Reoperation for dilatation of the pulmonary autograft after the Ross procedure

Thoralf M. Sundt, MD,[a] Marc R. Moon, MD,[a] and Haodong Xu, MD, PhD,[b] St Louis, Mo

The pulmonary autograft or "Ross procedure" offers hope for a durable, anticoagulant-free replacement option for a dysfunctional aortic valve. As a viable graft, it has the potential for growth. The distinction between growth and passive dilatation, however, may be problematic. We recently encountered a case that graphically demonstrated dilatation of the wall of the autograft both radiologically and grossly.

From the Division of Cardiothoracic Surgery, Department of Surgery,[a] and the Department of Pathology,[b] Washington University, St Louis, Mo.

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Address for reprints: Thoralf M. Sundt, MD, Mayo Clinic, 200 First St SW, Rochester, MN 55905.

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Clinical Summary

A 19-year-old man with a history of aortic regurgitation caused by a bicuspid aortic valve had undergone a Ross procedure at age 13 years. The inflow suture line of the pulmonary autograft had not been reinforced with felt. A 24-mm pulmonary allograft valve had been placed in the pulmonic position. At recent echocardiographic follow-up, asymptomatic dilatation of the neo-aortic root without valvular regurgitation or stenosis was identified. Magnetic resonance imaging demonstrated dilatation of the neo-aortic root to 6.4 cm with a sharp transition to normal diameter (Figure 1). The pulmonic allograft was functioning normally.

At surgery, external inspection of the aorta suggested a "neck" at the transition between autograft and native aorta (Figure 2, *A*). This was confirmed at aortotomy (Figure 2, *B* and *C*). The autograft leaflets and anulus appeared normal (Figure 2, *C*). As suggested previously by T. E. David (personal communication, October 1999), the root was reconstructed with a scalloped graft preserving the autograft leaflets after the manner described by Yacoub. Transesophageal echocardiography demonstrated a competent valve at the end of the procedure. The

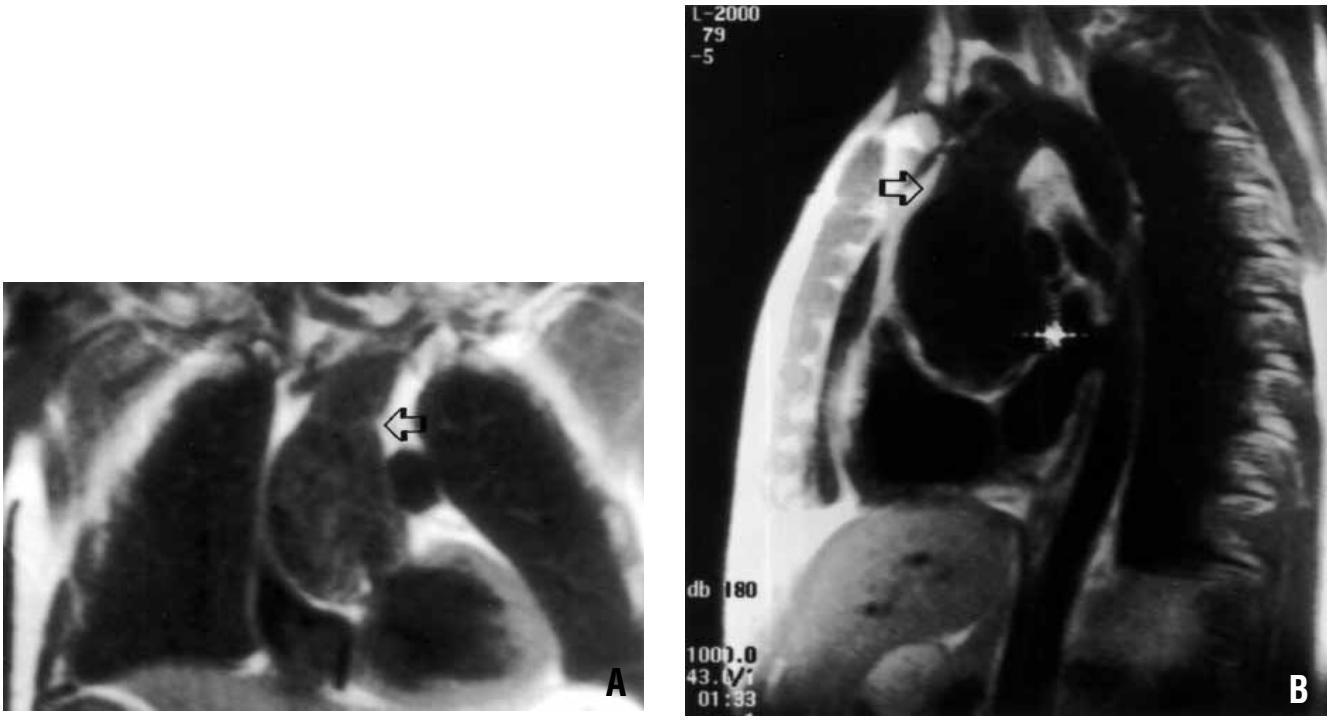

Figure 1. Magnetic resonance images of the neo-aortic root demonstrating dilatation of the pulmonary autograft and an abrupt transition to normal aortic diameter at the distal suture line *(arrow)* in the coronal section (A) and sagittal section (B).

excised autograft tissue demonstrated medial myxoid degenerative changes and fibrosis by hematoxylin-eosin stain and severe fragmentation of elastin fibers by Verhoeff–van Gieson stain.

Discussion

Inclusive of the patient presented above, we have reoperated on 4 patients late after the Ross procedure for neo-aortic valve malfunction (Table 1). In 1 patient, a 34-year-old woman with Goldenhar syndrome and a history of ventricular septal defect and aortic regurgitation, moderate aortic regurgitation developed as a result of a torn leaflet 7 years after the Ross procedure. No dilatation of the neo-aortic root was observed. The neo-aortic valve was replaced with a mechanical valve within the pulmonary autograft. Of the remaining 3 patients, all had aneurysmal dilatation of the neo-aorta and all had bicuspid aortic valves with aortic regurgitation as their original pathologic condition. One of these requested a mechanical valve and underwent conventional composite root replacement; a second received a pericardial xenograft valve conduit because of contraindications to anticoagulation; and the third is the subject of the current clinical summary.

The potential for growth makes the pulmonary autograft uniquely appealing for the pediatric population, and the promise of durability without anticoagulation makes it attractive to the young adult. Unfortunately, the most common indication for surgery among young adults is bicuspid aortic valve disease, a condition thought to be associated with intrinsic abnormality of the aortic wall. It has been recognized for a decade that the aortic root may be enlarged in patients with a bicuspid aortic valve even without hemodynamically significant functional abnormality. More recently, histochemical studies have demonstrated an increased rate of smooth muscle cell apoptosis in the aortic media of patients with bicuspid aortic valves, even in the absence of gross dilatation.[1] Biochemical analysis has also demonstrated a decrease in collagen content strikingly similar to that observed in Marfan syndrome.[2] Elastin fragmentation, alterations in smooth muscle cell orientation, and the changes characterized by the term *cystic medial necrosis* have been observed in both the ascending aorta and pulmonary artery of patients undergoing the Ross operation for bicuspid aortic valve disease.[3]

The clinical correlate of these laboratory observations has been reported previously. David and colleagues[4] identified a statistically significant increase over time in the diameter of the sinuses of Valsalva, the aortic anulus, and the sinotubular junction among patients with bicuspid aortic valves undergoing the Ross procedure. Histologic analysis of the pulmonary autograft wall from such patients showed degenerative changes of cystic medial necrosis.[4,5] Our case graphically demonstrates the radiologic and gross pathologic correlate of these histologic findings.

Dilatation of the anulus after the Ross procedure has led to the recommendation by some that the inflow suture line be reinforced with prosthetic material. We observed significant dilata-

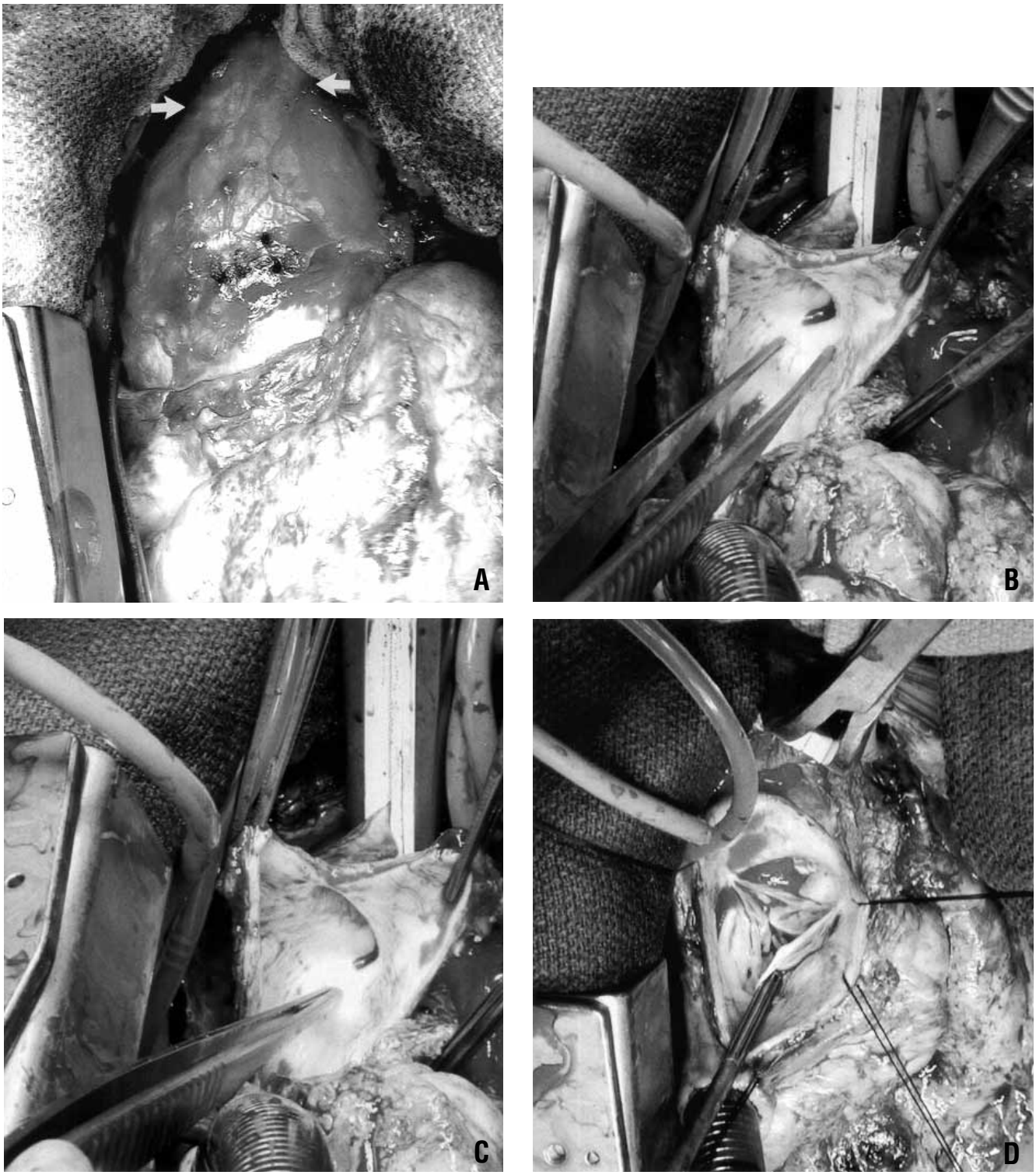

Figure 2. Intraoperative photographs confirming the transition to normal diameter aorta at the distal suture line. A, External appearance of the neo-aortic root with a perceptible narrowing at the distal anastomosis *(arrows).* B and C, Internal appearance of the transition between autograft and aorta demonstrating a "neck." D, The normal appearing autograft valve cusps that were preserved.

TABLE 1. Patients undergoing reoperation after failed Ross procedure

Age (y)	Sex	Original disease	Interval after Ross procedure	Diameter of neo-aorta	Valve disease at reoperation	Procedure at reoperation
34	F	VSD + VSD	7 y	Normal	Moderate AR and severe PS	Mechanical AVR and pulmonary allograft
39	M	BAV with AR	9 y	6.0 cm	Moderate AR and severe PS	Composite root replacement and pulmonary allograft
30	M	BAV with AR	6 y	5.0 cm	Severe AR and severe PS	Pericardial valve conduit and pulmonary allograft
19	M	BAV with AR	7 y	6.4 cm	No AR or PS	Yacoub root reconstruction

AR, Aortic regurgitation; *AVR,* aortic valve replacement; *BAV,* bicuspid aortic valve; *PS,* pulmonic stenosis; *VSD,* ventricular septal defect.

tion of the wall of the autograft, however, a complication unlikely to be affected by annular reinforcement. David and colleagues[4] have suggested that the root inclusion technique be considered in cases of bicuspid aortic valve. Our observations support this suggestion.

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