Ρευματικές νόσοι και καρδιακές βαλβίδες: τι πρέπει να γνωρίζει ο κλινικός Καρδιολόγος;

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The Rheumatic Heart Disease pathway

**Acute Rheumatic Fever** is an autoimmune response to a group A streptococcus (GAS) bacterial infection.

**Social determinants** such as poor living conditions are key drivers for onset of ARF and RHD.

**Symptoms** such as fever, joint pain, sore throats and rashes are non-specific to the infection.

**Diagnosis** should be based on assessment of symptoms combined with clinical history.

**Misdiagnosis** occurs due to complexity of diagnosis may result in delayed or no treatment.

**Recurrence** rate of ARF is greatest in first year after first episode.

**Rheumatic Heart Disease** occurs when untreated, recurrent ARF episodes cause heart valve damage.
History

• 1898 W. Cheadle – first description of disease
• 1904 Aschoff - histopathological lesions
• 1944 Jones diagnostic criteria
• 1992 AHA modified Jones diagnostic criteria
• 2015 modified Jones diagnostic criteria (low and high risk population)
Epidemiology of RF

- Virtually eliminated from high income countries (mainly in the ageing population) – 3.4/100.000
- In low income countries remains a leading disease of the young - 444/100.000
Risk Factors

• AGE
  • 5-14 ys initial episode
  • Recurrent episodes older children – young adults

• GENDER
  females>>males (unknown reason)

• ENVIROMENTAL FACTORS
  • Household crowding
  • low socioeconomic status
  • Insufficient nutrition
Pathogenesis

- Few weeks after GAS infection of pharynx
- Streptolysin S, O toxic effect
- Autoimmune reaction II against host tissues (molecular mimicry)
Cardiac involvement

- **Pancarditis**
  - Periarteritis nodosa -> lymphocytic inflammation -> coronary artery disease

- **Vascular involvement**
  - Endocarditis
  - 1-2 mm excrescences on the endocardium, subvalvular mechanism

- **Inflammation and phlegmon** of the valves
  - MacCallum patch
  - Erosion -> coagulation and formation of valvular insufficiency or regurgitation
  - Stenosis/insufficiency of the valve.
**Clinical manifestations**

**Acute illness** within weeks (1-5) of a group A streptococcal (GAS) tonsillopharyngitis (or streptococcal pyoderma in patients from tropical regions)

**Major manifestations**
- Carditis and valvulitis (eg, pancarditis) that is clinical or subclinical (50-70%)
- Arthritis (usually migratory polyarthitis predominantly involving the large joints) (35-66%)
- Central nervous system involvement (eg, Sydenham chorea) (10-30%)
- Subcutaneous nodules (0-10%)
- Erythema marginatum (<6%)

**Minor manifestations**
- Arthralgia
- Fever
- Elevated ESR, CRP
- Prolonged PR interval on electrocardiogram
# Symptoms of febrile illness versus neurologic illness in patients with acute rheumatic fever

<table>
<thead>
<tr>
<th>Acute febrile illness</th>
<th>Neurologic illness (25 to 30%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>ONSET two to four weeks after GAS infection</td>
<td>Later onset</td>
</tr>
<tr>
<td>Fever is common</td>
<td>• Two to six months after GAS infection</td>
</tr>
<tr>
<td>Acute joint symptoms and signs</td>
<td>No fever</td>
</tr>
<tr>
<td>Carditis</td>
<td>• Joint manifestations are not a feature</td>
</tr>
<tr>
<td>• Clinical and subclinical</td>
<td>• Behavioral disorder and distinctive chorea</td>
</tr>
<tr>
<td>Skin manifestations and subcutaneous nodules (both are rare)</td>
<td>Carditis &gt;30%</td>
</tr>
<tr>
<td>Raised inflammatory markers</td>
<td>• Often subclinical</td>
</tr>
<tr>
<td>Evidence of preceding GAS infection (elevated ASO and anti-DNase B titers)</td>
<td>Often normal inflammatory markers</td>
</tr>
<tr>
<td>Dramatic symptomatic response to aspirin and NSAIDS</td>
<td>ASO often unhelpful, anti-DNase B may be raised</td>
</tr>
<tr>
<td>Duration usually &lt;6 weeks</td>
<td>Followed by RHD in approximately 50%</td>
</tr>
<tr>
<td>Followed by RHD in approximately 75%</td>
<td></td>
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</tbody>
</table>
Revised Jones criteria

**Low Risk:** <2/100,000 cases of acute RF in School age children
OR
<1/1000 patients with RHD at any age during one year

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2015 AHA Update to Jones Criteria for diagnosis of RF. Circ 2015
Rheumatic Heart Disease

- RHD is the permanent heart valve damage resulting from ≥ 1 attacks of RF
- Prevalence increases with age (peak 25-35ys)


Acute rheumatic fever.
Carapetis JR¹, McDonald M, Wilson NJ.

*Figure 1: Incidence of ARF in 2002 and prevalence of RHD in 2003 by age in Aboriginal Australians from the top end of the Northern Territory (personal communication, Top End RHD Control Program, Department of Health and Community Services, Darwin, Australia)*
Symptoms of RHD
Types of RHD

MR in younger patients
MS in older patients
Mitral Regurgitation

- Cause of primary MR
- Long asymptomatic period (10-20 ys)
- Holosystolic murmur (apex and L axillary)
- Mid- diastolic murmur (Carey – Coombs)
ECHO characteristics

Morphological

Chronic MR
• MV leaflets thickening
• Thickening of chordae tendineae with fusion
• Restricted movement --> doming (elbow deformity of anterior leaflet)
• Leaflet calcification seen in older patients

Acute MR
• Dilatation of mitral annulus
• Elongation of chordae tendineae
• Prolapse (often anterior) leaflet
• Nodular lesions on leaflets

Doppler
• Pathological mitral regurgitation
  • Visible at least in 2 views
  • Regurgitation jet length >2cm (at least 1 view)
  • Regurgitation peak velocity >3m/sec
  • Pansystolic Doppler signal

*** Quantitative methods to quantify MR (PISA, VC, EROA, RV) also apply

Revision of the Jones Criteria for the Diagnosis of Acute Rheumatic Fever in the Era of Doppler Echocardiography
A Scientific Statement From the American Heart Association
Endorsed by the World Heart Federation

(Circulation. 2015;131:1806-1818. DOI: 10.1161
Management

• Medical therapy
• Acute MR
  • Nitrates and diuretics --> reduce filling pressures.
  • SNP reduces afterload and regurgitant fraction.
  • Inotropic agents and an intra-aortic balloon pump are of use in hypotension and haemodynamic instability.
• Chronic MR
  • Good ventricular function, there is no evidence to support the prophylactic use of vasodilators, including ACE inhibitors.
  • When HF --> OMT
Indications for intervention

• MV repair is preferred method, when feasible.
• However challenging in rheumatic lesions
• When not feasible MVR with preservation of the subvalvular apparatus.
• Transcatheter MV repair (edge to edge technique) is NOT recommended in RHD
Mitral Stenosis

- Secondary to RF in 99% of cases
- 2/3 females
- Variable period before symptoms appear
- Slow progression (reduction of MVA $0.01 \text{cm}^2$/year)
- Symptoms develop when MVA $< 1.5 \text{cm}^2$
- Mitral facies (plethoric cheeks with bluish patches)
- Mid-diastolic low pitched murmur (apex)
- Opening snap
- Loud 1st heart sound

The murmur of mitral stenosis

Mild mitral stenosis
Severe mitral stenosis
Severe mitral stenosis with atrial fibrillation

Opening Snap
ECHO characteristics

• Morphological
  • Thickened MV (>5mm thickness of anterior leaflet)
  • Restricted motion-->doming
  • Commisural fusion
  • Thickening and shortening of chordae
  • Calcifications
  • Dilatation of LA
  • LA thrombus not uncommon
Evaluation of severity of MS

- IMPORTANT for management decisions
- Transmitral gradients less useful (depend on HR, rhythm, CO)
- Planimetry (mid-diastole, narrowest orifice, tips of the leaflets)
- PHT
Management

• Medical therapy
  • Diuretics
  • Bb/digoxin/heart rate regulating CCB
  • Anticoagulation (VKA for patients with medium to severe MS)
  • Secondary prophylaxis to prevent recurrence of ARF

• Intervention
  • PMC in symptomatic pts with no contraindications

• Mitral valve surgery +/- Maze procedure
- Old age
- History of commissurotomy
- NYHA class IV
- Permanent Afib
- Severe pulmonary hypertension (RVSP + RAP >60mmHg)

Anatomical characteristics:
- Echocardiographic score >8
- Cormier score 3 (calcification of mitral valve of any extent as assessed by fluoroscopy)
- Very small mitral valve area
- Severe tricuspid regurgitation.
• 69 ys female
• NYHA III - multiple admissions due to APE
• Hx of RF, Afib, DM II, HTx, TAVR
• ECHO: severe MS, mean gradient 13mmHg
Mean gradient post 8mmHg
No re-admissions post NYHA II
Case

- 80 yrs male, NYHA IV
- Hx of RF, TAVI (Sapien XT 23mm), DM, CKD
- Mixed mitral valve disease
Baseline
3D-Printing: Implantation of Sapien XT in model
Transapical Sapien 3 #29mm Implantation
More than one valves?

- Common combinations are MR/AR and MS/AR
- Tricuspid involvement secondary to PHTx

**PROXIMAL LESIONS MAY MASK THE SEVERITY OF DISTAL LESIONS**

- Pt with severe AR may not show dilated LV if severe MS present
- Severity of MS may be underestimated when severe AS present
Conclusions...

• RHD is a preventable heart condition
• Endemic among vulnerable groups
• Prevention is the key for disease control and elimination
• Clinical awareness and knowledge of disease progression and symptoms
• Echocardiographic surveillance
• Optimal medical therapy
• Surgical and percutaneous treatment when indicated
Thank you for your attention