



Erhebungseinheit für seltene pädiatrische Erkrankungen in Deutschland

Forschungsstelle für pädiatrische Epidemiologie bei der Deutschen Gesellschaft für Kinder- und Jugendmedizin e.V.

Acute rheumatic fever and post-streptococcal glomerulonephritis in Germany

Objectives: The primary study objectives are to determine the incidence of acute rheumatic fever (ARF) and post-streptococcal glomerulonephritis (PSGN) in children and adolescents. Secondary study objectives are the recording of regional differences, (familial) risk factors and ARF/PSGN-associated morbidity and mortality.

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Study duration: 1 year, Start: 12/2023

Background: The estimated pre-pandemic incidences of ARF and PSGN in Germany were 0.05/100,000 and 0.7/100,000 respectively. From winter 2022, there was a massive increase in non-invasive and invasive infections caused by group A streptococci in Germany and many other European countries. The post pandemic incidence for non-suppurative immunogenic sequela are neither known for children and adolescents in Germany nor in many other European countries. The aim of this survey is therefore to determine the incidence and disease burden of these post-streptococcal diseases in Germany to provide evidence for futurous treatment recommendations.

Questions:

1. What is the current incidence of ARF and PSGN in Germany?
2. Are there regional differences or other risk factors associated with the incidence of ARF or PSGN in Germany?
3. What are the ARF- and PSGN-associated morbidity & mortality rates in Germany?

Case definition:

Please report all patients aged 0 to 18 years with one of the following diseases:

Acute rheumatic fever

OR Post-streptococcal glomerulonephritis

WITH confirmed preceding GAS infection according to ≥ 1 of the following criteria:

- (1) Cultural evidence of group A streptococci,
- (2) Positive streptococcal antigen detection,
- (3) Elevated or rising streptococcal antibody titre.

Case definition 1: Diagnosis of acute rheumatic fever (ARF)¹⁻⁵

- 1.1) ARF according to the Jones criteria,
if 2 major criteria or 1 major criterion and 2 minor criteria are positiv
Major criteria: Carditis clinical (or echocardiographic) - polyarthrits - chorea minor - erythema anulare - subcutaneous nodules
Secondary criteria: Arthralgias - fever - elevated erythrocyte sedimentation rate (ESR) and/or elevated C-reactive protein (CrP) - prolonged PQ time
- 1.2) ARF outside the Jones criteria if ≥ 1 of the following criteria² is/are fulfilled:
 - Carditis (with clear anamnestic reference to previous streptococcal infection)
 - Chorea minor (after exclusion of other CNS diseases)
 - Recurrence of rheumatic fever

Falldefinition 2: Diagnose der Poststreptokokken-Glomerulonephritis

- 2.1) highly clinically suspected diagnosis of glomerulonephritis due to Nephritic syndrome:
micro/macropaematuria, pathological proteinuria, increase in renal retention parameters, C3 decrease (transient), hypertension, increase in ASL and/or anti-DNaseB titres
- 2.2) histologically confirmed glomerulonephritis (rarely performed)

Logistics: Please report all patients with ARF or PSGN to ESPED.

Please answer a short digital questionnaire (completion time < 5 minutes) on the ESPED study platform.

In addition, you will receive an information letter including a declaration of consent. We would be very gratefull, if this will be forwarded to the care-givers/patients. They can send us their contact details at their own discretion so that we can contact them in the context of follow-up surveys and further examination offers.

If you have any questions, please feel free to contact us:

ESPED-Studie zu Poststreptokokken-Erkrankungen

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Literature:

1. Report of a WHO Expert Consultation (2001) Rheumatic Fever and Rheumatic Heart Disease. WHO Technical Report Series 923; 2, www.who.int.
2. AWMF (2012) Rheumatisches Fieber – Poststreptokokkenarthritis im Kindes- und Jugendalter. S2k Leitlinie, Register Nr. 023/027, www.awmf.org.
3. AWMF (2015) Therapie entzündlicher Erkrankungen der Gaumenmandeln – Tonsillitis. S2k-Leitlinie, Register-Nr.017/024, www.awmf.org.
4. 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 (1992) Guidelines for the diagnosis of rheumatic fever. Jones Criteria, 1992 update. JAMA 268:2069–73.
5. Gewitz MH, Baltimore RS, Tani LY et al. (2015) 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. Circulation 131:1806–18.