Guideline: (Acute) **Rheumatic Fever and** post-streptococcal Arthritis

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„Rheumatisches Fieber - Poststreptokokkenarthritis im Kindes- und Jugendalter
Registernummer 023 - 027
Klassifikation S2k

Stand: 31.08.2013 , gültig bis 31.08.2018“

According to AWMF

Revision 2016

Autoren: R. Trauzeddel, U. Neudorf
Parachute use to prevent death and major trauma related to gravitational challenge: systematic review of randomised controlled trials.

Guidelines, Recommendations, Evidence?

Is it necessary?????

„Parachutes reduce the risk of injury after gravitational challenge, but their effectiveness has not been proved with randomised controlled trials“
Is this guideline still necessary in Western Europe in 2017?
Yes,
for two reasons:

- The disease still exists
- New „Jones-criteria“
Case Report
History:

2015, January
Leon B., 12 years old, German origin
Fever
Pain + swollen right ankle, both knees

2-3 weeks before:
Pharyngitis, exanthema
⇒ symptomatic therapy (No antibiotics!!!)
2015, January

Leukocytosis 23/nl,
Thrombocytosis 680/nl

CrP 15.4 mg/dl, BSG 84/114 mm

BNP 157 pg/ml,
LDH 480 U/l (141-231)

Antistreptolysin 2210 IU/ml

Ferritin 324 µg/l (9-159)
Microbiology throat culture: negative
**ECG:**
SR, long PR-Intervall

**ECHO:**
Enlarged left ventricle, normal systolic function
Enlarged left atrium
moderate mitral regurgitation
Diagnosis according to the Jones-criteria 1992:

Acute rheumatic fever

(2 major – 5 minor criteria)
Major criteria

Subcutaneous nodules
**Polyarthritis**
Erythema anulare or marginatum
Chorea minor

Carditis

Minor criteria

Fever
Arthralgia
Elevated ESR/high CrP-Level
Leucozytosis
ECG with long PR-Intervall
June, 2015  6 month later

For 5 days uncontrolled movements of arms, hands, legs and facial muscles

No fever, no trauma, no drugs
Diagnosis: Chorea minor – Sydenham (Chorea meens Dance)

Therapy approaches described:

- Corticosteroids 1 mg/kg/KG
- Valproat 10 mg/kg
- Carbamazepin
- Haloperidol
- Phenobarbital
- Diazepam
- Chlorpromazine
- Plasmapheresis
- ivIGG
Reasons for revision
Jones TD.
Diagnosis of rheumatic fever.
JAMA. 1944;126:481–484

Guidelines for the diagnosis of rheumatic fever. Jones Criteria, 1992 update. Special Writing Group of the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young of the American Heart Association

2015 update in the echo-era
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

Michael H. Gewitz, MD, FAHA, Co-Chair; Robert S. Baltimore, MD, Co-Chair; Lloyd Y. Tani, MD, FAHA; Craig A. Sable, MD, FAHA; Stanford T. Shulman, MD; Jonathan Carapetis, MBBS; Bo Remenyi, MBBS; Kathryn A. Taubert, PhD, FAHA; Ann F. Bolger, MD, FAHA; Lee Beerman, MD; Bongani M. Mayosi, MBChB; Andrea Beaton, MD; Natesa G. Pandian, MD; Edward L. Kaplan, MD, FAHA; on behalf of the American Heart Association Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young

Background—Acute rheumatic fever remains a serious healthcare concern for the majority of the world’s population despite its decline in incidence in Europe and North America. The goal of this statement was to review the historic Jones criteria used to diagnose acute rheumatic fever in the context of the current epidemiology of the disease and to update those criteria to also take into account recent evidence supporting the use of Doppler echocardiography in the diagnosis of carditis as a major manifestation of acute rheumatic fever.

Methods and Results—To achieve this goal, the American Heart Association’s Council on Cardiovascular Disease in the Young and its Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee organized a writing group to comprehensively review and evaluate the impact of population-specific differences in acute rheumatic fever presentation and changes in presentation that can result from the now worldwide availability of nonsteroidal anti-inflammatory drugs. In addition, a methodological assessment of the numerous published studies that support the use of Doppler echocardiography as a means to diagnose cardiac involvement in acute rheumatic fever, even when overt clinical findings are not apparent, was undertaken to determine the evidence basis for defining subclinical carditis and including it as a major criterion of the Jones criteria. This effort has resulted in the first substantial revision to the Jones criteria by the American Heart Association since 1992 and the first application of the Classification of Recommendations and Levels of Evidence categories developed by the American College of Cardiology/American Heart Association to the Jones criteria.

Conclusions—This revision of the Jones criteria now brings them into closer alignment with other international guidelines for the diagnosis of acute rheumatic fever by defining high-risk populations, recognizing variability in clinical presentation in these high-risk populations, and including Doppler echocardiography as a tool to diagnose cardiac involvement. 

(Circulation. 2015;131:1806-1818. DOI: 10.1161/CIRCULATIONAHA.115.019803)

Key Words: AHA Scientific Statements • acute rheumatic fever • Doppler echocardiography • Jones criteria • rheumatic heart disease • subclinical carditis
AHA 2015 – Scientific statement: What has changed?

- Risk stratification
- Subclinical carditis
- Criteria minor and major
- Criteria for recurrences
• Risk stratification
Genetics

- familial - higher Incidence

- HLA Association
  (HLA-DR 2, 4, 1, 3, 7
  DRB1*16, B-cell marker D8/17)

Virulence

certain Core proteins
(1, 3, 5, 6, 18, 19, 24)

Environment

- Social and economic circumstances
  More cases in fall, winter and spring
High risk Incidence > $\frac{2}{100000}$ school-aged children
> 1/1000 population/year

Low risk Inzidence $\frac{2}{100000}$ school-aged children
< 1/1000 population/year
Rheumatisches Fieber
Risikoeinteilung

ARF Inzidenz (5-14 J)

All age prevalence

Genetischer Einfluss
Regionale Herkunft

Niedriges individuelles Risiko

Moderates bis hohes individuelles Risiko

Germany: low risk Population (Inzidence <2/100000 School age children)
• Criteria
Major criteria

Before 2015

- Arthritis
  - Polyarthritis
- Carditis
  - Carditis, clinical +/- subclinical (Echo Valvulitis!)
- Chorea
- Erythema marginatum
- Subcutaneous nodules

Population low risk

Major Criteria since 2015

Population high risk

- Mon- o. Polyarthritis/ Polyarthralgia
- Carditis, clinical +/- subclinical (Echo Valvulitis!)
- Chorea
- Erythema marginatum
- Subcutaneous nodules
Minor criteria 2015

Low risk population

- Polyarthralgia
- Fever $\geq 38.5^\circ C$
- ESR $\geq 60$ mm/h $+/-\ CrP \geq 30$ mg/l
- Prolonged PR Interval unless carditis is a major criterion

High risk population

- Monarthralgia
- Fever $\geq 38^\circ$
- ESR $\geq 30$ mm/h $+/-\ cCP \geq 30$ mg/l
- Prolonged PR Interval unless carditis is a major criterion
• Criteria for recurrences
Revision AHA 2015
Recurrences - Criteria

after ARF or established RHD and group A-Streptococcal infection

• 2 Major or
• 1 Major + 2 Minor or
• 3 Minor criteria may be sufficient
• Subclinical carditis
1. Pathological **mitral** regurgitation (all 4 criteria met)
   - Seen in at least 2 views
   - Jet length $\geq 2$ cm in at least 1 view
   - Peak velocity $> 3$ m/s
   - Pansystolic jet in at least 1 envelope

2. Pathological **aortic** regurgitation (all 4 criteria met)
   - Seen in at least 2 views
   - Jet length $\geq 1$ cm in at least 1 view
   - Peak velocity $> 3$ m/s
   - Pandiastolic jet in at least 1 envelope
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 Lawsonson, Graeme Maguire, Éloi Marjion, Mariana Mirabel, Ana Olga Mocumbi, Cleanice Mota, John Paar, Anita Saxena, Janet Scheel, John Stirling, Satupaita Viall, Vijayalakshmi I. Balekundri, Gavin Wheaton, Liesl Zühlke and Jonathan Carapetis

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.

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).
Therapy
## Therapy - primary prevention of RF (Treatment of streptococcal Tonsillopharyngitis)

<table>
<thead>
<tr>
<th></th>
<th>Daily dosages kgKG</th>
<th>Dosages</th>
<th>Therapy length (days)</th>
<th>max. 2 Mio/IE (adult. 3 Mio/IE)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Phenoxy-penicillin</strong></td>
<td>100 000 IE</td>
<td>2 – (3)</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>- Penicillin V</td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td><strong>Penicillin – Benzathin</strong></td>
<td>50 000 IE</td>
<td>2</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>(Infectobicillin)</td>
<td></td>
<td></td>
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**For individuals allergic to Penicillin**

| **Makrolide**          | 10-15mg            | 2       | 10                    |                                 |
| - (Clarithromycin)     |                    |         |                       |                                 |
| **Cephalosporin**      | orally              |         | 10                    |                                 |

*Gerber MA, et al, Circulation 2009*

*DGPI Handbuch*
## Prevention of recurrent attacks of RF (Secondary prevention)

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dosage</th>
<th>Application/Intervall</th>
</tr>
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</table>
| Benzathin Penicillin| 600 000 IE \(<27\text{kg}\)  
1,2 Mill IE \(>27\text{kg}\) | i.m. 28 days  
21 days (in high risk situations) |
| Penicillin V        | 2 x 200 000 IE/day  
2 x 250 mg           | orally                                 |
| Erythromycin        | 2 x 250mg/day              | orally                                 |

<table>
<thead>
<tr>
<th>Category</th>
<th>Duration</th>
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<tbody>
<tr>
<td>ARF without Carditis</td>
<td>5 y / to 21.y of age*</td>
</tr>
<tr>
<td>ARF with Carditis without RHD</td>
<td>10 y / to 21.y of age*</td>
</tr>
<tr>
<td>ARF with Carditis and RHD</td>
<td>10 y / to 40.y of age*</td>
</tr>
</tbody>
</table>

*The longer period is taken*