CASE REPORT

Treatment of trivalvular rheumatic heart disease: why it matters where we live

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SUMMARY

A 42-year-old woman who had recently emigrated to the UK from Ethiopia presented to her general practitioner with increasing breathlessness and palpitations. Transoesophageal echocardiogram revealed rheumatic heart disease affecting three valves, and severe aortic stenosis. ECG demonstrated atrial fibrillation. She was treated operatively with mitral and aortic valve replacement and tricuspid valve repair and put on lifelong warfarin. Four months later she remains well, with substantially increased exercise tolerance. This case report summarises treatment options for severe rheumatic heart disease and discusses differences in prevention and management between patients presenting in the UK and in the developing world.

BACKGROUND

Rheumatic heart disease (RHD) is rare in the developed world but remains a significant global burden, leading to 250 000 premature deaths worldwide annually, many of which are in sub-Saharan Africa.¹ Most patients initially present with shortness of breath and, without appropriate intervention, will progress to congestive heart failure. In the presence of a valvular murmur, RHD should therefore be considered as a significant differential diagnosis by clinicians working in areas with large immigrant populations, particularly in patients originating from sub-Saharan Africa or south-central Asia.

CASE PRESENTATION

A 42-year-old female teacher, recently arrived in the UK from Ethiopia, presented to her general practitioner (GP) for a repeat prescription of drugs started 4 years previously in Addis Ababa, which included warfarin, furosemide, propranolol and spironolactone. She reported increasing breathlessness and occasional palpitations. She could walk for 5 min before becoming breathless, less when uphill. Her ankles were occasionally swollen but she denied any chest pains or paroxysmal nocturnal dyspnoea. Her palpitations lasted for roughly 10 min and occurred up to five times a day. She recalled having 'tonsillitis' with 'flu-like' symptoms and then shortness of breath 12 years previously, in Ethiopia. Following this episode, she was given monthly injections of benzylpenicillin and prednisolone for 3 years, before being discharged from the cardiologist's care. She had sought further help from the cardiologist, for breathlessness, 8 years later, and was put on the four drugs previously mentioned. She was told that this was the optimal

treatment which could be offered to her in Ethiopia.

INVESTIGATIONS

Following referral by her GP in the UK, the patient was further investigated.

ECG demonstrated atrial fibrillation and trans-thoracic and transoesophageal echocardiogram revealed RHD with severe tricuspid regurgitation, dilated superior vena cava, moderate mitral stenosis with mild regurgitation and moderate aortic regurgitation with rolled-up and thickened aortic leaflets (figures 1–3).

Mean pulmonary artery pressure was >50 mm Hg (normally <25 mm Hg) and pulmonary artery systolic pressure was 65–70 mm Hg (normally <40 mm Hg).²

TREATMENT

Surgery consisting of mechanical valve replacement of the aortic and mitral valves and repair of the tricuspid valve was proposed to the patient.

She was counselled that the risk of death was 20% and, of stroke, 5%. She was also told she would need to continue lifelong anticoagulation and warned of the risks of pregnancy while taking warfarin. The patient chose to undergo the surgery.

A median sternotomy was performed. Cardiopulmonary bypass was initiated and the patient was cooled to 32°C. The aortic valve was replaced with a 20 mm ATS-advanced performance valve and the mitral valve with a 29 mm St Jude valve. For the tricuspid valve, an anterior leaflet extension with pericardial patch was made and an annuloplasty performed.

OUTCOME AND FOLLOW-UP

There were no significant postoperative complications and on day 8, the patient was discharged, reporting only slight breathlessness on climbing stairs. Four months later, the patient remains well, with significantly reduced breathlessness.

DISCUSSION

This case was unusual in that significant trivalvular disease, involving aortic, mitral and tricuspid valves is rarely seen in the developed world.³

The decisions for the patient, relating to surgery, are complex. Having a single-valve replacement carries a mortality rate of less than 5%, which can rise to 19% for triple-valve replacement.⁴ The patient was counselled that her risk was around this 20% level. However, a recent study describes an operative mortality in some centres of 12%,⁵ and

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Figure 1 Parasternal short axis view, showing the trileaflet, but calcified aortic valve. RVOT, right ventricular outflow tract; RA, right atrium; LA, left atrium; (A) right coronary cusp, aortic valve; (B) left coronary cusp, aortic valve; (C) non-coronary cusp, aortic valve; arrow, aortic valve, calcified.

when her age and ejection fraction of 55% are taken into account, in an individualised risk calculator such as Euroscore II, an even lower figure of around 4% is suggested.⁶

The choice of prosthesis is also difficult, particularly, as in this case, in a woman of childbearing age. Implantation of a bioprosthetic mitral valve carries the strong possibility of the valve degenerating in the patient's lifetime. It is controversial whether pregnancy itself may expedite the degenerative process.⁷ However, the use of a mechanical valve will mean treatment

with teratogenic warfarin, all the more necessary in our patient who had concurrent atrial fibrillation. Research has recommended the use of bioprosthetic valves in Indigenous Australian and New Zealand populations because of poor anticoagulation control, thought to be the result of population transience and lack of access to pathology services.⁸

These considerations apply in this case, as although the patient hopes to remain working in this country for some time, she may return to Ethiopia in the future. It is possible that

Figure 2 Mitral inflow continuous wave (CW) Doppler profile. This image shows the severity of the mitral stenosis (MS). Maximum and mean pressure gradients across the mitral valve are shown in the top left of the image (MV maxPG and MV meanPG, respectively). A mean pressure gradient of almost 14 mm Hg is recorded (>10 mm Hg indicates severe MS). The five-chamber view of the heart at the top of the image demonstrates an enlarged left atrium (LA) consistent with mitral stenosis.

Figure 3 Continuous wave (CW) Doppler profile demonstrating the severity of the aortic stenosis (below the baseline), recorded from a five-chamber view, shown at the top of the picture. The calculated maximum and mean pressure gradients across the aortic valve (AV maxPG and AV meanPG, respectively) are demonstrated in the top left-hand corner of the image. The increased pressure gradient seen here demonstrates severe aortic stenosis. The arrow shows peak blood velocity through the aorta.

optimal management of her anticoagulation will then become more challenging. Given the challenges for this patient, who was affluent enough to seek cardiology help in Ethiopia and also to come to the UK, we can assume that there are many more patients for whom anticoagulation therapy would present too great a barrier to the use of a mechanical valve.

For many patients in the developing world, the option of surgery may not be available at all, either because of advanced heart failure, or the cost of having to travel to an overseas centre. If recent proposals for levying a National Health Service charge on temporary migrants come to fruition, this option may be pushed even further from the reach of patients like this one.⁹

This necessitates a switch in focus from treatment to less costly preventative strategies. There is some debate about how best prevention can be achieved. RHD occurs as a consequence of autoimmune-mediated valvular damage usually following *Streptococcus pyogenes* pharyngitis.¹

Primary prevention involves antibiotic treatment of streptococcal sore throat, but may be difficult without recourse to laboratory facilities for confirming the causative organism. The aim of secondary prevention is to stop recurrence of acute rheumatic fever by penicillin prophylaxis and is advocated by the WHO, but it does not alter the course of the disease once valvular damage has occurred.¹⁰

Those arguing against secondary prevention say that poor compliance is also an issue: 5 years of benzylpenicillin injections are required following an attack of acute rheumatic fever. Completing this course is rendered challenging by population mobility, understaffing of programmes and remote settings.¹¹ Even this patient, living in the major city of Addis Ababa, followed the regimen for only 3 years.

A proposed third way is to adopt a 'prevention plus' approach, where the 'plus' means that medical interventions

are accompanied by efforts to educate, improve treatment access and actively detect cases. The efficacy of combined strategies consisting of education and primary and secondary prophylaxis has been shown in Costa Rica, Cuba and Santa Lucia, where rheumatic fever has been almost completely eradicated.¹

Learning points

- ▶ Rheumatic heart disease (RHD) is rare in the UK but needs to be considered in patients presenting with breathlessness, heart failure or a valvular murmur, and in particular those from high-risk groups.
- ▶ The lack of definitive treatment for RHD in the developing world may prompt those who can afford it to seek surgery in the developed world.
- ▶ Decisions relating to choice of valve prosthesis may well differ between the UK and the developing world due to issues of cost, lack of surgical facilities or appropriate follow-up, and difficulty maintaining adequate thromboprophylaxis.
- ▶ Primary and secondary prevention of RHD in the developing world are not without their problems; a 'prevention plus' strategy may be the solution.

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Competing interests None.

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