# Priorities in Cardiac Surgery for Rheumatic Heart Disease

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## ABSTRACT

This review outlines a philosophy of surgical cardiac care for rheumatic heart disease, which has evolved over the past 2 decades, in the young in the Oceania region. Topics covered include the optimal timing of surgery, recommended strategies for mitral and aortic valve disease, and the importance of the team approach to these patients. There is a global priority for more cardiac surgeons to become skilled in repair of the rheumatic mitral valve. Surgeons operating on patients from remote regions with RHD are encouraged to audit outcomes and help these communities develop their health services to optimize continued RHD care.

As has been highlighted in other reports in this issue of *Global Heart,* 80% of the world's countries still have rheumatic fever and its important long-term sequel, rheumatic heart disease (RHD). Those with severe RHD that do not have access to cardiac surgery will die. The mean age of death in some regions of Africa is as young as 25 years [1] and for the indigenous Aborigine population of Australia, 22 years [2].

The particular challenge for a cardiac surgeon dealing with severe RHD is to repair as many valves as possible rather than replace them. The populations that suffer from rheumatic fever, and the affected families within those ethnic populations, have the least resources, lowest levels of education, and worst access to health care. There is good evidence to demonstrate how poorly people in these circumstances do with mechanical valve insertion and anticoagulation [3]. Unfortunately, rheumatic disease damages the valve leaflets and subchordal apparatus, making repair difficult in comparison to other mitral and aortic lesions. The surgeon and the cardiology unit need a team approach to achieve good outcomes. It is not just about what happens in the operating theater, but it also involves good triage, timely intervention, echocardiographic detailed assessment, outreach clinics, nursing input for family education, post-operative case audit, and more.

This review outlines some of the approaches we have used in Starship Hospital Auckland for the New Zealand and Pacific Island pediatric rheumatic population over the past 2 decades to improve the outcomes of these children and teenagers.

## EVIDENCE FOR THE SURVIVAL VALUE OF MITRAL VALVE REPAIR

Antunes [4] in 1990 in South Africa was among the first to publish the dangers of using mechanical or bioprosthetic valves in the mitral position in the rheumatic population, both adult and pediatric. This included both mortality rate per patient-year and the mortality rate of reoperation (Table 1) [4].

In 1999, a similar message was reported from New Zealand in a study of women of child-bearing age requiring valve surgery, the commonest indication being rheumatic. The outcome for those who had mechanical valve replacements was significantly worse than those with repairs or bioprostheses, many of which were aortic homografts (Fig. 1) [5]. Pacific Islanders and the indigenous Māori women with mechanical valves had over 6 to 8 times higher mortality than other ethnic groups did (Table 2).

Our unit has recently reported a survival advantage following mitral valve (MV) repair compared with mitral valve replacement (MVR) for the young with RHD [6]. This retrospective study of 81 patients showed that from the time of patient discharge, the long-term durability of mitral repair was equal to that of MVR. Despite the need for early reoperation in 11% who underwent repair, freedom from reoperation was equal in the 2 groups for the duration of the follow-up. Analysis of those with MVR reveals that 50% of the patients with MVR had a significant hemorrhagic, thrombotic, or embolic event within 11 years, and rates of endocarditis were also high. There was 100% freedom from embolic, thrombotic, or hemorrhagic events in those with MV repairs in this and other studies in the young [7–10].

For adults, despite the absence of a randomized comparison between the results of valve replacement and repair, it is widely accepted that valve repair is the optimal surgical treatment in patients with severe mitral regurgitation (MR) [11–13]. Compared with valve replacement, MV repair has a lower perioperative mortality, better preservation of post-operative left ventricular function, improved survival, and lower long-term morbidity [13]. In adults who are in sinus rhythm, repair avoids the need for anticoagulation and risks of thromboembolism with prosthetic valves, makes pregnancy safe, allows for continued participation in contact sports such as rugby, and avoids the sudden deterioration that can occur with bioprosthetic valves in the mitral position.

These data challenge the assumption that if there is funding for 1 operation only, it is best to replace a valve rather than repair it. This needs to be confirmed with more studies specifically in these populations that are vulnerable to medication shortages, lack of monitoring, endocarditis,

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**TABLE 1.** Incidence of late mortality

| | Total %/ Patient-Year | Valve-Related %/ Patient-Year | Ratio* % |
|---|---|---|---|
| Bioprosthetic MVR | 7.4 | 4.2 | 56.8 |
| Mechanical MVR | 5.7 | 2.5 | 43.1 |
| Mitral valve repair | 2.6 | 1.0 | 38.5 |

MVR, mitral valve replacement.
*Valve-related/total.
Adapted, with permission, Antunes [4].

**TABLE 2.** Relative risk of death in women with bioprosthetic or mechanical valves

| | RR (95% CI) |
|---|---|
| Mechanical valve | 2.17 (0.78–5.88) |
| Age at valve replacement, yrs | 1.00 (1.00–1.01) |
| Number of concurrent valves | 0.86 (0.39–1.92) |
| Years of operation | 1.04 (0.93–1.16) |
| Maori | 8.45 (1.82–39.3) |
| Pacific Islander | 6.54 (1.16–36.7) |
| Pregnancy | 0.38 (0.17–0.84) |
| Valve site* | 0.50 (0.11–2.39) |

*Aortic or tricuspid versus mitral.
Adapted, with permission, from North et al. [5].

and medically unsupervised pregnancies, usually in early adulthood. Following cardiac surgery, the challenge is to achieve good follow-up by optimizing outreach clinics, rheumatic fever registries, and nursing networks and linking with the local staff who deliver secondary prophylaxis.

## MITRAL VALVE REGURGITATION

### Indications for cardiac surgery

There are many published guidelines addressing indications for cardiac surgery for adult patients [11,13,14] that are largely based on symptoms and echocardiographic assessment of left ventricular size and function (Table 3). Cardiologists, physicians, and pediatricians should use these guidelines to refer RHD cases in a timely fashion for surgery whether or not the patient is in a high-income or low-income country setting. Patients with chronic RHD who develop cardiac failure and impaired ventricular function may be too late for effective cardiac surgery due to failure of the myocardium to recover normal function. Physicians working in remote settings historically refer the sickest patients, but it may be appropriate for the cardiac unit to decline such patients when ventricular function is irrevocably impaired. A decision to not offer surgery may be more humane after assessment in the local setting rather than after the patient has traveled to an overseas cardiac unit.

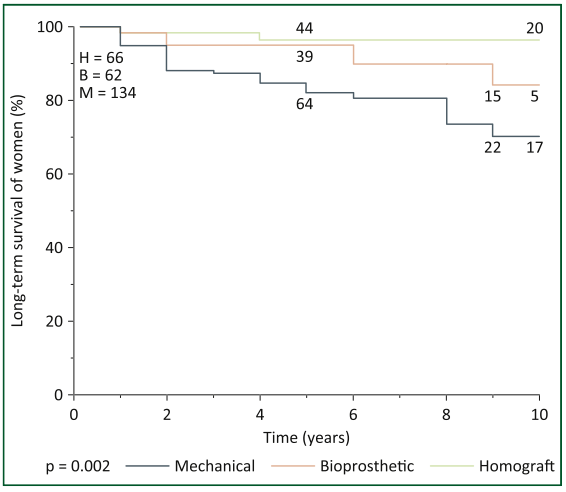


**FIGURE 1. Long-term survival of women according to type of valve replacement**. Among 232 women, 35 had >1 valve state included. Test of difference in survival between 3 valve types: p = 0.002. Test of difference between mechanical and bioprosthetic valve types: p = 0.04. B, bioprosthetic; H, homograft; and M, mechanical. Adapted, with permission, from North et al. [5].

## MITRAL VALVE REPAIR

This complex topic is beyond the scope of this review, but we have included several tables and figures outlining the

**TABLE 3.** Class I indications for mitral valve surgery in adults

| | |
|---|---|
| 1 | MV surgery is recommended for the symptomatic patient with acute severe MR. (Level of Evidence: B) |
| 2 | MV surgery is beneficial for patients with chronic severe MR and NYHA functional class II, III, or IV symptoms in the absence of severe LV dysfunction (severe LV dysfunction is defined as ejection fraction <0.30) and/or end-systolic dimension >55 mm. (Level of Evidence: B) |
| 3 | MV surgery is beneficial for asymptomatic patients with chronic severe MR and mild to moderate LV dysfunction, ejection fraction 0.30 to 0.60, and/or end-systolic dimension ≥40 mm. (Level of Evidence: B) |
| 4 | MV repair is recommended over MV replacement in the majority of patients with severe chronic MR who require surgery, and patients should be referred to surgical centers experienced in MV repair. (Level of Evidence: C) |

LV, left ventricular; MR, mitral regurgitation; MV, mitral valve; NYHA, New York Heart Association.