THE NEW ZEALAND MEDICAL JOURNAL
Journal of the New Zealand Medical Association

Undetected rheumatic heart disease revealed using portable echocardiography in a population of school students in Tairawhiti, New Zealand

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Abstract

Aim The aim of this programme was to find undetected rheumatic heart disease (RHD) in students from selected schools in the Tairawhiti region (eastern part of the North Island) of New Zealand.

Method Portable echocardiography was used to scan students in 5 urban and rural schools in Tairawhiti where the population is predominantly Māori. The age range of students in the urban schools was 10–13 years and in the rural schools 5–17 years. Those with abnormal echocardiograms were referred for a paediatric consultation, with hospital-based echocardiography if required for the clarification of diagnoses and further management.

Results A total of 685 students, representing over 95% of the schools' students, consented to having echocardiographic scanning. After repeat hospital based echocardiography for 11 students, a total of 52 scans were regarded as abnormal. In this population definite (n=4) or probable (n=7) RHD was found in 11 students a prevalence of 1.61% (95%CIs 0.80–2.85). Possible RHD was found in 19 students. Previously undetected confirmed (n=1) or probable (n=7) RHD was found in 8 students a prevalence of 1.17% (95%CIs 0.51–2.29). Congenital heart defects (CHD) were found in 22 students a prevalence of 3.21% (95%CIs 2.02–4.83).

Conclusion Echocardiography was a popular modality and detected a significant burden of previously unknown RHD in this young Māori population who are now receiving penicillin. However, echocardiography detected a greater prevalence of possible RHD for which optimum management is at present uncertain. Echocardiography also detected students with a range of severity of CHD. Screening with echocardiography for RHD would involve a significant use of public health, paediatric and cardiac resources with 7.6% of students and their families requiring clinical consultations and ongoing management of the abnormal echocardiographic results.

Many children affected by rheumatic fever have no history of the classical symptoms of arthritis but have episodes of undetected inflammation of the heart leading to permanent rheumatic heart disease (RHD).[1,2] Untreated RHD leads to progressive cardiac valve changes, heart failure in early adulthood and decreased life expectancy.[1,2]

Many adults present in their 3rd or 4th decade with heart failure due to RHD, many with no recorded history or memory of having had ARF.[3] There are 150–180 deaths annually from RHD in New Zealand.[2] However, if RHD is detected early, progression

to serious RHD related morbidity can be prevented by penicillin.[4] A recent innovative approach to allow rapid, detection of RHD for childhood populations is this use of portable ultrasound (echo) screening.[5,6]

The first New Zealand study using portable echocardiography was undertaken in South Auckland in 2007–08.[7] This study revealed a prevalence of 2.4% RHD in a population of 1174 10–13 year old children mainly of Pacific ethnicity from education decile 1 and 2 schools.

An opportunity arose in Tairawhiti District Health to undertake a programme of echocardiographic scanning for undetected RHD for a limited period of two weeks in schools with high numbers of children already known to have had had episodes of ARF. The programme was entitled *Let's get rid of rumatiki in Tairawhiti* and was based on the evidence from the South Auckland Study. [7]

Methods

The echocardiography or scanning programme was planned and coordinated by the Public Health Service of Tairawhiti District Health Board and the Paediatric Department in Gisborne Hospital in conjunction with the cardiology department of Starship Children's Hospital in Auckland.

The programme of scanning was undertaken over a two week period in March 2009. Consultations took place with schools and Māori health providers. As the project proposal was based on the South Auckland protocols, which had received ethics approval the Northern Y Regional Ethics Committee stated that no separate ethics committee review was required for this project. Of the childhood population just under 8% were screened.

Due to time and resource constraints the programme could not be offered to all the intermediate schools in Gisborne City. For the first week of the programme of the three intermediate schools (age range from 10–13 years) the school that was situated in an area known to have a high number of children who have had ARF was selected.

For the second week, the decision was made to scan a predominately Māori coastal community also with a high number of children who have had ARF. The four schools in this township community comprised two full primary schools for students aged 5–13 years and 2 composite schools for students aged 5–18 years.

The project team liaised with school staff and families and made educational presentations to the school children in their classrooms. A locally devised and detailed information sheet and consent form were sent home from school, outlining information about ARF and RHD, the scanning procedure, consent for the procedure and allowing parents/caregivers the option to be present for the procedure. Public Health and Rural Health nurses assisted with this process by conducting home visits and telephoning parents if consent forms were not returned.

School scanning procedures—The programme was based on echocardiography alone as it had been shown that auscultation was not sensitive in the South Auckland study[7] or internationally.[5] Vivid™e (GE Healthcare) portable cardiac ultrasound machines were used. Gain settings were optimised by the sonographers as the large variation in body habitus precluded the use of standardised settings. The echocardiography protocol consisted of 2-dimensional and colour Doppler images in parasternal and apical four-chamber views, with careful attention to record colour sweeps across any mitral or aortic regurgitant jets.

Pulse-wave Doppler interrogation of regurgitant jets was undertaken. Valve leaflet morphology was assessed in parasternal long axis (PSLA) (mitral valve and aortic valve) and parasternal short axis (PSSA) views (aortic valve). A variable range 2.5–5 MmHz probe was used for all studies. When indicated, echocardiographic measurements were interpreted according to body surface area. Considerable effort was undertaken to preserve modesty especially for adolescent girls. Private areas were available to change into gowns. All scanning was performed by female echocardiographers and the students were behind screens lying down on plinths. The echocardiograms were reported by the Starship Paediatric Cardiology Department. Criteria for the diagnosis of RHD is shown in Appendix 1, this process was the same as used in the South Auckland study.[7]

The parents or guardians of all the students were sent a letter after the scanning. If the results were normal this was stated on the letter, if the results were abnormal the letter stated "Your child … will be seen with you by the children's doctors from Gisborne to discuss this result and to see if there is a need for any further tests or treatment". For each child a letter was also sent to the family doctor stating either that there was a normal result or an abnormal that required further paediatric assessment.

The programme took place over a 2-week period with a week in the urban school and a further week in the rural schools.

Any scans that were abnormal were reviewed by two paediatric cardiologists with a third opinion available and the classification made available to the project paediatrician.

Paediatric consultation and assessment—Clinical consultation allowed a full history, in particular any history of episodes of arthralgia or arthritis or other illness that could have been an episode of ARF. Examination was used to exclude abnormal body habitus or mitral valve clicks which would indicate that mitral regurgitation was congenital rather than rheumatic in aetiology. The presence of a diagnostic murmur enabled a clinical case definition of definite RHD rather than probable RHD. Repeat echocardiography was only utilised where there was diagnostic doubt about the echocardiogram.

Students with definite or probable RHD were recommended to commence long-acting intramuscular benzathine penicillin G (BPG) prophylaxis whereas observation with repeat consultation and echocardiography was recommended for those with possible RHD.

Results

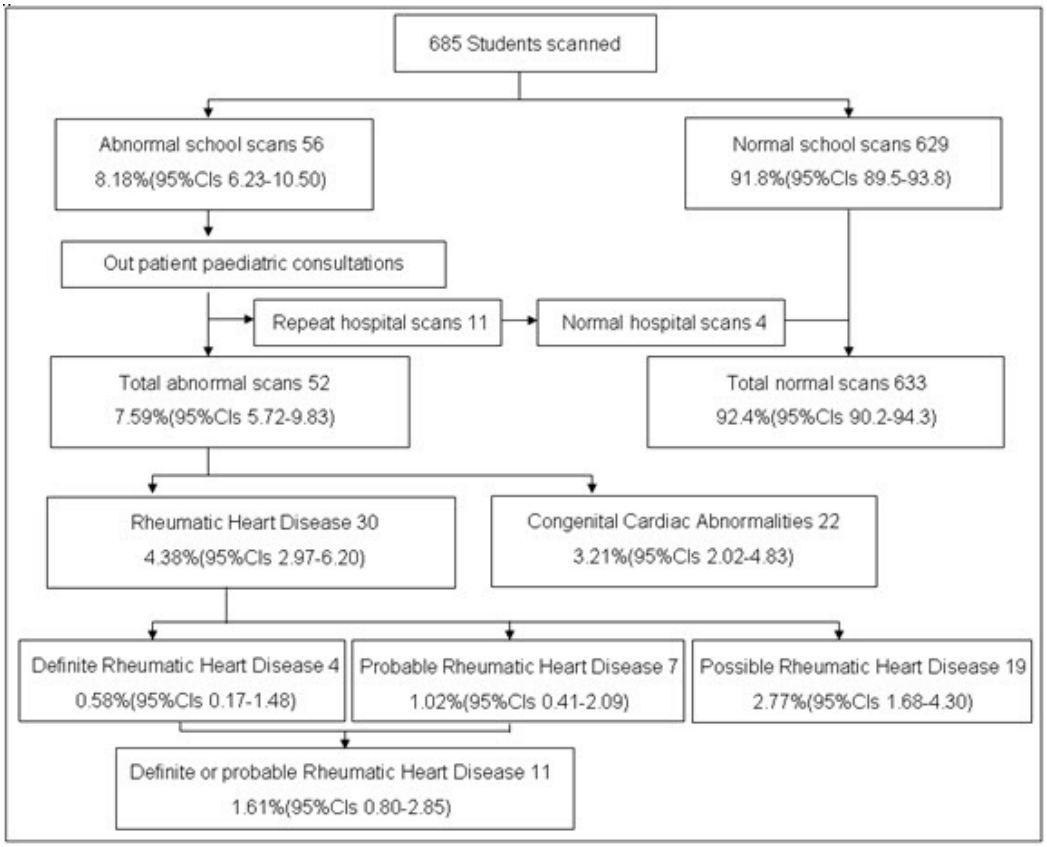
For the urban school the return rate of forms was 92% (437/474), with 96% consenting for echocardiography an overall consent rate of 88%. In the rural region the consent rate was 99% (366/369) with all those consenting wishing to be scanned.

Over the 2 weeks of the programme 362 echocardiograms were performed in the urban school and 323 in the rural locations, giving a total of 685 scans. The two echocardiographers performed on average 37 scans each per day, equating to one scan every 8 minutes.

Of the 685 echocardiograms performed 629 (91.8%) were classified as normal and 56 (8.2%) echocardiograms were classified as abnormal. Of the students with abnormal echocardiograms 11 (1.6% of the total performed) also underwent repeat hospital based echocardiography as the portable echocardiogram was not of sufficient quality to be diagnostic.

Students with abnormal scans were subsequently seen for a full consultant paediatric clinical assessment based on the echocardiographic findings. One child with probable RHD remains lost to follow up. After consultation by the paediatrician and repeat echocardiography a total of 52 students were found to have a cardiac abnormality, 30 due to RHD and 22 due to congenital heart disease (CHD). (Figure 1)

Figure 1. Flow diagram showing the number of students scanned, the numbers and percentages with 95% confidence intervals (CIs) of normal and abnormal scans found at the schools and at subsequent repeat hospital scans and for the final diagnostic classification after clinical consultation



Of the 30 students with rheumatic valvular changes 11 had probable (n=7) or definite (n=4) RHD requiring prophylactic penicillin. Three of these 11 children were already known to have confirmed RHD on previous echocardiography and were under follow up and on BPG. Thus, the programme identified 8 students with previously undetected RHD, with a prevalence in this population sub group of 1.17% (95%CIs 0.51–2.29).

Only one of these had a typical murmur of mitral regurgitation Overall, the RHD prevalence for the cohort was 1.6% (95%CIs 0.80–2.85). There was no statistical difference between the prevalence of RHD in the urban compared to the rural regions.

Nineteen children had valvular changes consistent with possible RHD representing 2.77% (95%CIs 1.68–4.30). Of these the cardiac lesions observed were Mitral regurgitation (n= 11), isolated aortic regurgitation (n=5) and structural RHD without significant regurgitation (n=3).

These families have been counselled about the importance of sore throats, and their general practitioner advised about the importance of acute Group A streptococcal

(GAS) sore throat management. Repeat echocardiography is also recommended after 2 years to ascertain if there is progression to definite RHD and this work is currently being undertaken.

Twenty-two children had a CHD (Table 1) ranging from minor lesions such as bicuspid aortic valve to a child with a symptomatic large atrial septal defect who underwent semi-urgent cardiac surgery. The six children diagnosed as congenital mitral valve prolapse did not have a mitral valve click on auscultation but three of them were tall with thin body habitus. Overall the prevalence of CHD was 3.21% (95%CIs 2.20–4.83).

Four thought to have abnormal echocardiograms were classified as normal after paediatric consultation and repeat echocardiography. One child had difficult echocardiographic windows due to pectus excavatum.

Table 1 Classification and number of congenital heart defects (N=22) discovered during the programme

Congenital heart defects	n
Congenital mitral valve prolapse	6
Bicuspid aortic valve ± aortic regurgitation or aortic annular dilatation	8
Atrial septal defect	1
Patent ductus arteriosus	4
Pulmonary stenosis	2
Tricuspid regurgitation	1

Discussion

We found 11 students with definite or probable RHD out of 685, a prevalence of 1.6% (95%CIs 0.80–2.85) in 5 urban and rural schools in the Tairawhiti region. Of these 11 students with RHD 8 had previously undetected RHD with a prevalence of 1.17% (95%CIs 0.51–2.29).

The prevalence of RHD was lower than the 2.4% detected in South Auckland region but statistically the difference is insignificant.[8] In Tairawhiti the population is mainly Māori whereas in South Auckland the population is mainly of Pacific Island ethnicity with known higher rates of ARF than Māori.[9]

The RHD prevalence in this population was also lower than some international reports[5,6,11] but direct comparisons are problematic due to the variations in case definitions used for rheumatic heart disease.

There was a high consent rate in all schools reflecting the high level of awareness of the consequences of ARF and well known cases of early death from RHD experienced in these close communities. In the rural region, all families were known to the Rural Health nurse who empowered the families to participate. Offsetting this was that consented students only underwent scanning if they were at school on the days when echocardiography team were present.

There was a lower rate of need for repeat hospital based echocardiography in this propgramme compared to that in the South Auckland study (1.6% vs 8%) reflecting

the accuracy of the portable echocardiography based on the experience of the South Auckland study.[7]

Echocardiography not only detects RHD but also detects CHD and in this programme 22 students were found with CHD which is 3.21% (95%CIs 2.02-4.83). Some of the students with CHD required medical and even surgical intervention but many did not. Thus, considerable paediatric outpatient workload is created for those with abnormal echocardiography results, with the need for clinical assessment of the child and counselling of the families. Should echocardiographic screening be more widely implemented in New Zealand, an increase in paediatric clinical workload will occur.

The natural history of possible RHD (usually mild mitral regurgitation) without morphological changes of RHD is unknown, particularly what percentage of possible RHD on echocardiogram progresses to definite RHD.

In New Zealand we are currently recommending follow up in the first instance to see if there is any progression of RHD changes. The Australian gECHO programme is following up those with possible RHD as a case control study.[10] A recent study in Nicaragua suggests that half of those defined as possible RHD may progress to definite RHD by echocardiography a decade later.[11]

Since this programme, a decile 10 school in New Zealand in a region without any cases of ARF was studied. No cases of definite or probable RHD were found in 397 children but 2 had mild pathological mitral regurgitation meeting the criteria of possible RHD.[8]

The use of screening for RHD using echocardiography challenges the traditional model of the epidemiology of RHD where symptomatic ARF is the precursor of RHD.

There are many clinical scenarios where RHD presents without a history of ARF: Indolent carditis and Sydenham's chorea in childhood[12] and in adulthood symptomatic mitral stenosis, heart failure with mitral or aortic regurgitation, arrhythmia with underlying RHD, endocarditis and strokes with underlying RHD.[2] Thus, GAS infection causing asymptomatic Recurrent Rheumatic Fever and RHD may be as common as episodes of clinical ARF.

The considerable academic and clinical endeavour to combat ARF and RHD in New Zealand over the past three to four decades has recently been summarised.[2] Despite these endeavours, the incidence of ARF and prevalence of RHD has not fallen since the 1980s. Indeed, ethnic disparities are actually widening,[9] and Māori and Pacific rates may be increasing.

Primary prevention of GAS pharyngitis is very important to reduce cases of ARF.[12] For many reasons this has not altered ARF rates in New Zealand. Children with a GAS sore throat may have difficulty accessing medical care, antibiotics may not be prescribed or a full course may not be taken. Importantly, ARF occurs without a history of sore throat in over a third of cases.[13]

There has been a renewed effort to reduce ARF rates through the production of New Zealand guidelines to raise awareness of ARF and for its appropriate management.[12,13] There are also recent innovations using GAS sore throat school clinics in the primary prevention of ARF[14] which can be effective when combined with community and

health professional awareness of the importance and correct treatment of GAS pharyngitis.[15–17]

Effective secondary prevention has been the mainstay of ARF control in New Zealand with well run ARF registers,[13,18,19] well audited in the Auckland region[19,20] which has 60% of the disease burden of ARF in New Zealand.[13]

It is known that regular intramuscular long acting benzathine penicillin (BPG) reduces the recurrence of ARF by 87–97%.[21] In Auckland ARF recurrences fell from 20% in 1980 to 5% in the period 1993–9.[20] Secondary prevention is currently considered to be the most cost effective clinical intervention for the control of RHD.[22]

The place of echocardiography screening in RHD disease control is still to be established in New Zealand and globally. Portable echocardiography as a utility was demonstrated to be both fast (an average of an echo every 8 minutes) and also acceptable with 97% of students willing to have this form of test. This programme demonstrated that previously undetected RHD and clinically significant CHD can be detected using methodology that would be similar to a screening programme.

Echocardiographic screening is likely to be complementary to the primary prevention of ARF and secondary prevention of RHD. There is little doubt that it is better to diagnose early RHD changes so that BPG can be commenced to reduce the progression of the RHD and the burden of heart failure and valve replacement for individuals and the health service. We found the echocardiographic scanning was very acceptable to the students and parents with consent rates of over 95%.

Limitations of the programme described include not meeting all the requirements of an ideal screening test.[24] In particular the natural history of subclinical RHD detected by echocardiography is unknown and required. Treatment thresholds are still based on best clinical practice for RHD following an episode of ARF.

There are no randomised controlled studies that prove that echocardiography will result in reduced mortality and morbidity for RHD. Cost-effectiveness was not addressed in this study.

Competing interests: None known.

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Acknowledgements: We thank the principal, staff and students of Ilminster Intermediate School, Gisborne, and the principals, staff and students of Hiruharama Primary School, Te Kura o Makarika, Te Kura Kaupapa Māori O Te Waiu O Ngati Porou, and Ngata Memorial College, Ruatoria for their willing participation and support in the programme.

In addition, we thank Ghillian Whalley, Jill Eastham, and Vicky D'Arth for performing the echocardiograms; Tom Gentles, John Stirling, and Clare O'Donnell (paediatric cardiologists) for reading abnormal echocardiograms, and Shaun Grant for performing follow up echocardiograms at Gisborne Hospital.

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