

The significance of RHD detected by echocardiography

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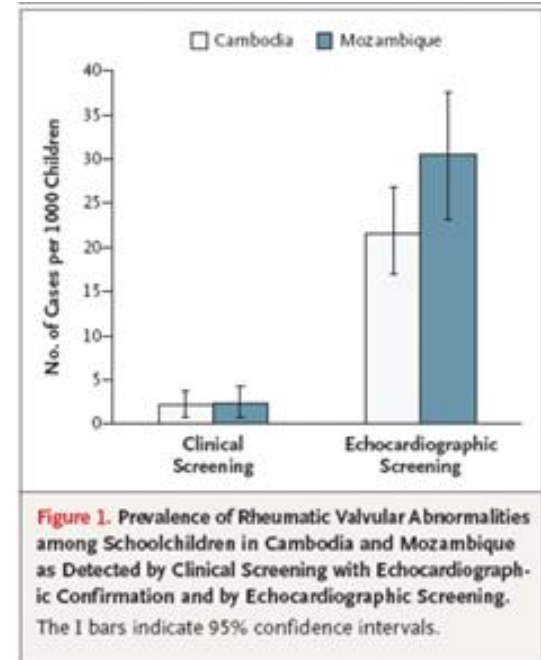
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RHD echo in high burden ARF/RHD populations

- Benzathine penicillin prevents recurrences & improves outcomes
- Globally, the majority of adults with RHD have not had prior ARF
- There is usually a latent phase in RHD

Echocardiography - a tool for case finding, initiating secondary prophylaxis, improving outcomes



Marijon et al. NEJM 2007

RHD echo research in NZ since 2007

- Disease burden
- Diagnostic criteria
- Natural history
- Perspective of screened population
- RHD prevalence in special population groups



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New Zealand echo screening 2007 onwards

High prevalence regions

Region	Surgery		Definite RHD	Possible/Borderline
South Auckland	1142	2	25 (2.4%)	30 (2.6%)
Tairāwhiti	685	1	8 (1.1%)	19 (2.7%)
Bay of Plenty	553		3 (0.5%)	15 (2.7%)
Kaitiaki	635	1	5 (0.8%)	16 (2.51)
Porirua (WHF)	621		8 (1.3%)	14 (2.3%)
South Auckland (adults, WHF)	465	3	10 (2.2%)	16 (3.4%)
Total	4,101	7	59 (1.4%)	110 (2.6%)

Low prevalence regions

North Shore	396	0	0	2 (0.5%)
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World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease—an evidence-based guideline

Bo Reményi, Nigel Wilson, Andrew Steer, Beatriz Ferreira, Joseph Kado, Krishna Kumar, John Lawrenson, Graeme Maguire, Eloi Marijon, Mariana Mirabel, Ana Olga Mocumbi, Cleonice Mota, John Paar, Anita Saxena, Janet Scheel, John Stirling, Satupaitea Viali, Vijayalakshmi I. Balekundri, Gavin Wheaton, Liesl Zühlke and Jonathan Carapetis



Definite RHD = regurgitation + morphologic changes

Borderline RHD = regurgitation

Remenyi et al. Nat Rev Cardiol 2012.

RHD Prevalence in high-risk NZ children (WHF)

	Borderline RHD		Definite RHD		RHD (Definite + Borderline)	
	Rate	Prevalence	Rate	Prevalence	Rate	Prevalence
NZ children 10 -13 years	90/3634	24.8 per 1000 20-30 per 1000	41/3634	11 per 1000 9 - 15 per 1000	131/3634	35.8 per 1000 30 - 42 per 1000

Prevalence of Definite RHD in Pacific children ~ 2%

Prevalence of Definite RHD in Māori children ~ 1%



Beth Tilton



Nikki Culliford-Semmens

Valvular Regurgitation Using Portable Echocardiography in a Healthy Student Population: Implications for Rheumatic Heart Disease Screening

Rachel H. Webb, MBChB, FRACP, Tom L. Gentles, FRACP, John W. Stirling, FRACP, Mildred Lee, MSc (Hons), Clare O'Donnell, MBChB, SM, FRACP, and Nigel J. Wilson, FRACP, Auckland, New Zealand

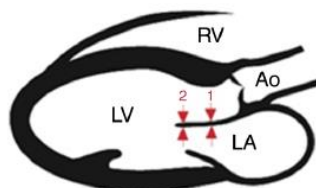
Table 2 Prevalence of valvular regurgitation detected by screening echocardiography ($n = 396$)

	<i>n</i>	HR (95% CI)
MR: all grades		
1. Nil	329	83.1% (79.1%–86.5%)
2. Physiologic MR*	59	14.9% (11.7%–18.7%)
3. Pathologic MR	5 [†]	1.3% (0.6%–2.9%)
AR: all grades		
1. Nil	388	98% (96.1%–99.0%)
2. Physiologic AR	8	2% (1.0%–3.9%)
3. Pathologic AR	0	0% (0.0%–1.0%)
TR: all grades		
1. Nil	107	27.0% (22.9%–31.6%)
2. Physiologic/mild	288	72.7% (68.1%–76.9%)
3. Pathologic	1	0.3% (0.04%–1.4%)
PR: all grades		
1. Nil	40	10.1% (7.5%–13.5%)
2. Physiologic/mild	353	89.6% (85.7%–91.8%)
3. Pathologic	1	0.3% (0.04%–1.4%)

- Prevalence of valvular regurgitation in healthy children is not well described
- N = 396 children at low risk for ARF/RHD
- Spectrum of echo findings described
- 1.5% had mild pathologic MR

Normal echocardiographic mitral and aortic valve thickness in children

Rachel H Webb,^{1,2} Nicola Culliford-Semmens,¹ Karishma Sidhu,³ Nigel J Wilson¹



1. Measurement for anterior mitral valve leaflet midpoint
2. Measurement for anterior mitral valve leaflet tip

3mm cut-off

Table 1 Comparison of valve leaflet measurements between normal and RHD (in mm)

Valve	Variable	Normal (n=288)		Abnormal (n=51)		p Value
		Median	IQR	Median	IQR	
Mitral valve	Maximal thickness AMVL tip PSLA	2.0	1.7–2.4	2.6	2.3–3.2	<0.0001
	Maximal thickness AMVL midpoint PSLA	2.0	1.7–2.4	2.8	2.5–3.2	<0.0001
	Maximal thickness PMVL midpoint	2.2	1.9–2.5	2.5	2.2–3.0	<0.0001
	PMVL length PSLA	11.1	10.0–12.4	11.2	10.7–12.4	0.2785
	Maximal thickness AMVL midpoint A4C	2.0	1.8–2.4	2.5	2.1–2.7	<0.0001
Aortic valve	Maximal thickness AV leaflet midpoint PSLA	1.5	1.3–1.6	1.9	1.7–2.1	<0.0001
	Maximal thickness AV leaflet midpoint PSSA	1.4	1.2–1.6	1.9	1.7–2.1	<0.0001

AMVL, anterior mitral valve leaflet; AV, aortic valve; PMVL, posterior mitral valve leaflet; PSLA, parasternal long axis; PSSA, parasternal short axis; RHD, rheumatic heart disease.

BPG adherence in RHD detected by echo screening in NZ

- BPG records for 57/62 individuals with echo-detected RHD and clinical recommendation to commence secondary prophylaxis
- Median duration of f/up = 5.8 years (range 16-95 months)

Figure 2: Adherence to BPG by year since commencement.

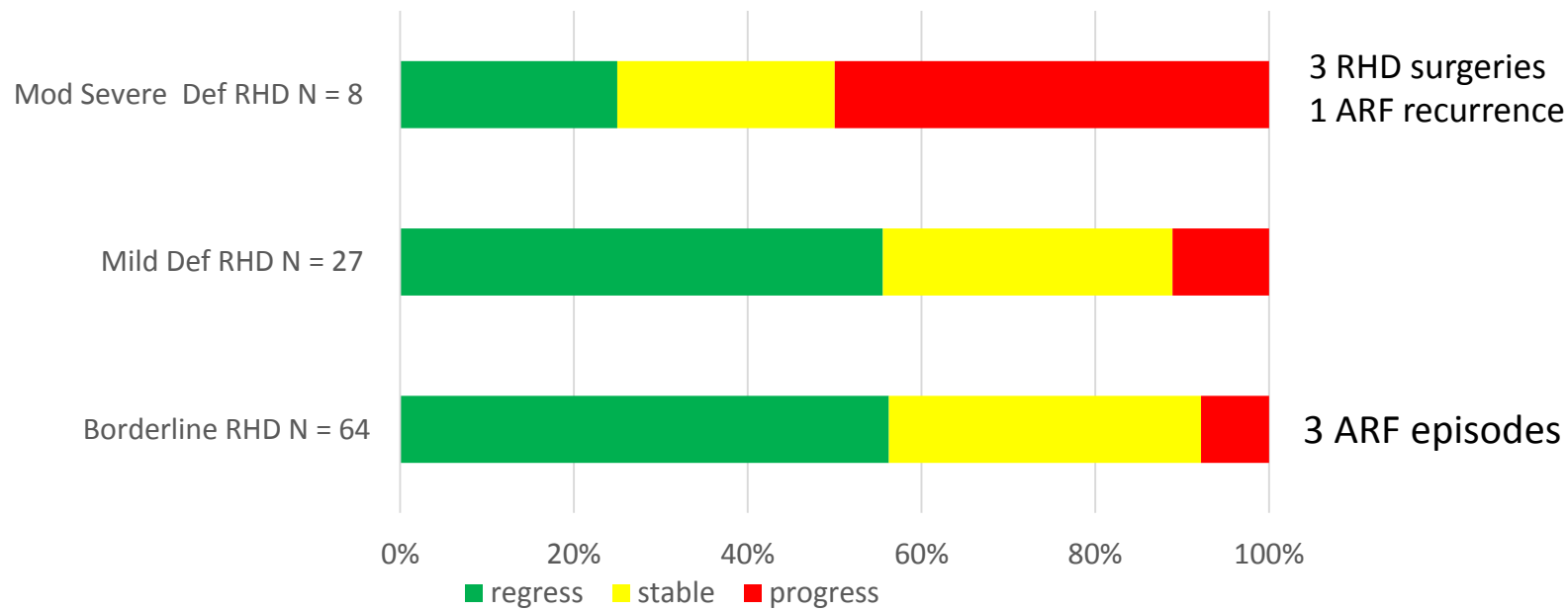
	Year 1	Year 2	Year 3	Year 4	Year 5	Year 6	Year 7	Year 8
n	57	57	53	46	45	38	23	12
Mean	90.4%	85.5%	82.6%	77.6%	75.7%	66.6%	74.7%	66.1%
Median	100.0%	100.0%	92.3%	84.6%	91.7%	84.0%	87.5%	87.5%

Screened population perspective

- Key findings from NZ
 - Strong family support / positive experience
 - High levels of rheumatic fever and sore throat awareness
 - Reduced physical activity and worry with abnormal result

Gurney et al. International Journal of Cardiology 2016; 221:734-740
Perelini et al. Journal of Paediatrics and Child Health 2015; 51:682-688.

Natural history N = 99 (borderline >12mo f/up)



Borderline RHD: Median f/up 57mo
Definite RHD: Median f/up 51mo

Definite RHD: 27/35 commenced prophylaxis
Borderline RHD: 23/64 commenced prophylaxis

Outcomes – pooled data

county	Number of patients	Person f/up days
Uganda	227	220190
New Zealand	94	157,826
Malawi	64	69,184
Australia	62	83,142
Fiji	37	70,189
Total	484	600,531

Preliminary analysis Dec 2018: Amy Sanyahumbi , Nigel Wilson et al

Average f/up 3.42 years

360 (74.7%) “good outcome”

122 (25.3%) “poor outcome”

Answering the Threshold Question:

Does penicillin prophylaxis improve outcomes for children with latent (echo detected) RHD?

- Randomized controlled trial comparing **2 year echo-outcomes** for children receiving every-4-week BPG and those not receiving prophylaxis
- ~ 900 children with borderline and mild definite RHD to be enrolled
- Progression vs regression?



GWOKO ADUNU PA LUTINO
PROTECT THE HEART OF A CHILD

Gulu, Uganda



RHD in NZ young adults: The South Seas Study

Aims

- Determine RHD prevalence in young adults from a high risk ARF/RHD population
- Explore feasibility of RHD echocardiography in an urban primary-care setting

Methods

- N = 465 young adults 16-40 years
- Stratified randomisation from PHO enrolment database
- Echos performed at GP clinic , Vivid Q[®] (GE) , scans reported according to WHF criteria

Results

- 8 (1.7%) had a previous clinical history of ARF/RHD.
- 10 had echocardiographic definite RHD, 6 were new diagnoses
- Definite RHD prevalence = 22 per 1,000 (2%)



The NZ familial echo study

Rationale: Family history ARF/RHD often reported, parental concern for siblings

Aim: Determine prevalence of RHD in first degree relatives of ARF patients

Methods:

- Recruitment from 3 Auckland region inpatient units Jan 2014 – Dec 2016
- Inclusion criteria: Biologic siblings of ARF cases ages ≥ 4 years & parents
- Questionnaire to determine family history of ARF/RHD.
- Standardised echocardiography protocol, hospital platform / VividQ
- Compared to demographically similar NZ populations

The NZ familial study: RHD Prevalence in siblings

12 RHD cases detected in 133 siblings (9 borderline & 3 definite)

	Total RHD (Definite + Borderline)	
	Rate	Prevalence
Siblings	12/133	90 per 1000 45 – 140 per 1000
NZ high-risk children	131/3634	36 per 1000 30 - 42 per 1000

**P
0.001**

SIBLINGS: 9% RHD (Definite + Borderline) compared to 3.5% in high-risk NZ children

Supports assessing siblings for RHD when a child is diagnosed with ARF

Echocardiography for enhanced case detection in a selected high-risk group

Summary:

the significance of RHD detected by echocardiography

- Epidemiology and advocacy
- Impact on disease severity under evaluation
 - Evolving f/up data from screened cohorts
 - RCT Uganda
- Shift in emphasis to active case finding in targeted groups

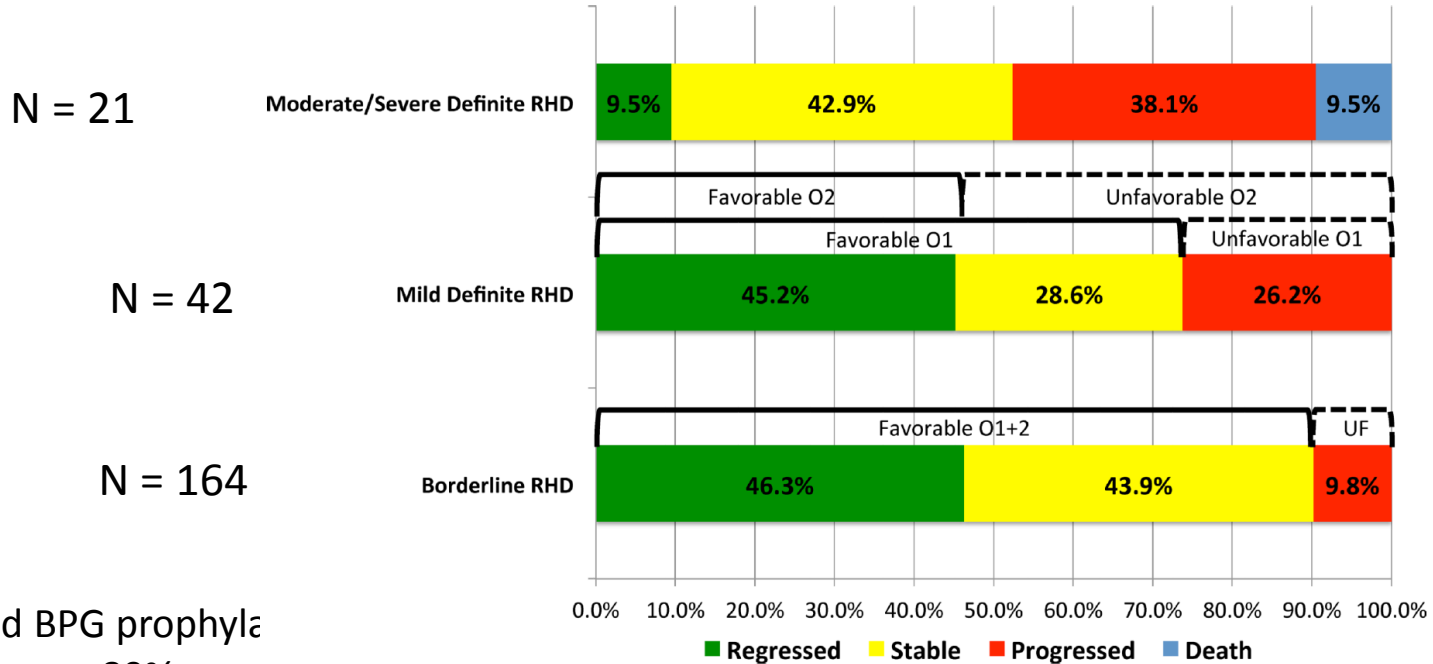
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Uganda:

Beaton et al. Circulation 2017; 136(23): 2233 – 2244



49% received BPG prophylaxis

80% adherence > 80%

Median follow-up duration 2.3 years