RHEUMATIC HEART DISEASE

What is the problem?

- Acute rheumatic fever is a pancarditis the main component of which is damage to the heart valves
- Define the problem

Why is it important?

- Morbidity: rheumatic heart disease is a common cause of acquired heart disease in children and is preventable. (see National Guideline: "Primary Prevention and Prophylaxis of Acute Rheumatic Fever and Rheumatic Heart Disease…" Rheumatic heart disease seriously disrupts childrens' lives, especially schooling, and monthly visits to health centres drain family resources.
- Mortality: children die both during episodes of acute rheumatic fever, and from the sequelae and complications of chronic rheumatic heart disease.

What is the severity?

- The severity of the illness depends on the severity of valve damage.

> Remember that after the acute episode there is often some degree of valve 'recovery'. But a second episode of acute rheumatic fever – because of prophylaxis failure – is devastating

What are the implications of severity?

- All children with acute rheumatic fever must be referred to a regional hospital DURING THE ACUTE EPISODE
- All children who have had an episode of acute rheumatic fever must be followed up in a cardiology service (even if chorea was the only manifestation). Monthly medication repeats can be done at clinic level.
- The markers of severity include:
  - Effort intolerance
  - Signs of heart failure
  - A mid-diastolic murmur at the apex
  - Any STENOSED/STENOSING valve

> Remember: heart failure must be diagnosed early, because sustained heart failure with inadequate treatment can lead to myocardial apoptosis, which is irreversible

What is the cause?

- Acute rheumatic fever follows streptococcal infection, including both sore throat and impetigo.
- Modified Jone's Criteria must be used to make the diagnosis DURING THE ACUTE EPISODE.

Evidence of preceding streptococcal infection	Culture	Rising ASOT	Scarlet Fever	None	Unknown
Major Criteria	Pancarditis	Flitting arthritis	Chorea	Erythema marginatum	Subcutaneous nodules
Minor Criteria	Long PR Interval	Arthralgia	Previous ARF/RHD	Fever	Raised ESR/CRP/WCC

- It is often extremely difficult to make the diagnosis retrospectively, especially in mild disease. It is important not to subject a child to long term monthly injections unnecessarily, but also to make sure that mild cases are NOT missed.
- Rheumatic fever can be diagnosed without supporting evidence of a recent streptococcal infection if Sydenham's chorea is the only manifestation of rheumatic fever

What is the management

- All patients with acute rheumatic fever must be admitted and referred to a specialist hospital. Use the EDL, page 78.

What is the follow up?

Once an episode of acute rheumatic fever has occurred, the most important intervention that you can make is to ensure proper monthly follow up for penicillin injections. You should use the 'Rheumatic Heart Disease Prophylaxis Patient held letter (Form Paed/25), AND make sure that the patient and caregiver understand its use and the importance of NEVER GETTING ACUTE RHEUMATIC FEVER AGAIN

- All patients with rheumatic heart disease, and Sydenham's Chorea, must be followed up in a cardiology service. Then:
- Level 1: for monthly penicillin, and other treatment
- Level 2: for cardiac assessment, 6 monthly to annually
- Level 3 if: for long term planning at disease onset, and if heart valve replacement is a possibility

Use the Rheumatic Heart Disease Follow Up Flowsheet (Form Paed/28), to track progress. Check the patient held letter ON EVERY VISIT and make sure penicillin prophylaxis is being given

What are the preventive measures?

- Home-based: prevent poverty and overcrowding!
- Health service-based: all children with sore throats from the ages of 3 – 15 years should receive penicillin (IM or oral for 10 days) or erythromycin for 10 days if penicillin allergic. (see National Guideline: page 11)

What information should be given to caregivers?

- The severity of the valve damage is variable. If it is mild, following the acute rheumatic fever episode, the child is LUCKY, and a repeat episode of ARF must never be allowed to happen
- Monthly penicillin (or erythromycin) must continue until 35 years
- At puberty, planned parenthood discussions must begin. Pregnancy is a potentially lethal condition in children with rheumatic heart disease
- Care dependency grants may be warranted, depending on the degree of disability

What has happened if a child with rheumatic heart disease gets very sick?

If a child with established rheumatic heart disease, the possible reasons are as follows:

1) Another episode of acute rheumatic fever (due to failed prophylaxis)
   - Check the diagnostic criteria (Jone's)
2) Infective endocarditis
   - Look for the classic signs, but especially pyrexia, splenomegaly, haematuria, leucopaenia and do three blood cultures within 1 hour, before starting antibiotics
3) Precipitous myocardial failure (irreversible – apoptosis, reversible – treatment problem)
   - Find out about adherence, and check previous myocardial function
4) Valve stenosis, especially mitral
   - Critical mitral stenosis is life-threatening and if present needs to be attended to URGENTLY. Check for a loud S1, a long diastolic murmur at the apex and severe LAH on ECG. If in doubt, refer
5) A non-cardiac condition (e.g. severe ARI, APSGN, pregnancy)
   - Look for the cardiac causes AND think about others

The inpatient management of a child with acute rheumatic fever can be challenging, but pales to insignificance compared to the challenge of managing the AMBULATORY care of a child with rheumatic heart disease. Make sure you get the ambulatory care right.