Rheumatic Fever

Pathogenesis: Pharyngeal infection with Lancefield Group A β-haemolytic streptococci of M serotype triggers rheumatic fever 2-4 weeks later, in the susceptible 2% of the pop. Due to cross-reactivity of a strep carbohydrate cell wall antigen & valve tissue. Common in the 3rd World/Aborignal/Islanders/Maori.

Incidence: Peak incidence: 5-15yrs. Tends to recur (10-50%) unless prevented.

Diagnosis: based on the *revised Jones criteria*. There must be evidence of recent streptococcal infection plus 2 major criteria, or 1 major + 2 minor criteria.

Evidence of streptococcal infection: (may have been asymptomatic)

- History of scarlet fever, positive throat swab, ↑ASOT >200U/mL or ↑DNase B titre

Major criteria: (ACESS)

- *Arthritis* - A migratory, 'flitting' polyarthritis; usually affects the larger joints (75%).
- *Carditis* - Tachycardia, murmurs (MR, AR, Carey Coombs' mid-dia murmur), pericardial rub, CCF, cardiomegaly, gallop rhythm, conduction defects (45-70%).
- *Erythema marginatum* - Geographical-type rash with red, raised edges and clear centre (never on face); occurs mainly on trunk, thighs, arms in 2–10%.
- *Subcutaneous nodules* - Small, mobile painless nodules on jt ext surfaces & spine (2-20%).
- *Sydenham's Chorea (St Vitus' dance)* - Occurs late in 10%. Unilateral or bilateral involuntary semi-purposeful movements. May be preceded by emotional lability and unusual behaviour.

***Minor criteria**: (HEAPP)*

- *History of previous rheumatic fever*
- *Elevated ESR or CRP*
- *Arthralgia* (but not if arthritis is one of the major criteria).
- *Pyrexia (>38°C)*
- *Prolonged PR interval* (but not if carditis is major criterion).

Management:

- Bed rest until CRP normal for 2 weeks (may be 3 months).
- **Benzylpenicillin** 0.6-1.2g IM stat then **penicillin V** 250mg/12h PO x 10 days.
- Analgesia for carditis/arthritis: **Aspirin** 100mg/kg/day PO in divided doses (maximum 8g/day) for 2 days, then 70mg/kg/day for 6 weeks. Monitor salicylate level. Toxicity causes tinnitus, hyperventilation, metabolic acidosis. Alternative: NSAIDs
- Steroids if fever/heart failure resistant.
- Immobilize joints in severe arthritis.
- **Haloperidol** (0.5mg/8h PO), **valproate** or **diazepam** for the chorea.

Prognosis: 60% with carditis develop chronic Rh disease. Acute attacks last an ave of 3 months. Recurrence may be precipitated by further streptococcal infections, pregnancy, or use of the OCP. Cardiac sequelae affect mitral (70%), aortic (40%), tricuspid (10%), and pulmonary (2%) valves. Incompetent lesions develop during the attack, stenoses years later.

Secondary prophylaxis: **Penicillin V** 250mg/12h PO until no longer at risk (30yrs). Alternative: **sulphadiazine** 1g daily (0.5g if <30kg). Give antibiotic prophylaxis for dental or other surgery.