**ACUTE RHEUMATIC FEVER**

- Acute rheumatic fever (ARF) is an inflammatory disease involving cardiac and joint tissue that develops due to inadequately treated group A streptococcal (GAS) infection.
- In cases of carditis, secondary prophylaxis requires monthly injections of penicillin G for a duration of ten years.
- Recurrence of ARF is associated with history of carditis and lack of adherence to secondary prophylaxis.

**PENICILLIN FOR SECONDARY PROPHYLAXIS**

- Patients with well-documented histories of rheumatic fever require continuous antibiotic prophylaxis for secondary prevention of recurrent ARF.
- The agent of choice is penicillin G benzathine 1.2 million U IM every four weeks (AHA evidence rating 1A).
  - Per guidelines, this therapy is particularly beneficial in cases where adherence to oral therapy is likely to be poor.
  - Less supported alternative therapies include penicillin V potassium (1B), sulfadizine (1B), macrolides (1C), or azalides (1C).
- The duration of secondary prophylaxis varies based on clinical presentation and age of onset (1C).
  - Rheumatic fever with carditis and residual valvular disease requires a duration of ten years or until age 40 (whichever is longer).
  - Rheumatic fever with carditis but no residual valvular disease requires a duration of ten years or until age 21 (whichever is longer).
  - Rheumatic fever without carditis requires a duration of five years or until age 21 (whichever is longer).

**A CASE OF RECURRENCE**

- A 47-year-old man Micronesian with history of acute rheumatic fever (ARF) complicated by carditis with inconsistent medical follow-up and nonadherence to secondary prophylaxis presented to the Emergency Department with acute-on-chronic, diffuse polyarthritis.
- History of throat culture positive for group A Streptococcus.
- ASO Ab titer elevated to 1066 IU/ml.
- DNase B Ab titer elevated to 1560 IU/ml.
- Found to meet:
  - **Two major Jones Criteria**
    - Polyarthritis/polyarthralgia
    - Sydenham chorea
  - **All minor Jones Criteria**
    - Polyarthralgias
    - Hyperpyrexia
    - Elevated ESR/CRP
    - Prolonged PR interval on ECG
- Given his persistent symptoms in the absence of alternative diagnoses, his well-documented history of ARF, and his history of nonadherence to secondary prophylactic therapy, the patient was treated based on standard-of-care with a single dose of penicillin G 1.2 million units IM.
- He was discharged with a plan for monthly injections for the recommended duration of ten years of secondary prophylaxis; the patient was lost to follow-up shortly after discharge.

**REVISED JONES CRITERIA FOR PRESUMPTIVE DIAGNOSIS OF RECURRENT ARF**

Presumptive diagnosis of recurrent ARF requires:

1. History of ARF or rheumatic heart disease
2. Evidence of preceding streptococcal infection, AND
3. Either
   a) Two major,
   b) One major and two minor, or
   c) Three minor manifestations

- Level C, Class IIb evidence (consensus of expert opinion based on clinical experience or case studies; usefulness is not well established).

**CASE HIGHLIGHTS**

- This case highlights:
  - The importance of secondary prophylaxis in prevention of recurrent ARF.
  - The importance of recognizing and managing the once common syndrome of recurrent ARF.
  - The need for further clinical research to aid in evidence-based decision-making in cases with high clinical suspicion for recurrent ARF.
  - The roles of therapeutic adherence and access-to-care in achieving successful outcomes in the management of chronic illnesses.

**References:**

3. https://www.masterfile.com