Rheumatic Heart Disease in New Caledonia

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For the New Caledonian
Working Group on Acute Rheumatic Fever and Rheumatic Heart Disease

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New Caledonia, South Pacific French Territory

350 km x 70
New Caledonia

- 256,275 inhabitants (2009 Census)
- Archipelago in South Pacific
- 1200 kilometers (750 mi) east of Australia and 1500 kilometers (930 mi) northwest of New Zealand, 5000 km (3500 mi) west from French Polynesia
- A main island (Grande Terre), Loyalty Islands, and several smaller islands
- 52% of population in Noumea and suburbs, 35% in others districts on main island and 13% in the other islands
- Half population < 30 yo and 25% < 15 yo
- 44% population belong to the “kanak” ethnicity (indigenous Melanesian), 34% to the European community, 12% to Polynesian community, 2.5 Indonesian community, 2% to Asiatic community, and 7.5 others

1. One of late complications of Acute Rheumatic Fever (ARF)

2. ARF secondary to Group A streptococci (GAS)

3. Most often through repeated untreated pharyngitis, but perhaps also from cutaneous infections (impetigo)

4. Auto immune disease, directed against joints, heart (valves most often, but also myocardium, pericardium)

5. Very dangerous disease, associating insidious and longtime asymptomatic heart lesions, in patients not easily consulting, often at advanced levels

6. Cardiac surgery often proposed, with their complications

7. Disappearance from developed countries within the 50 last years, including North Africa

8. High prevalence in developing countries (Africa, Asia, Central America, South Pacific), or in high income developed countries with specific subpopulations (some South Pacific populations)

9. Great difficulty to fight this severe disease in developing countries, because of lack of health care resources for prevention, detection and treatment

10. Echocardiography is the only detection method of RHD

Rheumatic Heart Disease: What is less known

1. Statistics and Epidemiology of RHD in New Caledonia

2. Exact risk factors: environment, genetics, GAS specificity

3. Exact echocardiographic criteria for diagnosis of RHD, either in asymptomatic children for systematic detection in high risk populations, or in patients presenting with symptoms of ARF or any other valvular disease

4. How to improve global prognosis: primary prevention, secondary prevention, and systematic detection of RHD in all children in high risk areas

Bongani Mayosi, Natural history, screening, and management of rheumatic heart disease. UpToDate, Dec 2012.
New Caledonia, a model of a developing country disease: Acute Rheumatic Fever in a high income country

- Disappearance of ARF in high income countries

- Still a high prevalence disease in developing countries with low outcomes: Africa, Asia and Pacific Islands (Fiji, Tonga, Samoa, Vanuatu, Wallis and Futuna…)

- And still a high prevalence in high income countries, in some specific (often native) populations: Australian Aborigine, New Zealand Maoris, New Caledonian Melanesians, French Polynesia, Wallis and Futuna population living in New Caledonia


Adapted with permission from N. Baroux et al. High prevalence of Rheumatic Heart Disease in schooled-age children detected by Echocardiography Screening in New Caledonia. In Press. J Pediatric Child Health
Development of an epidemiologic, prevention and management program for RHD since 2007 in New Caledonia

- Health and Social Agency NC (Agence Sanitaire et Sociale NC)

- A government public agency

- In charge of epidemiologic survey, prevention, and management of some major health issues (Diabetes, Tobacco, Cannabis, Obesity, Contraception, most frequent cancers, tuberculosis, ARF and RHD, …)

- In 2007, study set up for RHD: annual exhaustive RHD screening for school-aged children in 4th year primary school (9-10 yo) in whole NC

Adapted with permission from N. Baroux et al. High prevalence of Rheumatic Heart Disease in schooled-age children detected by Echocardiography Screening in New Caledonia. In Press. J Peadiatric Child Health
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Directions Provinciales ASS, (Sud, Nord, Iles)
CAFAT (Local Social Security)
Centres Médicaux-Sociaux de NC,
Médecins libéraux
- Docteur Jean-Michel MEUNIER, cardiologue libéral
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- Docteur Baptiste NOEL, cardiologue CHT
- Docteur Gwendoline PHILLIPPEAU, cardiologue CHT
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- Docteur Eric D’ORTENZIO, médecin épidémiologiste INSTITUT PATEUR
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- Docteur Pierre BOURGOIN, cardiologue libéral
- Madame Corinne ROBILLARD Infirmière prestataire ASS NC
By far the most important form of acquired heart disease in children and young adults living in developing countries (inhabited by 80 percent of the world’s population); about a quarter of all patients with heart failure in endemic countries


- 62-78 million individuals worldwide, with a potential death rate of 1.4 M from RHD and its complications for others

- High prevalence rates in different countries

### Rheumatic Heart Disease worldwide

**Prevalence rates in developing countries**

<table>
<thead>
<tr>
<th>Country</th>
<th>Prevalence rate (‰)</th>
<th>Method</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cambodia</td>
<td>21.5 (in 6-17 yo)</td>
<td>Echo</td>
<td>Marijon, NEJM 07</td>
</tr>
<tr>
<td>Mozambique</td>
<td>30.4 (in 6-17 yo)</td>
<td>Echo</td>
<td>Marijon, NEJM 07</td>
</tr>
<tr>
<td>Nicaragua</td>
<td>48 (34 in urban, 80 non urban, 5-15 yo) (20 in 20-35 yo)</td>
<td>Echo</td>
<td>Paar, AJC 2010</td>
</tr>
<tr>
<td>Kenya</td>
<td>27</td>
<td>Echo</td>
<td>Anabwani, EAMJ 1996</td>
</tr>
<tr>
<td>Average in school age children</td>
<td>40</td>
<td></td>
<td>Bongani, Uptodate, 2012</td>
</tr>
<tr>
<td>South Pacific</td>
<td>7.6</td>
<td></td>
<td>WHO, 2005</td>
</tr>
</tbody>
</table>

## Rheumatic Heart Disease Prevalence rates in In South Pacific (high income countries)

<table>
<thead>
<tr>
<th>Country</th>
<th>Prevalence rate (‰)</th>
<th>Method</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fiji</td>
<td>4.1 (5-15 yo)</td>
<td>Echo</td>
<td>Steer, J Heart Valve Dis, 2009</td>
</tr>
<tr>
<td>Tonga</td>
<td>33.2</td>
<td>Echo</td>
<td>Carapetis, NCPCM, 2008</td>
</tr>
<tr>
<td>New Caledonia</td>
<td>8.9 (13.4 in melanesian, 5 in polynesian)</td>
<td>Echo</td>
<td>Baroux, In Press, JPCH</td>
</tr>
<tr>
<td>French Polynesia</td>
<td>22 (18 school age child.)</td>
<td>Echo</td>
<td>Costes. PF 2006</td>
</tr>
<tr>
<td>American Samoa</td>
<td>0 on 140 (7-18 yo)</td>
<td>Echo</td>
<td>Barnes, PHMT 2011</td>
</tr>
<tr>
<td>Global for South Pacific</td>
<td>7.6</td>
<td>Echo</td>
<td>WHO, 2005</td>
</tr>
</tbody>
</table>

Development of an epidemiologic, prevention and management program for RHD since 2007 in New Caledonia

1. High Prevalence of Rheumatic Heart Disease in School Children detected by Echocardiography Screening in New Caledonia. 2008-2010
Baroux N (IPNC), Rouchon B (ASSNC), Huon B, Germain A (ASSNC), Meunier JM, D’Ortenzio E (IPNC).

2. Ethnic disparities in the risk of rheumatic heart disease among schoolchildren in New Caledonia: a case-control study
D’Ortenzio E. (IPNC), Baroux N. (IPNC), Rouchon B. (ASSNC)
Submitted for publication
Main results:

1. Prevalence of RHD in school children (9-10 yo) in New Caledonia: 8.9/1000 (IC 95% 7.3-10.6)

2. Higher prevalence on the Main Inland (“Grande Terre”) outside Noumea and its suburbs: 13.7 / 1000 (IC 95%, 9.8-17.5) and in Loyalties Islands: 14.6 / 1000 (IC 95%, 8.4-20.9) than in Noumea and suburbs: 5.8 / 1000 (IC 95% 4.1-7.5)

3. RHD more prevalent among Melanesian children (13.5 / 1000; IC 95%, 10.9-16.1) than in European children (1.8 / 1000; IC 95%, 0.4-3.1)

→ Confirmation of this important Public Health Problem of RHD in New-Caledonia
RHD prevalence rate in NC in 2010

From Agence Sanitaire et Sociale NC 2010 Annual report (website)
Comments

1. Prophylactic antibiotic therapy and regular medical follow up initiated if RHD validated by child’s general practitioner.

2. Between 2008 and 2010, 13,635 children, fourth year primary school, 93.5% had an echocardiography screening at school, 4.5% were absent and 1.9% had no signed consent forms. Mean age of screened children: 9.6 ± 0.6 yo (7.0 – 13.6)

3. Possible underestimation of global prevalence rate: 14% unschooled children, and results concern 10 yo children

4. Difficulty cases: mild echocardiographic findings: normal or abnormal
1. Increase of screening rate in grade 4, and to cardiologic consult in 2011

2. 300 children screened in 2011, like in 2010, 600 in 2009

3. 40 children with RHD among 4308 screened, added to the 27 already known: (16‰ for 9 yo children)

Main conclusions:

1. Study Objective: to identify risk factors of RHD in the school children 2010 subpopulation (9-10 yo) of the previous study in New Caledonia

2. Case control observational study

3. 40 cases and 145 controls included

4. Average age: 9.8 yo and sex-ratio M/F 1

5. Melanesian children are 6.5 more at risk to develop RHD than non-melanesian children (single risk factor by multivariate conditional logistic regression analysis)
Main conclusions:

6. Hypotheses, to explain this discrepancy:

1- A way of life favoring untreated *Group A streptococci* (GAS),

2- A genetic susceptibility or more probably the association of both conditions

These results should encourage 1) primary prevention, 2) treatment of GAS pharyngitis and detection of RHD, in these populations at high risk

A genetic study could either exclude or confirm the hypothesis of the genetic susceptibility of RHD in the melanesian population
2. Ethnic disparities in the risk of rheumatic heart disease among schoolchildren in New Caledonia: a case-control study.
D’Ortenzio E. (IPNC), Baroux N. (IPNC), Rouchon B. (ASSNC)
Submitted for publication

4371 children underwent echocardiography screening* in 2010

4326 children without RHD

45 children were diagnosed with RHD and were selected for the study

200 control subjects randomly selected*  

42 cases contact details available

145 control contact details available

1 excluded†  

1 refusal

40 case-subjects enrolled  

145 control-subjects enrolled
2. Ethnic disparities in the risk of rheumatic heart disease among schoolchildren in New Caledonia: a case-control study. D’Ortenzio E. (IPNC), Baroux N. (IPNC), Rouchon B. (ASSNC) Submitted for publication

![Graph showing proportions of cases among Melanesian and Non-Melanesian schoolchildren with different income levels.](image)
2. Ethnic disparities in the risk of rheumatic heart disease among schoolchildren in New Caledonia: a case-control study.  
D’Ortenzio E. (IPNC), Baroux N. (IPNC), Rouchon B. (ASSNC)  
Submitted for publication

Comments

1- 74% melanesian, 15% polynesian, 10% mixed, no european

2. 62% in rural areas, with a tribute way of life,

3. 46% household income < 1600 $ US/month (NS in melanesian)

4. Household income in melanesian NS: way of life probably more important than economic status?  
Genetic susceptibility in Melanesian population? (HLA class II alleles)

5. Proportion of people at risk for ARF seems unchanged in all ethnic groups, except in most developed countries where GAS infections are decreased (Bryant et al: Some of the people, some of the time: susceptibility to acute rheumatic fever. Circulation 2009, 119(5):742-753)
Echocardiographic Criteria of RHD


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

Echocardiographic Criteria of RHD different if:

- patient presenting with symptoms of RHF, or valvular disease
- or systematic screening in high risk population (‘borderline RHD’ echocardiographic category established to improve sensitivity of the test at the expense of specificity in high risk populations)


Echocardiographic Criteria of RHD

For systematic screening, inadequacy of previous Echo WHO criteria for RHD have been advocated:


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

Echocardiographic Criteria of RHD
NC cardiologists working group with ASSNC
before 2011 WHF GDL

Table 1. Echocardiographic criteria for confirmation of rheumatic heart disease, New Caledonia, 2008-2010,

<table>
<thead>
<tr>
<th>Criteria for rheumatic mitral regurgitation</th>
</tr>
</thead>
<tbody>
<tr>
<td>(a) Stenosis</td>
</tr>
<tr>
<td>OR</td>
</tr>
<tr>
<td>(b) Two of Doppler-detected signs:</td>
</tr>
<tr>
<td>- Regurgitation identified in at least two planes</td>
</tr>
<tr>
<td>- Regurgitant jet greater than 2 cm</td>
</tr>
<tr>
<td>- Pansystolic regurgitation</td>
</tr>
<tr>
<td>- Mosaic color jet with peak velocity greater than 2.5m/s</td>
</tr>
<tr>
<td>Accompanied by one of the following signs:</td>
</tr>
<tr>
<td>- Valvular thickening &gt; 5 mm in TM mesodiastolic</td>
</tr>
<tr>
<td>- Abnormal mobility of valve layers</td>
</tr>
<tr>
<td>- Abnormal subvalvular thickening</td>
</tr>
<tr>
<td>- Thickened “dog leg” mitral valve leaflet</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Criteria for rheumatic aortic regurgitation</th>
</tr>
</thead>
<tbody>
<tr>
<td>(c) Stenosis</td>
</tr>
<tr>
<td>OR</td>
</tr>
<tr>
<td>(d) All Doppler-detected signs:</td>
</tr>
<tr>
<td>- Regurgitant jet greater than 1 cm</td>
</tr>
<tr>
<td>- Pandiastolic regurgitation</td>
</tr>
<tr>
<td>- A mosaic color jet with peak velocity greater than 2.5m/s</td>
</tr>
</tbody>
</table>

Adapted with permission from N. Baroux et al. High prevalence of Rheumatic Heart Disease in school-age children detected by Echocardiography Screening in New Caledonia. In Press. J Paediatric Child Health
Echocardiographic Criteria of RHD: WHF 2012 guidelines = “Bangkok criteria”

Box 1 | 2012 WHF criteria for echocardiographic diagnosis of RHD

Echocardiographic criteria for individuals aged ≤20 years
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 ≥4 mmHg*
- C) Pathological AR and at least two morphological features of RHD of the AV†
- D) Borderline disease of both the AV and MV§
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 of A, B, C, and D):
- A) MR that does not meet all four Doppler echocardiographic criteria (physiological MR)
- B) AR that does not meet all four Doppler echocardiographic criteria (physiological AR)
- C) An isolated morphological feature of RHD of the MV (for example, valvular thickening) without any associated pathological stenosis or regurgitation
- D) Morphological feature of RHD of the AV (for example, valvular thickening) without any associated pathological stenosis or regurgitation

Echocardiographic criteria for individuals aged >20 years
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 ≥4 mmHg*
- C) Pathological AR and at least two morphological features of RHD of the AV, only in individuals aged <35 years†
- D) Pathological AR and at least two morphological features of RHD of the MV
*Congenital MV anomalies must be excluded. Furthermore, inflow obstruction due to nonrheumatic mitral annular calcification must be excluded in adults. †Bicuspid AV, dilated aortic root, and hypertension must be excluded. ‡Combined AR and MR in high prevalence regions and in the absence of congenital heart disease is regarded as rheumatic.
Abbreviations: AR, aortic regurgitation; AV, aortic valve; MR, mitral regurgitation; MS, mitral stenosis; MV, mitral valve; RHD, rheumatic heart disease; WHF, World Heart Federation.
Echocardiographic Criteria of RHD: WHF 2012 guidelines = “Bangkok criteria”

Box 2 | Criteria for pathological regurgitation

Pathological mitral regurgitation
(All four Doppler echocardiographic criteria must be met)
- Seen in two views
- In at least one view, jet length ≥2 cm*
- Velocity ≥3 m/s for one complete envelope
- Pan-systolic jet in at least one envelope

Pathological aortic regurgitation
(All four Doppler echocardiographic criteria must be met)
- Seen in two views
- In at least one view, jet length ≥1 cm*
- Velocity ≥3 m/s in early diastole
- Pan-diastolic jet in at least one envelope

* A regurgitant jet length should be measured from the vena contracta to the last pixel of regurgitant color (blue or red).

**Important considerations**

*AMVL* thickness should be measured during diastole at full excursion. Measurement should be taken at the thickest portion of the leaflet, including focal thickening, beading, and nodularity. Measurement should be performed on a frame with maximal separation of chordae from the leaflet tissue. Valve thickness can only be assessed if the images were acquired at optimal gain settings without harmonics and with a frequency $\geq 2.0$ MHz.

- Abnormal thickening of the AMVL is age-specific and defined as follows: $\geq 3$ mm for individuals aged $\leq 20$ years; $\geq 4$ mm for individuals aged 21–40 years; $\geq 5$ mm for individuals aged $> 40$ years. Valve thickness measurements obtained using harmonic imaging should be cautiously interpreted and a thickness up to 4 mm should be considered normal in those aged $\leq 20$ years.

- Restricted leaflet motion of either the anterior or the posterior MV leaflet is usually the result of chordal shortening or fusion, commissural fusion, or leaflet thickening.

- Excessive leaflet tip motion is the result of elongation of the primary chords, and is defined as displacement of the tip or edge of an involved leaflet towards the left atrium resulting in abnormal coaptation and regurgitation. Excessive leaflet tip motion does not need to meet the standard echocardiographic definition of MV prolapse disease, as that refers to a different disease process. This feature applies to only those aged $< 35$ years. In the presence of a flail MV leaflet in the young ($\leq 20$ years), this single morphological feature is sufficient to meet the morphological criteria for RHD (that is, where the criteria state “at least two morphological features of RHD of the MV” a flail leaflet in a person aged $\leq 20$ years is sufficient).

- In the parasternal short axis view, the right and noncoronary aortic cusp closure line often appears echogenic (thickened) in healthy individuals and this should be considered as normal.

Abbreviations: AMVL, anterior mitral valve leaflet; AV, aortic valve; MV, mitral valve; RHD, rheumatic heart disease.
Echocardiographic Criteria of RHD: WHF 2012 guidelines = “Bangkok criteria”


Box 4 | Echocardiography machine settings

- Nyquist limits for color-Doppler echocardiography should be set on maximum to avoid overestimation of jet length
- Images for assessment of valvular and chordal thickness should be acquired with harmonics turned off and probes with variable frequency set on ≥2.0 MHz; low frequency settings and harmonics exaggerate valve and chordal thickness
- Gain settings should be adjusted to achieve optimal resolution; images acquired with an excessive gain setting will not be suitable for objective valve thickness measurements
- All other settings (including depth, sector size, and focus) should also be optimized to achieve maximal frame rate (ideally 30–60 frames per second) and resolution
Conclusion (1)

1- High prevalence rate of Rheumatic Heart Disease in New Caledonia, remaining a major health burden. Exact numbers still not known for global population.

2- Existence of a very active control program of ARF and RHD in New Caledonia with the Health and Social Agency (ASSNC) with an active register ++ (prevention and early detection).

3- **Primary prevention**: overcrowding, socio-economic status, urbanization, distance between home and health centre and access to medical service, GAS pharyngitis treatment…

**Secondary Prevention**: IM Extencilline, echo follow up.

Systematic campaigns in progress

**Epidemiologic research**: genetics and specific streptococci (emm)

4- Early detection with systematic echo screening in schools set up since 2007.
Conclusion (2)

5- Still important difficulties in managing RHD in NC
   - imperfect compliance of patients to follow-up
   - primary prevention difficult, although high income country

6- Importance of echocardiography in any screening program using
   Bangkok WHF 2012 guidelines (training ++)

7- Criteria different from those used to diagnose RHD in RHF
   suspicion or follow up, or the cause of a valvular heart disease

8- Discussion: applicability of this methodology in developed low
   income countries with numerous populations and limited health
   care resources? (cf all studies in Africa, India, Central America).
9- Study project in New Caledonia:

- Register of all patients presenting with RHD, either new or known, coming for consult, echocardiography at the hospital (Cardiology, Pediatrics, Internal Medicine)

- More difficult in private practice