

# Valve Repair for Mitral Insufficiency Secondary to Idiopathic Juvenile Polyarthrititis

Stéphane Aubert<sup>1</sup>, Gilles Hayem<sup>2</sup>, Marie-Claude Aumont<sup>3</sup>, Eric Brochet<sup>3</sup>, Christophe Acar<sup>1</sup>

<sup>1</sup>Cardiovascular Surgery, Institute of Cardiology, Hôpital Pitié Salpêtrière, Paris, Departments of <sup>2</sup>Rheumatology and

<sup>3</sup>Cardiology, Hôpital Bichat, Paris, France

A female patient in whom idiopathic rheumatoid polyarthrititis was diagnosed at the age of 8 years required surgery for severe mitral valve insufficiency 16 years later. Intraoperative analysis revealed a fibrotic endocarditis involving mainly the posterior leaflet. Granulomatous vegetations as well as a large thrombus which filled the left ventricular apex and simulated endomyocardial fibrosis were noted. Valve repair was achieved using an anterior leaflet

augmentation with a patch of mitral homograft associated with a prosthetic ring annuloplasty. Postoperatively, a severe pericardial effusion required surgical drainage. Eight years later, the patient had no cardiac symptoms and echocardiography confirmed a normally functioning mitral valve.

The Journal of Heart Valve Disease 2007;16:324-327

Cardiac valve disease during the course of idiopathic juvenile polyarthrititis is rare, and exceptionally requires surgical correction. The originality of the present case relies on its anatomic presentation, which simulated endomyocardial fibrosis, and the proposed surgical treatment.

## Case report

A 23-year-old woman was hospitalized for heart failure related to a severe mitral valve insufficiency. Her medical history revealed an idiopathic juvenile polyarthrititis, the first symptoms of which occurred when she was aged 8 years. The physical examination showed a bilateral rheumatoid disease which affected mainly the upper limbs, causing severe hand, wrist, and elbow deformities. The patient also had right knee pain, and X-radiography identified erosion of the metatarsophalangeal joints. A nasal polyposis with multiple episodes of sinusitis suggested Wegener disease, but this was disproved by histology of mucous biopsies. The diagnosis of juvenile polyarthrititis was

confirmed by high-level rheumatoid factors (Latex 81 UI/ml; Waaler-Rose 32 UI/ml) with an elevation in anti-keratin antibodies IgG (100); the dosage of native antiDNA and antiphospholipid antibodies remained negative. Previously, various therapies had been instituted, including plaquenil, cortisone, gold salts and D penicillamine.

At three months prior to the hospitalization a progressively worsening dyspnea on exercise occurred, together with an inflammatory syndrome which resolved with cortisone therapy. Biologically, the sedimentation rate was accelerated (35 at the first hour), the leukocyte count was elevated (17,000/mm<sup>3</sup>) with <3% eosinophils, and signs of hepatic insufficiency (prothrombin time 50%, and a 12-fold increase in transaminases).

Transesophageal echocardiography (TEE) revealed a severe mitral insufficiency due to posterior leaflet restriction with severe retraction (Carpentier type IIIa), with a normal left ventricular function and an elevated systolic pulmonary pressure (65 mmHg). The aortic valve was morphologically and functionally normal. The hemodynamics was stabilized using diuretics and vasodilators associated with low-dose cortisone, and surgery was planned for three months later.

Using a standard left atriotomy approach, the inspection revealed fibrotic involvement of the entire mitral valve apparatus. The posterior leaflet and its

---

Address for correspondence:

Dr. C. Acar, Département de Chirurgie Cardiovasculaire, Institut de Cardiologie, Hôpital Pitié Salpêtrière, 50-52 Bd. Vincent Auriol, 75013 Paris, France  
e-mail: c.acar@psl.aphp.fr

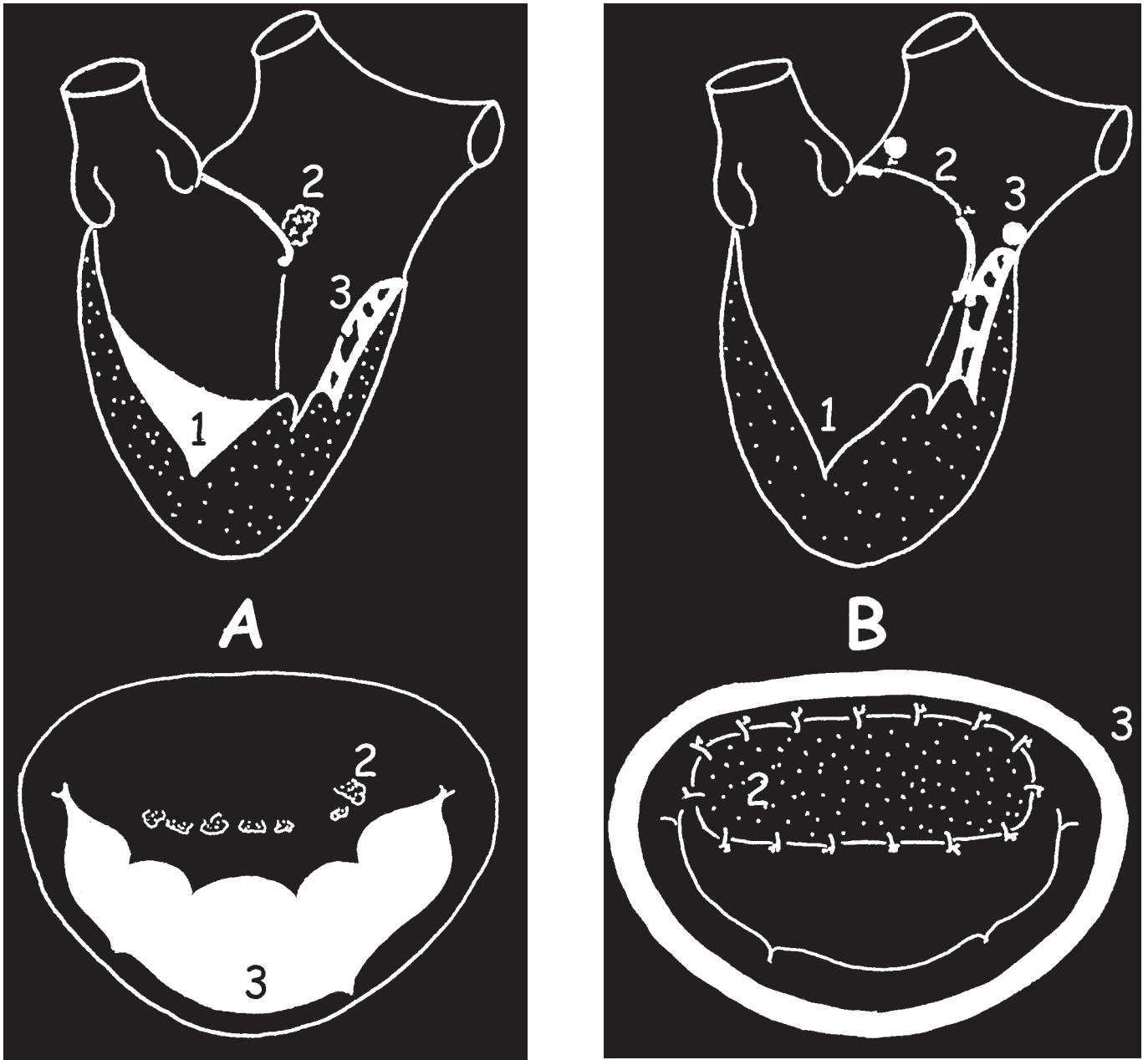


Figure 1: A) Operative findings. 1, apical thrombus; 2, granulomatous vegetations; 3, endocarditis with fibrosis incorporating the posterior leaflet and its chordae. B) Mitral repair. 1, thrombus removal; 2, anterior leaflet augmentation with a homologous leaflet patch; 3, prosthetic ring annuloplasty.

chordae were completely incorporated in the fibrotic process, with complete adhesion to the lateral wall of the left ventricle, as is commonly observed in endomyocardial fibrosis (Fig. 1A). A large organized thrombus filled the apex of the left ventricle. Fine granulomatous vegetations were attached at the free edge of the anterior leaflet (Fig. 1A). Although restricted by the scleroretractile process, leaflet mobility remained satisfactory and the chordae inserted on this leaflet were spared; thus, mitral valve repair was attempted.

Following removal of the thrombus and vegetations, the anterior leaflet was disinserted and its size enlarged using a piece of a cryopreserved homologous mitral leaflet (partial homograft) (Fig. 1B). Intraoperative TEE showed a normally functioning valve repair.

Pathological examination of the operative samples showed fibrin deposits including red blood cells, but no bacterial contamination. During the postoperative course, a marked inflammatory syndrome occurred

together with a pericardial effusion which required surgical drainage (800 ml of clear fluid). Throughout the follow up period the patient remained asymptomatic from a cardiac standpoint, and received no anticoagulation therapy. Treatment of the polyarthritis comprised etanercept and cortisone.

A TEE performed eight years postoperatively showed a stable mitral repair with no residual leak, a mean transvalvular gradient of 6 mmHg with a functional area of 1.97 cm<sup>2</sup>, and a slightly dilated left ventricle (end-diastolic diameter 59 mm) with normal contractility (ejection fraction 72%).

## Discussion

The echocardiographic detection of a degree of valvular insufficiency is a frequent occurrence in patients suffering from rheumatoid polyarthritis, with the mitral valve being involved in 60 to 80% of cases (1,2). A thickening of the mitral leaflets was noted in 20% of these patients (3). However, the intensity of valve dysfunction seldom requires surgical treatment. Those occasional patients with rheumatoid polyarthritis who required surgery were mainly females in their fifth decade of life, who had been followed for over 10 years and whose valve disease was most often aortic regurgitation (4,5). Isolated cases of mitral stenosis or regurgitation, with or without associated aortic valve disease, have been reported (6-8). From a pathologic standpoint, the presence of rheumatoid nodules is infrequent; tumor-like forms have been reported (8), but the most frequent finding is a non-specific fibrosis (4,9).

Juvenile idiopathic polyarthritis constitutes a genuine independent pathological entity, and valve disease which required aortic and/or mitral valve replacement has been described (10,11). Fibrotic lesions, located mainly at the aortomitral junction as observed in spondyloarthritis, have also been reported (12). In the present patient, the valve anatomy was original, but the posterior leaflet and its chordae had become incorporated in the endocarditic fibrotic process that involved mainly the lateral wall of the left ventricle. In addition, a large apical thrombus was noted (Fig. 1). Although these lesions closely mimicked idiopathic endomyocardial fibrosis, the lack of an elevated eosinophil count, and of endocarditis of the right ventricle, disproved this hypothesis. Thus, it seemed logical to impute the mitral disease to the sequelae of multiple episodes of inflammatory reactions generated by the juvenile polyarthritis. The presence of granulomatous vegetations free from microorganisms also suggested Liebman-Sachs endocarditis as a possible diagnosis, but all immunological markers of systemic lupus were negative. Likewise,

the possibility of a primary antiphospholipid syndrome was excluded after repeated dosages.

The surgical treatment comprised a mitral valve repair with removal of the ventricular thrombus. In order to compensate the tissue retraction, the anterior leaflet was augmented by means of a patch of cryopreserved homologous leaflet (Fig. 1). The postoperative course was complicated by pericardial effusion of clear fluid, which is a known cardiac localization of rheumatoid polyarthritis in the inflammatory phase (3). The use of a partial homograft or autologous pericardial patch for anterior leaflet augmentation has offered excellent mid-term results in mitral insufficiency secondary to acute rheumatic fever (13,14). In the present authors' experience, the same technique applied to mitral disease induced by systemic lupus or by primary antiphospholipid syndrome has in general not been successful, and several reoperations have been necessary due to the continuously evolving valvular fibrosis. Although the idiopathic juvenile polyarthritis is an evolving disease, the time frame between the diagnosis of the rheumatoid affection and the onset of cardiac symptoms in the present patient was 15 years, which suggested a slow progression of the endocarditic involvement. The result of the mitral repair has remained remarkably stable for eight years, with the need for long-term anticoagulation being avoided.

## References

1. Nomeir AM, Turner RA, Watts LE. Cardiac involvement in rheumatoid arthritis. Follow-up study. *Arthritis Rheum* 1979;22:561-564
2. Guedes C, Bianchi-Fior P, Cormier B, Barthélémy B, Rat AC, Boissier MC. Cardiac manifestations of rheumatoid arthritis: A case-control transoesophageal echocardiography study in 30 patients. *Arthritis Rheum* 2001;45:129-135
3. Corrao S, Salli L, Arnome S, et al. Cardiac involvement in rheumatoid arthritis: Evidence of silent heart disease. *Eur Heart J* 1995;16:253-256
4. Baldé MD, Ecke JE, Ortega L, et al. Remplacement valvulaire aortique pour insuffisance aortique associée à la polyarthrite rhumatoïde: À propos de 5 cas. *Arch Mal Coeur Vaiss* 2006;99:569-574
5. Moini C, Paemelaire JM, Aupart M, et al. Insuffisance aortique rhumatoïde. À propos d'un cas traité par remplacement valvulaire mécanique. *Ann Cardiol Angiol* 1996;45:329-333
6. Bortolotti U, Casaratto D, Gallucci V, Gasparotto G, Thiene G. Mitral and aortic valve replacement in valvular rheumatoid heart disease. *Chest* 1978;73:427-429
7. Panwar RB, Aggrawal RP, Rajvanshi P, Misra SN. Mitral stenosis and aortic regurgitation in rheumatoid arthritis. *J Assoc Physicians India* 1991;39:776-

778

8. Mounet F, Soula P, Concina P, Baradat G, Cérène A. Les cardio-pathies valvulaires spécifiques dans la polyarthrite rhumatoïde. À propos de deux observations. *Arch Mal Coeur Vaiss* 1997;90:987-989
9. Delahaye JP, Milon H, Loire R, Lantelme P. Etiologies et lésions anatomiques des valvulopathies acquises. In: Acar J, Acar C (ed.), *Cardiopathies valvulaires acquises*. Flammarion, Paris 2000:3-27
10. Ozer S, Alehan D, Ozme S, Bakkaloglu A, Soylemezoglu O. Mitral and aortic insufficiency in polyarticular juvenile rheumatoid arthritis. *Pediatr Cardiol* 1994;15:151-153
11. Chen YS, Yang YH, Lin YT, Chiang BL. A patient diagnosed with pauciarticular juvenile rheumatoid arthritis after mechanical prosthetic valve replacement due to aortic regurgitation. *J Microbiol Immunol Infect* 2004;37:200-202
12. Lee SJ, Im HY, Schueller WC. HLA B27 positive juvenile arthritis with cardiac involvement preceding sacroiliac joint changes. *Heart* 2001;86:e19
13. Acar C, de Ibarra JS, Lansac E. Plastie d'élargissement de la valve antérieure dans l'insuffisance mitrale rhumatismale. *Arch Mal Coeur* 2004;97:875-880
14. Ali M, Iung B, Lansac E, Bruneval P, Acar C. Homograft replacement of the mitral valve: Eight-year results. *J Thorac Cardiovasc Surg* 2004;128:529-534