RHEUMATIC HEART DISEASE IN CHILDREN

ACUTE RHEUMATIC FEVER

Autoimmune consequence of infection with Group A streptococcal infection

Results in a generalised inflammatory response affecting brains, joints, skin, subcutaneous tissues and the heart.

Currently, the modified Duckett-Jones criteria form the basis of the diagnosis of the condition.

RHEUMATIC HEART DISEASE

Rheumatic Heart Disease is the permanent heart valve damage resulting from one or more attacks of ARF.

It is thought that 40-60% of patients with ARF will go on to developing RHD.

The commonest valves affecting are the mitral and aortic, in that order. However all four valves can be affected

PATHOPHYSIOLOGY

In 0.3-3% of cases, infection leads to rheumatic fever several weeks after the sore throat has resolved.

The organism spreads by direct contact with oral or respiratory secretions, and spread is enhanced by crowded living conditions.

Patients remain infected for weeks after symptomatic resolution of pharyngitis and may serve as a reservoir for infecting others.

Group A Streptococcus is a gram-positive coccus that frequently colonizes the skin and oropharynx.

It also may be associated with nonsuppurative disease, such as rheumatic fever and acute poststreptococcal glomerulonephritis.

After an incubation period of 2-4 days, the invading organisms elicit an acute inflammatory response with 3-5 days of sore throat, fever, malaise, headache, and an elevated leukocyte count.

Acute rheumatic heart disease often produces a pancarditis characterized by endocarditis, myocarditis, and pericarditis.

Endocarditis is manifested as valve insufficiency.

The mitral valve is most commonly and severely affected (65-70% of patients), and the aortic valve is second in frequency (25%).

The tricuspid valve is deformed in only 10% of patients and is almost always associated with mitral and aortic lesions.

The pulmonary valve is rarely affected.
Pericarditis, when present, rarely affects cardiac function or results in constrictive pericarditis.

Chronic manifestations due to residual and progressive valve deformity occur in 9-39% of adults with previous rheumatic heart disease.

Fusion of the valve apparatus resulting in stenosis or a combination of stenosis and insufficiency develops 2-10 years after an episode of acute rheumatic fever, and recurrent episodes may cause progressive damage to the valves.

**EPIDEMIOLOGY**

**Race**

**Sex**

Rheumatic fever occurs in equal numbers in males and females, but the prognosis is worse for females than for males.

**Age**

Rheumatic fever is principally a disease of childhood, with a median age of 10 years, although it also occurs in adults (20% of cases).

**INCIDENCE**

The incidence of rheumatic fever (RF) varies from 0.2 to 0.75/1000/year in schoolchildren 5–15 years of age (2001 Govt. Census)

(Anil Grover, Padamavati S et al, et.al INJ 2002)

**CLINICAL PRESENTATIONS**

**HISTORY**

The modified Jones criteria (revised in 1992) provide guidelines for the diagnosis of rheumatic fever.

The Jones criteria require the presence of 2 major or 1 major and 2 minor criteria for the diagnosis of rheumatic fever.

The major diagnostic criteria include carditis, polyarthritis, chorea, subcutaneous nodules, and erythema marginatum.

The minor diagnostic criteria include fever, arthralgia, prolonged PR interval on ECG, elevated acute phase reactants (increased erythrocyte sedimentation rate, presence of C-reactive protein, and leukocytosis.

One of the following must be present:

- Positive throat culture or rapid streptococcal antigen test result
- Elevated or rising streptococcal antibody titer
- History of previous rheumatic fever or rheumatic heart disease

**TESTS**

On ECG, sinus tachycardia most frequently accompanies acute rheumatic heart disease. Alternatively, some children develop sinus bradycardia from increased vagal tone.
First-degree atrioventricular (AV) block (prolongation of the PR interval) is observed in some patients with rheumatic heart disease.

First-degree AV block is a nonspecific finding and should not be used as a criterion for the diagnosis of rheumatic heart disease.

**MANAGEMENT OF RHD**

**TREATMENT AND MANAGEMENT**

Medical therapy in rheumatic heart disease includes attempts to prevent rheumatic fever (and thus rheumatic heart disease).

In patients who develop rheumatic heart disease, therapy is directed toward eliminating the group A streptococcal pharyngitis (if still present), suppressing inflammation from the autoimmune response, and providing supportive treatment for congestive heart failure.

Following the resolution of the acute episode, subsequent therapy is directed towards preventing recurrent rheumatic heart disease in children and monitoring for the complications and sequelae of chronic rheumatic heart disease in adults.

**Prevention of rheumatic fever in patients with group A beta hemolytic streptococci (GABHS) pharyngitis**

Oral (PO) penicillin V remains the drug of choice for treatment of GABHS pharyngitis, but ampicillin and amoxicillin are equally effective.

When PO penicillin is not feasible or dependable, a single dose of intramuscular benzathine penicillin G or benzathine/procaine penicillin combination is therapeutic.

For patients who are allergic to penicillin, administer erythromycin or a first-generation cephalosporin. Other options include clarithromycin for 10 days, azithromycin for 5 days, or a narrow-spectrum (first-generation) cephalosporin for 10 days.

**Surgical Care**

When heart failure persists or worsens after aggressive medical therapy for acute rheumatic heart disease, surgery to decrease valve insufficiency may be life-saving.

Forty percent of patients with acute rheumatic heart disease subsequently develop mitral stenosis as adults.

In patients with critical stenosis, mitral valvulotomy, percutaneous balloon valvuloplasty, or mitral valve replacement may be indicated.

Due to high rates of recurrent symptoms after annuloplasty or other repair procedures, valve replacement appears to be the preferred surgical option.
RHYTHM DISORDER IN CHILDREN

Rates

Bradyarrhythmias
  Sinus Bradycardia
  Sick Sinus Syndrome

AV Nodal Blockade
  First Degree
  Second Degree
    Mobitz I
    Mobitz II
  Third Degree
    Complete Heart Block

Tachyarrhythmias
  Supraventricular
    Originate from foci above or within the atrioventricular node
    Main players in outpatient setting
    All the favorites
      AV nodal reentrant tachycardia (SVT)
      Atrial flutter
      Atrial fibrillation

Location

Supraventricular Arrhythmias
  Originate from foci above or within the atrioventricular node

Ventricular Arrhythmias
  Non-sustained ventricular tachycardia
  Sustained ventricular tachycardia
    Stable
      Know the neighborhood
      Do no harm
    Unstable
      ACLS
  Ventricular fibrillation
    Never a stable rhythm
    Immediate ACLS