Pediatrics TeamWork

437

Acquired Heart Diseases

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Notes Previous Notes Book Important!

Rheumatic Fever

Most common cause of acquired heart disease in developing countries

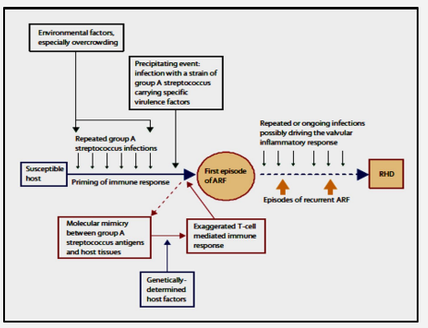
- 150 in 100,000 in developing countries
- 1 in 100,000 in developed countries

Rheumatic heart disease (RHD) inflammatory changes to cardiac valves and myocardium.

Precipitated by Group A Streptococcal (GAS) pharyngitis (not other types of GAS infections)

2-4 weeks after untreated GAS Pharyngitis but most commonly is due to tonsillitis, that is why when you get tonsillitis you have to take antibiotics 7-10 days

T-cell and B-cell lymphocytes produce antibodies against some GAS antigens that cross-react with antigens on myocytes or cardiac valve tissue.

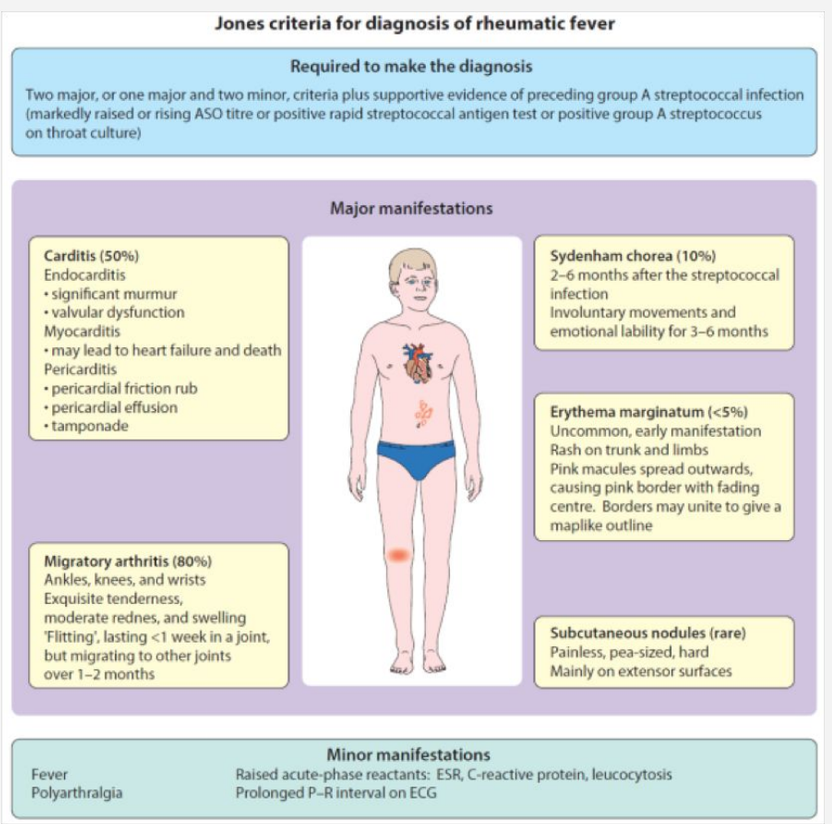


JONES Criteria

Two majors or one major and two minors plus **evidence of antecedent** GAS infection

Exceptions:

- Chorea & indolent carditis do **NOT** require evidence of antecedent GAS infection
- Recurrent episode requires:
- Only one major **OR** Several minor manifestations
- Plus evidence GAS infection



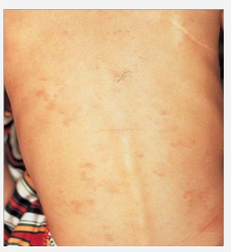
Major Manifestations

Carditis :
-Valvulitis (MV / MV plus AoV /Ao V)
• Early disease leads to valvular regurgitation,
• prolonged or recurrent attacks lead stenosis
• +/- Pericarditis and myocarditis

Polyarthritis :
Most common symptoms (75%)
Migratory
Asymmetrical
Polyarthritis
Large joints
– Typically: extremely painful
– Highly responsive to NSAID therapy, it is an arthritis that significantly improves on the next day

Chorea :
• Emotional lability
• Jerky, uncoordinated movements could be manifested as worsening hand writing
• Hands, feet, tongue and face.
• Disappear during sleep
• More common in adolescence female
• May appears very late after ARF episode (6 wks - 3 yrs following GAS infection)
• Strong association with carditis

Erythema marginatum:
• Rare
• Difficult to detect in dark-skin
• Circular patterns of pink macules
• Blanch under pressure
• Trunk and proximal extremities
• Almost never on face
• Not itchy or painful



Subcutaneous nodules:
-rare <2% of cases
-HIGHLY specific manifestations of ARF
-Round firm, mobile and painless nodules
-1-2 weeks after onset of other symptoms
-Last 1-2 weeks (rarely > 1 month);
-Strongly associated with carditis

Minor Manifestations

Arthralgia if you used arthritis as a major criteria, you can't use arthralgia as a minor

Fever

Raised ERS or CRP

Prolonged PR interval on ECG if you used carditis as a major criteria, you can't use ECG as a minor

Evidence of antecedent GAS infection

Positive throat culture or rapid antigen test for GAS

Raised or rising streptococcal antibody titre

Treatment

• Antibiotics to eradicate GAS bacteria
• Anti-inflammatory agents
– High dose aspirin
– Steroids for severe carditis
• CHF therapy as indicated
• Sydenham Chorea:
– haloperidol

Aspirin has been used traditionally to suppress the inflammatory response of the joints and heart, but has largely been superseded by naproxen as it has fewer side-effects.

Rheumatic Fever

SECONDARY PROPHYLAXIS

- **Benzathine penicillin G (BPG) :** IM , Q4 wks **OR** Q3 wks if confirmed recurrent ARF despite adherence to 4 wks injection . Dose 1,200,000 U ≥ 20kg **OR** 600,000 U < 20kg
- **If the patient has Penicillin allergy** give 40mg/kg/day (children) Orally from **Erythromycin** BID –QID
- We usually avoid the oral route because of patients compliance is poor

DURATION OF SECONDARY PROPHYLAXIS:

- **ARF with no or mild carditis :** Min of 10 years after most recent episode ARF **OR** until age 21 years (whichever is longer)
- **ARF with moderate carditis :** Min of 10 years after most recent episode ARF **OR** until age 30 years (whichever is longer)
- **ARF with severe carditis :** Min of 10 years after most recent episode ARF **OR** until age 30 years (whichever is longer) , May need life long prophylaxis

Infective Endocarditis

- Infection of the endocardial lining of the heart or cardiac vessels
- Rare but with high mortality
- Usually affect abnormal cardiac structure:

Valvular disease	Septal defects	Presence of foreign material such as mechanical valves and patch material after surgical repair

Etiology

- **Gram- positive bacteria:**

– > 90 % 0f bacterial case
– Streptococci Viridans : most common you have to know that viridans is the most common cause
– Staphylococcal species – esp. prosthetic valves.
– Enterococci : less common in children

- **Gram-negative bacteria:**

– < 10% of bacterial cases
– Example: HACEK group

- **Fungal** – uncommon

– immunocompromised patients, prolonged Abx

- **10% IE case:** organisms cannot be identified.

Infective Endocarditis

Clinical Features

- **General:**

– Fever
– Nonspecific manifestations : myalgias, arthralgias, headache, malaise

- **Cardiac:**

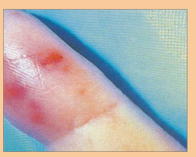
– New onset or worsening valvular regurgitation
– Congestive heart failure
– Heart block

- **Extracardiac - septic embolism:**

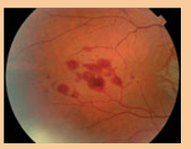
– CNS: infraction, brain abscess
– Renal: proteinuria, hematuria, pyuria
-Skin: necrotic skin lesions

- **STIGMATA OF SBE:**

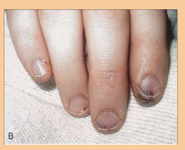
– Janeway lesions. Painless, on palms and soles
– Osler's nodes painful, on fingertips and toes
– Roth's spots
– Splinter hemorrhages
– Splenomegaly
– Microscopic Hematuria



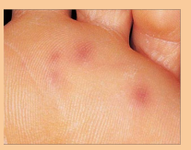
Osler's nodes



Roth's spot



Splinter hemorrhage



Janeway lesions

Investigations

- **Blood cultures: <u>most</u> important lab test**

– **<u>Three</u>** blood cultures at least
– Collected over a 24-h period
– Preferably from different site to rule out contamination of the samples

- **CBC:**

– Anemia

- **Elevated ESR/CRP**
- **Positive rheumatoid factor**
- **Echo:** Vegetation

Treatment

- **Supportive medical therapy**
- **Prolonged antibiotic therapy**

– Initially empiric broad spectrum Abx (high dose penicillin + Aminoglycosides)
– Abx adjusted according to organism sensitivity
– 4-8 weeks course depending on organism and resistance.

- **Removal of infected line**
- **Surgical intervention:** may be required in cases of prosthesis

Infective Endocarditis

Prophylaxis

- **Only for dental procedures and invasive ENT procedures for high risk patients**
- **No longer recommended for GI or GU procedures**
- **Prophylaxis regimens:**

– Oral amoxicillin 50 mg/kg (up to 2 g) OR
– IV/IM ampicillin 50 mg/kg

- **Patients allergic to penicillin**

– Cephalexin 50 mg/kg PO (up to 2 g) OR
– Clindamycin 20 mg/kg (up to 600 mg) OR
– Azithromycin 15 mg/kg (up to 500 mg)

- **For who ?**

– Prosthetic cardiac valve
– Previous IE
– Complex congenital heart disease :

- Unrepaired cyanotic CHD, including palliative shunts and conduits
- Repaired CHD with prosthetic material or device for first 6 months after the procedure
- Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device

– Cardiac transplantation recipients with cardiac valvulopathy

Myocarditis

- **Inflammation of myocardium with necrosis**
- **Viral infections: most common causes**

– Coxsackievirus type B
– Adenovirus
– Parvovirus B19
–Others: CMV, EBV, HIV, Hep C

- **Others: bacteria, rickettsiae, protozoa.**
- **Non-infectious causes:**

– Rheumatologic disease: SLE, rheumatoid arthritis
– Drugs and Toxins: chemotherapy

Presentations

- Variable initial presentation
- **Viral prodrome:** fever, URTI or GI symptoms
- **Hx:**

– Lethargy, poor feeding, irritability
– Respiratory distress
– Exercise intolerance

- **PHx:**

– Signs of CHF :

Tachycardia	Tachypnea	Gallop rhythm	Murmur of mitral regurgitation	Hepatomegaly

Myocarditis

Investigations

CXR:
– Cardiomegaly
– Pulmonary congestion

ECG:
– Sinus tachycardia
– Low voltage ORS
– Non-specific T wave changes

Echo:
– Dilated LV with reduced systolic function
– MR
– Pericardial effusion

Myocardial biopsy:
– Historically used to be the gold standard test
– Currently not routinely performed **why?**
- Invasive
- low sensitivity of the procedure (3–63%)
- patchy involvement of the myocardium

Treatment

- **Supportive**

– Inotropes, Ventilation, ECMO
– Anti-CHF therapy
– IVIG and Steroid (Not supporting evidence for benefit)
– Antiviral agents and interferon (Need further studies)

Cardiomyopathy

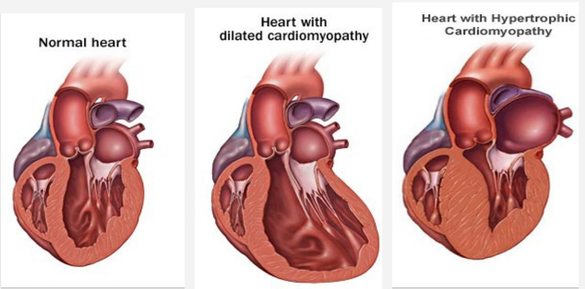
- **Cardiomyopathy:** myocardial disease resulting in thickening of myocardial fibers or fibrosis.
- **Pediatric cardiomyopathy:** almost exclusively non-ischemic
- **Types:**

Dilated cardiomyopathy (58%)

Hypertrophic cardiomyopathy (30%)

Restrictive cardiomyopathy (5%)

Arrhythmogenic right ventricular cardiomyopathy (ARVC)(5%)



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Arrhythmia

- **Supraventricular tachycardia** is the most common childhood arrhythmia, heart rate is rapid 250-300 beats per minute.
- It can cause poor cardiac output and pulmonary edema and Heart failure symptoms.
- ECG→ Narrow complex tachycardia.
- Management;

1. Circulatory and respiratory support
2. Vagal stimulation maneuver.
3. IV adenosine, if fails: electrical cardioversion (0.5 - 2 J/kg body weight) with a synchronized direct current shock.
4. Once sinus rhythm is restored, maintenance therapy will be required (e.g. flecainide or sotalol). **When to stop?** At 1 year of age if there is no WPW, if there is, then atrial pacing in adulthood.
5. If they relapsed >> percutaneous radiofrequency ablation or cryoablation

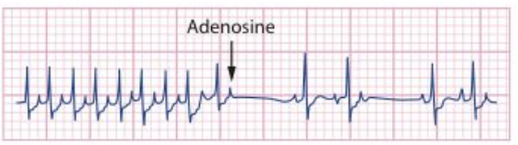

Figure 18.18 Rhythm strip showing supraventricular re-entry tachycardia, in which there is a narrow complex (<120 ms or three small squares) tachycardia of 250–300 beats/min, and response to treatment with adenosine.

- **Congenital complete heart block:** due to the presence of maternal anti-Ro or anti-La antibodies. All children with symptoms (presyncope or syncope) require insertion of an endocardial pacemaker

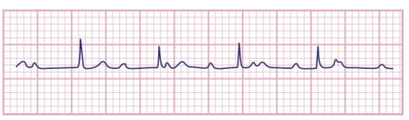

Figure 18.19 Electrocardiography of congenital complete heart block. The P waves and QRS complexes are dissociated.

- **Long QT syndrome:** it can be associated with sudden loss of consciousness during exercise (can be mistaken as epilepsy) , if untreated can cause death due to Vtach. This condition can be associated with erythromycin therapy, electrolytes disorders and head injury
- Vasovagal syncope **vs** cardiac syncope:

	Vasovagal syncope	Seizure	Cardiac syncope
Trigger	Common (upright posture, hot environment, pain, fear)	Rare (flashing lights, hyperventilation)	Rare, exertional
Prodrome	Almost always (presyncope)	Common (aura)	Uncommon or brief
Onset	Gradual (often minutes)	Usually sudden	Usually sudden
Duration	1-30 s	1–3 min	Variable
Convulsive jerk	Common (brief), occurs after LOC has set in	Common, prolonged, occurs during LOC	Common (brief), occurs after LOC
Incontinence	Uncommon	Common	Uncommon
Tongue bite	Very rare	Common	Very rare
Injury	Less likely	Likely	Less likely
Color	Very pale, cold skin	Pale, flushed or blue	Very pale, cold skin
Postictal confusion	Rare (wakes on floor), more in elderly	Common (wakes in ambulance)	Rare (wakes on the floor)
Recovery	Prompt (if lie flat quickly)	Slow (confused)	Prompt
	Fatigue (minutes to hours)	Fatigue (minutes to hours)	No fatigue

LOC=Loss of consciousness

- Features suggestive of cardiac syncope: **symptoms on exertion** (potentially dangerous), **family history of sudden unexplained death**, and **palpitations**

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- Chest pain in pediatrics is rarely due to heart disease. Those occurring at rest usually require no further investigations, but if they occur in a child with known heart disease or on exercise, then they should be taken more seriously
- Healthy child with enlarged heart and heart failure >> you should suspect dilated cardiomyopathy which is the most common. Other cardiomyopathies are RARE in children, and are usually related to systemic disorder (Hurler, Pompe or noonan syndrome)
- The diagnosis of dilated cardiomyopathy is done by echo, and treatment is symptomatic with diuretics and ACE inhibitors and carvedilol
- Medical therapy of PHTN: Cyclic guanosine monophosphate pathway (inhaled nitric oxide, IV Mg sulphate and oral PDE inhibitors), endothelin receptor antagonists (oral bosentan), cyclic adenosine monophosphate pathway (IV prostacyclin or inhaled iloprost). These medications allow transplantation to be delayed for many years

Box 18.4 Causes of pulmonary hypertension

- Pulmonary arterial hypertension
  - Idiopathic: sporadic or familial
  - Post-tricuspid shunts (e.g. VSD, AVSD, PDA)
  - HIV infection
  - Persistent pulmonary hypertension of the newborn
- Pulmonary venous hypertension
  - Left-sided heart disease
  - Pulmonary vein stenosis or compression
- Pulmonary hypertension with respiratory disease
  - Chronic obstructive lung disease or bronchopulmonary dysplasia in preterm infants
  - Interstitial lung disease
  - Obstructive sleep apnoea or upper airway obstruction
- Pulmonary thromboembolic disease
- Pulmonary inflammatory or capillary disease.

AVSD, atrioventricular septal defect; HIV, human immunodeficiency virus; PDA, persistent ductus arteriosus; VSD, ventricular septal defect.

KAWASAKI DISEASE

Kawasaki disease is an **acute inflammatory vasculitis** of medium sized arteries.

CLINICAL FEATURES OF TYPICAL KAWASAKI DISEASE

Diagnostic criteria: **fever for ≥ 5 days** with ≥ 4 out of 5 of the following features:

- **Conjunctivitis** (bilateral, bulbar, non-purulent)
- **Rash** (any rash except vesicular)
- **Extremities**
  - Acute: palmar/plantar erythema and/or edema
  - Subacute: periungual desquamation
- **Adenopathy** (cervical, ≥ 1.5 cm, unilateral)
- **Mucosal changes** (strawberry tongue, cracked and erythematous lips, etc.)

Incomplete (atypical) Kawasaki disease: fever for ≥ 5 days with ≥ 2 of the above criteria. Assess for supplemental laboratory criteria.

Memory aid: **"WARM CREAM"**

OTHER POTENTIAL CLINICAL FINDINGS

- **Cardiovascular**: coronary artery aneurysms, ↓ left ventricular function, pericarditis, myocarditis, mitral regurgitation, shock
- **Gastrointestinal**: abdominal pain, diarrhea, nausea, vomiting
- **Neurological**: irritability, facial nerve palsy, hearing loss
- **MSK**: arthritis, arthralgia (pleocytosis of synovial fluid)

KD PHASES

- Acute: 1-2 weeks
- Subacute: 3-4 weeks
- Convalescent: 6-8 weeks

INVESTIGATIONS

- **Basic labs**: CBC+diff, lytes, Cr, CRP, ESR, ALT, albumin, urinalysis
- **Transthoracic ECHO** (rule out coronary artery aneurysms) at diagnosis and follow-up as indicated
- **ECG** (ST changes, pericarditis, myocarditis, etc.)
- **CXR** (rule out pulmonary pathology, rule out cardiomegaly)
- **Blood culture**, if clinically indicated
- Other **bacterial** and **viral testing** as clinically indicated
- **Multisystem inflammatory syndrome in children (MIS-C)**: above tests + COVID-19 PCR and serology, CK, ferritin, LDH, fibrinogen, D-dimer, PTT, INR, troponin, NT-proBNP

LAB FINDINGS

- Anemia
- ↑ platelets
- ↑ WBC
- Hyponatremia
- ↑ CRP
- ↑ ESR
- ↑ liver enzymes
- ↓ albumin
- Sterile pyuria

TREATMENT

- **Intravenous immunoglobulin (IVIG)**: 2g/kg x 1 dose, reassess after 24 hours, then consider a second dose.
  - ✓ If available, consider sending COVID-19 serology prior to IVIG administration.
- **ASA as per institutional guidelines** until normal follow-up ECHO.

COMPLICATIONS

Coronary artery aneurysms

- Untreated KD: 20-25%
- Treated KD: 1-3 %

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