Original Article

Medical Management of Chronic Rheumatic Heart Disease

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Keywords. Rheumatic heart disease; Mitral valve regurgitation; Mitral valve stenosis; Aortic valve regurgitation; Aortic valve stenosis

Introduction

There are many barriers to optimal medical management of chronic rheumatic heart disease in the Indigenous population [1]. In Australia there are major geographical barriers as many Aboriginal and Torres Strait Islander people live in remote areas of the country. In these regions Indigenous people usually must travel significant distances to access major health centres for specialist cardiac services, which are often hospital based. The lack of and, when available, the cost of public transportation become major barriers to accessing these mainstream cardiac services. Another barrier to optimal management is the mobility of the Indigenous population. Many Aboriginal people travel significant distances for extended visits to their relations in other communities and this poses a major difficulty in providing continuity of care. The client's medical record is usually not available in these communities, which may be serviced by quite different Health Services. The lack of continuity of care with the same provider hinders the development of trust in the doctor–client relationships, which is so important in maintaining the continuity and quality of health care.

The cost of medications is an issue for Aboriginal and Torres Strait Islander patients, especially in urban and rural environments. Remote Australian communities usually have access to free medications through the S100 scheme but this is not available in the urban or rural areas. In these situations the cost of the PBS medicines is a significant issue. Medications may not be renewed, because other financial priorities may be more important. Cessation of cardiac medications may lead to exacerbation of symptoms and even admission to hospital, e.g. heart failure.

Specialist cardiology outreach services to rural and remote areas in Australia are very limited due to funding and workforce shortages. There have been some increases in specialist outreach services but given the enormous need and the reluctance of Aboriginal and Torres Strait Islander people to travel to mainstream regional health centres, the availability is grossly inadequate for the need. This makes it more difficult to maintain continuity of care for patients with chronic rheumatic heart disease who live in these areas. Limited specialist outreach services also means that echocardiography is often not available since this procedure requires a skilled sonographer or cardiologist/physician to perform the studies. The ability to perform regular Echocardiographic studies in patients with chronic rheumatic heart disease is essential and the recent availability of portable cardiac ultrasound units has been a major step forward. However, the units are expensive, often more than $100,000 and require skilled operators, who are not always available, either medical or technical.

Problems with medication adherence are not uncommon in Indigenous communities because of socioeconomic and educational disadvantage in addition to language, cultural and geographic barriers. The need for regular monitoring of Warfarin therapy is an example of this challenge. Monitoring anticoagulation in Aboriginal communities can be quite difficult. Again the more recent availability of point of care INR units has made this problem easier to deal with, not requiring specimens to be sent to regional pathology centres for INR measurement. However not all communities have access to point of care machines.

The need for a very high adherence rate to monthly Penicillin prophylaxis cannot be over-emphasised to prevent recurrence of acute rheumatic fever. One of the principal causes of progression of valvular disease is recurrence of rheumatic fever, which may not always be clinically apparent or reported to health care workers. We know that if Penicillin adherence is very high the likelihood of recurrence occurring is very low. However the adherence rate for Penicillin prophylaxis varies greatly across northern Australia and in many regions is unacceptably low. The development and implementation of effective systems of Penicillin prophylaxis is beyond the scope of this review.

Because of the many barriers patients with rheumatic heart disease may present rather late in the course of their disease. Their first presentation may be to the local hospital with acute heart failure due to rheumatic valve disease. These patients do not necessarily have a past history of acute rheumatic fever. Because of the often advanced valve disease in these situations these patients are often less

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1443-9506/04/$36.00
doi:10.1016/j.hlc.2010.04.130

suitable for more conservative therapeutic interventions such as valve repair or balloon mitral valvuloplasty. Therefore the only therapeutic option may be valve replacement with the need for long-term anticoagulation with Warfarin.

Mitral regurgitation

Mitral regurgitation [2,3] is the most common valve lesion in rheumatic heart disease. In one series from the Northern Territory 41% of Aboriginal clients with RHD had pure mitral regurgitation, whereas in those aged under 10 years the prevalence was more than 90%. Mitral regurgitation causes left ventricular volume overload which, if significant, will over time cause impairment of left ventricular systolic contractile performance. It is important to note that left ventricular outflow resistance is reduced in mitral regurgitation by emptying into the low pressure left atrium so the left ventricular function may appear to be normal, even when contractility is intrinsically impaired. This has implications for timing of mitral valve surgery in mitral regurgitation. There is wide variation in the rate of progression of mitral regurgitation but many cases tend to progress, especially if there is a recurrence of acute rheumatic fever.

Patients may remain asymptomatic for many years, even with moderate to severe mitral regurgitation. The initial symptoms are those of exertional dyspnoea and fatigue. The classic murmur of mitral regurgitation is loud pansystolic maximal at the apex. However patients with mild mitral regurgitation may not have an audible murmur. The diagnosis of mitral regurgitation is confirmed by echocardiography, showing most typically overriding or prolapse of the anterior mitral valve leaflet, due to elongation of the chordae and in more severe cases dilation of the posterior mitral annulus. This usually results in an eccentric posteriorly directed jet, of variable severity, depending on the degree of prolapse. There may be some thickening or tenting in the parasternal window of leaflets resulting in a so-called *elbow or dog leg appearance* of the anterior mitral valve leaflet consistent with a degree of coexisting mitral stenosis. Echocardiography is also important in measuring left ventricular chamber size and systolic function. The severity of mitral regurgitation is assessed by estimating the area of the regurgitant jet in the left atrium. In addition pulmonary artery systolic pressure can also be calculated if tricuspid regurgitation is present.

Transoesophageal echocardiography may be used to obtain more detailed assessment of the severity and nature of the rheumatic valve disease. Its greater availability especially in rural and remote regions must be encouraged.

Vasodilator therapy such as dihydropyridines or ACE inhibitors have been suggested as potentially beneficial for the volume overloaded left ventricle by virtue of their afterload reducing action. However, there is very little data available on the efficacy of vasodilator therapy for chronic mitral regurgitation and given the fact that afterload is already reduced because of the low resistance leak into the left atrium, vasodilator therapy is not currently recommended for medical management of mitral regurgitation.

Indications for surgery in mitral regurgitation

Patients should be referred for surgical assessment if they are symptomatic and/or have signs of clinical heart failure. In addition they should also be referred if there left ventricular systolic function is reduced, e.g. ejection fraction <60%, or if left ventricular end systolic diameter is >40 mm on echocardiography in an adult. Critical left ventricular end systolic dimension have not been identified in children. There is controversy about referral for surgery in patients who are asymptomatic or only minimally symptomatic with moderate or moderate to severe mitral regurgitation. In many cases the mitral valve is amenable to repair, particularly in younger patients and this would mean that the indication would not be as stringent as in those whom mitral valve replacement is likely to be required. Another argument in favour of early valve repair is that borderline or normal systolic function may indicate LV dysfunction is present and therefore early repair would be beneficial, provided it can be performed at low risk. On the other hand early reoperation after repair may be required in up to 10% of cases. In addition in some cases it may not be technically possible to repair the valve and hence the patient will receive a mitral valve prosthesis. If patients develop signs of clinical heart failure they should be treated with standard therapy with loop diuretics and ACE inhibitors and/or angiotensin receptor blocker therapy.

Mitral stenosis

Approximately 30% of Aboriginal rheumatic heart disease patients in the Northern Territory, aged 10–19, have mitral stenosis [3]. The mean age of those with mitral stenosis was 33 years, which is older than those with pure mitral regurgitations. Many patients at the time of diagnosis do not recall a history of acute rheumatic fever. Mitral stenosis may progress at a variable rate, even in the absence of documented episodes of acute rheumatic fever. Progressive obstruction of the left ventricular inflow develops, leading to a significant diastolic gradient between the left atrium and the ventricle [4]. This gradient is increased by faster heart rates, for example with exercise or atrial fibrillation with a rapid ventricular rate. Patients usually do not develop symptoms until the mitral valve area has decreased to <2 cm^2. Initially symptoms are exertional dyspnoea and fatigue but these may progress to clinical heart failure over time, particularly if the mitral valve area decreased to <1.5 cm^2.

Physical examination may reveal the classic mid diastolic murmur with pre-systolic accentuation if the patient is in sinus rhythm. However, the murmur can be missed, particularly in obese patients or with less experienced auscultators. A chest X-ray and ECG are usually not helpful in diagnosis. Echocardiography is diagnostic demonstrating thickened and restricted anterior and posterior mitral valve leaflets, tethered together, often with doming motion of the anterior leaflet in the parasternal view. There may be associated mitral regurgitation and annular and valvular calcification. The mitral valve gradient can be quantitatively measured with continuous wave Doppler echo