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Quadruple valve replacement for rheumatic valvular disease

Excellent durability for 20 years

Valvular heart disease progresses slowly and generally remains asymptomatic. There is currently no medical cure for valvular disease. As lifestyle modifications and medications can only reduce the associated symptoms, surgery is generally required [1]. When valve repair techniques are not an option, valve replacement with mechanical or biologic prosthetic valves is inevitable. Despite the development of new operative and myocardial protection techniques, multiple valve surgery for advanced rheumatic heart disease (RHD) remains challenging. These patients are usually in the end stage of disease, and are often subjected to a long duration of cardiopulmonary bypass and prolonged myocardial ischemia during a complex operation [2, 3]. Furthermore, compared with the replacement of a single valve, the rates of long-term morbidities, such as thromboembolism, anticoagulation-related hemorrhage, endocarditis, and paravalvular leakage, are higher after multiple valve replacement [4]. Here, we report a rare case of a patient who underwent quadruple valve replacement for RHD 20 years ago. To the best of our knowledge the patient's survival time after surgery is the longest described among the current reports of quadruple valve replacement.

Case report

A 58-year-old woman presented with decreased exercise tolerance with a medical history of quadruple valve replacement for RHD 20 years earlier. Physical examination showed no lower extremity edema, ascites, orthopnea, or paroxysmal nocturnal dyspnea. Blood pressure was 110/60 mmHg and the pulse rate was 70 bpm. Auscultation revealed normal, crisp prosthesis sounds with no third and fourth heart sounds. Physical examination of the lungs showed normal results. An electrocardiogram showed normal sinus rhythm (**Fig. 1**) and a chest radiograph showed four mechanical valves (**Fig. 2**). Laboratory data revealed no abnormalities in liver and kidney functions, the white blood cell count was 6,700/µl, platelet count was 209,000/µl, and the international normalized ratio was 3.19.

Transthoracic echocardiography demonstrated all four valves were mechanical (**Fig. 3, 4**) and also showed that all mechanical prostheses were functioning normally.

Discussion

In 1952, the first mechanical heart valve was implanted by Dr. Charles Hufnage [5]. Improvements in myocardial protection and operative techniques and the evolution of mechanical valve designs have dra-

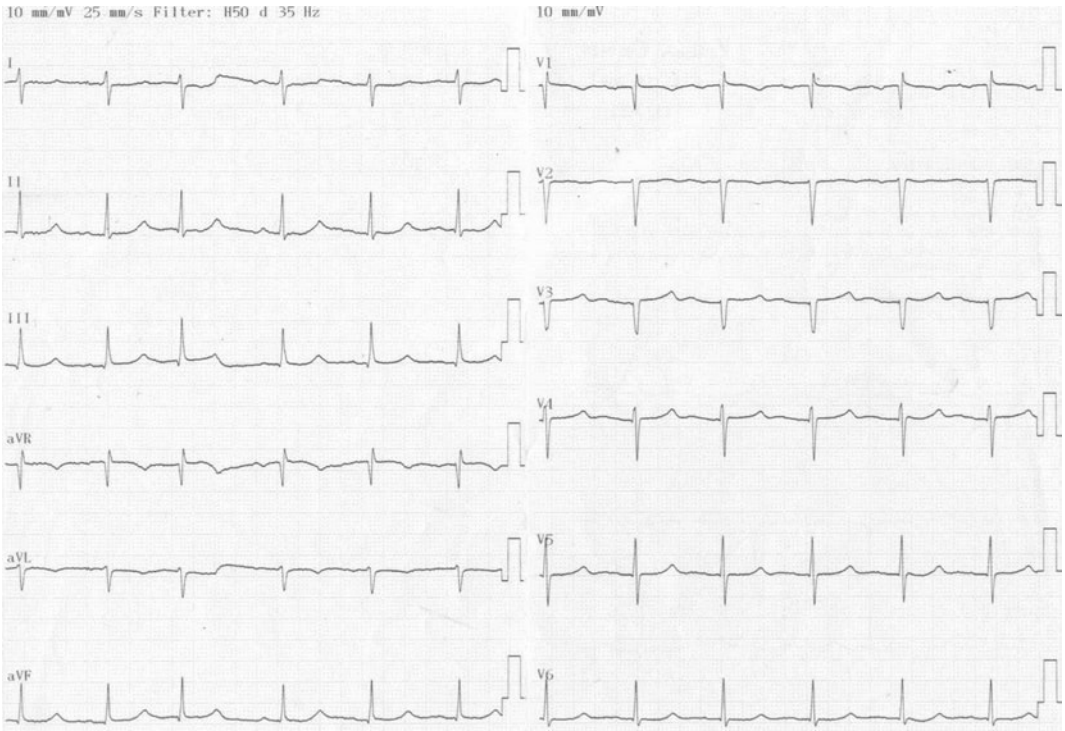

Fig. 1 ◀ Electrocardiogram shows normal sinus rhythm

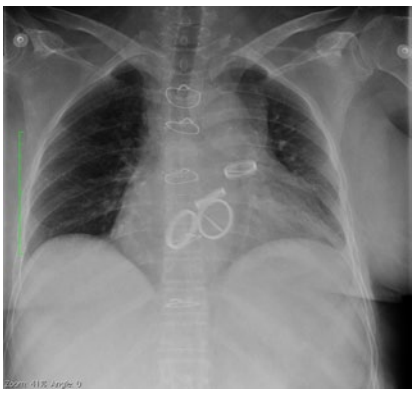

Fig. 2 ▲ Chest x-ray film reveals four mechanical prostheses and clear lung fields

matically decreased the risk of operative complications. Mechanical valves have the advantage of potentially lasting a lifetime (>30 years) and bileaflet mechanical prostheses have demonstrated excellent long-term results [6].

Mechanical valves are mostly preferred in aortic and mitral positions. Triple valve replacement is still challenging for the more advanced stages of the disease and complexity of the operation. Furthermore, associated cardiovascular diseases and concomitant operative procedures complicate the outcome but a large series is reported in the literature with operative mortality rates ranging from 8 to 31% [2, 4, 7]. Alsoufi et al. [7] reported that the 5-year and 10-year survival rates after triple valve replacement were 75 and 61%, respectively. In the tricuspid position, one of the major risks of valvular replacement is the necessity for permanent pacemaker implantation due to atrioventricular block ranging from 15 to 20% [8, 9]. Pulmonary valve replacement (PVR) is rarely performed in the adult population and has generally been performed in patients with congenital heart disease, mostly after the correction of tetralogy of Fallot [10]. Quadruple mechanical valve replacement is very rare in the literature and the case reported here has the longest follow-up without complications. After detailed examination of the heart and lungs, it was concluded that the patient's complaints were not related to the circulatory or respiratory system.

Involvement of all four heart valves can be observed in several situations. In 1987, Hossack et al. [11] reported the case of a patient with rheumatic involvement of all valves treated with aortic, mitral, and pulmonary valve replacement followed by tricuspid valve replacement. In 2006, Chiappini et al. [12] reported on a patient who underwent replacement of the tricuspid and pulmonary valves and repair of the aortic and mitral valves due to carcinoid heart disease. In 2008, Castillo and colleagues reported on a patient who underwent tricuspid and pulmonary bioprosthetic valve replacement, mitral and aortic valve reconstruction secondary to carcinoid heart disease. Echocardiographic follow-up after 25 months showed competent mitral and aortic valves with mild regurgitation [8]. In 2009, Seeburger et al. reported on a patient with endocarditis of the aortic, tricuspid, and pulmonary valves with severe insufficiency and moderate mitral insufficiency with suspected vegetations; they successfully replaced all heart valves in a single operation [13]. In 2010, Arghami et al. [14] described their experiences with seven patients who underwent quadruple valve replacement because of carcinoid heart disease and concluded that quadruple valve replacement in symptomatic patients is a reasonable option that can result in functional improvement. In 2012, Cao and colleagues reported a 11-year follow-up study of a patient with subacute rheumatic endocarditis who underwent quadruple valve replacement with mechanical valves combined with the correction of complex congenital heart disease. No complications occurred during the follow-up [3].The results of the case reported here suggest that replacement of all cardiac valves with me-

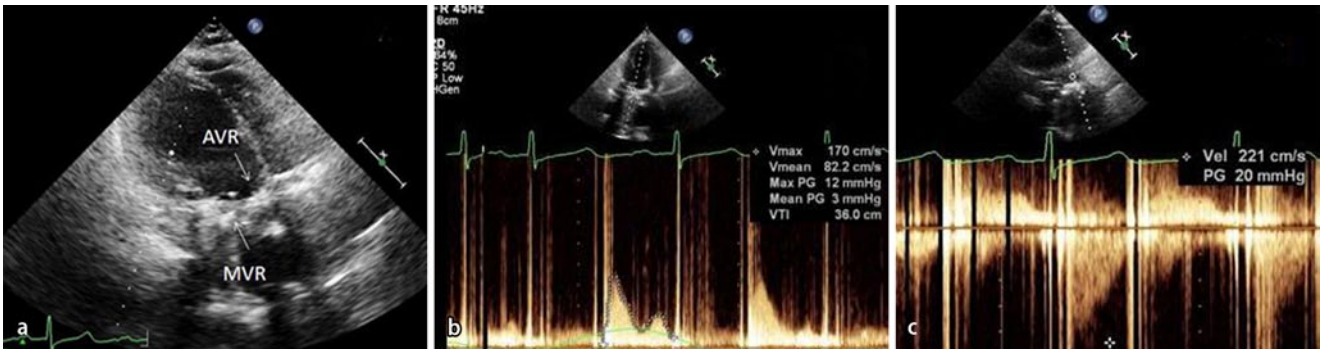

Fig. 3 ▲ Transthoracic echocardiography showing **a** aortic and mitral mechanical prostheses, **b** aortic valve prosthesis pressure gradient, and **c** mitral valve prosthesis pressure gradient. *MVR* mitral valve replacement, *AVR* aortic valve replacement

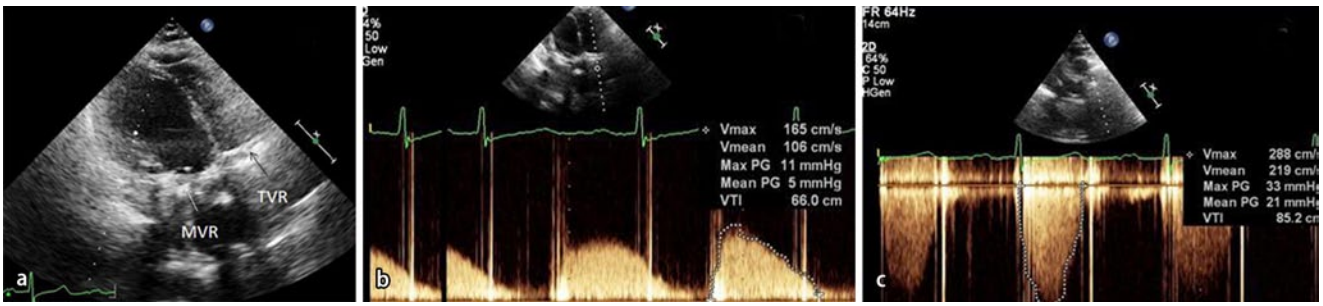

Fig. 4 ▲ Transthoracic echocardiography showing **a** mitral and tricuspid valve prostheses, **b** tricuspid valve prosthesis pressure gradient, and **c** pulmonary valve prosthesis pressure gradient. *MVR* mitral valve replacement, *TVR* tricuspid valve replacement

chanical prostheses can be accomplished in selected patients with quadruple valve disease. This case is the longest follow-up study of a patient who underwent quadruple valve replacement without any complications.

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Compliance with ethical guidelines

Conflict of interest. A.B. Budak, K. Korkmaz, H.S. Gedik, S.B. Genç, A.Ü. Yener, İ. Alur, and K. Çağlı state that there are no conflicts of interest. Consent was obtained from all patients identifiable from images or other information within the manuscript. In the case of underage patients, consent was obtained from a parent or legal guardian.

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