

Ross procedure after aortic balloon valvuloplasty: the youngest case in Turkey

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In this study, we report a Ross procedure which was performed on an emergent basis for severe aortic regurgitation following balloon aortic valvuloplasty in an infant with congenital critical aortic stenosis. To the best of our knowledge, this is the youngest case who was applied the Ross procedure in Turkey.

Keywords: Balloon valvuloplasty; congenital heart disease; left ventricular outflow tract obstruction; Ross procedure.

Left ventricular outflow tract obstruction (LVOTO) remains a significant challenge in neonates and infants in pediatric cardiology and cardiac surgery.^[1] Residual stenosis or severe aortic regurgitation may develop in neonates and infants with critical aortic stenosis undergoing surgical valvotomy or balloon aortic valvuloplasty (BAV).^[2-4] The Ross procedure and Ross-Konno procedure are one of the treatment of choices for such small babies with severe left ventricular dysfunction and heart failure.^[1,5]

In this study, we report a Ross procedure which was performed on an emergent basis for severe aortic regurgitation following BAV in small baby with congenital critical aortic stenosis. To the best of our knowledge, this is the youngest case who was applied the Ross procedure in Turkey.

CASE REPORT

A two and half-month-old premature male infant with a weight of 2.5 kg was referred to our clinic due to severe heart failure. The patient had previous BAV, when he was 30 days old. Echocardiographic examination revealed severe aortic regurgitation and valvular aortic stenosis with a gradient of 44 mmHg. Anatomy of aortic annulus, pulmonary annulus and pulmonary valve were normal. Severe left ventricular dysfunction was detected (fractional shortening: 17%). The patient with a poor health status underwent mechanical ventilation via endotracheal intubation in intensive care unit. The patient was stabilized using inotropic agents, diuretics, and blood gas analysis, and then urgently operated.

Operative technique

Standard cardiopulmonary bypass (CPB) was initiated through median sternotomy, aortic, and

bicaval cannulation. Hypothermia at 28 °C and intermittent hypothermic blood cardioplegia were applied. Aortic valve was unicuspid and complicated by rupture of the aortic annulus, clinging to one side of the valve. Pulmonary artery was transected proximal to its bifurcation and pulmonary autograft was prepared. Right and left coronary buttons were excised and the pulmonary autograft was implanted in the aortic position using aortic root replacement technique.^[2] Sutures were ligated over the three thin autologous pericardial strips to strengthen neo-aortic annulus and ensure blood management. Right ventricular outflow tract was reconstructed using 14 mm Bovine Jugular Vein (Contegra, Medtronic Inc., Minneapolis, Minnesota, USA). Aortic cross-clamp time was 146 minutes and CPB time was 235 minutes. On Day 7, postoperative echocardiography revealed normal left ventricular function (fractional shortening: 36%) and neo-aortic valve (very mild regurgitation) (Figure 1). No pulmonary conduit stenosis was observed.

The patient was discharged at 25 days following surgery and followed every six months. At 42 months, his motor-mental development was very good with a NYHA Class I functional capacity. He remained under follow-up without any medication excluding aspirin. Repeated echocardiography every six months also revealed normal growth of autograft

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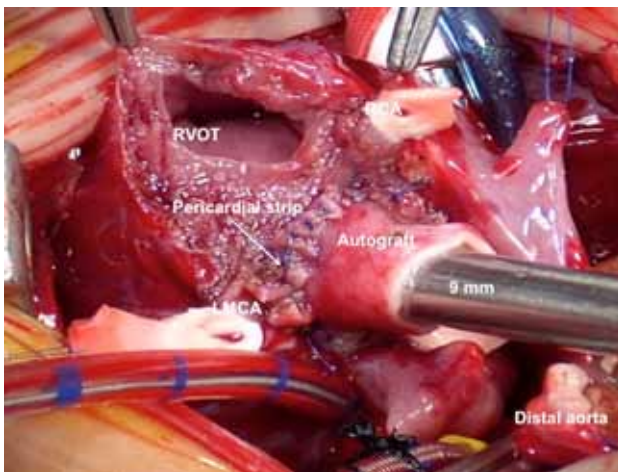


Figure 1. Intraoperative view. RVOT: Right ventricular outflow tract, RCA: Right coronary artery; LMCA: Left main coronary artery.

(annulus: 2.2 cm) without annular and sinotubular dilatation. There was no progress in the aortic regurgitation. The left ventricular function was very good (fractional shortening: 42%). However, valvular stenosis in the right ventricle to pulmonary artery (RV-PA) conduit was observed with a gradient of 62 mmHg.

DISCUSSION

Open or closed surgical valvotomy or BAV may offer interim palliation in infants with critical aortic stenosis with normal biventricular heart. However, severe aortic regurgitation or residual aortic stenosis may subsequently develop.^[3,4] Therefore, a rapid and radical management approach may be required, as in our case. The main disadvantages of prosthetic valve replacement in neonates and infants with critical aortic stenosis include implanting an appropriate prosthetic valve by expanding aortic annulus, need for re-do surgery in the following years, and challenges in using anticoagulants.^[6] On the other hand, aortic valve replacement (AVR) using homografts is not a reasonable alternative due to the lack of growth potential and rapid degeneration of allografts in pediatric population, as well as difficulty in homograft supplying.^[7]

The Ross procedure is the most reasonable treatment of choice in neonates and infants requiring AVR. However, an optimal compliance between pulmonary autograft and aortic annulus, growth potential, acceptable durability profile of pulmonary autograft in the mid- and long-term and no need for anticoagulants

are the main advantages of the procedure. In addition, it can be combined with the Konno procedure in the presence of complex LVOTO.^[1,5,8-10] In our case, aortic annulus was within normal size (9 mm). We would also perform the Ross-Konno procedure in case of annulus hypoplasia.^[9]

Although the Ross procedure is a technically challenging and time-consuming intervention, it can be safely applied with a mortality rate below 5%.^[1,4,8-10] However, there are still concerns regarding putting at a risk of both valves, autograft dilatation in the long-term and need for re-do surgery of the right ventricular outflow tract conduit.^[11-13] There are several studies showing less annular and sinotubular dilatation of the autograft and aortic regurgitation in neonates and infants.^[1,4] Maeda et al.^[1] reported that 74% (7±12.9) of infants had very mild aortic regurgitation at five years following the Ross-Konno procedure. Shinkava et al.^[5] also reported that 95.2% of infants had excellent autograft functions which tended to grow with increasing age in a 10-year follow-up period. This may be explained by an ongoing histological structuring of semilunar valves (mucopolysaccharide and collagen balance) in neonates and infants. Pulmonary arteries and valves may release histological adaptation against systemic pressure, thereby leading to less autograft dilatation and dysfunction in this population.^[5] In addition, improved neo-aortic valve functions in the long-term following arterial switch operation support this assumption.

Nonetheless, the most important disadvantage of the Ross procedure is the risk of re-do surgery due to the possible RV-PA conduit-related complications. The Contegra valved bovine jugular vein graft (Contegra; Medtronic, Inc., Minneapolis, Minn) have a very low early re-do surgery rate for RVOT reconstruction and could be therefore used in neonates and children under the age of three years, unless a blood group-compatible homograft can be found.^[13,14]

There are several reports demonstrating functional RV-PA conduit in 50-70% of the patients at 10 years.^[5,7,10-12] A large-size conduit as much as possible should be placed during the initial operation to ensure a more durable RV-PA conduit. It is well-established that conduits less than 14 mm in diameter may result in early re-do surgery. In our study, we implanted a 14 mm Contegra conduit (Medtronic Inc., Minneapolis, Minn, USA), as his weight was 2.5 kg. At 42 months of surgery, conduit stenosis was

observed preserving its function. We believe that we can increase the lifespan of the conduit by pulmonary balloon angioplasty.

In conclusion, the Ross procedure and Ross-Konno procedure are among the first treatment of choices in the management of aortic valve pathologies and LVOTO in neonates and infants.

Declaration of conflicting interests

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