—Case Reports—

Repair of Recoarctation and Aortic Valve Replacement with the On-X Valve in a Pediatric Patient: A Case Report

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Abstract

A 10-year-old boy had undergone conventional two-stage surgery that includeed repair of aortic coarctation with pulmonary artery banding as an initial operation followed by closure of a ventricular septal defect as a secondary procedure. Following these procedures, aortic coarctation recurred, and balloon angioplasty was performed three times. Aortic recoarctation, however, persisted, and aortic stenosis and regurgitation and pulmonary stenosis gradually developed. An operation was therefore performed to repair recoarctation, aortic stenosis and regurgitation, and pulmonary stenosis. Patch aortoplasty was performed for recoarctation. The aortic valve was replaced with a 19 mm On-X valve, with Konno root enlargement. Pulmonary arterial patch plasty was also performed. During aortic arch reconstruction, selective cerebral perfusion was performed via the innominate artery and left carotid artery, and the lower half of the body was perfused through a cannula in the left femoral artery. The postoperative course was uneventful, and aortography revealed a well-reconstructed aortic arch free of obstruction. There were no valve-related complications.
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Key words: recoarctation of aorta, aortic valve replacement, Konno procedure, On-X valve

Introduction

After repair of aortic coarctation, associated cardiovascular diseases are the most common cause of repeat surgery. Simultaneous repair of recoarctation and associated lesions requires complex surgical techniques. We report herein a rare case of recoarctation with aortic stenosis and regurgitation being repaired simultaneously.

The On-X valve, a new mechanical valve substitute, has not previously been used in the pediatric patients. We used this prosthesis for aortic valve replacement (AVR) in this patient.

Case

A 10-year-old boy weighting 29 kg presented to our hospital for surgical treatment of recoarctation and aortic stenosis and regurgitation. He had undergone a two-stage operation for coarctation complex. He was delivered at 38 weeks' gestation. On day 5, the patient became tachypneic and cyanotic. On day 9, he was referred to our hospital,

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where coarctation of the aorta (CoA), a subpulmonary ventricular septal defect (VSD), and a closing patent ductus arteriosus (PDA) were diagnosed. Angiography revealed that the CoA was due to tubular hypoplasia of the aortic arch between the left subclavian artery and the PDA. Emergency surgery was performed. Through a left thoracotomy, extended resection and end-to-end anastomosis for CoA, and pulmonary artery banding constricting the pulmonary artery circumference to 22 mm were performed. At 7 months, patch closure of the VSD and removal of the pulmonary artery band were performed. After the pulmonary band was cut, a vertical incision was added to the pulmonary artery, and the VSD was closed with a Gore-Tex Patch. As the size of the pulmonary artery was adequate for a patient of this age, patch augmentation was not considered necessary. After VSD closure, coarctation between the left carotid artery and left subclavian artery recurred, necessitating balloon angioplasties, three times from the age of 8 months to 3 years. The site of coarctation was the same each time, and the pressure gradient across the recoarctation improved from 30 mmHg to 15 mmHg at 8 months, 47 mmHg to 37 mmHg at 2 years, and 34 mmHg to 27 mmHg at 3 years. Although coarctation persisted after these treatments, balloon angioplasty could not be performed again because the site was approaching the top of the aortic arch. Aortic stenosis (AS) with aortic regurgitation (AR) and pulmonary stenosis (PS) had been observed since the patient was 4 years old, and the stenosis was gradually becoming more severe.

On this admission, a chest roentgenogram showed a cardiothoracic ratio of 46%. Electrocardiography showed left ventricular hypertrophy. Echocardiography demonstrated valvular AS with a pressure gradient of 40 mmHg, mild AR, CoA with a pressure gradient of 50 mmHg, and supravalvular PS with a pressure gradient of 40 mmHg. The opening of the commissure between the right coronary cusp (RCC) and the noncoronary cusp (NCC) was limited, although the RCC and NCC were approximately equal in size. Angiographic measurements of the aortic valve annulus, ascending aorta diameter, recoarctation diameter, and descending aorta diameter were 16 mm, 17 mm, 6 mm, and 11 mm, respectively.

Under general anesthesia, median sternotomy was performed, and cardiopulmonary bypass was established with the ascending aortic and bicaval cannulations. The left ventricle was vented via the left atrium. During arch reconstruction, selective cerebral perfusion was performed via the innominate artery and left carotid artery, and the lower half of the body was perfused through a cannula in the left femoral artery. With the moderate hypothermic (27℃) cardiopulmonary bypass technique, cardiac arrest was achieved by an antegrade infusion of blood cardioplegia solution. CoA was present from the innominate artery to the proximal descending artery. To relieve this condition, patch aortoplasty using a Xenomedica patch (Edwards Lifesciences, LLC, Irvine, California, U.S.A.) lined with knitted polyester fabric extending into the proximal arch was performed. Next, AVR was performed with a 19-mm On-X prosthetic valve (Medical Carbon Research Institute, LLC, Austin, Texas, U.S.A.) using the Konno root enlargement. The aortic valve was tricuspid, and three cusps were thickened. There was no adhesion between the RCC and NCC. At the end of the reconstruction, pulmonary arterial patch plasty was performed to enlarge the supravalvular PS. The surgical time, extracorporeal circulation time (selective cerebral perfusion time), and aortic cross-clamp time were 385, 172 (28), and 121 minutes, respectively. Pathological examination of the aortic valve showed myxoid degeneration and fibrosis. The postoperative echocardiogram showed no aortic arch stenosis, with a pressure gradient of 10 mmHg, no CoA flow pattern in the abdominal aorta, and right ventricular outflow tract stenosis with a pressure gradient of 25 mmHg. Aortography 7 months after surgery revealed a well-reconstructed aortic arch free of obstruction (**Fig. 1**). The patient was maintained on permanent anticoagulation with warfarin with an international normalized ratio target of 1.8 to 2.0. The On-X valve was implanted safely and effectively and there were no valve-related complications.

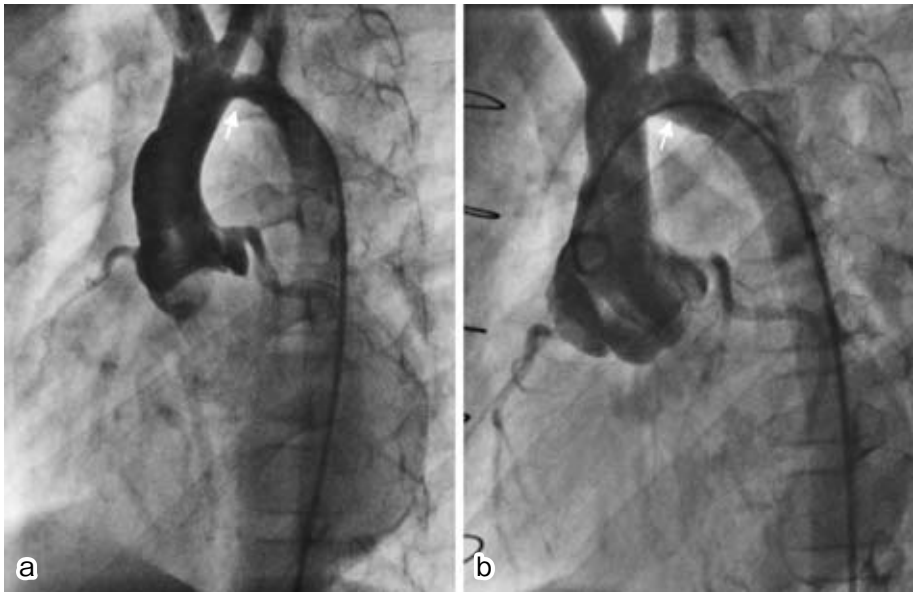

Fig. 1
a: Preoperative aortography. There was a significant obstruction **(white arrow)** between the innominate artery and the left subclavian artery (length, 10 mm; diameter, 6 mm), with a pressure gradient of 40 mmHg. Moderate regurgitant flow from the aortic valve into the left ventricle on the anterior side of the aortic root was confirmed.
b: Aortography after repair of recurrent aortic obstruction and after AVR using the Konno procedure. The aortic arch was widely reconstructed without any site of obstruction **(white arrow)**.

Discussion

Backer et al. have reported that the rate of recoarctation after extended end-to-end anastomosis is 4% to 23%[1]. DiBardino et al. have reported that this rate is influenced largely by patient's age at operation, averaging 20% when the operation is performed in neonates. The recoarctation rate decreases to approximately 15% at age 6 months, and to less than 5% after 1 year of age[2]. In the present case, recoarctation was first detected at 8 months. This delay may have been caused by a lack of growth of the suture line or remnants of ductal tissue or damage of the aorta from the vascular clamps used at repair. Balloon angioplasty has been increasingly used to treat recurrent aortic coarctation and has recently been recommended by several centers[3]. At our institution, balloon angioplasty is also preffered for recurrent coarctation. When the results of balloon angioplasty are unsatisfactory, surgery is considered. Surgical reconstruction techniques for recurrent aortic obstruction have historically included resection and reanastomosis, patch aortoplasty, and interposition grafting[2]. We selected patch aortoplasty because tension-free anastomosis was considered to be a possibility with direct anastomosis reconstruction due to the long-segmental stenosis.

CoA sometimes coexists with AS, but this patient had not only AS but also AR, and their severity had increased. The VSD was a subarterial defect located beneath the RCC of the aortic valve. The echocardiography revealed restricted opening of the commissure between the RCC and NCC. The VSD patch sutured to the aortic valve annulus might contribute to development of the AS and AR.

The On-X prosthetic heart valve was introduced in 1996. Its design was intended to address existing deficiencies of mechanical valves, including inadequate hemodynamics in small aortic sizes, occasional incidents of unexplained hemolytic anemia, tissue interference or excessive pannus overgrowth, and thrombotic complications. Owing in part to the improved hemodynamics, the valve was expected to have low thromboembolism rates that

appear to justify further study at reduced anticoagulant levels, especially in patients with AVR in sinus rhythm[4,5]. In fact, the incidence of thrombosis and thromboembolism in poorly anticoagulated patients has been similar to that in well-controlled patients[6]. At our institution, the On-X valve is the preferred choice for pediatric patients because of the expectation of less need for anticoagulation. We have controlled warfarin-based anticoagulation at a level slightly lower than that recommended for bileaflet valves in the aortic position. The orifice area of the 19-mm On-X valve used in the present case is 2.22 cm^2, which is larger than that of 21-mm St. Jude Medical Standard valve (orifice area=2.10 cm^2). The effective orifice area of the 19-mm On-X valve is 1.5 cm^2, which is theoretically usable for patients with a body surface area up to 1.7 m^2 without prosthesis-patient mismatch according to the following criterion: indexed effective orifice area (effective orifice area/ body surface area)>0.85[4,5]. After 18 months of the follow-up, the On-X valve exhibited improved hemodynamics, no thromboembolism, and no hemolysis.

In conclusion, we simultaneously performed repair of recoarctation and AVR using an On-X valve in a pediatric patient. The operation was complicated, but the results were satisfactory. Longer follow-up is needed to allow a more definitive comparison with other established treatments.

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