Early cardiac morbidity of rheumatic fever in children in New Zealand

Olwen Gilbert, Nigel Wilson, Kirsten Finucane

Abstract

Aim The aim of this study was to review the severity and morbidity of acute rheumatic fever (ARF) and rheumatic heart disease (RHD) for children with the most significant cardiac disease in the current era in New Zealand.

Method Retrospective 2-year review of children with ARF and RHD admitted to Starship Children’s cardiology ward. Medical and surgical admissions were classified. Echocardiographic severity of cardiac disease and cardiac surgical data were analyzed. Using length of stay data and 2009 District Health Board costings, admission costs were calculated.

Results 36 children had 49 admissions. Mean age 11.8 ± 2.4 years. All but one child was of Māori or Pacific Island ethnicity. 10 children had symptoms and signs of congestive cardiac failure on admission. The average length of stay was 23 days, but the subset of children with ARF requiring cardiac surgery at the same admission had an average of 54 days (range 36–78 days) in hospital. The total hospital costs over the 2-year period was $1,918,600.

Conclusion Failure to prevent rheumatic fever in New Zealand means that there is significant cardiac sequelae for those children who develop severe RHD. The early morbidity includes heart failure, need for cardiac surgery, and prolonged hospital stay.

Acute rheumatic fever (ARF) continues unabated in New Zealand in Māori and Pacific Island populations.¹ ² The cardiac sequelae of ARF includes rheumatic heart disease (RHD) which can lead to impairment of exercise, need for cardiac operations and premature mortality. There are still over 200 deaths per year attributed to RHD in New Zealand, mainly in adults.³

In New Zealand, children with severe cardiac disease from ARF or RHD that may require operation are referred to the sole cardiac surgical unit at Starship Hospital, Auckland. The aim of this study was to review the severity and morbidity of ARF and RHD for children with the most significant cardiac disease in the current era.

Method

This study was a retrospective chart review of those children with ARF or RHD admitted to the Paediatric and Congenital Cardiology Ward between July 2007 and June 2009 inclusive. Demographic analysis was by patient. Length of stay and cost analysis was by each admission.

Definitions of groups:

- ‘ARF-medical’ was defined as children fulfilling the diagnosis of ARF by the NZ rheumatic fever guidelines criteria 2 which have greater sensitivity for ARF than the Jones criteria.⁴ These patients had carditis as the major criterion of ARF, raised inflammatory markers and elevated streptococcal titres.
‘ARF-surgical’ was defined as those with ARF who proceeded to cardiac surgery during the same admission.

‘RHD-surgical’ was defined as those who had elective cardiac valve surgery beyond the acute phase of ARF.

‘RHD-medical’ was defined as any patient who was admitted with known RHD but not for the purpose of cardiac surgery.

Echocardiographic results analyzed were severity of valve regurgitation and assessment of ventricular function. Cardiac dilatation was assessed by relating the left ventricular end systolic and diastolic diameters and volumes to body surface area, expressed as a Z score, which represents the number of standard deviations from the normal population mean value. Left ventricular ejection fraction less than 55% was regarded as abnormal.

Admission costs were calculated by using the 2009 Auckland District Health Board (ADHB) costing, used for inter-DHB charging. (S. Adams, personal communication) Cardiac ward cost is $1200/night, paediatric intensive care unit $4700/night, theatre costs $2700/hour. The average time for single valve replacement was assigned 4 hours, aortic root replacement 5 hours, single valve repair 5 hours, double valve repair 5 hours and triple valve repair 6 hours. The cost of valve replacement was $3600 per prosthetic valve, $2000 per homograft and $6000 per porcine valve. Analysis of the continuous variables hospital costs and length of stay was expressed as mean and standard deviation.

The study received ethical approval from the New Zealand Northern Y Regional Ethics committee. Patients referred from the Pacific Islands for RHD cardiac surgery were excluded from the analysis.

Results

Patient demographics—Over the 2-year period 36 children had a total of 49 admissions. Twenty-four children had a single admission, 11 children had a second admission, and one child had three admissions in the time period studied. There were 21 male and 15 female patients.

Recorded ethnicity: Māori (13), Samoan (11), Cook Island Maori (7), Tongan (2), other Pacific not specified (1), Niuean (1) and Indian (1). All but one child were domiciled in the North Island. 22 of the 49 admissions were from the Auckland region with most of the disease burden in South Auckland (Figure 1).

Diagnostic Admission Groups—There were 23 admissions with ARF of which 16 were ARF-medical and seven ARF-surgical. There were 26 admissions with RHD, 18 were RHD-surgical and eight RHD-medical. Reasons for admission for the RHD-medical group were readmission after cardiac surgery with wound infection (n=2) or pleurisy (1). Two children with RHD and sepsis were admitted to exclude bacterial endocarditis. One had a final diagnosis of Salmonella typhi septicemia and the second had reactivation of Hepatitis B. One child was admitted for non-cardiac surgery, two were admitted for diagnostic work up of valve disease aetiology or RHD disease severity requiring transoesophageal echocardiography under anaesthetic.
The mean age of admission was 11.8±2.4 years with an age range 5 to 15 years. The mean body mass index was 25.4±6.1 with a range 12.4–38.5.

**Cardiac disease severity**—Ten children had symptoms and signs of congestive cardiac failure on admission (Six of the 16 children in the ARF-medical group had cardiac failure on admission but this was controlled without the need for early cardiac surgery).

Four of the seven children in the ARF–surgical group had fulminant ARF requiring early cardiac surgery during their initial presentation. One of these was the youngest child of 5.4 years who was cachectic with a BMI of 12.4. Her heart failure symptoms were recent, but judging by the degree of cachexia and chronic rheumatic changes of the valve by echocardiography, the length of illness was likely several months duration. Two of the 18 in the RHD-surgical group had signs of cardiac failure. No child showed mitral stenosis.

One child was admitted with ARF had complete heart block which resolved spontaneously. She was discharged with mild residual pathological mitral regurgitation.

Left ventricular size and function was recorded on 46 admissions. The left ventricular size related to body surface area was significantly increased for the cohort: left ventricular end diastolic dimension Z score+4.8±2.8, left ventricular end systolic dimension Z score+3.6±3. Left ventricular function was depressed in 10 patients and normal in 36. The type of cardiac valve with regurgitation is shown in Figure 2.
Cardiac surgery—Twenty-five patients underwent cardiac surgery, mean age 11.6 ± 2.6 years. Ten had single valve surgery, 14 had double valve surgery and one had triple valve surgery. Of those who underwent mitral valve surgery, 17 had a mitral valve repair and two received a mitral valve replacement as the valve changes were too severe to achieve a repair. Four of 15 patients who underwent aortic valve surgery had a repair and 11 had aortic valve replacement, usually an aortic homograft for which warfarin anticoagulation is not required.

Complications in hospital—There was no mortality for this cohort during the study period. One child developed acute cerebral cortical ischaemia perioperatively leading to hemiplegia and seizures following cardiac surgery. One child developed Stevens Johnson Syndrome as an allergic response to penicillin. Erythromycin was started for ongoing antibiotic prophylaxis to prevent ARF recurrence. Other complications included Klebsiella pneumonia (n =1), children who had transient heart block (2), pericarditis (1) and epistaxis (3).

Complications for those undergoing cardiac surgery included left lower lobe collapse (n=4), arrhythmia requiring either pacing or anti-arrhythmic medication (4) post operative pulmonary oedema (3), and pericardial effusion (4). All of these had resolved by discharge.

All children continued to receive 28 day intramuscular benzathine penicillin as secondary prophylaxis to prevent recurrences of ARF except the one child who was continued on erythromycin prophylaxis.

The mean length of stay for all admissions was 22.7±20.3 days with a range of 2–78 days. Length of stay by admission type is shown in Figure 3. The ARF-surgical group had the longest length of stay with a mean of 54 days±16, range 36-78 days. The shortest stay group was RHD medical with a mean of 5.4 days.
Figure 3. Length of stay for each admission group.

![Mean Length of Stay diagram]

Figure 4. Cost of admission for each patient group

![Approximate Cost of Admission diagram]
Six children with ARF who did not require early operation were transferred back to their local paediatrician for further inpatient management. Four of the children with RHD who had surgery were transferred back to their own hospital for management of moderate pericardial effusions. The two children who had wound infections were transferred back for ongoing antibiotic treatment.

Cost of admissions—The average costs of admissions by group are shown in Figure 4:

The most expensive category was ARF-Surgical patient group, with an average cost of $90,157±$6,388, range $66,500-113,300. The total hospital costs over the 2 year period was $1,918,600 or nearly $1,000,000 per year.

Discussion

This study has examined the subset of children with ARF or RHD who had the most significant cardiac sequelae of the disease. It reveals considerable early morbidity with heart failure, need for cardiac surgery, and prolonged length of hospital stay. Rheumatic fever continues to affect almost exclusively Māori and Pacific populations in New Zealand, confirmed in this study.

There was a need for cardiac surgery in this group of children due to cardiac symptoms or cardiac dilatation that resulted from cardiac valve regurgitation, with the risk of long term myocardial damage and premature mortality. Mitral valve repair is preferred to valve replacement in children as the morbidity of prosthetic valves and need for warfarin anticoagulation is significant. There has been extensive use of aortic valve homografts for aortic valve replacement in children, as these do not require warfarin. Tricuspid valve surgery can usually be achieved by an annuloplasty ring and valve repair, again without the need for warfarin.

Prolonged hospitalisation is part of the morbidity of ARF. Traditionally, those with ARF were managed with strict bed rest, due to the known ongoing inflammation which can last months, judged by acute inflammatory markers, active inflammation found at the time of cardiac surgery or at post-mortem.

At KidsFirst Hospital, South Auckland, Nicholson and colleagues showed that those with ARF without carditis could be safely mobilized within 2 weeks and this has lead to earlier mobilization of ARF patients nationally. However, currently at KidzFirst hospital, the average length of stay for those with severe valve regurgitation is 45 days (R Nicholson, personal communication) similar to the average 29 days in the current study when allowance is made that some children returned for inpatient care to their referral hospital.

The calculated costs were direct in-patient care costs, not the total medical costs of this disease. North and colleagues calculated that costs for rheumatic fever in Auckland in 1992 to be $3.6m per year. As Auckland has approximately half the disease burden of ARF and RHD in New Zealand, a simple doubling of these costs, with an inflation factor allowance to 2010, gives current national medical costs to be $9.5–10.5m per year.
There is considerable recent endeavour to reduce the disease burden of ARF and RHD, outlined in New Zealand guidelines\(^1,\!^2,\!^10,\!^11\) with renewed leadership by the New Zealand Rheumatic Fever Steering group. The aetiology of ARF in New Zealand shows striking ethnic differences, which reflects health and social inequality.\(^12\) Primordial and primary prevention is possible.\(^13,\!^14\)

The development of severe RHD may be reduced by detecting mild disease by echocardiography, then treating with penicillin.\(^15,\!^16\) Until all these measures take effect, there will continue to be children and young adults with severe cardiac consequences of ARF and RHD in New Zealand.

**Competing interests:** None.

**Author information:** Olwen Gilbert, Paediatric Registrar; Nigel J Wilson, Paediatric Cardiologist; Kirsten Finucane, Cardiothoracic Surgeon; Green Lane Paediatric and Congenital Cardiac Services, Starship Children's Hospital, Auckland

**Correspondence:** Nigel Wilson, Paediatric Cardiologist, Green Lane Paediatric and Congenital Cardiac Services, Starship Children's Hospital, Private Bag 92024, Auckland 1142, New Zealand. Fax: +64 (0)9 3757026; email: nigelw@adhb.govt.nz

**References:**


