References

Ross procedure with a quadricuspid pulmonary autograft

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Aortic valve replacement using the pulmonary autograft as described by Ross in 1967 is an excellent permanent therapy for aortic valve disease, particularly in young patients. This procedure facilitates the omission of anticoagulation. The pulmonary autograft displays a superior longevity and excellent hemodynamic properties in combination with a low incidence of thromboembolism compared with all other aortic valve replacement procedures. However, its success is dictated by the suitability of the pulmonary autograft before the switch to the aortic position; a normal pulmonary valve (PV) without anatomic abnormalities is a prerequisite for the achievement of a satisfying operative and hemodynamic result.

We report on the postoperative outcome and midterm follow-up (4 years) of a 48-year-old female patient who underwent an aortic valve replacement with a quadricuspid pulmonary autograft. A quadricuspid PV is a rare anatomic feature with an incidence of 1 in 1100 individuals. To our knowledge, the use of a quadricuspid autograft in the Ross procedure has not been reported before.

Clinical Summary
A 48-year-old female patient presented with signs of congestive heart failure such as peripheral edema and fatigue. Cardiac auscultation revealed a typical systolic murmur with punctum maximum in the third right intercostal space and projection into the carotid arteries. Angiography and echocardiography revealed good left ventricular function (left ventricular ejection fraction 60%) and severe left ventricular hypertrophy caused by advanced valvular stenosis with a calculated orifice area (aortic valve area) of 0.45 cm² and increased transvalvular gradient of 121/84 mm Hg (peak/mean). The ascending aorta showed poststenotic dilation with a maximal diameter of 40 mm. The aortic root was normal with a diameter of 25 mm.

The patient underwent aortic autograft valve replacement. Preparation of the pulmonary and aortic valve was performed in a standard technique during cardiac arrest on extracorporeal circulation. After the pulmonary valve was excised, a quadricuspid morphology became evident (PV), which was undetected before surgery. Vigorous irrigation tests demonstrated a patent PV without any sign of regurgitation. The autograft was implanted in a free-root replacement technique. The postoperative course was uneventful, and the patient was discharged on postoperative day 12 in excellent condition. Echocardiography performed intraoperatively and transthoracic echocardiogram performed 4 years after Ross procedure.

Figure 1. Transthoracic echocardiography of the quadricuspid autograft 4 years after Ross procedure. The quadricuspid valve is superimposed by lines to demonstrate the margins of the 4 leaflets.
phy performed 1 month after surgery demonstrated a morphologically and functionally normal autograft.

Four years later, a routine follow-up echocardiography was performed in the clinically asymptomatic patient and revealed good left ventricular function with an ejection fraction of 65%. Although no recurrence of aortic stenosis was detectable, a second-degree aortic regurgitation (Figure 1) was evident in color Doppler echocardiography. The patient denied any symptoms, despite her physically demanding profession.

Discussion

Donald Ross1 introduced the use of the pulmonary autograft for aortic valve replacement in 1967. This technique allows the replacement of a diseased aortic valve with the autologous PV. Thus, systemic anticoagulation is circumvented, and the risk of thromboembolism or cerebral bleeding is negligible. In comparison with all other valve substitutes, the free-root technique shows the best hemodynamic outcome and consequently is the most frequently used approach.3 However, the normally tricuspid PV must be of regular anatomy and function. To our knowledge the use of a quadricuspid pulmonary autograft has not been described before. The early postoperative result with the quadricuspid autograft was promising. However, the development of autograft regurgitation of second degree (echocardiographic guidelines) as early as 4 years after surgical treatment identifies quadricuspid autografts as potentially problematic for this procedure. Preoperative echocardiographic evaluation of PV function and morphology are therefore mandatory.

References


Aortic root replacement in a patient with pulmonary dysfunction caused by severe chest deformity associated with Marfan syndrome

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Great advances in respiratory management have recently widened the indication for cardiovascular surgery for patients with pulmonary dysfunction. We describe a patient with an extremely low vital capacity caused by severe chest deformity who tolerated aortic root replacement as a result of adequate respiratory management, including bilevel positive airway pressure (BiPAP) ventilation.

Clinical Summary

A 39-year-old man with Marfan syndrome was scheduled to undergo aortic root replacement for anulaoartic ectasia and aortic insufficiency. He had been receiving home oxygen therapy for 10 years and nocturnal use of nasal continuous positive airway pressure for 1 year because of frequent episodes of respiratory failure.

On admission, he showed respiratory distress at grade IV of the Hugh-Jones classification. Spirometry showed a severe restrictive pattern: vital capacity of 1080 mL (28% of predicted value) and forced expiratory volume in 1 second of 890 mL. Arterial blood gases (ABGs) under 1L of oxygen inhalation revealed metabolically compensated hypercapnia: pH, 7.41; PaCO₂, 60.8 mm Hg; PaO₂, 80.4 mm Hg; base excess, 10.9 mmol/L. Chest radiography showed cardiomegaly with mild lung congestion and severe scoliosis (Figure 1). Computed tomographic (CT) scans revealed the dilated aortic root (62 mm in diameter) and marked elongation of the descending aorta along the vertebral bodies (Figure 2). Trans-thoracic echocardiography revealed a redundant tricuspid aortic valve with severe regurgitation. The left ventricular ejection fraction was 45%, as estimated by ventriculography.

At the time of sternotomy, we paid particular attention to avoiding lung injury and to not opening the pleural spaces. After establishment of cardiopulmonary bypass, the aortic root was replaced with a composite graft by use of the Carrel patch technique because aortic leaflets were severely degenerated. Weaning from bypass was uneventful.

ABGs on admission to the intensive care unit were favorable: pH, 7.38; PaCO₂, 50.1 mm Hg; PaO₂, 167.2 mm Hg; base excess, 4.4 mmol/L (synchronized intermittent mandatory ventilation; fraction of inspired oxygen of 0.5; respiratory rate of 12 breaths/