complete cricopharyngeal myotomy and division of the septum between the esophagus and diverticulum were performed. The myotomy could lower the upper esophageal sphincter pressure and intrabolus pharyngeal pressure. Furthermore, the division of the common septum between the esophagus and diverticulum created a common cavity that could facilitate the emptying of the large pouch into the esophagus. It is reasonable to assume that the relief of the patient’s symptoms was due to both these reasons. The efficacy of this surgical procedure should be tested with a larger series of cases and with esophageal manometry data included.

To our knowledge, this case represents the first report of a giant Zenker’s diverticulum treated successfully with video-assisted thoracoscopic surgery.

References

Ross Procedure for Patient With Marfan Syndrome
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The most prominent long-term complication after the Ross procedure is the risk of autograft dilatation, and therefore its application in patients at increased perceived risk of autograft dilatation (those with bicuspid aortic valve disease, aortic insufficiency [AI] with dilated aorta, collagen vascular diseases such as Marfan syndrome) has been discouraged. We reported a modified Ross procedure in 2005 in which the autograft was completely encased in nonexpandable polyester (a combination Ross with “valve-sparing” root replacement). We described the technique in 2005 and again, with modifications, in 2010. We describe a case of a successful modified Ross procedure in a patient with Marfan syndrome associated with a dilated ascending aorta and bicuspid aortic valve with AI.

A 47-year-old man with Marfan syndrome was referred for evaluation of his aortic valve and aortic root. Echocardiography revealed moderate AI from a bicuspid aortic valve and mild mitral insufficiency (Fig 1). His aortic root measurement showed progressive dilatation to 4.8 cm in diameter (Fig 2A). He reported having 2 siblings, also with Marfan syndrome, and each had undergone aortic root and valve replacement at other institutions—1 with a mechanical valve and 1 with a bioprosthetic valve. He preferred a Ross procedure using a pulmonary autograft for his aortic valve, if it was possible. He did not want a mechanical valve or a bioprosthetic valve if it could be avoided. He acknowledged that although there was uncertainty as to the viability of

Fig 1. Preoperative transthoracic echocardiogram. Apical long-axis view shows a central jet of moderate aortic insufficiency (AD) (Ao = aorta; LV = left ventricle.)
an autograft in his circumstances, he wanted to try the procedure and accept the risk of failure before using an alternative prosthetic valve.

He underwent a modified Ross operation (under institutional review board approval) in July 2005. His aortic valve was bicuspid, with calcification and thickening, and performing a valve-sparing aortic root replacement was a less attractive option. We used the technique that we have previously described [5, 6]. After harvesting a pulmonary autograft from the right ventricular outflow tract, his autograft was sewn into a 32-mm polyester tube. His aortic root was replaced with this autograft construct and his ascending aorta was replaced with a portion of the same graft material. His postoperative course was uneventful and he was discharged and was to be followed by his referring physician. His most recent echocardiogram 6 years and 4 months after the operation demonstrated a stable aortic root, measuring 39 mm in diameter, and no aortic stenosis or insufficiency (Fig 2B). He had no interval change on his most recent (March 2013) outpatient office echocardiogram [personal communication with patient and physician]. The pulmonary valve that was replaced with allograft had no obstruction and only mild insufficiency. He has had no limitations on activity for almost 8 years since the operation and reports feeling in excellent health (personal communication, June 2013).

Comment

The pulmonary autograft operation was introduced by Donald Ross in 1967, and the Ross procedure has become an attractive option for aortic valve replacement. However, the application of Ross procedure aortic valve replacement in adults has been declining over the past decade, possibly as a result of numerous factors, including a variety of new prosthetic options, the technical challenge of the procedure itself, and the well-recognized risk for autograft dilatation with progressive valve insufficiency. To prevent potential significant autograft dilatation and AI, we created a modification to the Ross procedure in 2004 [6] that combined the hemodynamic and lifestyle advantages of a pulmonary autograft with the security of nonexpandable aortic root replacement.

Our procedure is similar to others reported in the literature [7, 8]. We have had no mortality in 36 patients in whom we have performed this procedure since October 2004. Two patients experienced autograft insufficiency and eventually had to undergo aortic valve replacement, and 1 patient experienced AI and was able to have the aortic valve repaired. All of these patients were early in our experience and before the technical modifications described in our more recent publication [6]. We have not seen any dilatation of the autograft in the patients in whom we have used this technique, including the patient with Marfan syndrome described in this report, and it is now 8 years after the operation.

Our experience indicates that this modification of the Ross procedure can be performed with dependable reproducibility. It should be offered only to patients in whom autograft growth is not necessary (ie, adults or full-grown teenagers). Intermediate-term data (out to 9 years of follow-up) are encouraging with respect to autograft function and stabilization of the neo-aorta. Our encouraging outcomes to date lead us to recommend consideration of this modified Ross procedure to patients who might otherwise not be considered candidates for pulmonary autograft aortic valve replacement and who might therefore be offered options that may not produce the same intermediate-term benefits—normal hemodynamics, no anticoagulation, and potential for a long-term viable aortic valve. We believe that this procedure is still "under investigation" and consider these results to be "short-term" but encouraging.

References

Acute Dilatation of the Ascending Aorta and Aortic Valve Regurgitation in Loeys-Dietz Syndrome

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Loeys-Dietz syndrome (LDS) is a recently recognized connective tissue disorder caused by mutations of the transforming growth factor (TGF)-β receptors. It is an autosomal dominant syndrome characterized by the triad of arterial tortuosity and aneurysms, hypertelorism, and bifid uvula or cleft palate. We treated an 18-year-old woman with a 100-mm-diameter aortic root aneurysm and severe aortic valve regurgitation. She underwent urgent aortic root replacement and bioprosthetic valve implantation. LDS was diagnosed by postoperative genetic screening results. Histopathologic examination of the aortic wall showed diffuse degeneration and elastin fragmentation in the media.

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Patients typically have aggressive arterial aneurysms (mean age of death, 26.0 years) and a high incidence of pregnancy-related complications [1]. It has been reported that patients with LDS undergo cardiovascular operations earlier (mean age, 16.9 years) and die earlier (mean age, 22.6 years) but have a lower intraoperative mortality rate compared with patients with vascular Ehlers-Danlos syndrome or Marfan’s syndrome [2].

An 18-year-old female patient was discovered to have a cardiac murmur during a school medical examination. Physical examination was otherwise unremarkable. There were no abnormal findings on a previous chest roentgenogram performed at age 15 years (Fig 1). She had a height of 166 cm, weight of 53.5 kg, and body mass index of 21.1 kg/m². There was no relevant family history of cardiovascular disease. She did not have any

Fig 1. Preoperative 3-dimensional computed tomographic image showing a 100-mm-diameter aneurysm of the ascending aorta (green arrow) and a 15-mm-diameter superior mesenteric artery (white arrow).