Rheumatic Heart Disease in Australia – a Dickensian Disease still prevalent in the top end

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Claire Boardman
Deputy Director, RHDAustralia
BN, Cert IC, MPH, CICP, Senior Lecturer Griffith University Qld.

Streptococcus pyogenes bacteria, Pappenheim’s stain
QUICK QUIZ

Who:
- Lives or works remotely?
- Has seen a case of ARF?
- Has given a shot of BPG (LA Bicillin)?
- Knows how often BPG should be given?
- Knows how to prevent RHD?
- Knows how/whom to refer to?
- Is aware of the National guidelines?
- Has downloaded the Diagnostic App?
- Has visited our NRHC stand?
Born in 1812 England
2nd of 8 children

Victorian England’s most popular novelists

What does Charles Dickens have in common with rheumatic heart disease?
What history tells us

• RF was leading cause of death in people aged 5-20 years in US 100 years ago.

• Mortality rate was 8-30% from carditis and valvulitis this decreased to 4% by 1930s.

• Following penicillin use mortality rate < 0% by 1960s but remains at 1-10% in developing countries.

• Before penicillin, 60-70% of patients developed valve disease, compared to 9-39% since penicillin was developed.
It's the wet season in Arnhem Land but it hasn't rained for days. The dancers lose down the hot red sand just so they can stamp their feet.

They raise their hands and cry out for this—a funeral for a footballer.

It has been the same song for weeks now. Part of the month-long mourning since the young man died of a Third World disease in a land that should be far removed from it.

He had paced well against his opponents that day but stumbled off the field in the final quarter, weeping and holding his chest.

He collapsed and died on the sidelines in a scene too often repeated across the Top End. Another life lost to rheumatic heart disease—the ugly plague that extends its reach each day like cracks through the dry earth.

The traditional sounds of a digeridoo have twisted through the trees each night since his death, drawing family and friends in Maningrida for their final farewells.

His body lies in state—a symbolic shelter of tin and leaves and branches.
Rheumatic heart disease is a sentinel condition of poverty and of health inequality; its persistence marks the failure of our health systems to address the NCDs of the poor.

Professor Bongani Mayosi | South Africa
5 fast facts

- ARF is entirely preventable - ie address the risk factors

- Mostly affects children 5 -14 years of age & majority are female

- Caused by upper respiratory GAS infection (or skin infections)

- RHD affects 15.6 to 19.6 million people worldwide & causes 233,000 to 492,000 deaths each year.

- Timely diagnosis of an initial ARF episode and subsequent use of antibiotic prophylaxis is the best method of preventing RHD in Australia, in NZ early treatment of a sore throat.
World importance

• ARF remains the most common cause of acquired heart disease in children around the world
• ARF incidence is a sensitive marker of childhood disadvantage

Local incidence

• ARF incidence & RHD prevalence in remote Indigenous communities of Northern and Central Australia, Pacific Islanders & Maori are among the highest reported in the world
• ARF tends to run in families & is more common in females
• Incidence peaks in the 5 to 14 year age group
• It has all but disappeared from people in wealthy societies
Pathogenesis

- ARF is an illness caused by a reaction to a bacterial infection with group A streptococcus (GAS)
- Not everyone is susceptible to ARF, and not all GAS strains are capable of causing ARF in a susceptible host
- The role of preceding GAS pharyngitis is undisputed but the role of preceding skin infection is less certain
- Preceding symptomatic GAS pharyngitis is rarely identified in Indigenous populations

Clinical

- ARF ‘licks the joints and bites the heart’
- Key clinical features: fever, acute joint pain and carditis.
- Carditis is usually manifest by prolonged PR interval on ECG & mitral regurgitation. Most episodes last < 6 weeks
- Sydenham’s chorea is a separate and delayed movement & mood disorder; it accounts for about 25% of cases in Indigenous Australians. Episodes last about 3 months, sometimes longer
- Presentation with subcutaneous nodules and erythema marginatum is rare, but highly specific, and a distinctive feature of ARF
Diagnosis

• ARF is a clinical diagnosis; there are no definitive diagnostic tests
• Jones and W.H.O criteria have been modified to form Australian guidelines to increase the sensitivity for ARF diagnosis in Australia’s unique high risk population
• Suspect ARF in any child with fever and joint pain, look for other signs of ARF (e.g. murmur), do relevant investigations (ECG and blood tests) & always discuss with experienced colleagues
• Most healthy children in high-risk settings also have persistently raised ASOT & anti-DNase B; this limits their diagnostic usefulness

Treatment

• Joint pain and fever usually settle within 24 hours with oral aspirin — this is a diagnostic pointer
• Penicillin is prescribed to clear GAS from the throat
• Corticosteroids are sometimes given for acute severe carditis
• Sydenham’s chorea usually resolves without treatment — sodium valproate is the drug of choice when symptoms are severe
• It is crucial that SPx (BPG) is commenced immediately
• Education for patient and families to highlight the importance of long-term adherence and regular review
Recurrence

• ARF tends to recur with subsequent GAS infections
• Recurrent ARF leads to cumulative heart valve scarring (RHD) – especially the mitral and aortic valves
• Progression causes valve regurgitation, stenosis or both – possibly leading to heart failure, strokes, infective endocarditis, disability and early death
• Echocardiographic screening of high-risk children shows that most early asymptomatic RHD goes clinically undetected

Primary prevention

• Treatment of streptococcal pharyngitis is effective in controlled temperate climate settings (e.g. the US military) but has not been successful in poorly-resourced high-risk populations
• It has been difficult to sustain healthy skin programs in many remote communities and a vaccine is many years way
Secondary prevention & RHD control programs

• Register-based programs providing 21 - 28 days BPG reduce recurrent ARF
• Regular clinical followup & echocardiograms - prioritised according to a disease severity scale. The guidelines also cover routine vaccination, dental care & endocarditis prophylaxis
• Delivery of secondary ARF prophylaxis in remote primary care settings can be extremely challenging
• ARF is a notifiable disease in QLD, NT and WA (SA almost)
• Australia guidelines for prevention, diagnosis and management & quick reference guides are available from: www.rhdaustralia.org.au

Primordial prevention

• Ultimately, primordial prevention (adequate housing, education, employment and access to health services) is the only sustainable way to prevent ARF and RHD
THE ARF/RHD PATHWAY

**PRIMORDIAL**
- Stop development of risk factors
- Prevent GAS infections

**PRIMARY**
- Target populations at risk
- Stop sore throats* & manage skin sores

**SECONDARY**
- Diagnose & manage ARF
- Secondary Px with BPG
- Adherence rates

**TERTIARY**
- Surgical intervention
- Valve replacement

PRIMARY HEALTHCARE MODEL - 1978 WHO ALMA ATA DECLARATION

Social approach to health founded on human rights framework
Based on economic and social justice
Affordable, accessible, appropriate
Considers culture, environment, ethnicity

SOCIAL DETERMINANTS OF HEALTH INCLUDE:

• Stress
• Social exclusion
• Unemployment
• Addiction
• Availability of healthy food
• Availability of healthy transportation
• Social support networks
• Early childhood development
• Social gradients (shorter life expectancy, the poorer you are > disease risk)
PRIMORDIAL PREVENTION of PYODERMA

• Healthy housing
• Education
• Hygiene
• Early detection & treatment of skin sores
• Scabies control
What we know & what we don’t

Delays in service delivery

Gaps in continuum of care

Lack of knowledge (patients, clinicians, and community members)

Large scale health system strengthening

Improvement in delivery of PHC (change at multiple levels)

How do we get here?
RHDAUST BACKGROUND

• Based at Menzies in Darwin, NCU established in 2009 to support control of RHD in Australia
• Funded under DoHA Rheumatic Fever Strategy
• Partners include: Baker IDI, JCU, National Heart Foundation, SAMRHI, Telethon

OVERALL PROGRAM AIM
To reduce death and disability from ARF/RHD in Australian Aboriginal and Torres Strait Islander people by:

• Supporting RHD jurisdictional programs
• Establishing a data collection & reporting system
• Disseminating evidence based practice guidelines
• Increasing community awareness of ARF/RHD & prevention
The Guidelines

- New recommended management for Probable ARF
- New algorithm for Management of Probable ARF
- Expanded discussion around short-course antibiotics for treatment of ARF

Quick reference guides

1. Primary prevention of ARF
2. Diagnosis of ARF
3. Management of ARF
4. Secondary prevention of ARF
5. Management of RHD
6. RHD in pregnancy
7. RHD control programs

Find the guideline on the RHDA website homepage here:
SMART PHONE APPS

This application is for general information only. The information contained here is derived from the Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition) and refers to high risk groups. In Australia, high risk groups include Aboriginal people and Torres Strait Islanders, particularly across central and northern Australia. Pacific Islanders and migrants from countries with high prevalence of RHD are also known to be at high risk.

For more information visit
www.rhdaustralia.org.au

Tap Here to read the full disclaimer detailed


Key information available on ipad, android and iphone
ARF diagnosis flowchart & App

https://www.youtube.com/watch?v=RsIQFeYOkAg&feature=player_embedded
CLINICAL & COMMUNITY
Develop best practice standardised resources:
• 5 self paced clinical education modules
• Presentation materials
• Posters, pamphlets, electronic audio, visual media, etc
NEW CLINICIAN MODULES

Advanced education package provides clinicians & senior health staff with a suite of online e-learning modules. Package has been designed by clinicians for clinicians to further improve preventative capabilities, control & management of acute rheumatic fever and rheumatic heart disease.

1. ARF/RHD: What is it?
2. Primordial and Primary Prevention
3. Diagnosis of ARF in Children
4. Diagnosis of ARF in Adults
5. Secondary Prevention
6. RHD Diagnosis: General Principles
7. RHD and dental care
8. Medical Management of Mitral Valve Disease
9. Medical Management of Aortic Valve Disease
10. RHD: Mitral Stenosis – The option of mitral balloon Valvuloplasty
11. Rheumatic Heart Disease (RHD) and Stroke
12. RHD Management Anticoagulation in the bush
13. RHD & Infective Endocarditis
14. Pregnancy in RHD
15. Screening for RHD
WHAT CAN YOU DO AS RURAL HEALTH PRACTITIONERS?

• Beware boiled frog analogy
• Treat skin sores, follow up sore throats
• Work with community to ensure kids are coming into clinic for their SPx every 21-28 days
• Do the modules
• Download the Apps
• Talk to families
### CURRENT RHD PROJECTS IN NT & BEYOND

<table>
<thead>
<tr>
<th>Project</th>
<th>Description</th>
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<tbody>
<tr>
<td>RHDAustralia</td>
<td>RHDAustralia works with RHD control programs &amp; other partners throughout Australia to reduce death &amp; disability from this disease among Aboriginal and Torres Strait Islander people.</td>
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<tr>
<td>RHD Secondary Prophylaxis</td>
<td>This project will implement and evaluate an intervention package aimed at improving health systems to increase delivery of Secondary Prophylaxis in NT health centres.</td>
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<tr>
<td>RHD Genetics</td>
<td>To understand why some people appear to be susceptible to RHD while others are not despite being exposed to GAS.</td>
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<tr>
<td>RHD in Pregnancy - AMOSS</td>
<td>This project aims to provide an evidence base to improve clinical care and outcomes for women with RHD in pregnancy and their babies.</td>
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<tr>
<td>ARF Immunology</td>
<td>This project aims to find markers in the blood that can be used to rapidly and accurately diagnose acute rheumatic fever so that people can get treatment they need as soon as they can.</td>
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| RHD Echo screening                           | To evaluate role of early detection of RHD via echo screening in RHD control.  
  - To evaluate the economics of echo screening & development of evidence-based diagnostic & treatment echo screening protocols.                           |
| WHF Pacific and International RHD programme | Direct programme support provided to 5 countries (Fiji, Tuvalu, Nauru, Solomon Islands and Kiribati) as well as several studies, evaluations and projects.                                                  |
The impact