CASE REPORT

Quadruple valve replacement in a patient with severe rheumatic heart disease

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Summary

We present a patient with rheumatic heart disease involving all the heart valves. An intraoperative transoesophageal echocardiography confirmed severe mitral stenosis, severe aortic regurgitation, severe tricuspid regurgitation and stenosis, and severe pulmonary stenosis. The patient underwent successful quadruple valve replacement during a single operation at the Groote Schuur Hospital, Cape Town, South Africa.

Key Words
▶ echocardiography
▶ rheumatic heart disease
▶ valve replacement

Learning points:

- Rheumatic heart disease can affect all the heart valves including the pulmonary valve.
- Intraoperative transoesophageal echocardiography is key for diagnosis, monitoring and confirmation of successful surgical result during heart valve surgery.
- Combined surgical procedure of all four valves is possible though associated with long procedural time.

Background

Rheumatic heart disease (RHD), a consequence of rheumatic fever, is the leading cause of cardiovascular morbidity and mortality in young patients. According to the 2015 Global Disease Burden study, RHD was associated with 319,000 deaths and 33.4 million cases globally (1). The majority of these cases occur in the poorest regions of the world. RHD commonly affects the mitral, aortic and occasionally tricuspid valves. Rheumatic pulmonary valve disease is very rare. During heart surgery, the use of intraoperative transoesophageal echocardiography (TOE) is key in confirmation of valvular heart pathology and as a monitoring tool during surgery. There is paucity of data on quadruple valve replacement.

Case presentation

A 32-year-old female presented with worsening dyspnoea (NYHA III, CCS 0), two pillow orthopnoea, pedal oedema, fatigue, polyarthralgia (both small and large joints), sore throat and a fever for 2 months. She was HIV positive with a CD4 count of 540, controlled with antiretroviral medication.

On examination, she had bilateral pedal oedema, wrist tenderness and no stigmata for infective endocarditis. She was 164 cm tall, 64 kg with BMI 23.8.

Her cardiovascular examination revealed distended neck veins (jugular venous pressure (JVP) 7 cm above sternal notch), normal apex as well as a parasternal heave. A precordial grade 3/6 pansystolic murmur was present increasing with inspiration over the pulmonic area as well

as an end diastolic murmur with a loud P2 over the aortic area. She had a pulsatile liver.

Investigations

Transthoracic echocardiography

Transthoracic echocardiography (TTE) examination demonstrated thickened mitral valve leaflets with severe mitral stenosis (MS), moderate-to-severe aortic regurgitation (AR), tricuspid valve leaflets thickened with severe tricuspid regurgitation (TR)/tricuspid stenosis (TS), and restricted mobility of pulmonary cusps with moderate pulmonary stenosis (PS). The patient's left atrium and right atrium (RA) were dilated. The ejection fraction of her left ventricle (LV) was 70%. All quantification of the valvular pathologies was made according to the 2017 ESC/EACTS Guidelines for the management of valvular heart diseases (2).

12 lead ECG

Normal sinus rhythm.

Intra operative TOE evaluation

Intraoperative TOE images were obtained using a GE Vivid E9 machine and 6 VT-D transducer. TOE confirmed calcified, thickened leaflets with severe MS, severe AR, severe TR/TS and severe PS.

Both atria were dilated and the interatrial septum was bowing to the left side. RV dilatation and a thickened free wall were visualised.

The LV systolic function was preserved. The aortic dimensions of this patient were normal with holodiastolic flow reversal demonstrated on pulse wave Doppler in the descending thoracic aorta indicative of severe AR.

TOE confirmed all preoperative TTE findings, although the pressure gradients were slightly lower on TOE due to changes in loading conditions associated with the general anaesthesia hemodynamic state of the patient.

Treatment and outcome

Median sternotomy was performed and routine cardiopulmonary bypass (CPB) was commenced after surgical aorto-bicaval cannulation, placement of right superior pulmonary vein vent and an anterograde blood cardioplegia cannula. After initiation of moderate hypothermia CPB (32°C), aortic cross clamping was performed and cardioplegia was administered. Thereafter, 25 mm ATS mitral mechanical prosthesis, 20 mm ATS aortic mechanical prosthesis and 23 mm pulmonary freestyle bioprosthesis were used to replace the native valves. The heart was de-aired and sinus rhythm resumed after cross clamp was released. Tricuspid valve was replaced with 27 mm mosaic bioprosthesis while the heart was beating. The total bypass time was 222 min and the cross clamp time 155 min. Temporary pacing leads as well as sequential permanent epicardial pacing leads were placed and secured. The patient had uncomplicated intensive care unit stay for 2 days and uneventful ward stay as well. She was discharged home on the 6th postoperative day with a haemoglobin of 9.3, normal kidney function and international normalised ratio of 2.45.

Discussion

RHD is a chronic disease affecting the heart valves. It occurs after a single or multiple bouts of acute rheumatic fever, which is an autoimmune response to group A *Streptococcus* infection (3). Penicillin with or without surgery remains the backbone for treatment of RHD. However, this disease remains a big challenge in low- and middle-income countries, which still have high prevalence and mortality (1). It usually presents with leaflet thickening, fibrosis, commissural fusion and often total destruction of the valve (Figure 1).

Rheumatic mitral and aortic valve disease has been extensively described in literature together with echocardiographic assessment of these valves.
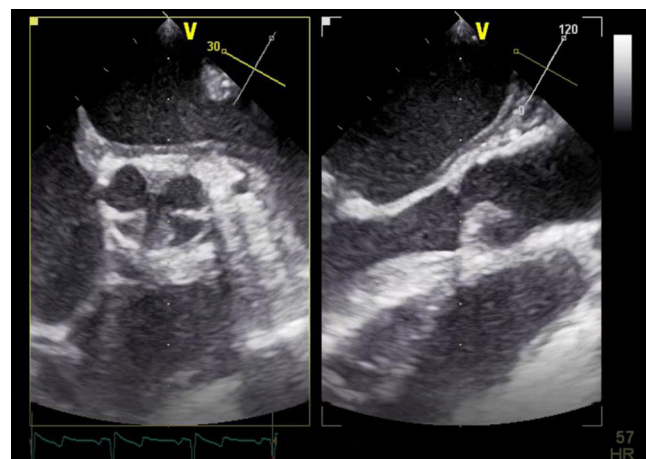

Figure 1
ME AV biplane view showing commissural fusion of aortic valve leaflets.

Quadruple valve replacement, however, is rare. Arghami *et al.* published a case series of seven patients at the Mayo clinic who underwent quadruple valve replacement for carcinoid heart disease between 1989 and 2010 (4). Cao *et al.* from China also published a case report on quadruple valve replacement in a patient with rheumatic endocarditis (5). In both cases, there was functional improvement in a majority of the patients. Our patient provided us with an opportunity to assess such a case using TOE according to recognised guidelines.

Quantification of severity of MS can be done by various methods. Planimetry is the original reference measurement of mitral valve area (MVA) (2). It has the advantage of being independent of loading conditions and associated heart pathologies, but it is not easily reproducible. The pressure half time (PHT) method is generally easy and reliable to perform and is therefore widely used (6) (Fig. 2). However, it may be misleading in cases of AR, in the presence of abnormal compliance of cardiac chambers or immediately after balloon mitral valvotomy (6). The use of the continuity equation to assess valve area is not valid in cases with concurrent significant MR or AR. Its accuracy and reproducibility are also limited given the number of measurements and calculations involved (5). The proximal isovelocity surface area (PISA) is technically demanding, requires multiple measurements and may be unreliable, if not accurately performed by an experienced operator. Mean mitral gradient, as assessed by pulse or continuous wave Doppler echocardiography, is not a reliable means to assess the severity of MS because it is highly dependent on flow conditions (5) (Fig. 3). However, the gradient value should
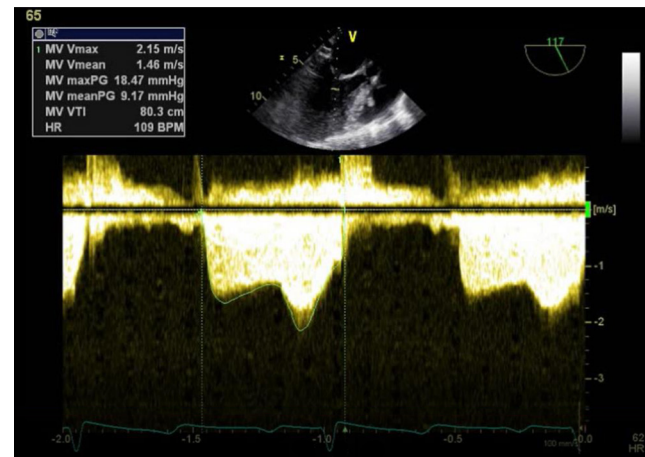

Figure 3
ME LAX CWD with calculated pressure gradients across mitral valve.

be consistent with the valve area value. In this particular patient, calculation of MVA by planimetry was difficult due to the extensive calcification of the valves with lots of artefacts hence difficult to delineate borders. However, MVA derived from PHT was 0.5 cm² with a mean gradient of 9.17 mmHg.

The severity of AR is assessed using colour Doppler flow imaging (vena contracta of regurgitant jet) and pulse wave Doppler imaging (holodiastolic flow reversal in the descending thoracic and abdominal aorta) (7). These indices are influenced by loading conditions and the compliance of the ascending aorta and LV. Quantitative Doppler echocardiography, using the continuity equation or analysis of PISA, is less sensitive to loading conditions and provides measures of regurgitant volume,
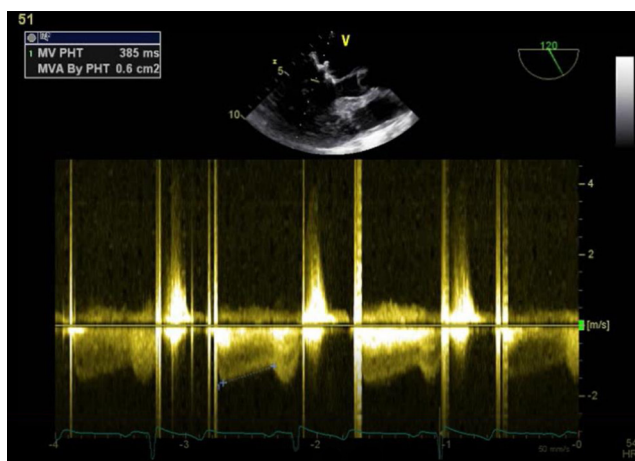

Figure 2
ME AV LAX MVA by PHT. Note the 'hockey stick' appearance of AMV leaflet. MVA, mitral valve area; PHT, pressure half time.

Figure 4
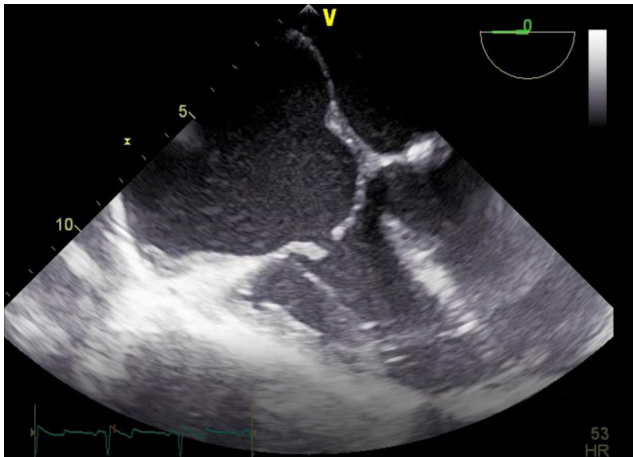ME RV inflow outflow with CFD and CWD across TV.

Figure 5
ME 4CH view showing dilated cardiac chambers and poor coaptation of tricuspid valve leaflets.

regurgitant fraction and effective regurgitant orifice (7). This patient had holodiastolic flow reversal in the descending thoracic aorta with AR vena contracta of 1 cm.

TR is most frequently functional in origin secondary to another disease process causing RV dilation, distortion of the subvalvular apparatus, tricuspid annular dilation or a combination of these (8) (Figs 4 and 5). Echocardiographic findings in patients with severe TR include inferior vena cava dilation >2 cm and systolic flow reversals in the hepatic veins (8) (Fig. 6). The effective regurgitant orifice area can be estimated by measuring the vena contracta on colour-flow Doppler imaging; a vena contracta larger than 0.7 cm indicates severe TR. Quantitative Doppler assessment is also feasible with use of PISA method (8). This patient had hepatic vein flow reversal with a systolic
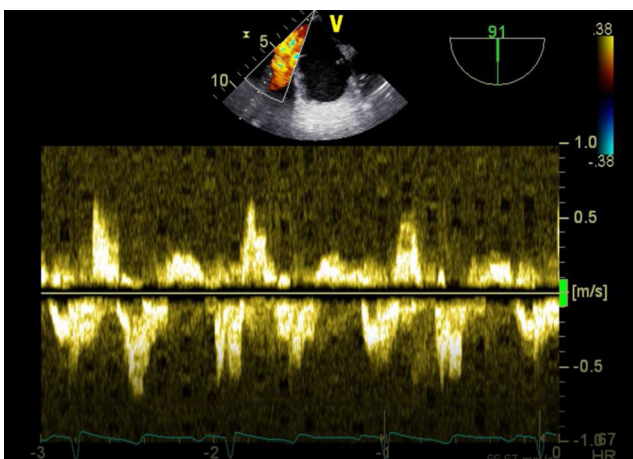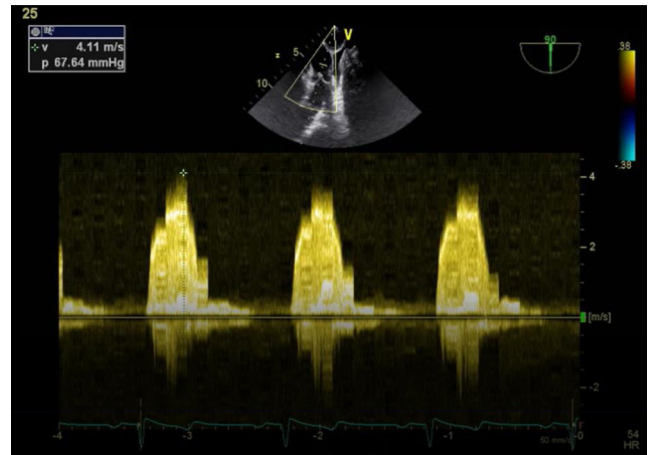

Figure 7
UE aortic arch SAX CWD across the pulmonary valve.

RV to RA regurgitant gradient across the tricuspid valve of 60 mmHg.

TS occurs infrequently and is essentially never seen in isolation. One of the commonest cause of TS is RHD. Evaluation of the degree of TS includes calculation of the mean pressure gradient and valve area. Tricuspid stenosis is considered haemodynamically significant when the mean gradient is 5 mmHg or greater, valve area is 1.0 cm^2 or less and the PHT is 190 ms or longer (8, 10).

The pulmonary valve is the least imaged of the cardiac valves because of its distance from the probe in adults (9). PS is related to a congenital or genetic disorder in 95% of cases (10). Rarely, carcinoid syndrome and RHD may cause PS, but these lesions essentially always occur in conjunction with other valve disease. Using the peak transpulmonary velocity measured with continuous wave
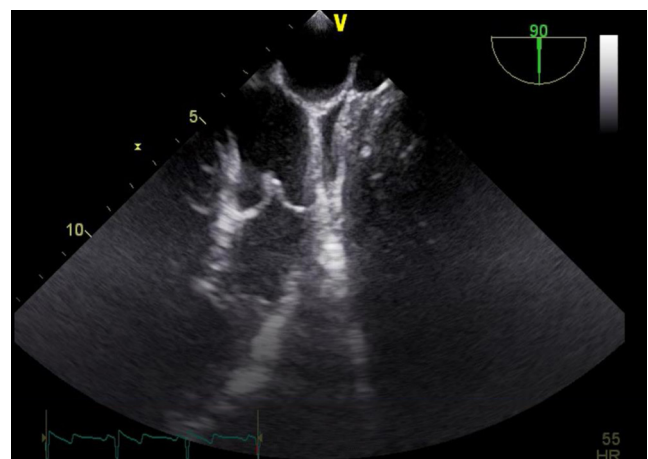

Figure 6
PWD of hepatic vein showing flow reversal.

Figure 8
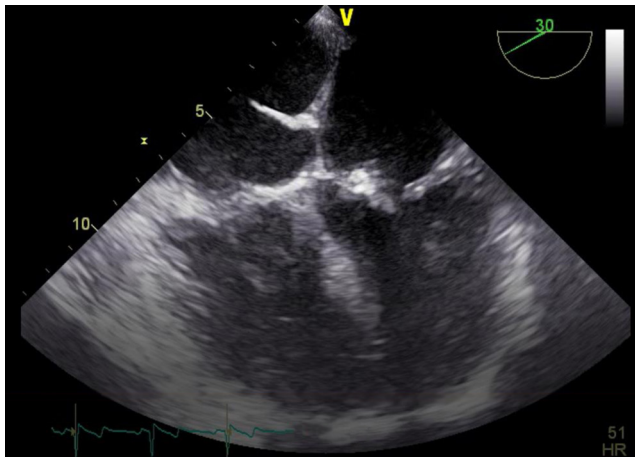Upper oesophageal aortic arch short axis view showing thickened pulmonary valve leaflets.

Figure 9
ME 4CH showing thickened valves and dilated heart chambers.

Doppler to calculate the transvalvular pressure gradients determines the severity of PS (Fig. 7). Severe PS is defined as a peak gradient greater than 60 mmHg, moderate as a peak gradient 36–60 mmHg, and mild as a peak gradient less than 36 mmHg (10). This patient had severe PS with pressure gradient of up to 67 mmHg across the valve. The leaflets were thickened in keeping with RHD (Fig. 8).

The response of the RV to chronic pressure overload differs from that of the LV. Although the initial response is an increase in wall thickness, ventricular dilation may occur and depends on the acuteness and severity of the pressure overload state (10) (Fig. 9). With a gradual increase in RV pressure, RV size and systolic function may remain normal with a compensatory increase in RV wall thickness (Fig. 7). Although few studies have analysed the extent of improvement in RV function after relief of PS, the improvement in RV dimensions and systolic function in most patients after lung transplantation supports the concept that systolic function improves with decreased afterload (10). For routine clinical assessment, tricuspid annular plane systolic excursion is a simple and reproducible measurement of RV longitudinal function that correlates well with RV ejection fraction measured with cardiac MRI. Other measures of RV function include the right-sided index of myocardial performance (Tei Index) (8).

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