterior and 30 degrees to the right of the pulmonary arterial root. The diameters of the aorta and the pulmonary trunk were 6 mm and 8 mm, respectively.

Clinical Summary

Balloon atrial septostomy was not performed because there was acceptable interatrial communication and an appropriately sized balloon catheter was not available. Prostaglandin E1 was given to maintain ductus arteriosus patency.

Surgery was performed at age 14 days, when the patient weighed 1103 g. Through a median sternotomy, cardiopulmonary bypass with aortic and bicaval cannulation was established. After aortic crossclamping, the aorta was transected 2 mm above the sinotubular junction. The pulmonary artery was transected at the same level. The right and left coronary ostial diameters were 1.0 mm and 0.5 mm, respectively. After the Lecompte maneuver, one quarter of the circumference of the aortic root located above sinus 2 was sutured to the apposing edge of the pulmonary arterial root (Figure 1, A). A longitudinal incision was made in the right anterior aspect of the distal aorta; the pulmonary arterial root and distal aorta were anastomosed, leaving an inverted V-shaped opening above sinus 2 (Figure 1, B). To create a neo-aorta–coronary artery tunnel, a glutaraldehyde-fixed autologous pericardium was sutured between the neopulmonary valve (the anatomic aortic valve) cusp at sinus 2 and the opening in the distal aorta; the upper half of the tunnel consisted of the pericardial hood and aortic opening, and the lower half of the tunnel consisted of sinus 2 and the accompanying valve cusp. Near the course of the intramural coronary artery, the stitches were placed on the valve tissue, not on

the aortic intima, so as not to distort the coronary artery (Figure 1, C). The neopulmonary arterial root was longitudinally incised between sinus 1 and the nonfacing sinus, and then enlarged using a pericardial patch. To prevent compression of the neo-aorta–coronary artery tunnel by the anteriorly relocated pulmonary arterial wall, a longitudinal incision was made in the pulmonary artery bifurcation; the edge of this incision was sutured around the suture line between the pericardial hood and the aorta (Figure 1, D). Finally, a distal pulmonary artery–neopulmonary arterial root anastomosis was made. The sternum was left open because of cardiac edema. The operation time was 320 minutes, cardiopulmonary bypass time was 190 minutes, and aortic crossclamp time was 90 minutes.

The sternotomy was closed on postoperative day 6. The patient recovered uneventfully otherwise. He was extubated at age 28 days and discharged at age 109 days when he weighed 2720 g. Postoperative echocardiography showed normal left ventricular function, a widely patent neo-aorta–coronary artery tunnel, and mild neopulmonary regurgitation.

Figure 1. The procedure is shown sequentially from A to D. See text for details of each stage. C, Area (arrow) where the pericardial patch was sutured to the aortic valve cusp to avoid distorting the adjacent intramural coronary artery.

From the Departments of Cardiovascular Surgery,^a Neonatology,^b and Pediatrics,^c Japanese Red Cross Medical Center, Tokyo, Japan.

Received for publication May 25, 2007; accepted for publication June 14, 2007.

Address for reprints: Yukihiro Kaneko, MD, Department of Cardiovascular Surgery, Japanese Red Cross Medical Center 4-1-22 Hiroo, Shibuya-ku, Tokyo 150-8935, Japan (E-mail: yukihirokaneko@hotmail.com).

J Thorac Cardiovasc Surg 2007;134:1064-5

0022-5223/\$32.00

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doi:10.1016/j.jtcvs.2007.06.009

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, sur-

gery-induced coronary insufficiency was eliminated by a large neo-aorta–coronary artery tunnel, the absence of coronary artery rotation, and a suture line placed on the valve tissue near the intramural coronary artery.

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

Priti M. Patel, MD,^a Heather L. Bartlett, MD, FACC,^a Thomas Scholz, MD, FACC,^a Jeffrey Burzynski, MD, FACP,^b Douglas M. Behrendt, MD,^c and Harold M. Burkhart, MD,^c Iowa City, Iowa

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 atrioventricular septal defect with a hypoplastic left-sided atrioventricular 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

From the Departments of Pediatric Cardiology,^a Pediatric Critical Care,^b and Pediatric Cardiac Surgery,^c University of Iowa Hospitals and Clinics, Iowa City, Iowa.

Received for publication April 12, 2007; accepted for publication April 23, 2007.

Address for reprints: Harold M. Burkhart, MD, Division of Cardiac Surgery, Mayo Clinic, 200 First St SW, Rochester, MN 55905 (E-mail: Burkhart.harold@mayo.edu).

J Thorac Cardiovasc Surg 2007;134:1065-6

0022-5223/\$32.00

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doi:10.1016/j.jtcvs.2007.04.066