A complex triple valve repair in a young rheumatic patient

Sandro Gelsomino, Carlo Rostagno*, Sergio Bevilacqua, Sabina Caciolli*,
Irene Capecchi*, Giancarlo Casolo*, Gian Franco Gensini*, Pierluigi Stefano

*Cardiac Surgery Unit, Careggi Hospital, *Department of Critical Care Medicine and Surgery,
University of Florence, Florence, Italy*

We report a case of a 22-year-old Ethiopian female presenting with multiple rheumatic valve disease. She was admitted to hospital because of dyspnea at rest. She underwent open mitral commissurotomy associated with splitting of the postero-medial papillary muscle, aortic right-coronary-left coronary commissural resuspension with resection of fibrous tissue from the free-edge cusps and open tricuspid commissurotomy of all three commissures completed with chordal shortening of the anterior leaflet. The postoperative course was uneventful. The patient was asymptomatic without recurrence of symptoms at 2 months. Echocardiography confirmed the satisfactory outcome of the multiple repair with no residual insufficiency. Multiple repair is advisable for patients living in many areas of the Third World, where the safety of long-term anticoagulation cannot be assured.

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In developing countries acute rheumatic fever is still a common and serious public health problem[1]. In the western world the incidence of rheumatic diseases has markedly decreased[2] and surgeons are usually faced with elderly patients with chronic calcified lesions requiring valvular replacement[3]. However, focal outbreaks of rheumatic fever in the United States renewed interest in this pathology in the developed countries as well[4]. Rheumatic fever remains the main cause of valvulopathy in non-industrialized countries[1,2] where the prevalence of the disease is still unacceptably high among children and adolescents[5] and where rheumatic fever is the main cause of cardiovascular death in the first decade of life[6].

Even though most groups sustained the usefulness of multiple valve replacement for such patients[7], nonetheless they are not ideal candidates for prosthetic valve replacement because of the limited durability of xenografts, particularly in the young population, and because of the well-known drawbacks of mechanical heart prostheses[8,9].

For these reasons surgeons were encouraged to resort to more conservative procedures. However, although the feasibility of mitral as well as of tricuspid valve repair have been well demonstrated in young rheumatic patients[10], aortic valve repair techniques such as pericardial aortic

cusp extension[11] or cusp resuspension, have still not gained the worldwide popularity of mitral and tricuspid repairs. These procedures are still not standardized and fully reproducible, are very technically demanding and the results of repair of the rheumatic aortic valve are still uncertain[12].

We report a successful repair of the mitral, aortic and tricuspid valves in a young patient who presented with complex lesions secondary to rheumatic fever.

Case report

A 22-year-old Ethiopian black female, who was a known case of multiple valve disease due to rheumatic fever in childhood, was admitted to hospital because of dyspnea at rest.

On cardiac examination, the point of maximal impulse was at the fifth intercostal space, medial to the left midclavicular line, S_1 was muffled at the apex and left sternal border, S_3 and S_4 were not audible and P_2 was accentuated; a pansystolic murmur (grade 3/6 Levine) radiating to the left axilla was best audible at the apex and left lower sternal border and a diastolic rumbling murmur was best audible at the apex. The laboratory data and blood cell count were normal. Chest X-ray showed cardiomegaly with a double atrial shadow. ECG was sug-

gestive of sinus rhythm with enlargement of the left atrium, right atrium and right ventricle. Transthoracic echocardiography and transesophageal echocardiography confirmed right atrial dilation (7.1 cm), left atrial dilation (5.4 cm) and mitral stenosis (mitral valve area 1.2 cm^2 calculated using the pressure half-time) with leaflet thickening, commissural fusion, diastolic "doming" of the anterior leaflet and systolic prolapse leading to mild mitral regurgitation (grade II). The aortic valve was tricuspid, with cusp thickening and moderate central aortic regurgitation (Fig. 1) due to lack of coaptation between the left coronary cusp and a prolapsing right coronary cusp. The tricuspid valve showed leaflet thickening and commissural fusion resulting in valve stenosis (valve area 1.5 cm^2); the anterior leaflet showed a systolic prolapse due to chordal elongation leading to valve incompetence (grade II).

The heart was exposed through a median sternotomy. Complete cardiopulmonary bypass was established by means of ascending aortic and bicaval cannulation. Antegrade and retrograde cold blood cardioplegia and systemic hypothermia were used for myocardial protection. The left atrium was incised behind the interatrial groove and open mitral commissurotomy of both commissures was performed, associated with splitting of the postero-medial papillary muscle and resection of the secondary chordae. A J-shaped incision was made on the aorta and fibrous tissue was resected from the free-edge cusps. The aortoplasty was completed by right-coronary-left coronary commissural resuspension performed as described in the literature[13]. The right atrium was incised and an open tricuspid commissurotomy of all three commissures was performed, associated with chordal shortening of the anterior leaflet. Tricuspid annuloplasty was completed as originally described by Kay et al.[14]. There were no intraoperative complications. At the end of surgery, intraoperative transesophageal echocardiography confirmed normal valvular function without residual insufficiency (Figs. 2 and 3). The postoperative course was uneventful. The
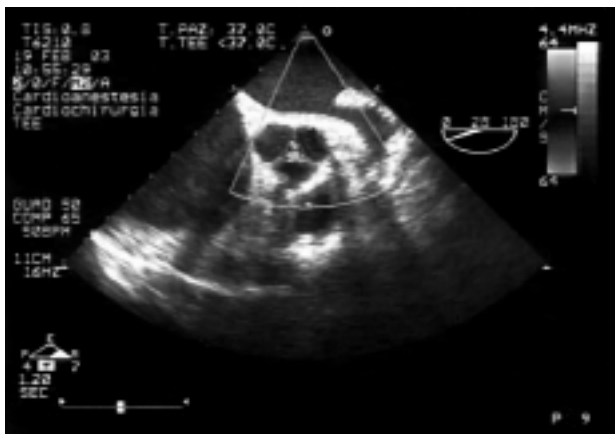

Figure 1. *Transesophageal echocardiography: preoperative image of the aortic valve showing moderate central regurgitation.*

Figure 2. *Transesophageal echocardiography: postoperative image of the aortic valve confirming no residual regurgitation.*
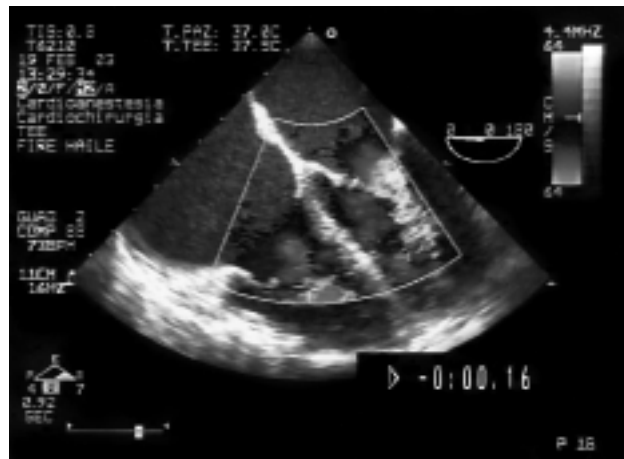

Figure 3. *Transesophageal echocardiography: postoperative image of the mitral and tricuspid valves confirming no residual regurgitation.*

patient was extubated within 6 hours of surgery, discharged from the intensive care unit on postoperative day 1 and from hospital on postoperative day 5. From the first postoperative day onwards, she started a 6-week regimen of warfarin sodium. The target INR was 2.0 to 2.5. The patient was asymptomatic without recurrence of symptoms at 2 months after the operation and a transthoracic echocardiographic control confirmed the excellent outcome of the multiple repair with no stenosis (mitral and tricuspid areas 2.7 and 3 cm^2 respectively) or residual insufficiency.

Discussion

Although the incidence of rheumatic fever in industrialized countries has declined to 0.05 per 1000 per year and rheumatic heart disease has declined in the United States (0.6 per 1000) and Japan (0.7 per 1000)[15,16], this is not so in developing countries where rheumatic fever and rheumatic heart disease still con-

stitute a challenging problem. High rates have been recorded in Zambia (12.6 per 1000), Sudan (10.2 per 1000), Bolivia (7.9 per 1000), Egypt (5.1 per 1000) and Kathmandu city, Nepal (1.2 per 1000) between 1986 and 1990[17,18]. Rheumatic valvular diseases are still a surgical challenge, especially in young patients in whom the use of bioprostheses is limited by their durability and that of mechanical prostheses is complicated by problems resulting from permanent anticoagulation. Furthermore, the operative risk of double valve replacement is still high, ranging between 5 and 12% and the simultaneous insertion of two prostheses has been demonstrated to be an incremental risk factor for late mortality[19]. The increasing awareness of the limitations of the use of prosthetic valves, especially in young individuals, has obliged cardiac surgeons to explore and apply more conservative procedures. Nonetheless, whereas mitral valve repair and tricuspid valve repair constitute widely accepted forms of surgical treatment[10], aortic valve repair, even though continuously evolving and more widely applied than in the past, is still far from having met the key requirements of a complete reproducibility and standardization[12,20]. However, although the advantages of mitral valve repair over mitral valve replacement are well documented[10,21], the durability of mitral valve repair is not 100%[10,12,22] and the 15-year freedom from mitral valve reoperation in patients with rheumatic disease is significantly lower than in patients with degenerative disease[10]. The durability of aortic valve repair is even less satisfactory. In patients with aortic regurgitation, the 5-year freedom from reoperation after repair of bicuspid aortic valves is 87%[22]. On the other hand, the repair of rheumatic aortic valves is associated with 30-month freedom from reoperation ranging between 77 and 94%[23]. Little information is available regarding the long-term results following multiple repair of rheumatic valves. Bernal et al.[12] documented a 25% 22-year freedom from aortic valve reoperation and a 21% 22-year freedom from mitral valve reoperation in patients with rheumatic disease. In this study, the authors concluded that conservative procedures for rheumatic aortic valve disease are not appropriate. Other studies demonstrated that mild rheumatic aortic regurgitation at the time of mitral valve surgery rarely progresses to severe aortic valve dysfunction; therefore, these authors concluded that prophylactic aortic valve surgery is not necessary in patients with mild rheumatic aortic valve disease[24,25]. Other data suggest a survival advantage for the strategy of aortic valve replacement combined with mitral valve repair[26,27]. In a recent report, Gillinov et al.[28] demonstrated a limited durability of double valve repair; they concluded that "valve repair should be reserved only for patients who cannot be anticoagulated, and should be used with caution in patients with aortic stenosis, rheumatic valve disease or anterior mitral leaflet pathology". In contrast to these groups, other authors have demonstrated a satisfactory durability of aortic

valve repair in children and young adults with severe rheumatic disease[20] and Grinda et al.[3] recently reported a 90% freedom from redo valvular surgery after 5 years in a cohort of 21 young rheumatic patients undergoing triple valve repair. We report a case of a 22-year-old Ethiopian female presenting with multiple valve disease due to rheumatic fever during childhood and undergoing complex triple valve repair. The postoperative course was uneventful and the young female was asymptomatic without recurrence of symptoms at 2 months.

Whereas it is true that the patient's young age may constitute a reasonable risk for reoperation in the future, a conservative approach has been preferred in this case for the following reasons: 1) the favorable surgical anatomy of the valve lesions; 2) the consent of the patient who, before considering double valve repair, was informed of the possibility of reoperation for recurrent valve dysfunction; 3) the patient's young age and the consequent reasonable likelihood that she would desire childbearing in the future; 4) the lack of adequate medical facilities in the country of origin, which does not ensure a safe and stable long-term anticoagulation.

In conclusion, in spite of its limited durability, a strategy of multiple valve repair in young rheumatic patients with severe aortic valve disease may be appropriate in selected cases. In our opinion, this option may become the technique of choice in case of patients living in areas of the Third World, in which poor sanitary conditions, the high rate of recurrent attacks of rheumatic disease as well as inadequate anticoagulation control oblige us to be more aggressive in the conservation of native heart valves.

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