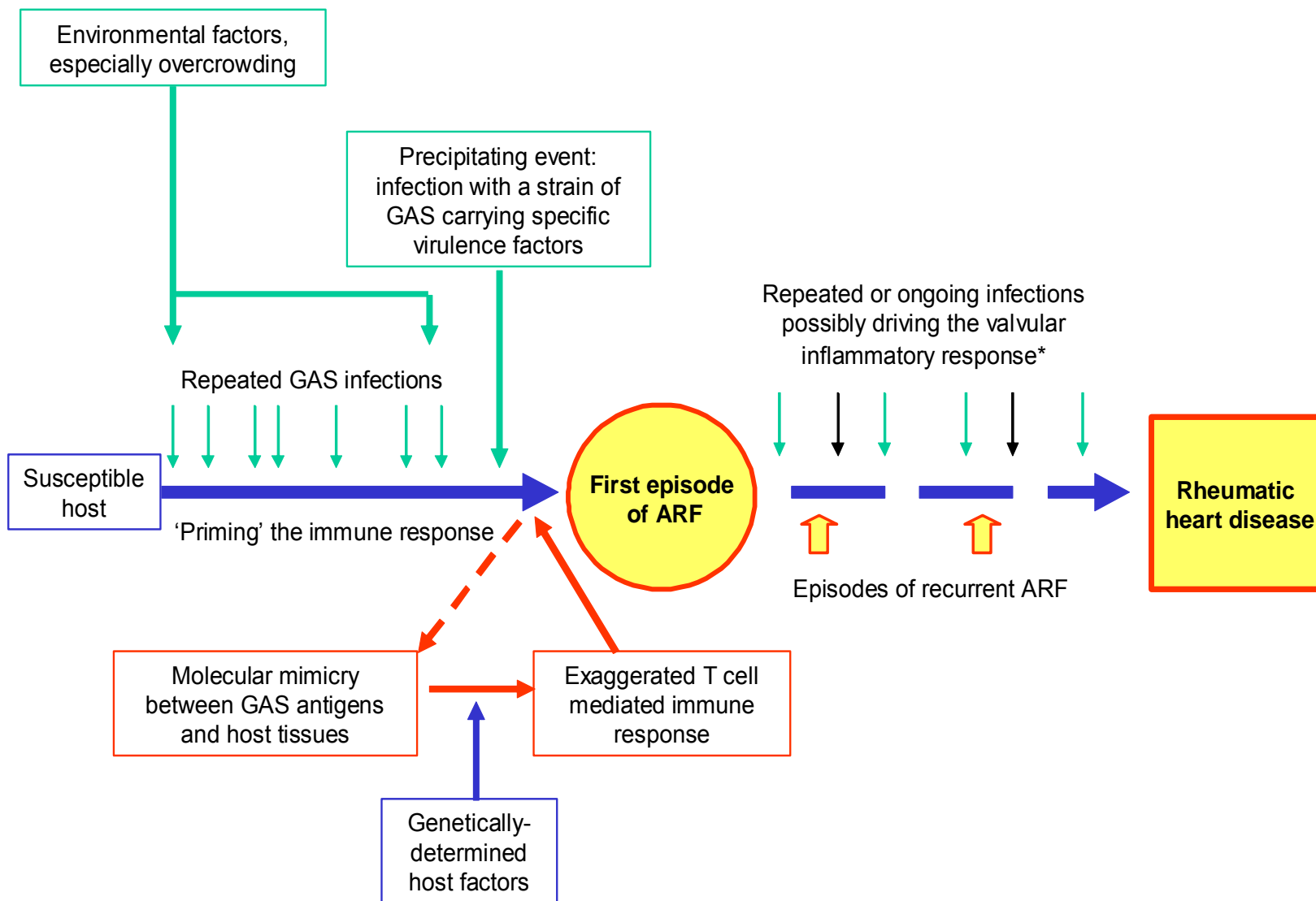


# **Echocardiographic Diagnosis of Rheumatic Heart Disease**

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GAS, group A streptococcus  
 \*GAS and possibly other infections<sup>143</sup>

# 3 pillars of Rheumatic fever knowledge

1. Without Group A streptococcus there would be no rheumatic fever
2. Recurrences of ARF can be prevented by the continuous administration of Benzathine penicillin
3. Severe RHD shortens life

# Severe RHD – shortens life

## **Without Penicillin**

20 yr mortality from RHD 30-80% most dying before aged 30

USA      Bland & Jones Circulation 1951

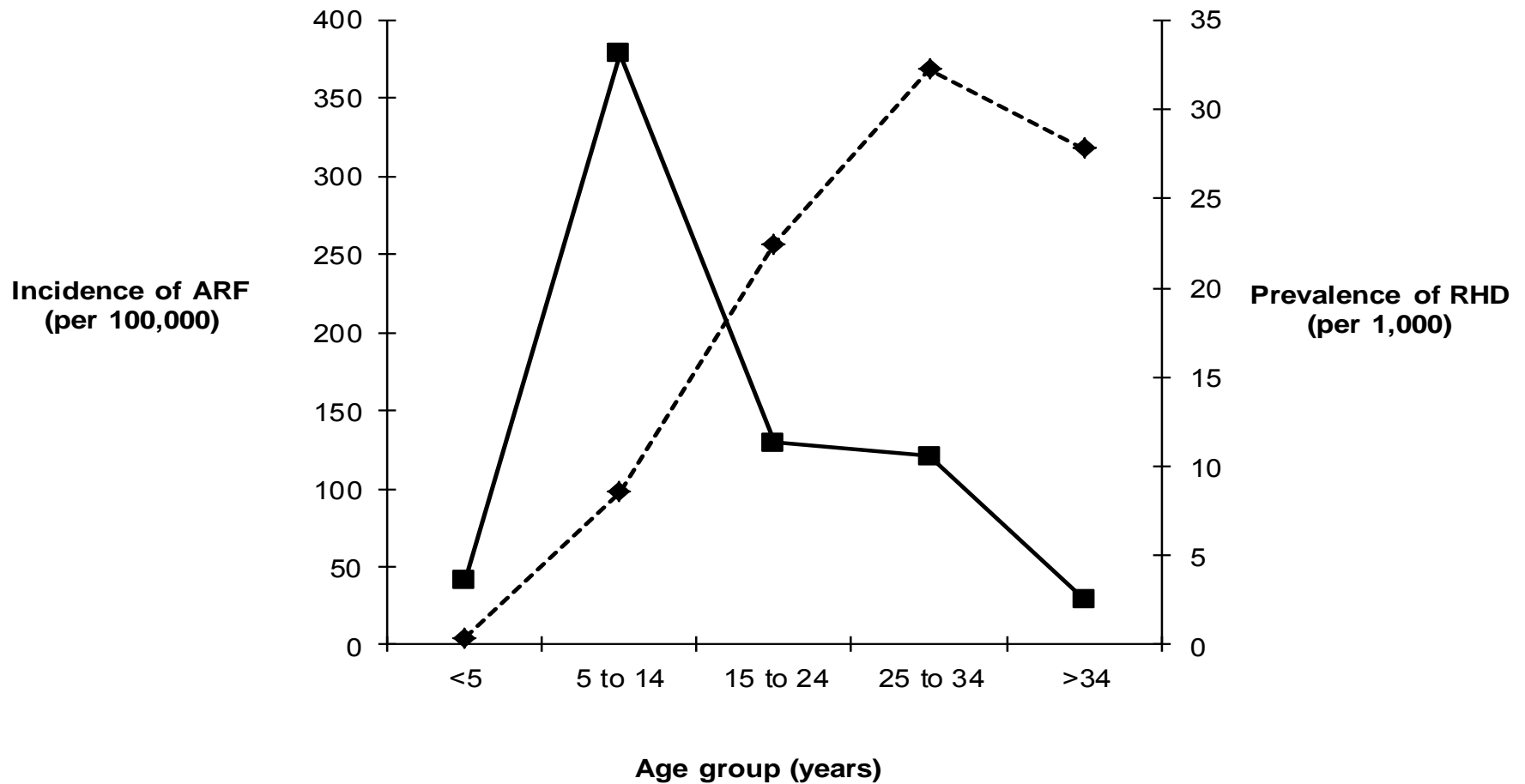
## **Mean age death from RHD < 25 years**

Nigeria   Trop Geogr Med 1981:33:8-13

Ethiopa   Lancet 2006:367:391

India      Indian Heart J 2002:54:54-58

# RHD: disease burden in adulthood



\* Data from the Top End Rheumatic Heart Disease Control Program.

# Pregnancy in pts with RHD and prosthetic heart valves

NWH-GLH surgical data base 1972-1992

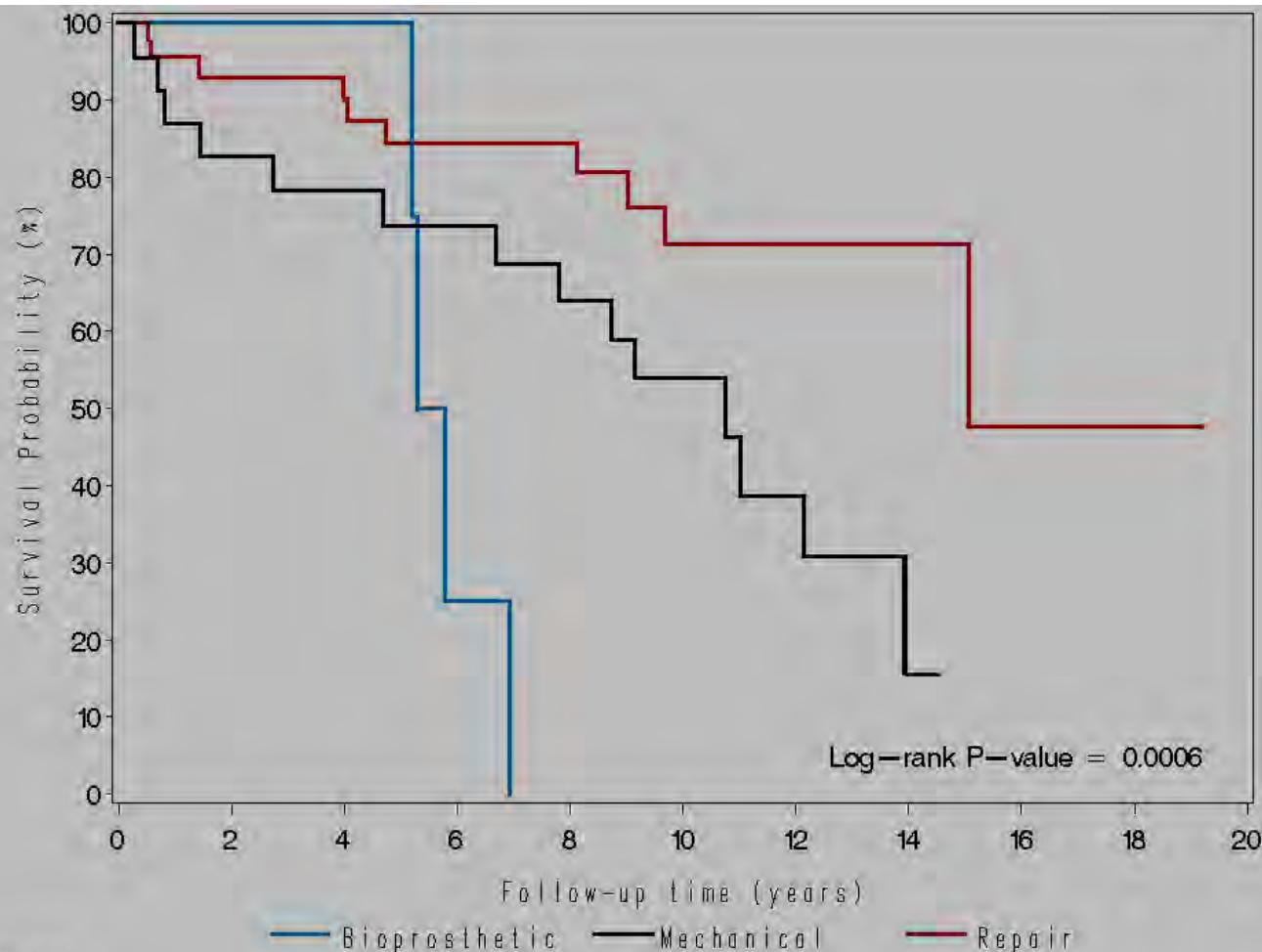
- **Disastrous results with warfarin for fetus** (teratogenic 1st trimester, fetal bleeding 2nd trimester)
- 59% fetal loss mechanical valve vs 7% bio prosthesis **versus disastrous maternal results without warfarin**
- high incidence of maternal valve complications (20%) if change to heparin including thromboembolism, maternal death, thromboembolism and major post partum haemorrhage

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# Freedom from all late valve related events



**At 14 yrs**

**Repair 71%**

**MVR 15%**

**Bioprosthetic 0%**



# Untreated RHD in childhood

- 44-80% of children untreated ARF & RHD progress to cardiac failure requiring medical and surgical treatment within 20 years  
Bland & Jones Circulation 1951: 4:836-41  
Cohn & Lingg JAMA1943:121:1-8
- ARF Recurrences lead to progressive worsening of RHD
- Hence importance of secondary prophylaxis

# Natural history of RHD with good secondary prophylaxis

**No, mild or moderate carditis:** very good

- no detectable heart disease after 10 years

**severe RHD**

- poor prognosis
- valves too damaged, cardiac muscle failure

Tompkins et al J Chronic disease 1972:45:543-551

Feinstein et al Annals Int Med 1964: 60 (suppl)87-123

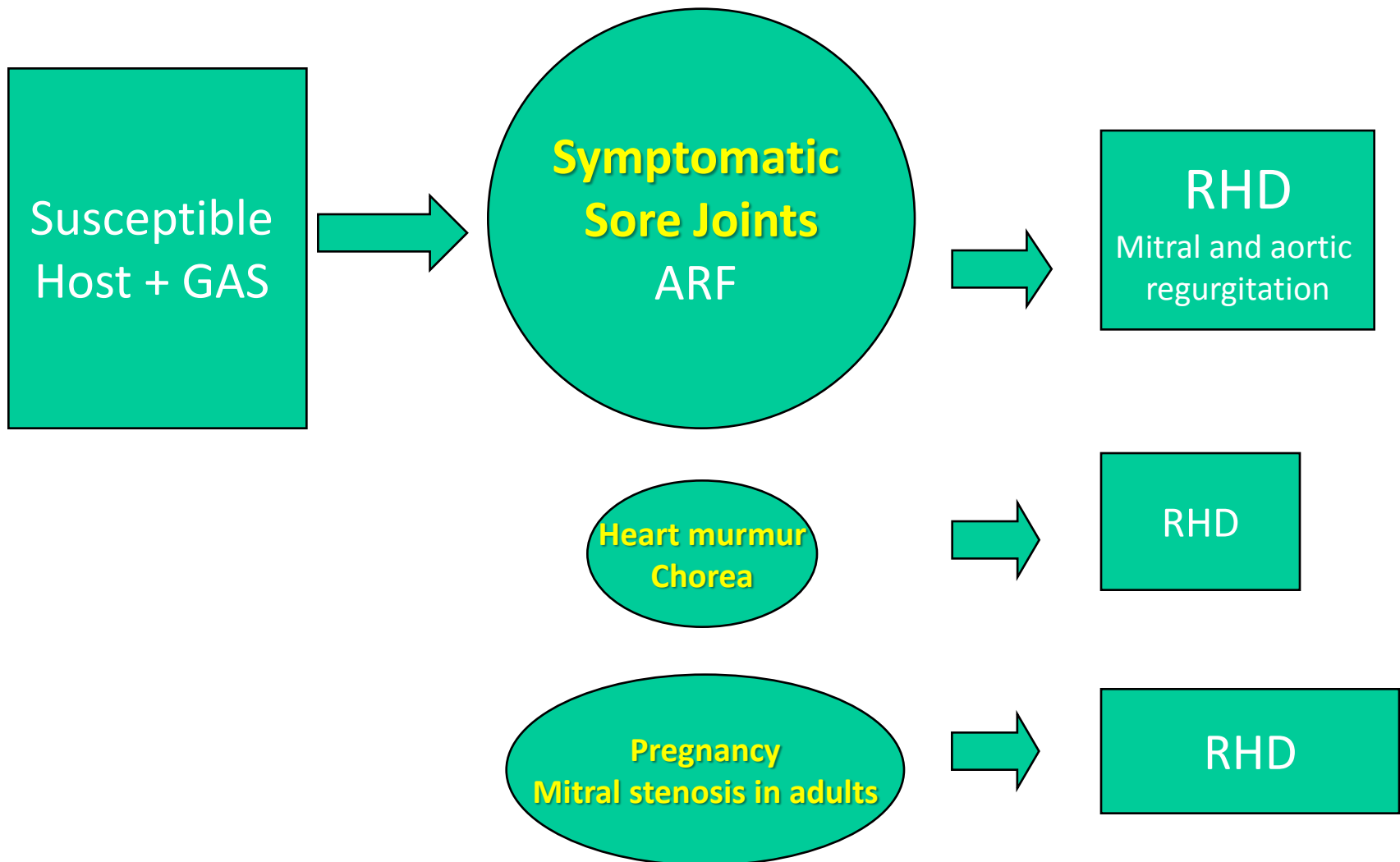
Majeed et al J Clin Epidemiol 1992:45:871-875

Kaseem et al Indian J Pediatr 1995:62: 712-723

# 40% of adults presenting with RHD never had an episode of ARF

- Tairawhiti, New Zealand 37%
- Why can ARF be silent ? - when there are no joint symptoms
  - because mild moderate and even severe RHD is **asymptomatic**. Only when get symptoms of cardiac failure (breathlessness etc)

# Pathogenesis of RHD past 50 years – USA model



# Evidence of clinically silent episodes of rheumatic fever

## Presentations with RHD

Child or adult heart murmur - uncommon

Indolent carditis – uncommon

Acute on chronic ARF - frequent

Chorea - common

Bacterial endocarditis in adults and children (18% GLH-SCH series) – not rare

Pregnancy and mitral stenosis (or CCF with AR/MR) – common

Older Adults with RHD/CCF – literature 40%

## Stroke in adults – uncommon

TOTAL: rheumatic fever without classical arthritis/ARF is common

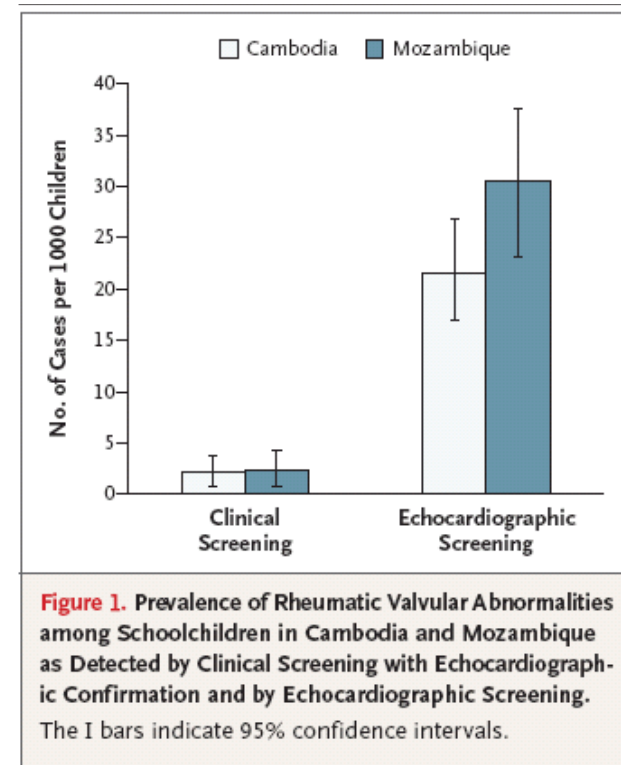
# Bland and Duckett Jones

*Circulation 1951*

- 1000 Pts followed from 1928 for 20 years
  - 347 with no murmur with ARF
- 53% (154/347) developed mitral stenosis (most without a recognized recurrence of RF)
- Seems very likely that these patients had subclinical carditis with mitral regurgitation that evolved to mitral stenosis

# Portable echocardiography in RHD

screening for secondary prevention / early detection

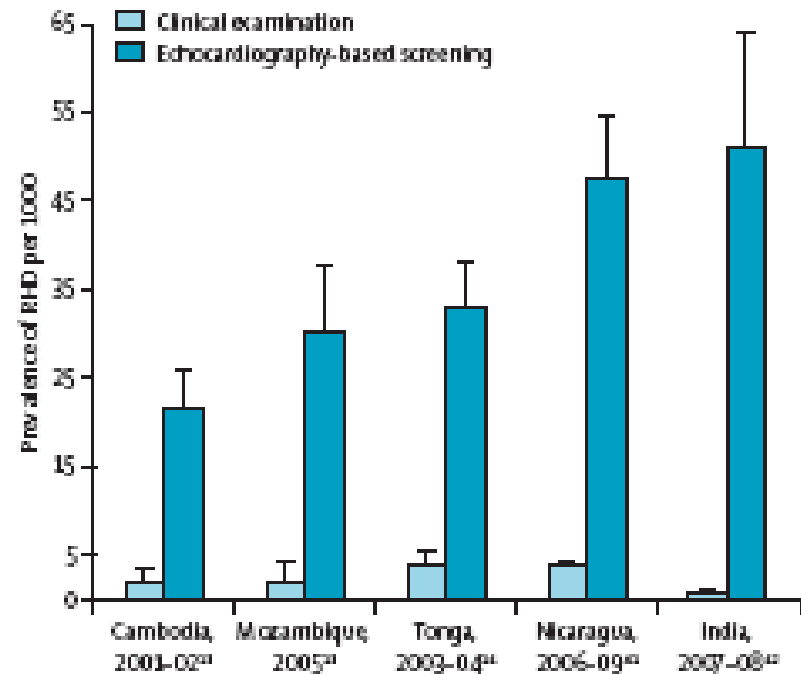


# Challenges of Screening for RHD using echo

## Different criteria used

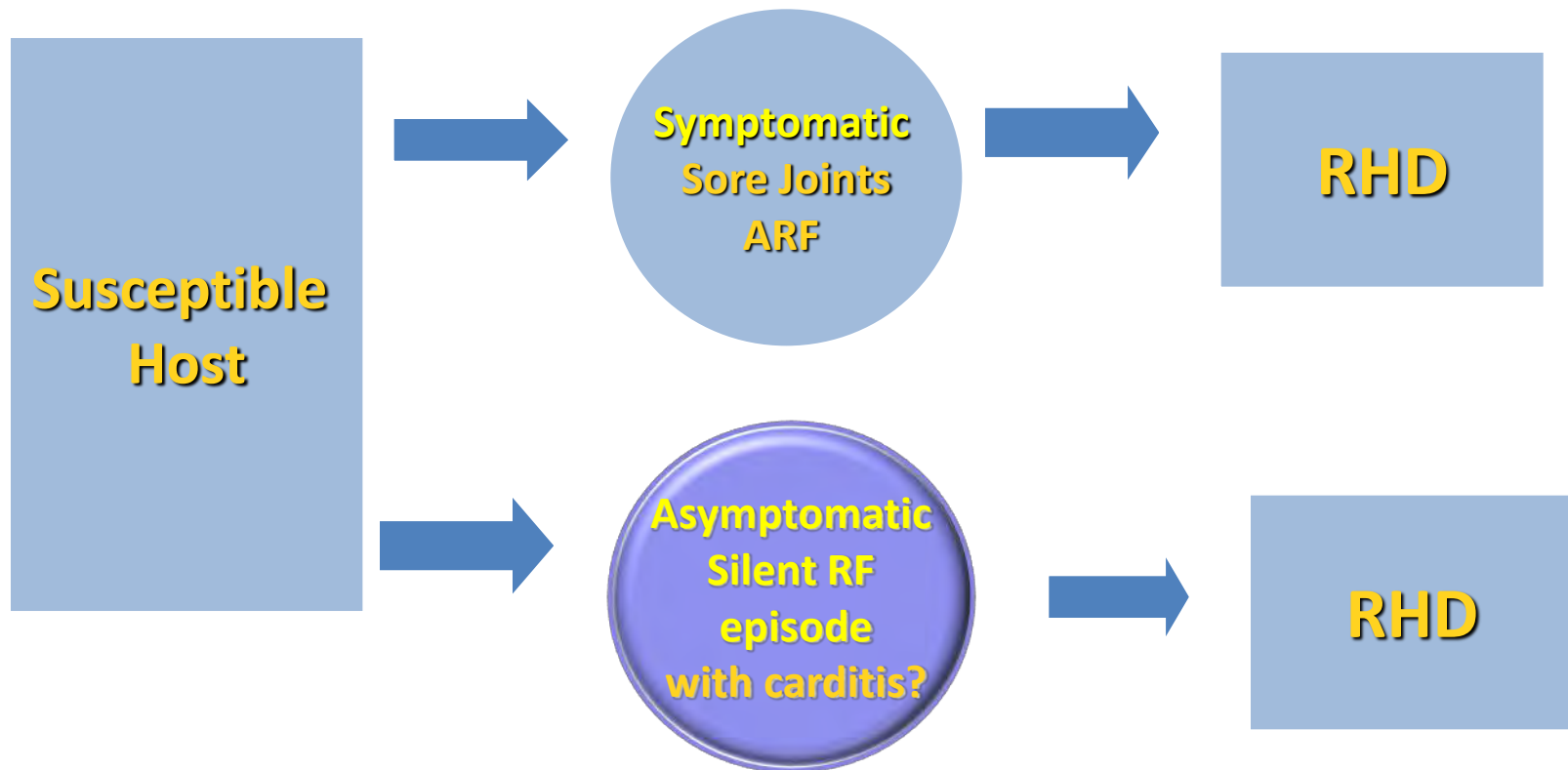
- NIH at Lancefield 2006
- WHO 2004
- Countries adapted

Over-diagnosis ?





# Pathogenesis of ARF to RHD



# Optimising echocardiographic screening for rheumatic heart disease in New Zealand: not all valve disease is rheumatic

Rachel H. Webb,<sup>1,2</sup> Nigel J. Wilson,<sup>1</sup> Diana R. Lennon,<sup>2,3,4</sup> Elizabeth M. Wilson,<sup>2</sup> Ross W. Nicholson,<sup>3</sup> Tom L. Gentles,<sup>1</sup> Clare P. O'Donnell,<sup>1</sup> John W. Stirling,<sup>1</sup> Irene Zeng,<sup>5</sup> Adrian A. Trenholme<sup>3</sup>

<sup>1</sup>Green Lane Paediatric and Congenital Cardiology Department, Starship Children's Hospital; <sup>2</sup>Paediatric Infectious Diseases, Starship Children's Hospital; <sup>3</sup>KidzFirst Children's Health, Counties Manukau District Health Board; <sup>4</sup>Department of Paediatrics, School of Population Health, University of Auckland; <sup>5</sup>Department of Cardiac Physiology, Green Lane Cardiac Services, Auckland City Hospital, Auckland, New Zealand

# Screening for previously undiagnosed RHD in NZ

2007- 2012

3,700 decile 1-2 students

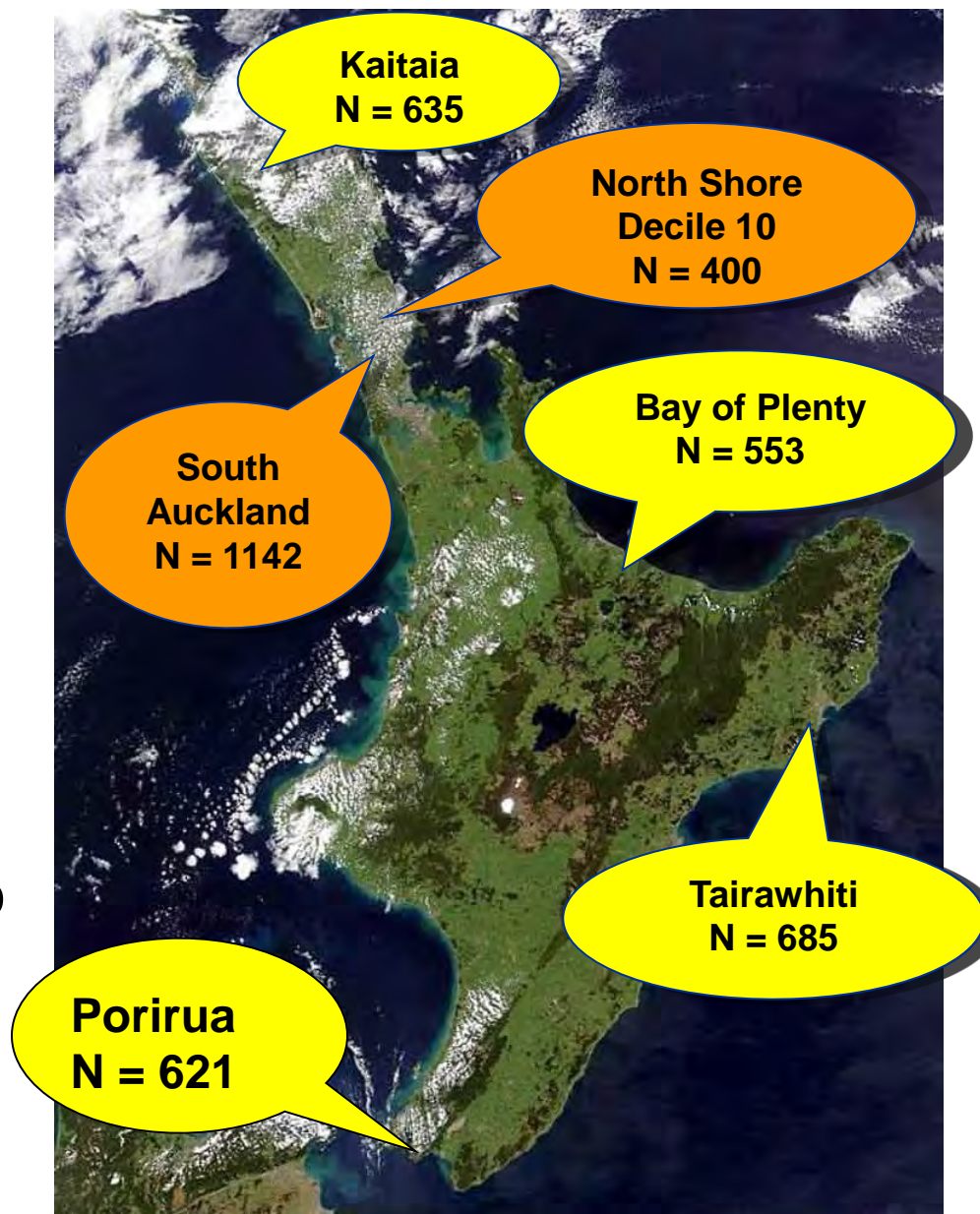
400 decile 10 students

**Auckland – research funding**

non Auckland DHBs

Paediatrician – Public Health –  
Nursing - Cardiology partnership

+ good community buy in



# NZ numbers: RHD echo findings

High prevalence regions				
	Heart Surgery		BPG	Possibles/Borderline
South Auckland	1142	2	25 (2.4%)	30
Tairāwhiti	685	1	8 (1.1%)	19
Bay of Plenty	553		3 (0.5%)	15
Kaitiaia	635	1	5 (0.8%)	16
Porirua (WHF)	621		8 (1.3%)	14
	<b>3665</b>			
Low prevalence regions				
North Shore (low prev)	400			2

# **Challenges for RHD diagnosis**

## **Need for Standardization of diagnostic criteria**

**Aim:** to define the minimal diagnostic criteria for RHD

**Evidence based**

should allow more consistent identification of individuals  
with RHD without a clear history of ARF



# International Standardisation of Echocardiographic Diagnosis of RHD

Bo Remenyi, Nigel Wilson and Jonathan Carapetis



**WORLD HEART  
FEDERATION®**



# Evidence -based guidelines for echo diagnosis of RHD

- An international advisory group was formed in 2009
- 21 investigators from six continents
- Web based system for echo review
- Combined their clinical experience with detailed systematic literature review



# World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease—an evidence-based guideline

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*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*



# WHF Echo criteria for RHD

## Echo criteria for children $\leq 20$ years of age

### **Definite RHD (either A, B, C or D):**

- A) Pathological MR and at least two morphological features of RHD of the MV
- B) MS mean gradient  $\geq$  to 4 mmHg (NB – exclude congenital MV anomalies)
- C) Pathological AR and at least two morphological features of RHD of the AV (NB – exclude bicuspid aortic valve and dilated aortic root)
- D) Borderline disease of both the aortic and mitral valves as defined below\*

### **Borderline RHD (either A, B or C):**

- A) At least two morphological features of RHD of the MV without pathological MR or MS
- B) Pathological MR
- C) Pathological AR

### **Normal Echocardiographic findings (all A, B and C):**

- A) MR that does not meet all four Doppler criteria (Physiological MR)
- B) AR that does not meet all four Doppler criteria (Physiological AR)
- C) An isolated morphological feature of RHD of the MV or the AV (e.g. valvar thickening) without any associated pathological stenosis or regurgitation

# RHD Echo screening 2007-2012 : Impact ?

Echocardiography has shown high levels of RHD prevalence, has increased **advocacy** for better RHD control in those regions

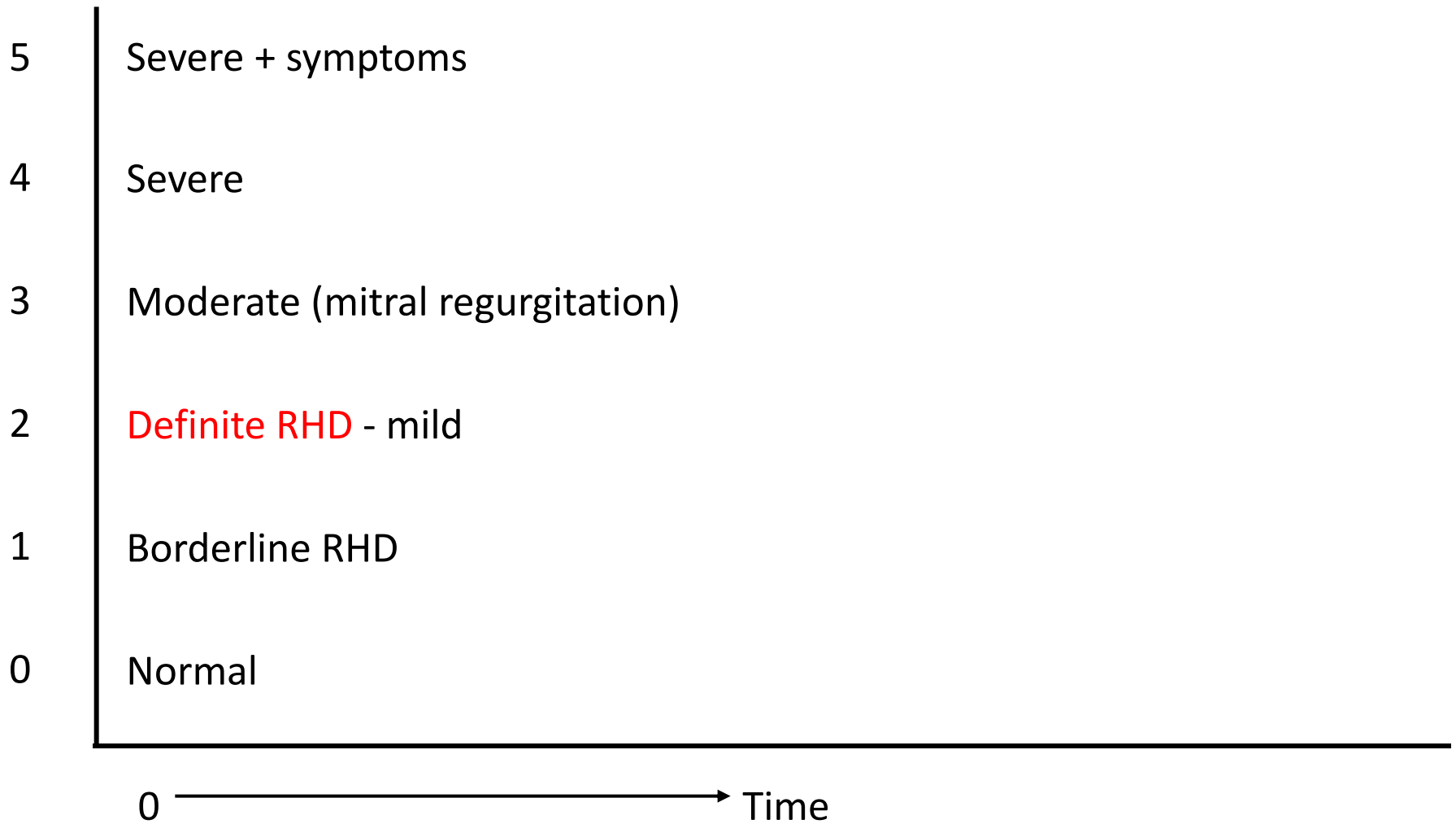
Impact on reduction in disease burden has not been evaluated except in some regions

- e.g. Tonga

# Natural history of echocardiographically detected RHD

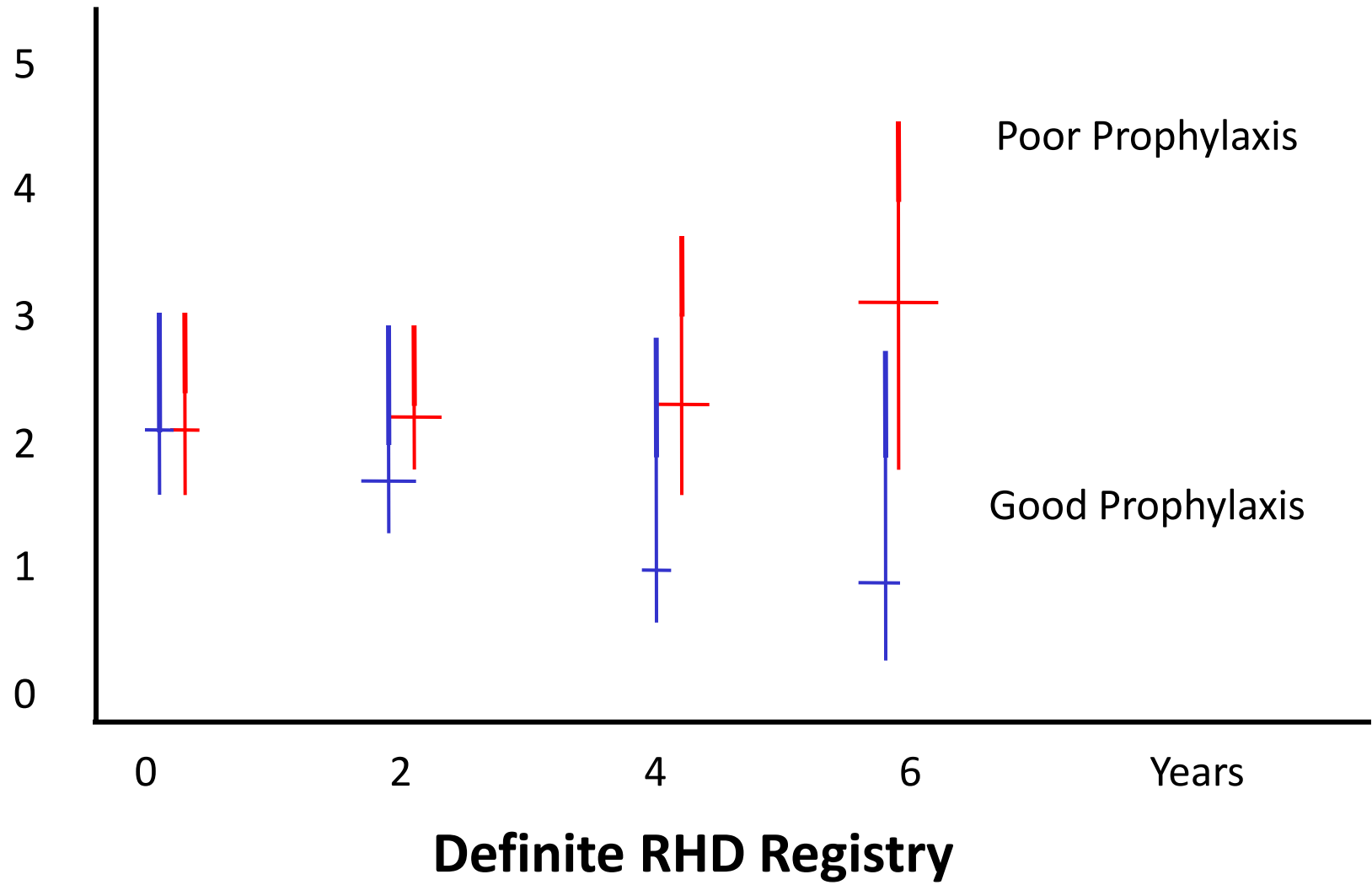
- Emphasis **has been** on mild/minor abnormalities
- **Emphasis must now shift to Definite RHD by WHF criteria**
  - the cardiology community need to show that latent definite RHD is the same as definite RHD following episode (s) of ARF in terms of
    - § 1) progression of RHD if untreated
    - § (and less likely) 2) recurrence of ARF

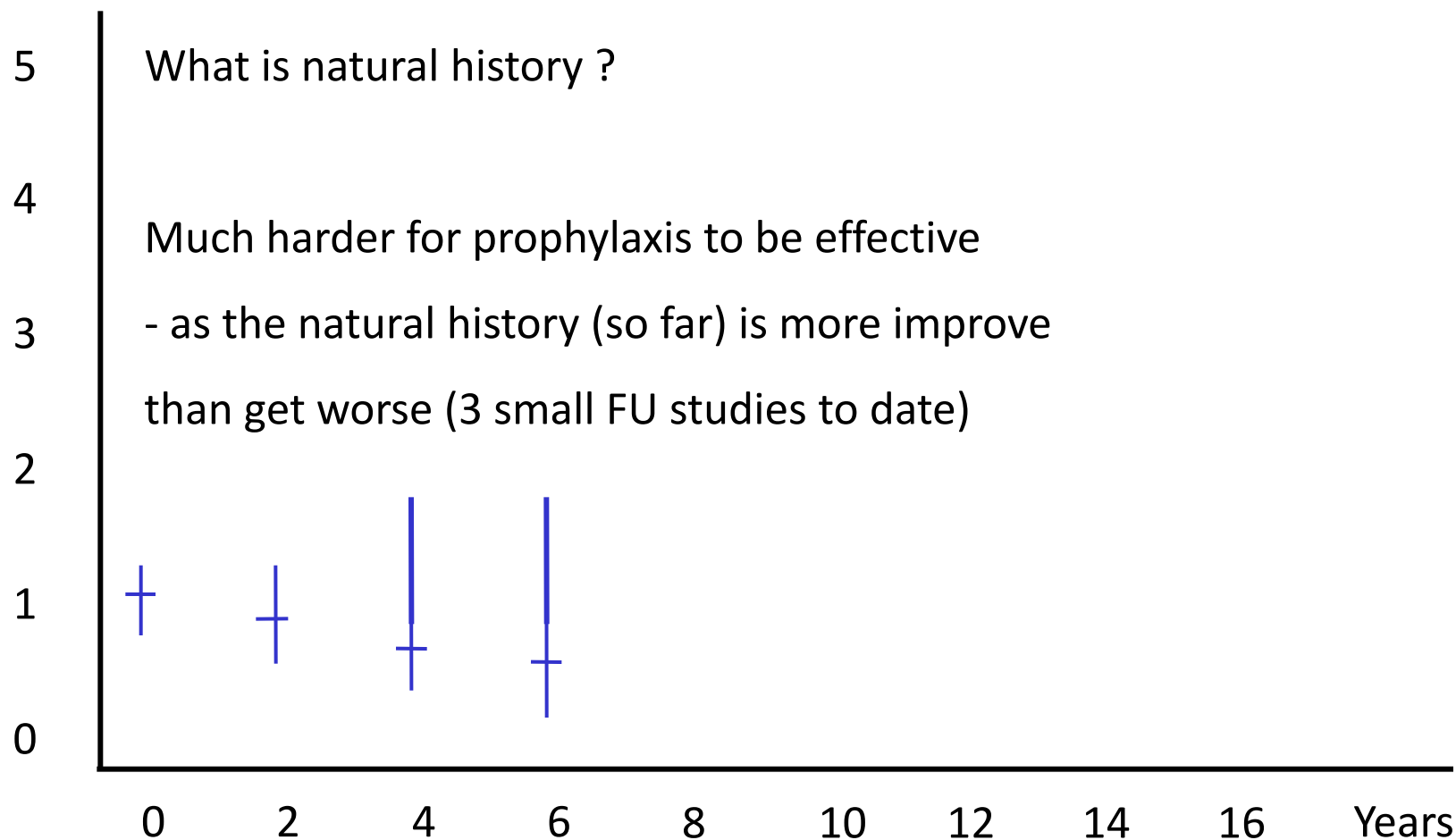
# Scale of RHD



**Definite RHD Registry**

# Prediction ?





## Borderline RHD Registry

# Summary and conclusions

## **Severe RHD shortens life expectancy**

- Morbidity high
- 600-800 admissions per year
- nearly 200 deaths per year

## **40% of adults with severe RHD do not have a history of ARF**

- echo screening for RHD is feasible but internationally there has been over-diagnosis
- reproducibility & accuracy improved by WHF criteria

# Conclusions: echo screening

Advocacy for RHD increased but internationally there has been over-diagnosis

- reproducibility & accuracy improved by WHF criteria
- new insights into pathogenesis of RHD **but need to define disease progression of definite RHD**
  - **they must be linked**

**40% of adults with severe RHD do not have a history of ARF**



# **New Zealand: conclusions**

## **Severe RHD shortens life expectancy**

Morbidity high

600-800 admissions per year

nearly 200 deaths per year

echo screening for RHD is feasible in New Zealand

Over 1% of decile 1-2 children in high prevalence ARF regions have definite RHD

MOH should consider further echo screening in targetted sites – there are decades of severe RHD even if no more ARF from now

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