

This is the **Submitted Version** of a paper published in the journal *Heart, Lung and Circulation*:

Rémond, M.G.W., Wheaton, G.R., Walsh, W.F., Prior, D.L., and Maguire, G.P.
(2012) *Acute rheumatic fever and rheumatic heart disease — priorities in prevention, diagnosis and management. A report of the CSANZ Indigenous Cardiovascular Health Conference, Alice Springs, 2011.* *Heart, Lung and Circulation*, 21 (10). pp. 632-638.

<http://dx.doi.org/10.1016/j.hlc.2012.05.006>

Title Page

Title: Acute Rheumatic Fever and Rheumatic Heart Disease - *Priorities in Prevention, Diagnosis and Management. A report of the CSANZ Indigenous Cardiovascular Health Conference, Alice Springs 2011*

Short Title: Priorities in ARF/RHD

Authors:

Rémond MGW¹

Wheaton GR²

Walsh WF³

Prior DL⁴

Maguire GP¹

Institutions and Affiliations:

¹Cairns Clinical School, School of Medicine and Dentistry, James Cook University, Cairns, Australia

²Cardiology Department, Women's and Children's Hospital, Adelaide, Australia, Australia

³Department of Cardiology, The Prince of Wales Hospital, Sydney, Australia

⁴Department of Medicine, St. Vincent's Hospital, The University of Melbourne, Melbourne, Australia

Corresponding Author:

Marc Rémond

School of Medicine and Dentistry

James Cook University,

Cairns Base Hospital, The Esplanade, PO Box 902, Cairns QLD 4870 AUSTRALIA

Fax: 07 4226 6831

Phone: 02 9976 5230

Email: marc.remond@my.jcu.edu.au

Abstract

Three priority areas in the prevention, diagnosis and management of acute rheumatic fever (ARF) and rheumatic heart disease (RHD) were identified and discussed in detail:

1. *Echocardiography and screening/diagnosis of RHD* - Given the existing uncertainty it remains premature to advocate for or to incorporate echocardiographic screening for RHD into Australian clinical practice. Further research is currently being undertaken to evaluate the potential for echocardiography screening.
2. *Secondary prophylaxis* – Secondary prophylaxis (long acting benzathine penicillin injections) must be seen as a priority. Systems-based approaches are necessary with a focus on the development and evaluation of primary health care-based or led strategies incorporating effective health information management systems. Better/novel systems of delivery of prophylactic medications should be investigated.
3. *Management of advanced RHD* - National centres of excellence for the diagnosis, assessment and surgical management of RHD are required. Early referral for surgical input is necessary with multidisciplinary care and team-based decision making that includes patient, family, local health providers. There is a need for a national RHD surgical register and research strategy for the assessment, intervention and long-term outcome of surgery and other interventions for RHD.

Keywords:

Rheumatic fever

Rheumatic heart disease

Australia

Indigenous population

Prevention and control

Text

Introduction

Any discussion of the prevention, diagnosis and management of acute rheumatic fever (ARF) and rheumatic heart disease (RHD) will highlight the complexity inherent in providing an effective response to a condition that extends from acute through to chronic disease. Given the underlying association between ARF/RHD and socioeconomic disadvantage [1-3] such discussion must, by extension, involve multiple dimensions across all levels of health care and society more generally.

In Australia, these diseases are almost exclusively borne by Aboriginal and Torres Strait Islander peoples, particularly those living in remote locations.[4-7] Geographical isolation and socioeconomic disadvantage, along with the need to provide long-term monitoring and care for those living with ARF/RHD, pose a number of major challenges to many patients, families, communities and health services. Delivery of ARF/RHD care in this setting is often less than optimal. Within this context, the aim of this workshop, undertaken as part of the CSANZ Indigenous Cardiovascular Health Conference in Alice Springs in 2011, was to identify priorities and provide guidance to inform the future response to the prevention, diagnosis and management of ARF and RHD in Australia and Oceania.

Ten priority areas were identified through working with health service organisations and health care providers both before and during the workshop (see Box 1). Whilst time constraints meant only three were discussed in detail, they all provide a valuable insight into how stakeholders in health care can inform the future response to prevention and disease management. The discussions involved over fifty stakeholders in Australian and New Zealand health care who outlined the current understanding of these issues, identified gaps in knowledge and current practice, and provided recommendations and guidance to CSANZ and Australian jurisdictions regarding how these gaps may be addressed to improve outcomes for people living with ARF and RHD in our region. The overview of these discussions detailed below provides a valuable local and clinical perspective on ARF/RHD prevention and management that will be important in informing the future Australian response to these conditions.

1. Is there a role for echocardiography in the screening of high-risk populations and better diagnosis of RHD?
2. Coordinating long-term care for people with RHD.
3. Better and more appropriate management of advanced RHD.
4. An appropriate and sustainable workforce.
5. Getting secondary prophylaxis to work.
6. Health determinants and the primordial prevention of ARF/RHD.
7. Primary prevention and Group A Streptococcus
8. Health promotion –communicating to patients, families, communities and health care.
9. Getting ARF/RHD on the national health agenda - why did it take so long to be recognized as a priority and how can we ensure that it remains on the national health agenda?
10. What are the systems issues that fail people living with ARF/RHD such that they do not receive the best practice care that they need?

Box 1 - Priorities for addressing ARF/RHD in Australia

Workshop Discussions and Recommendations

The three priority areas addressed in detail were:

1. Is there a role for echocardiography in screening and better diagnosis?
2. Getting secondary prophylaxis to work.
3. Better and more appropriate management of advanced RHD.

For each of these issues a brief background was provided, gaps in current systems identified and potential solutions for addressing these gaps highlighted.

1. Is there a role for echocardiography in screening and better diagnosis?

Background

Echocardiography is a crucial tool in diagnosing and assessing the severity of RHD.[8,9] With the availability of portable and relatively affordable echocardiography machines it is now possible to provide this to small and very remote communities as part of specialist outreach services. Nonetheless, there is ongoing debate regarding the details of valvular

morphologic change and the degree of functional impairment (regurgitation or stenosis) that are necessary to make a definitive diagnosis of RHD.[10] In particular, the question of whether potentially minor abnormalities of heart valve appearance or function represent the earliest signs of RHD remains unclear. Given this limitation the possible role of echocardiography in screening for early RHD cannot yet be fully addressed.



Figure 1 – Screening echocardiography – portable and non-invasive but is it effective?

Gaps

In discussing the use of echocardiography in screening for, and the better diagnosis of, RHD a number of gaps in knowledge were identified including:

- Based on existing uncertainty regarding **interpretation**, what should be done when echocardiography reveals minor changes in valve morphology? What are appropriate clinical algorithms for management of such minor abnormalities?
- **Valve (and particularly mitral valve) thickness** as a morphologic feature of RHD – measures of valve thickness are dependent on machine settings (gain, focus, transducer frequency) which are difficult to standardise. It seems unlikely that it will be possible to identify early disease through an objective echocardiographic measure of leaflet thickness.
- **Are lower cost and more portable echocardiography machines comparable to those that are more expensive in the diagnosis and assessment of RHD?** Anecdotal

evidence suggests lower cost portable machines may overstate the severity of valve lesions, especially for early disease. This is particularly important if the deployment of echocardiography-based screening programmes for RHD were considered with an attendant focus on early disease.

- **How could an echocardiography-based RHD screening programme be funded?**
Could it be resourced within existing service frameworks?
- **Can an echocardiography screening programme fulfil the criteria** for “screening” if delivery of secondary prophylaxis remains poor?
- What would be an **appropriate service/workforce model of care if a screening programme were implemented**. Options could include an expanded scope of practice for primary health care staff to undertake screening echocardiography, delivery by specialist-led teams or outreach echocardiographers, and/or telemedicine for review of acquired echocardiography studies and discussion of management.

Solutions

The response to some of the issues highlighted above is already underway. An extensive Australian screening study, the **gECHO** (getting Every Child’s Heart Okay) study, is nearing completion. This project (a collaboration between Baker IDI (Alice Springs), James Cook University (Cairns), Menzies School of Health Research (Darwin) and the University of Western Australia (Broome) supported by the Australian Department of Health and Ageing) undertook screening echocardiograms in 4000 Aboriginal and Torres Strait Islander children and 1000 non-Indigenous Australian children across northern Western Australia and Queensland, and the northern Top End and Centre of the Northern Territory. Preliminary results of gECHO identified a significant proportion of children with mild morphologic abnormalities, particularly of the mitral valve, of doubtful significance. In order to clarify the significance of these results, a follow up study is being undertaken.

RhFFUS (Rheumatic Fever Follow-Up Study) is a prospective cohort study of children with non-specific mitral and/or aortic valve abnormalities that will examine whether such children are more likely to have an episode of ARF or develop RHD than children with normal heart valves. Supported by the National Health and Medical Research Council

(NHMRC), the findings of RhFFUS will provide clarity regarding the echocardiographic diagnosis of early RHD, help clinicians to better understand the significance of subtle changes on echocardiography, and inform the health service response for children with minor valve abnormalities. If such children are shown to have an increased risk of ARF and/or progression to RHD, then a case may be made for identifying high risk children earlier through screening echocardiography and offering them regular secondary prophylaxis to prevent progression to more severe RHD.

Given the existing uncertainty it remains premature to advocate for, or incorporate, echocardiographic screening for RHD into Australian clinical practice. If results of gECHO and RhFFUS indicate that screening may be viable then the next step will be to undertake a detailed scoping and impact study focusing on how such a programme would be delivered and sustained, its cost and comparative cost-benefit, and how it would impact on the existing primary and specialist workforce. If a case cannot be made, or support obtained, for a national RHD screening programme, there may remain a rationale for screening on a quasi *ad hoc* basis within high risk communities and areas.

2. Getting secondary prophylaxis to work

Background

Repeated episodes of ARF increase the likelihood that a person will develop RHD or will cause progression of RHD in those with minor disease.[11] If such repeated episodes of ARF can be prevented then the possibility of the development of severe RHD, with the attendant requirement for surgery to repair or replace damaged heart valves or other interventions, is reduced. For this reason, secondary prophylaxis in the form of four-weekly long-acting benzathine (LAB) penicillin injections is recommended for those who have had an episode of ARF or who have RHD.[9] The rationale for this treatment is the prevention of further GAS infections that may in turn lead to recurrent ARF. It should be noted that while there is good evidence that secondary prophylaxis for ARF/RHD is effective, oral antibiotics are inferior to intramuscular LAB in preventing recurrent ARF. The use of oral antibiotics is therefore only encouraged in patients with clear hypersensitivity to penicillin.[12]



While the effectiveness of secondary prevention is proven, achieving effective delivery and uptake of this has often been difficult. There is no agreed benchmark for the uptake of secondary prophylaxis, and indeed anything less than 100% of doses is suboptimal. However, a generally utilised target for adequate uptake utilised in Australia is 80% of recommended LAB injections over a 12-month period. Unfortunately, data shows that relatively few Aboriginal and Torres Strait Islander individuals living with ARF/RHD achieve this level of secondary prophylaxis uptake.[13,14] While there is much anecdote regarding why the system is failing, there remains no clear evidence regarding how best to respond to this clear service gap. It is likely that one particular issue with secondary prophylaxis for ARF/RHD is the longevity and inconvenience of treatment. Clients accessing secondary prevention treatment usually have to undergo 10-20 years of painful four-weekly injections that may be perceived as having little benefit. The consequences of ARF/RHD are, like hypertension or kidney disease, only apparent once the disease is advanced at which time secondary prevention is often futile.

Gaps

Potential issues and service gaps influencing the uptake of effective secondary prevention for ARF/RHD were identified including:

- While **oral penicillin is not recommended**, too many health professionals prescribe it in place of LAB injections. The protection provided by the variable use of oral antibiotics is not sufficient.[12]
- **Centralised RHD register and recall programmes** are important in coordinating care. Nonetheless the Northern Territory experience would indicate that such systems alone cannot achieve the required levels of secondary prophylaxis uptake.
- There are great **disparities in the uptake of secondary prophylaxis** in different communities. Successes should inform programme development.
- In a primary health care environment faced with acute health care needs, secondary prophylaxis, like chronic disease management, is sometimes not seen as a priority.
- **Mobility** of some Aboriginal and Torres Strait Islander patients can make it difficult for the health system to effectively deliver regular prophylaxis.

Solutions

- Develop a sense of **urgency and priority** for the delivery of secondary prophylaxis in primary health care. Whilst primary care providers are faced with a broad range of health issues, all placing demands on finite time and resources, it is necessary to prioritise the delivery of secondary prophylaxis. Potential strategies include:
 - marketing (patient, family, community, health providers - “we're talking about children/the future”).
 - education (including utilising ‘clinical champions’/opinion leaders such as cardiologists and cardiac surgeons; introducing health provider training and professional development for all relevant primary health care providers and specialists).
- **Systems based approaches** are required to ensure ARF/RHD fits within established chronic disease frameworks and systems. Active recall and follow-up is vital with effective health information management systems that allow the sharing of health data so that patients can access care at different health care centres and care items received are notified to a central database that can be widely accessed. There needs to be integration between central ARF/RHD registers and primary health care health information management systems.
- Development and evaluation of primary health care-based or led strategies for delivery of secondary prophylaxis including:
 - **Whose job is it?** Is it important to have a dedicated person within the team who is responsible for ensuring prophylaxis is delivered? Does opportunistic delivery work when provided by all members of the primary health care team? The most effective and appropriate model for primary health care-based delivery of secondary prophylaxis should be a priority for future research.
 - **Work flow** - fast-tracking individuals presenting for their injection at clinics.
 - Basing **timing** of secondary prophylaxis on patient/community concepts of time – e.g. seasonal or community events (e.g. injections due on the full moon).
 - **Communication and recall** – what are patient, family, community needs and preferences? Is the concept of self-management appropriate in this setting?

- **Continuous quality improvement** initiatives to ensure better delivery of services. Focus on what the health service is doing rather than on what the patient is not doing.
- **Patient control and information ownership** - Hand-held records for patients so that they can access secondary prophylaxis at any primary health care site. Participation in Australian national shared electronic health record (eHealth) initiative.
- **Incentives** – Is there are role for reward system to encourage clients to achieve high levels of secondary prevention uptake?
- **Community-based delivery** – alternate modes of delivery including the New Zealand model of secondary prophylaxis delivery by community-based public health nurses in schools and homes.
- **Smart recall systems** – explore innovate methods for supporting clients and families and providing reminders through schools or workplace, or by using technology including SMS messaging, email and other internet based platforms.
- **Better methods of delivery** - it is arguable that the delivery of secondary prophylaxis by 4-weekly LAB injections is a failed treatment model. A paradigm shift in the mechanism for delivery or a means of improving the delivery of intramuscular long-acting penicillin is required. Investment in the development of innovative delivery systems for secondary antibiotic prophylaxis of ARF/RHD which are more convenient, less painful and longer-acting should be a priority. Given the small potential market, relying on commercial imperatives alone is unlikely to achieve this and strategic relationships with device and drug development organisations with a cost and risk-sharing model will most likely be required.

3. Better and more appropriate management of advanced RHD

Background

When the heart is no longer able to compensate for the abnormal functioning of damaged valves, heart failure results. This is most common in young adults, but is also sometimes seen in children.[4] Once valve damage is severe there are a broad range of options available dependent



both on patient circumstances and the degree and type of valve damage. Some options will not require the patient to be on lifelong anticoagulation (warfarin), a desirable outcome given the risks of bleeding and inconvenience of regular blood test monitoring. Other options will require warfarin therapy with its inherent inconvenience and complications including bleeding, valve thrombosis and embolisation. Furthermore, some surgical interventions rarely require repeat intervention (mechanical valves) while others may eventually require later operations (bioprosthetic valves, valve repair, balloon valvuloplasty). The choice and timing of intervention therefore needs to be carefully balanced taking into account patient preferences, the safety of anticoagulation and the risk of later reoperation before proceeding with any particular course of action.

In Australia, patients with RHD who require surgery are routinely transferred to one of approximately thirty city-based cardiothoracic surgical units. Given the number of units involved it is hardly surprising that the surgical management of RHD varies widely. For example, patients with mitral regurgitation, the most common valve damage seen in RHD, may undergo valve repair or a valve replacement with a mechanical or bioprosthetic valve; which occurs is often more dependent on where the operation is undertaken rather than on the application of consistent and objective criteria. This is perhaps why, once the patient returns to their usual health care providers, questions may arise regarding whether the intervention undertaken was the most appropriate option.

A similar situation exists for mitral stenosis. While percutaneous balloon valvuloplasty is an effective and comparatively safe treatment for mitral stenosis, particularly in younger and pregnant patients, there are few centres in Australia that undertake this in large numbers. If patients with mitral stenosis are referred to cardiothoracic surgeons anecdotal reports would indicate there can be a tendency to operate and replace the valve rather than to undertake balloon valvuloplasty.

The problem of inconsistency in surgical and other interventions (e.g. percutaneous balloon valvuloplasty) for the management of advanced RHD is further exacerbated by the fact that most patients undergo surgery in major city centres far removed from the realities of the

remote communities or regional centres where they usually live. This often entails a disconnection between the decisions being made by tertiary hospital-based specialists and surgeons, local primary and specialist health care providers, and the practical aspects of life and health service access in regional and remote Australia.

Gaps

- **Mitral valve repair** – The use of mitral valve repair versus mitral valve replacement for RHD varies greatly between different cardiothoracic surgical centres in Australia. Overall in Australia there is a general lack of experience with surgical repair as opposed to valve replacement.
- **Delay in presentation** - patients with RHD can first present for primary and specialist health care with symptomatic and advanced disease that requires early and occasionally urgent surgical intervention. This has particular implications for the suitability for mitral valve repair as late referral often means valve damage is extensive and mitral valve repair is not possible.
- **Consistency and leadership in the surgical management of RHD** – the diversity of the management of advanced RHD across Australia has been noted. There are no national centres of excellence for specialist RHD diagnosis, severity assessment and management.
- **Multidisciplinary team management of advanced RHD** – decisions regarding the details of management of advanced RHD are frequently undertaken by cardiothoracic surgical teams. This can often occur without broader input from the patient/family and other health care providers (including local primary health care providers and regional and visiting specialists) particularly with regard to the implications for local follow-up, the need for anticoagulation, future pregnancy, re-operation and infective endocarditis risk.

Solutions

- **Improved understanding of health care access and uptake of secondary prophylaxis** - issues pertaining to secondary prophylaxis for ARF/RHD are discussed above. A greater understanding is also required in relation to why patients are lost to

follow-up, how to identify RHD in women before they may become pregnant and how to encourage the early presentation and appropriate investigation of patients with unexplained shortness of breath.

- **National centres of excellence** for the diagnosis, assessment and surgical management of RHD are required. Health staff, particularly those at primary health care sites where most health care for people with ARF/RHD occurs, should be able to easily contact clinical experts who can provide consistent evidence-based advice that reflects the realities of regional and remote Australian life and clinical practice.
- **Multidisciplinary care and team-based decision making** for the planning of intervention for RHD. Decision-making needs to involve those who will be faced with the aftermath of intervention/surgery (i.e. patients, families and local primary and specialist health care providers).
- **Early referral for surgical input** would allow a broader range of options for intervention to be considered. Multidisciplinary and team-based decision making would encourage this particularly if such input could be provided locally either by telemedicine or through cardiothoracic surgical outreach to regional centres.
- **National RHD surgical register and research strategy** for the assessment, intervention and long-term outcome of surgery and other interventions for RHD. A priority is the development of a surgical management and outcome register that incorporates details regarding a standardised baseline assessment, documents the rationale for the intervention chosen, and allows short and long-term follow-up which includes re-operation, readmission, morbidity (including stroke and endocarditis) and survival. Where possible, additional measures incorporating objective assessments of function (six minute walk test) and quality of life should be included. This will enable the development of evidence-based recommendations for surgical and other interventions in the management of advanced RHD in Australia and have significant implications for international practice.
- **Improving the use of warfarin** – warfarin is likely to remain a reality of RHD management. Research to enhance the understanding of how patients and primary health care providers perceive long-term anticoagulation and how monitoring and regular use of warfarin can be enhanced should be a priority.

Additional issues pertaining to ARF/RHD care discussed in less detail

Whilst there were at least seven other areas that were not discussed in any detail the primordial and primary prevention of ARF/RHD was a recurring theme.

Primary prevention and Group A Streptococcus – where to from here?

Whilst not all patients presenting with ARF have a history of pharyngitis[15], the early treatment of Group A Streptococcus (GAS) associated pharyngitis provides an effective opportunity to prevent the development of ARF.[16,17] New Zealand experience would indicate that there is limited awareness of the importance of the early management of pharyngitis in high-risk (Māori and Pacific Islander) populations both in community members and health care providers. Research investigating the understanding of how communities at high risk of ARF/RHD and local primary health services perceive and respond to pharyngitis (including seeking health care review) is required. This should inform community and health provider education initiatives to ensure pharyngitis prompts primary health care review and that primary health care providers have clear and consistent protocols for confirming a diagnosis of GAS-associated pharyngitis or treatment protocols for empiric management.

Conclusion

The Cardiac Society of Australia and New Zealand's Alice Springs 2011 Indigenous Cardiovascular Health Conference provided a unique and valuable opportunity for experts with 'on the ground experience' in primary and specialist health care delivery and planning to gather and identify shared priorities in the Australian response to ARF/RHD (see Box 2). Although time was limited this group provided clear recommendations to inform the local, jurisdictional and national response to ARF/RHD. There remains much to be done and many unanswered questions. Nonetheless, it is hoped this document helps chart a course for addressing what is a complicated health issue in regional and remote Australia and for Aboriginal and Torres Strait Islander peoples.

As one participant noted:

'I work in the primary health area. I've been in Aboriginal health for a long time. To think that rheumatic fever has been around for a very long time, and is only just got on the agenda, and it's making me think, yeah, I've... got relatives and family who's got rheumatic fever. We talk about diabetes, we talk about HIV / AIDS... but to me this has just come on the agenda.'

It is imperative that ARF/RHD now remains on the national and international health agenda. With the support and advocacy of CSANZ and the ongoing advice and commitment from patients, communities and health providers, ARF/RHD can be largely eradicated as it has been for non-Indigenous Australians.

1. **Echocardiography and Screening/Diagnosis of RHD** - Given the existing uncertainty it remains premature to advocate for or to incorporate echocardiographic screening for RHD into Australian clinical practice. Further research is currently being undertaken to evaluate the potential for echocardiography screening.
2. **Secondary Prophylaxis** – Secondary prophylaxis (LAB injections) must be seen as a priority. Systems-based approaches are necessary with a focus on the development and evaluation of primary health care-based or led strategies incorporating effective health information management systems. Better/novel systems of delivery of prophylactic medications should be investigated.
3. **Management of Advanced RHD** - National centres of excellence for the diagnosis, assessment and surgical management of RHD are required. Early referral for surgical input is necessary with multidisciplinary care and team-based decision making that includes patient, family, local health providers. There is a need for a national RHD surgical register and research strategy for the assessment, intervention and long-term outcome of surgery and other interventions for RHD.

Box 2. Summary of recommendations from the CSANZ Indigenous Cardiovascular Health Conference 2011 – ARF/RHD workshop.

References

1. Kaplan EL. Epidemiological approaches to understanding the pathogenesis of rheumatic fever. *Int J Epidemiol* 1985; 14: 499-501.
2. Steer AC, Carapetis JR, Nolan TM, Shann F. Systematic review of rheumatic heart disease prevalence in children in developing countries: the role of environmental factors. *J Paediatr Child Health* 2002; 38: 229-34.
3. Kerdelmidis M, Lennon DR, Arroll B, Peat B, Jarman J. The primary prevention of rheumatic fever. *J Paediatr Child Health* 2010; 46: 534-48.
4. AIHW: Field B. Rheumatic heart disease: all but forgotten except among Aboriginal and Torres Strait Islander peoples. Australian Institute of Health and Welfare, Canberra: Bulletin no. 16. AIHW Cat. No. AUS 48.; 2004.
5. Carapetis JR, Wolff DR, Currie BJ. Acute rheumatic fever and rheumatic heart disease in the top end of Australia's Northern Territory. *Med J Aust* 1996; 164: 146-9.
6. Rothstein J, Heazlewood R, Fraser M. Health of Aboriginal and Torres Strait Islander children in remote far north Queensland: findings of the paediatric outreach service. *Med J Aust* 2007; 186: 519-21.
7. Richmond P, Harris L. Rheumatic fever in the Kimberley region of Western Australia. *J Trop Pediatr* 1998; 44: 148-52.
8. Marijon E, Ou P, Celermajer DS, Ferreira B, Mocumbi AO, Jani D et al. Prevalence of rheumatic heart disease detected by echocardiographic screening. *N Engl J Med* 2007; 357: 470-6.
9. National Heart Foundation of Australia (RF/RHD guideline development working group) and the Cardiac Society of Australia and New Zealand. Diagnosis and management of acute rheumatic fever and rheumatic heart disease in Australia - an evidence-based review; 2006.
10. Marijon E, Celermajer DS, Tafflet M, El-Haou S, Jani DN, Ferreira B et al. Rheumatic heart disease screening by echocardiography: the inadequacy of World Health Organization criteria for optimizing the diagnosis of subclinical disease. *Circulation* 2009; 120: 663-8.
11. Bland EF, Duckett Jones T. Rheumatic fever and rheumatic heart disease; a twenty year report on 1000 patients followed since childhood. *Circulation* 1951; 4: 836-43.

12. Manyemba J, Mayosi BM. Penicillin for secondary prevention of rheumatic fever. *Cochrane Database Syst Rev* 2002: CD002227.
13. Mincham CM, Mak DB, Plant AJ. The quality of management of rheumatic fever/heart disease in the Kimberley. *Aust N Z J Public Health* 2002; 26: 417-20.
14. Eissa S, Lee R, Binns P, Garstone G, McDonald M. Assessment of a register-based rheumatic heart disease secondary prevention programme in an Australian Aboriginal community. *Aust N Z J Public Health* 2005; 29: 521-5.
15. Veasy LG, Tani LY, Hill HR. Persistence of acute rheumatic fever in the intermountain area of the United States. *J Pediatr* 1994; 124: 9-16.
16. Denny FW, Wannamaker LW, Brink WR, Rammelkamp CH, Jr., Custer EA. Prevention of rheumatic fever; treatment of the preceding streptococcal infection. *J Am Med Assoc* 1950; 143: 151-3.
17. Catanzaro FJ, Rammelkamp CH, Jr., Chamovitz R. Prevention of rheumatic fever by treatment of streptococcal infections. II. Factors responsible for failures. *N Engl J Med* 1958; 259: 53-7.