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The Ross-Konno Procedure as Reoperative Treatment in a Young Adult with Congenital Aortic Stenosis

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ABSTRACT

Mechanical and biological prostheses are valid options when aortic valve replacement is necessary. The Ross procedure is also an alternative solution, especially for young patients.

We describe the case of a young patient with congenital aortic stenosis and bicuspid aortic valve who presented with dyspnea on exertion. An open commissurotomy was performed, and within 8 months the patient developed recurrent symptoms of severe aortic stenosis. He underwent redo sternotomy and a Ross-Konno procedure with an uneventful recovery.

INTRODUCTION

There are several surgical treatment modalities for the management of left ventricular outflow tract (LVOT) obstruction in adult and pediatric patients. The choice of each surgical procedure depends on the pathology, the location of the obstruction, and the experience of the surgeon. In childhood, either commissurotomy of the aortic valve or resection, as in subvalvular stenosis, may solve the problem at least in a midterm period. The latter procedures can be accompanied by other modifications such as LVOT interventricular septum enlargement and annular enlargement as described by Konno [Konno 1975]. Mechanical and biological prostheses are valid options when aortic valve replacement is necessary. Valve choice depends on patient characteristics such as age, presence of arrhythmias, concomitant diseases, and contraindications to anticoagulation therapy. The ideal prosthetic valve should restore normal valvular function, require minimal alteration in lifestyle, and provide durability and low cost. Within the last years, significant experience has accumulated with the Ross procedure. The initial skepticism gave way to enthusiasm after the publication of midterm results with the Ross procedure and turned it into a valid operation for young patients with aortic valve pathology [Konno 1975; Elkins 1998].

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We describe the case of a young patient with congenital aortic stenosis and bicuspid valve who underwent a redo sternotomy and a Ross-Konno procedure. To the best of our knowledge, this is the first reported Ross-Konno procedure as reoperative treatment in Greece.

CASE REPORT

A 27-year-old man with congenital aortic stenosis and bicuspid aortic valve presented with dyspnea on exertion. His past medical history included percutaneous valvuloplasty 16 years prior. Transthoracic echo study showed a bicuspid aortic valve with stenosis and severe calcification of the leaflets with left ventricular hypertrophy (peak pressure gradient of 100 mmHg). Cardiac catheterization confirmed the echo findings, and the patient was referred to surgery. In the operating room, he underwent open commissurotomy as a surgical treatment because the calcified commissure was too close to the coronary ostia, and the surgeon was reluctant to proceed with something more radical. Postoperatively, the patient had still dyspnea on exertion. Echo study and cardiac catheterization revealed a residual peak pressure gradient of 80 mmHg and a relatively small aortic annulus (Figure 1). Based on these findings, after a period of 8 months, the patient was referred to our department for further evaluation and management. It was decided to perform a reoperation with the Ross-Konno procedure.

In the operating room, the aortic valve was adherent to the aortic wall in a fixed semi-closed position, and fused adherent leaflets were detached from the aortic wall. Subsequently, the coronary arteries were better visualized and mobilized as buttons in the usual fashion. The Konno incision was performed, and then the pulmonary autograft was prepared. The autograft was sewn in place with an interrupted technique. Afterward, the right and then left coronary buttons were implanted into the neo aorta. The pulmonary artery was reconstructed with the use of 27 mm Shelhigh xenograft (Millburn, NJ, USA). The patient resumed normal sinus rhythm and was weaned from cardiopulmonary bypass with moderate inotropic support. Intraoperative transesophageal echo showed a neo-aortic valve without stenosis or regurgitation. He was transferred to the intensive care unit where he stayed for 2 days. His postoperative course was uneventful, and he was discharged from the hospital in 8 days.

After 24 months, cardiac catheterization (Figure 2) showed no stenosis and no regurgitation in the neo-aortic valve and resolution of the left ventricular hypertrophy. The pulmonary xenograft had to be dilated once.

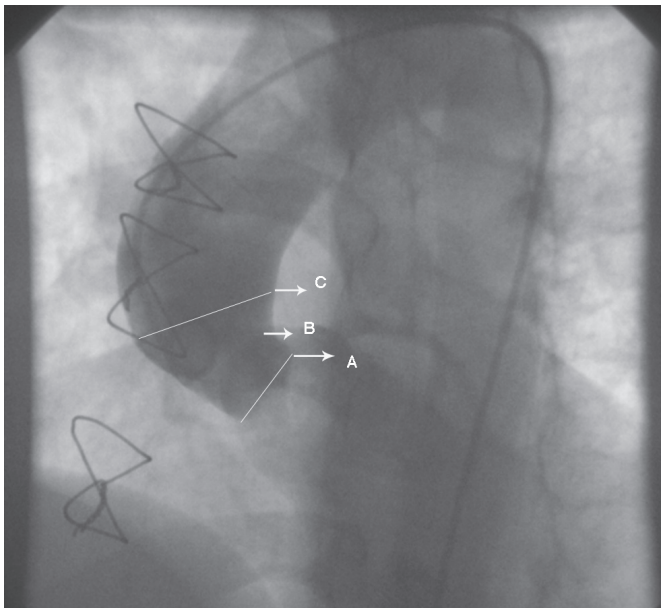

Figure 1. Cardiac catheterization before the Ross procedure, which depicts the aorta with the small aortic root. A, Small aortic annulus; B, bicuspid aortic valve; C, post-stenotic aortic dilatation.

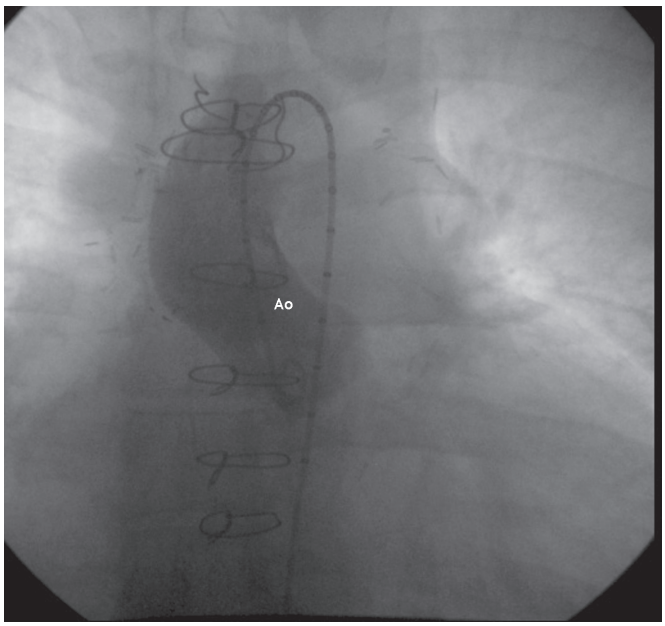

Figure 2. Cardiac catheterization, 2 years after the Ross procedure, which depicts the normal neo-aortic with the aortic root, without insufficiency. Ao indicates aorta.

DISCUSSION

The Ross procedure is a widely available procedure all over the world. In 1967, Ross performed the first successful clinical application of this procedure. The first series reported by Ross consisted of 14 young adults, two of whom received the pulmonary autograft in the mitral position [Ross 1967]. Some years later, Geens et al studied the anatomic relationships between the coronary arteries and the pulmonary root, establishing critical points for autograft extraction [Geens 1971].

The surgical technique and the experience have matured, and the pulmonary autograft seems to approximate what has been called "ideal" valve [Gerosa 1990; Kouchoukos 1994; Ross 1994; Elkins 1998]. However, the technical complexity of the procedure and the insufficient knowledge of this technique by surgeons and cardiologists deprive many potential candidates of the beneficial results of this operation. In fact, our country is still far from the yearly incidence of this operation in comparison with other counties in Europe and North America.

The Ross procedure is a technically demanding operation, but many authors have documented its safety, reporting no hospital deaths and no serious postoperative complications [Kouchoukos 1994; Elkins 1998]. Furthermore, despite the extra time of cardiac ischemia and longer cardiopulmonary bypass, patients have an impressive recovery with short intensive care unit and hospital stay [Raja 2004]. Hraska et al stated that the Ross and Ross-Konno procedures can be performed with a mortality rate approaching zero in both simple and complex left heart lesions even in the neonates and infants [Hraska 2004]. Jaggers and colleagues demonstrated that it can be accomplished with minimal perioperative risk and consider it a more cost-effective treatment modality [Jaggers 1998]. Another significant advantage of this procedure is the minimal risk for the development of thromboembolism and endocarditis [Kouchoukos 2004; Doss 2005].

The principal indication for the Ross procedure is the treatment of aortic valve disease. Young adults, especially with an active lifestyle, patients with contraindications for anticoagulation, women who wish to bear a child after the operation, infants, and adolescents are optimal candidates for pulmonary autograft replacement. Connective tissue disorders, autoimmune rheumatic diseases, and pulmonary valve abnormalities should contraindicate the performance of the Ross procedure [Concha 2004].

Raja et al reported relatively stable autograft function in treated patients with bicuspid aortic valve in their 7-year follow-up period [Raja 2004]. The risk of increased dilation of the autograft in these patients remains a concern [Concha 2004; Kouchoukos 2004]. Whether progressive neo-aortic valve regurgitation and dilation of the autograft can be eliminated by alternative techniques for implantation is not known [Kouchoukos 2004]. Modification of the root inclusion technique by fixation of the aortic annulus and the sinotubular junction may improve the outcome [Skillington 1999; David 2000]. In the literature, the long-term survival of these patients is excellent

[Elkins 1998; Kouchoukos 2004; Luciani 2005]. However, the need for reoperation is a possibility, and it is not dependent on the subtype of the procedure. Injury to the neo-aortic valve at the time of implantation, geometric mismatch, false aneurysm infection, immunological mediated injury, progressive dilation of the autograft, and regurgitation of the neo-aortic valve are potential long-term problems and some of them may mandate a reoperation. The latter is the most common indication for reoperation and usually occurs in the second 5-year interval of follow-up [Kouchoukos 2004]. Kouchoukos and colleagues reported that the actuarial freedom from reoperation on the pulmonary autograft was 75% in 10 years [Kouchoukos 2004]. Furthermore, Luciani et al supported that freedom from reoperation observed in young adults who underwent the Ross procedure is superior to young adults who underwent mechanical replacement [Luciani 2005], and Doss et al showed that young patients with aortic valve stenoses had better hemodynamic performance with the pulmonary autograft compared to mechanical prosthesis [Doss 2005]. Moreover, Pibarot et al showed that the pulmonary autograft provides excellent hemodynamics in aortic position either at rest or during exercise, although moderate gradients were observed during the exercise across the implanted pulmonary homograft [Pibarot 2000].

In the long term, the pulmonary homografts may develop stenoses or insufficiency. Small conduits placed in children have limited durability, and transcatheter interventions will probably be needed to prolong the lifespan of homografts [Forbess 2001]. However, patients older than 10 years who receive cryopreserved homografts have optimal conduit survival [Forbess 2001]. Kouchoukos et al reported that freedom from reoperation on the pulmonary allografts was 86% at 10 years [Kouchoukos 2004]. Lack of reoperation for pulmonary homograft dysfunction even after long-term follow-up is unlike aortic homograft; the pulmonary one lasts longer due to less intrinsic calcium and thickness. Furthermore, the recent development of interventional techniques may extend the life of the homograft without reoperation. In case of pulmonary insufficiency, the threshold for surgical treatment is high enough, considering that it is tolerated well for many years before right ventricular dilatation sets in [Raja 2004]. We elected to proceed with a Shelhigh stentless valved conduit, which is glutaraldehyde cross-linked, detoxified, and heparin-treated with No-React (Millburn, NJ). It ensures stable cross-linking and shows promising hemodynamic properties that make it an attractive alternative to homografts [Carrel 2003].

In conclusion, the Ross procedure is a technically demanding operation that has been established worldwide after a cumulative experience of more than 30 years as a viable alternative, especially in young patients who wish to continue an active lifestyle free from anticoagulation therapy. Late complications may be limited by careful operative planning, new techniques, and regular follow-up. Patients should be informed about this surgical procedure and have the opportunity to choose it.

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