Australian Paediatric Surveillance Unit
STUDY PROTOCOL
Acute Rheumatic Fever

COMMENCING October 2007

BACKGROUND

Acute rheumatic fever (ARF) is a multi-system disease caused by an immunological response to group A streptococcal (GAS) infection. Children who have had ARF are susceptible to recurrent episodes when subsequent GAS infections occur. These recurrences often cause accumulated damage to heart valves (rheumatic heart disease - RHD), and consequent cardiac failure, the need for valve surgery or death. GAS infections and subsequent recurring ARF can be prevented by regularly administering penicillin as a regimen of *secondary prophylaxis*.

The highest documented rates of ARF and RHD in the world are found in Aboriginal Australians, and Maori and Pacific Islander people in New Zealand and Pacific Island nations. Aboriginal and Torres Strait Islander people are reportedly up to 8 times more likely than non ATSI people to be hospitalised for ARF and RHD, and nearly 20 times as likely to die.[1] Approximately 43% of Aboriginal people with ARF or RHD in the Top End of the Northern Territory first present with established RHD.[2,3] As most rheumatic valve lesions are the result of repeated or prolonged episodes of ARF in childhood and adolescence, [4] these data suggest that the early episodes of ARF are not being diagnosed in many children in the Top End of NT.

ARF is predominantly, but not exclusively, a problem among Indigenous communities and our understanding of the epidemiology and impacts of ARF is currently restricted to the NT and QLD. However, there are no data on the incidence, management and outcomes for this debilitating condition for the southern regions of Australia where an estimated 57% of the Indigenous community lives. Furthermore, there are no data on the impact of ARF on children in other communities eg. immigrants and refugees.

This study aims to provide national data of ARF in Australian children and to determine where and in whom ARF is currently occurring. This study will also document recurring episodes of ARF and the use of secondary prophylaxis. Using the information we will make recommendations on where ARF and RHD programs should be established to reduce the level of sickness and death that results from ARF and RHD.

STUDY OBJECTIVES

This study aims to:

- Estimate the incidence of ARF in the child population of Australia, particularly in regions from which there are currently no data, or only poor quality data.
- Determine the proportion of all ARF episodes that are recurrences.
- Identify populations, groups and regions at highest risk of ARF.

REPORTING INSTRUCTIONS

Please report any new episode of Acute Rheumatic Fever (even if there is a history of previous episodes) in any child <15 years of age and diagnosed according to the criteria provided in the **case definition**.

CASE DEFINITION

According to the National Heart Foundation Guidelines for Diagnosis and Management of ARF and RHD [5].

<table>
<tr><th></th><th>High Risk Groups</th><th>All Other Groups</th></tr>
<tr><td>Initial episode of ARF</td><td colspan="2">2 major or 1 major and 2 minor manifestations
Plus
Evidence of a preceding GAS infection</td></tr>
<tr><td>Recurrent attack of ARF (in a patient with known past ARF or RHD)</td><td colspan="2">2 major or 1 major and 2 minor or 3 minor manifestations
Plus
Evidence of a preceding GAS infection</td></tr>
<tr><td></td><td></td><td></td></tr>
<tr><td>Major Manifestations</td><td>▪ Carditis (including subclinical evidence of rheumatic valve disease on echocardiogram)
▪ Polyarthritis or aseptic mono-arthritis or polyarthralgia
▪ Chorea
▪ Erythema marginatum
▪ Subcutaneous nodules</td><td>▪ Carditis (including subclinical evidence of rheumatic valve disease on echocardiogram)
▪ Polyarthritis
▪ Chorea
▪ Erythema marginatum
▪ Subcutaneous nodules</td></tr>
<tr><td>Minor manifestations</td><td>▪ Fever
▪ ESR ≥30 mm/hr or CPR ≥30 mg/l
▪ Prolonged P-R interval on ECG</td><td>▪ Fever
▪ Polyarthralgia or aseptic mono-arthritis
▪ ESR ≥30 mm/hr or CPR ≥30 mg/l
▪ Prolonged P-R interval on ECG</td></tr>
<tr><td>Evidence of a preceding GAS infection</td><td colspan="2">▪ Elevated or rising antistreptolysin O or other streptococcal antibody, or a positive throat culture or rapid antigen test for GAS.
▪ Upper limits of normal for streptococcal antibody titres in Australia:
<table>
<tr><th>AGE GROUP</th><th colspan="2">UPPER LIMIT OF NORMAL (IU/ML)</th></tr>
<tr><th>(years)</th><th>ASO titre</th><th>Anti-DNase B titre</th></tr>
<tr><td>4–5</td><td>120</td><td>100</td></tr>
<tr><td>6–9</td><td>480</td><td>400</td></tr>
<tr><td>10–14</td><td>320</td><td>380</td></tr>
</table></td></tr>
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FOLLOW UP OF NOTIFICATIONS: A brief questionnaire requesting details about the diagnosis will be sent to clinicians who notify a case of ARF to the APSU. In the event that Sydenham's chorea is identified, a brief questionnaire on chorea will also be sent to clinicians who agree to participate in collection of chorea data (optional).

INVESTIGATOR CONTACT DETAILS (*Principal investigator)

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REFERENCES

1. AIHW: Field B. Rheumatic heart disease: all but forgotten in Australia except among Aboriginal and Torres Strait Islander peoples. Canberra: AIHW, 2004.
2. Carapetis J, Wolff D, Currie B. Acute rheumatic fever and rheumatic heart disease in the Top End of Australia's Northern Territory. Med J Aust 1996;164:146-149.
3. Carapetis J, Currie B. Clinical epidemiology of rheumatic fever and rheumatic heart disease in tropical Australia. Adv Exp Med Biol 1997;418:233-236.
4. ABS, National Aboriginal and Torres Strait Islander Health Survey: Summary Booklet,
5. NHFA & CSANZ (2006) Diagnosis and management of acute rheumatic fever and rheumatic heart disease in Australia: An evidence-based review. *http://www.heartfoundation.org.au/document/NHF/PP-590_Diagnosis-Management_ARF-RHD_Evidence-Based%20Review_Sep06Update_FINAL.pdf*