acquired heart disease

Objectives:

- Acquired heart disease in children including acute rheumatic fever, Kawasaki disease,infective endocarditis, myocarditis, cardiomyopathy.

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Special thanks to team 437 & Faisal alsaif

Notes

Important

Book

acquired heart disease

just read about Kawasaki and Rheumatic fever

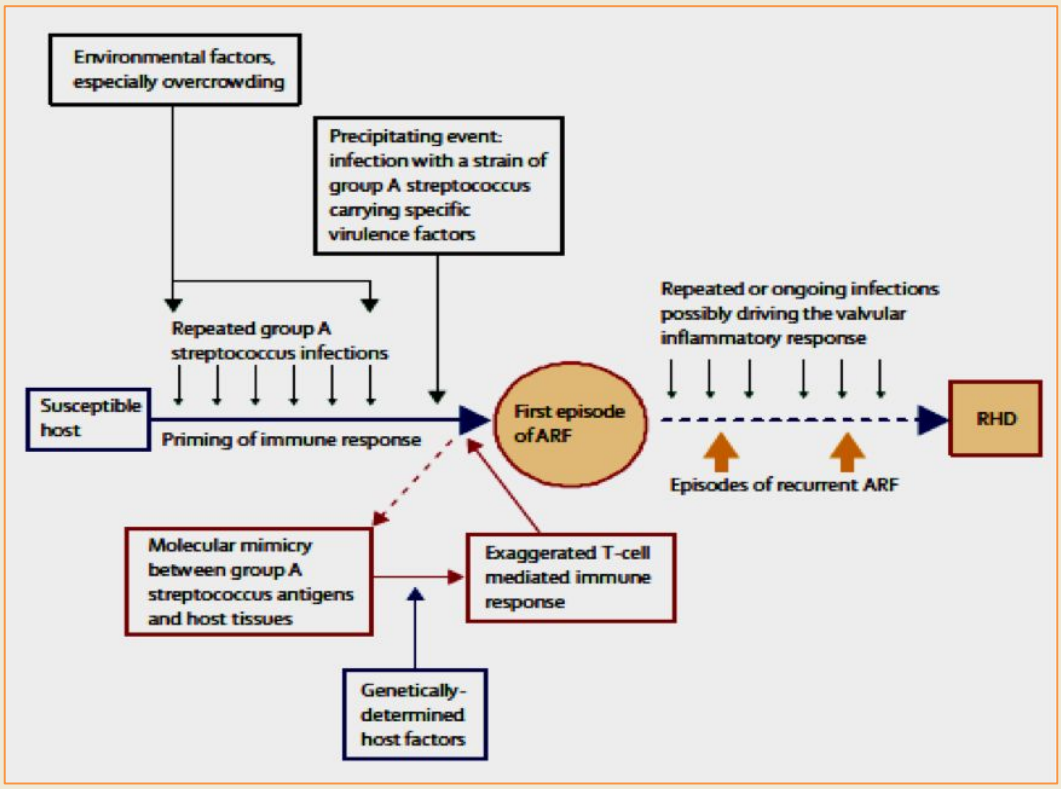
- **Rheumatic Fever**
- **Kawasaki Disease** very imp for mcq & osce
- Infective Endocarditis know the organism
- Pericarditis
- Myocarditis
- Cardiomyopathy
- Cardiac Arrhythmias

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



Acute rheumatic fever vs RHD:

- you can have ARF with no or mild heart involved = complete recovery
- RHD: cardiac complication secondary to ARF, could be permanent or not.
- What is the problem with ARF?

-Risk of heart complication from the second attack is way higher than the first

-So you need prophylaxis to avoid the second attack, the problem is prophylaxis means long term (up to 21, 40 years of age or even life long) monthly injection.

Rheumatic fever

JONES CRITERIA (1992):

Major manifestation	Minor manifestations
Carditis	Arthralgia [3]
Polyarthritis [1]	Fever
Chorea	Raised ESR or CRP
Erythema marginatum [2]	Prolonged PR interval on ECG [4]
Subcutaneous nodules [2]	

- **Evidence of antecedent GAS infection** [5]
  1. Positive throat culture or rapid antigen test for GAS
  2. Raised or rising streptococcal antibody titre

to diagnose:

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

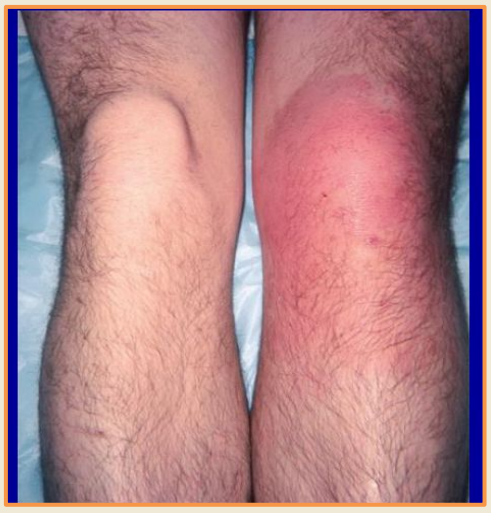
– Recurrent episode requires:

- Only one major OR
- Several minor manifestations
- Plus evidence GAS infection

1- most common
2- Highly associated with cardiac involvement (RHD)
3- Cannot be used as minor if polyarthritis is used as major
4- AKA 1st degree heart block, cannot be used as minor if carditis is used as major
5- ASO titer can be negative if taken too early, repeat in 2 weeks.
- High ASO titer means previous GAS infection, any infection (e.g. pharyngitis, scarlet fever, ARF, impetigo)
- Brucellosis gives you false negative

Rheumatic fever

ARTHRITIS:



- Most common symptoms (75%)
  - Migratory
  - Asymmetrical
  - Polyarthritis
  - Large joints
  - Typically: extremely painful
  - Highly responsive to NSAID therapy

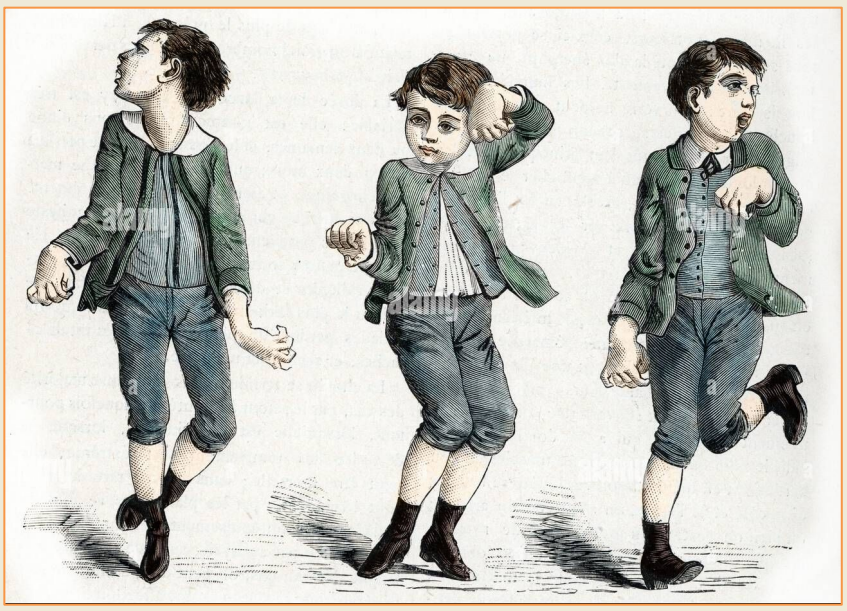
Carditis:

- **Valvulitis:**
- Involvement of mitral valve or mitral valve **plus** aortic valve or aortic valve alone rarely
  - Early disease leads to valvular regurgitation,
  - prolonged or recurrent attacks lead stenosis
  - +/- Pericarditis and myocarditis

SYDENHAM'S CHOREA:

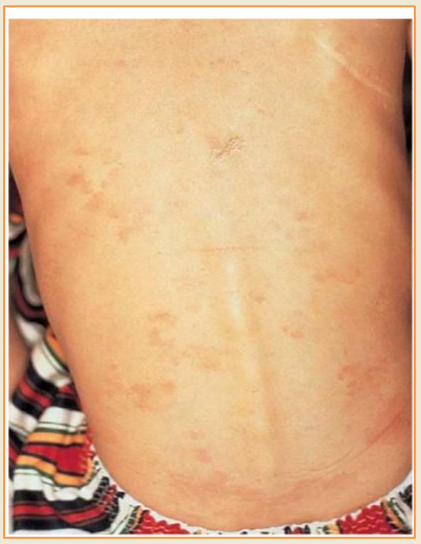
- Jerky, uncoordinated movements involving Hands, feet, tongue and face.
  - Disappear during sleep
  - More common in adolescence female
- May appears very late after acute rheumatic fever episode
  - 6 wks - 3 yrs following GAS infection
- Strong association with carditis

watch this video



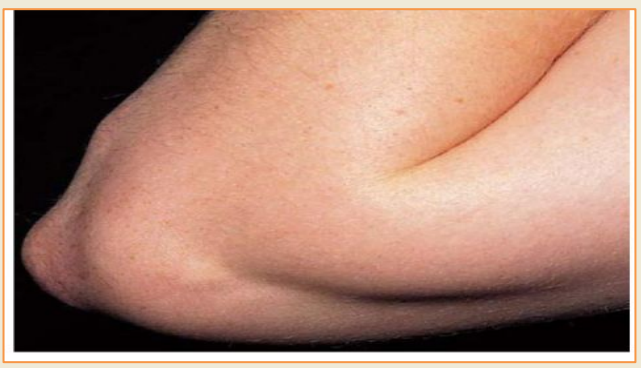
Rheumatic fever

ERYTHEMA MARGINATUM:



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

SUBCUTANEOUS NODULES



- Rare < 2% of cases
- Highly specific manifestation of acute rheumatic fever
- 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

Jones criteria for diagnosis of rheumatic fever

Required to make the diagnosis

Two major, or one major and two minor, criteria plus supportive evidence of preceding group A streptococcal infection (markedly raised or rising ASO titre or positive rapid streptococcal antigen test or positive group A streptococcus on throat culture)

Major manifestations

Carditis (50%)
Endocarditis
- significant murmur
- valvular dysfunction

Myocarditis
- may lead to heart failure and death

Pericarditis
- pericardial friction rub
- pericardial effusion
- tamponade

Migratory arthritis (80%)
Ankles, knees, and wrists
Exquisite tenderness, moderate rednes, and swelling
'Flitting', lasting <1 week in a joint, but migrating to other joints over 1–2 months

Sydenham chorea (10%)
2–6 months after the streptococcal infection
Involuntary movements and emotional lability for 3–6 months

Erythema marginatum (<5%)
Uncommon, early manifestation
Rash on trunk and limbs
Pink macules spread outwards, causing pink border with fading centre. Borders may unite to give a maplike outline

Subcutaneous nodules (rare)
Painless, pea-sized, hard
Mainly on extensor surfaces

Minor manifestations

Fever	Raised acute-phase reactants: ESR, C-reactive protein, leucocytosis
Polyarthralgia	Prolonged P–R interval on ECG

Rheumatic fever

Treatment:

- Antibiotics to eradicate GAS bacteria 10 days of penicillin
- Anti-inflammatory agents
  – High dose aspirin
  – Steroids for severe carditis. Required if fever and inflammation do not resolve rapidly with aspirin
- chronic heart failure therapy as indicated. With diuretic and ACE inhibitors

Sydenham Chorea:

- haloperidol Or sodium valproate

- Significant pericardial effusion may require pericardiocentesis

Secondary prophylaxis			
Antibiotics	**Dose**	**Route**	**Frequency**
Benzathine penicillin G (BPG)	1,200,000U >= 20 KG 600,000U < 20 KG	IM	Q4 wks OR Q3 wks if confirmed recurrent ARF despite adherence to 4 wks injection
Penicillin allergy			
Erythromycin	40mg/kg/day (children)	Oral	BID-QID
	400mg (adults)	Oral	BID

DURATION OF SECONDARY PROPHYLAXIS	
CATEGORY	**DURATION OF PROPHYLAXIS**
ARF with no or mild carditis	Min of 10 years after most recent carditis episode ARF OR until age 21 years (whichever is longer)
ARF with moderate carditis	Min of 10 years after most recent carditis episode ARF OR until age 30 years (whichever is longer)
with ARF with severe carditis	Min of 10 years after most recent carditis episode ARF OR until age 30 years (whichever is longer) May need lifelong prophylaxis

Kawasaki Disease

- KD: inflammatory disease of unknown etiology
- Acute, diffuse vasculitis of medium size blood vessels (mainly coronary arteries)
- Most common cause of acquired heart diseases in developed countries.
  – More common in Japanese population
  – Higher in males than females
  – Typical age: 6 months - 5 years

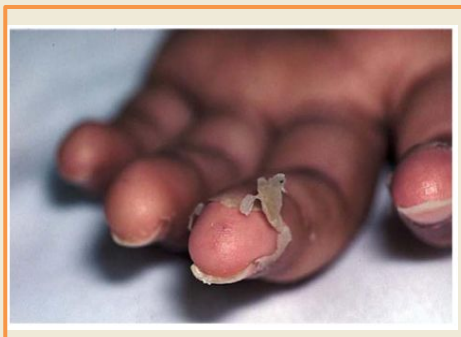
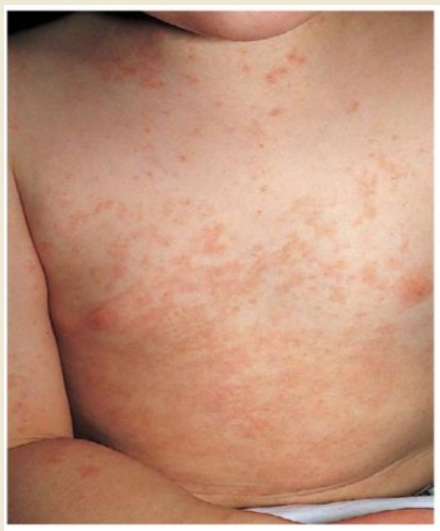
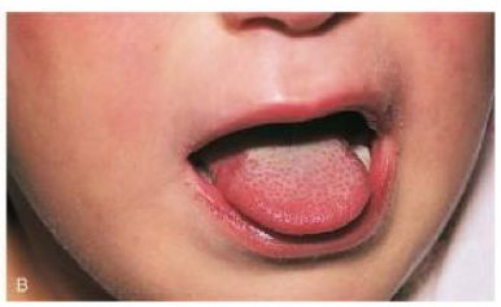
Cardiac Involvement:

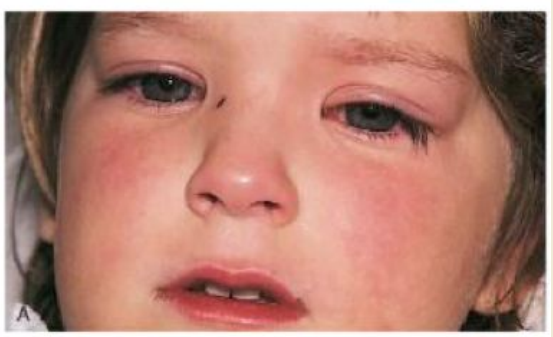
- **Coronary arteries:**
  - Dilation and aneurysm formation
  - Thrombus formation
  - Fibrosis and stenosis
  - Myocardial infarction
- **May cause myocarditis and endocarditis**

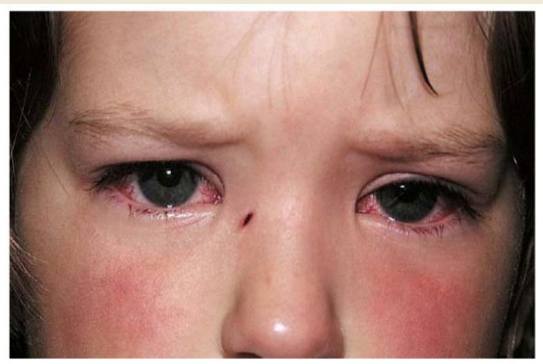
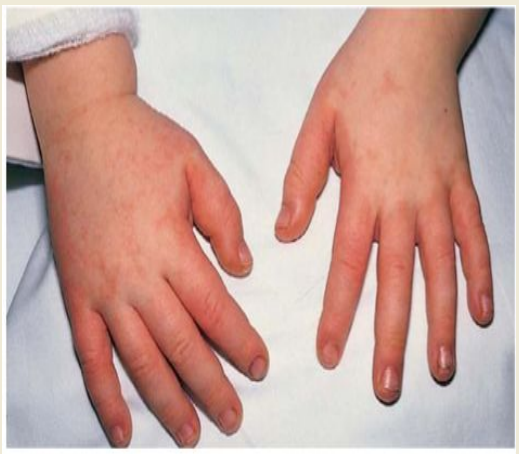
- Other medium size arteries include:
  - Auxiliary, femoral, iliac, and renal arteries

Diagnostic criteria

- **Fever:** at least 5 days duration **and** at least 4 of the following:
- **Conjunctivitis**: bilateral, non-purulent
- **Skin Rash:** polymorphous skin rash
- **Mucous membrane changes:** red, dry, and cracked lips, strawberry tongue
- **Extremities changes:** palms/soles erythema, hands/feet edema, Skin peeling
- **Cervical lymphadenopathy:** unilateral and >1.5 cm in diameter



Kawasaki Disease

Differential diagnosis:

- Scarlet fever
- EBV infection
- Adenovirus infection
- Staphylococcal scalded skin syndrome
- Drug reactions
- Stevens–Johnson syndrome.

ATYPICAL (INCOMPLETE) KD

- Cases of KD that do not fulfill diagnostic criteria.
- More common in infants – Children with Fever greater than 5 days
- Two or three classic symptoms
- Supporting laboratory abnormalities
- Echo evidence of coronary involvements have higher risk of coronary artery disease than typical

  Should be treated as KD

LABS ABNORMALITIES IN KD

- Elevated ESR > 50 (70%)
- Elevated C-reactive (50%)
- CBC: Neutropenia, leukocytosis (50%), Nonspecific anemia, and **Thrombocytosis** which is:
  - Marker of KD
  - Not seen until 2nd week of the disease
- Elevated liver transaminases (40%)
- Low serum albumin level
- Sterile pyuria (33%)
- Aseptic meningitis (up to 50%).

Kawasaki Disease

TREATMENTS:

IVIG 2g/kg single dose
– 2nd dose if persistent fever within 48 h of the initial dose

• High dose of aspirin of 30–100 mg/kg/day.
– Once afebrile: aspirin is decreased to 3–5 mg/kg/day

Echo at base-line

Repeat echocardiogram at 6–8 weeks
– If normal coronary arteries : aspirin can be stopped
– If coronary artery abnormalities: long-term Rx with aspirin

Other anticoagulants if giant aneurysm of coronary arteries

INFECTIVE ENDOCARDITIS

- infection of the endocardial lining of the heart or cardiac vessels
- Rare but with high mortality
- Usually affect **abnorma**l 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
– 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:

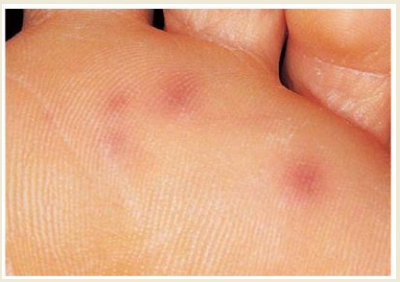
- **General:**
  Fever
  Nonspecific manifestations such as: myalgias, arthralgias, headache, malaise

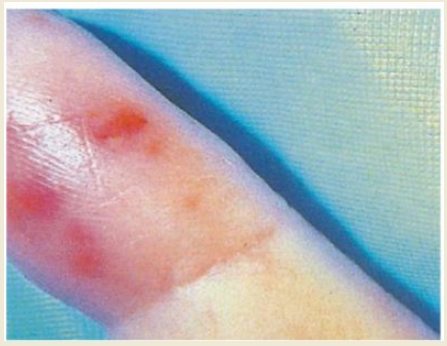
- **Cardiac**
  – New onset or worsening valvular regurgitation
  – Congestive heart failure
  – Heart block

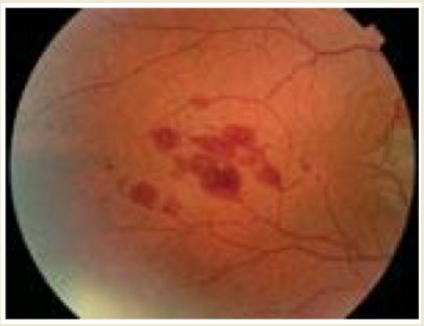
- **Extracardiac**
  - septic embolism: to CNS: infraction, brain abscess, to Renal: proteinuria, hematuria, pyuria

PHX - STIGMATA OF SBE:

- Janeway lesions
- Osler's nodes
- Roth's spots
- Splinter hemorrhages
- Splenomegaly
- Microscopic Hematuria







Investigation :

- **Blood cultures:** most important lab test
  – Three blood cultures
  – Collected over a 24-h period
  – Preferably from different site
- CBC: Anemia
- Elevated ESR/CRP
- Positive rheumatoid factor
- Echo: **Vegetation** Positive echo confirms the diagnosis, but negative echo can't exclude it

INFECTIVE ENDOCARDITIS

TREATMENT:

- Supportive medical therapy
- Prolonged antibiotic therapy High dose penicillin + aminoglycoside
  - – Initially empiric broad spectrum Abx
  - – Abx adjusted according to organism sensitivity
  - – 4-8 weeks course depending on organism and resistance. IV therapy
- Removal of infected line
- Surgical intervention: may be required In prosthetic valves

subacute bacterial endocarditis prophylaxis:

- Only for dental 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)

High risk patients for prophylaxis:

Previous IE

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

PRESENTATION:

- 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

Investigation:

- **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

Myocarditis

TREATMENT:

Supportive:

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

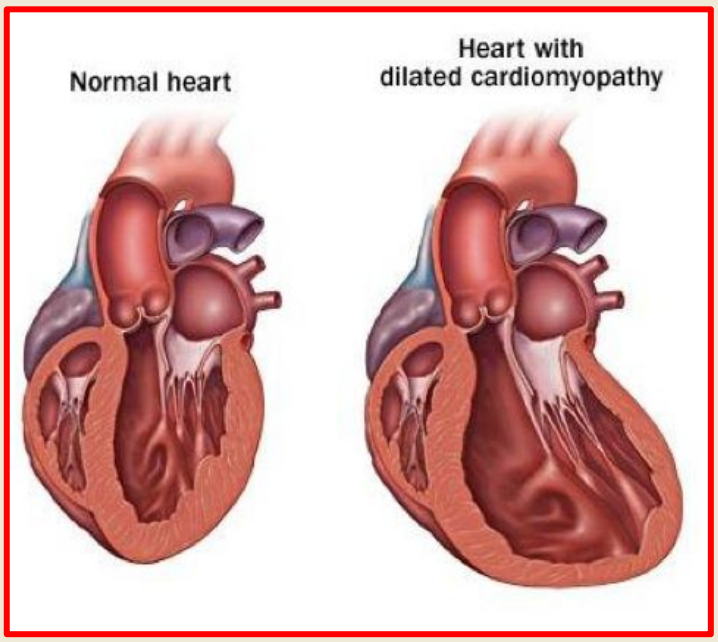
Cardiomyopathy

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

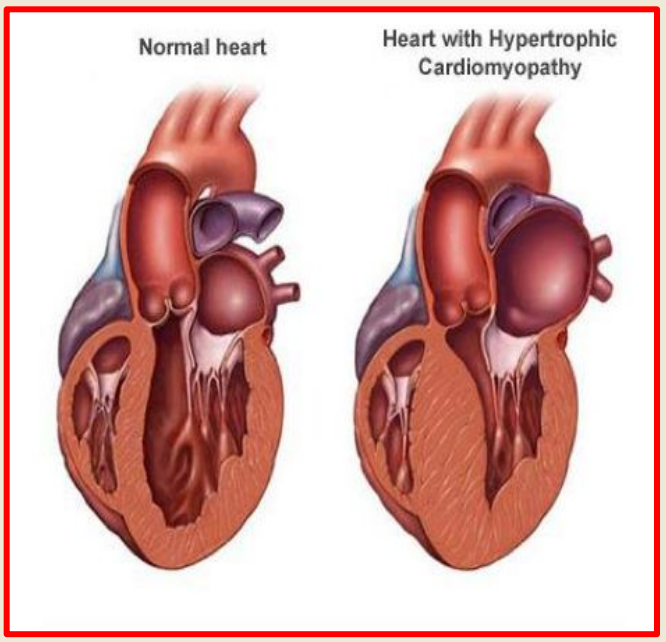
Types:

– Dilated cardiomyopathy (58%)
– Hypertrophic cardiomyopathy (30%)
– Restrictive cardiomyopathy (5%)
– Arrhythmogenic Right Ventricular Cardiomyopathy ARVC (5%)

Dilated Cardiomyopathy



Hypertrophic Cardiomyopathy



Extra from Nelson

TABLE 142.2 Dysrhythmias in Children

TYPE	ELECTROCARDIOGRAM CHARACTERISTICS	TREATMENT
Supraventricular tachycardia	Rate usually >220 beats/min (range, 180-320 beats/min); abnormal atrial rate for age; P waves may be present and are related to QRS complex; normal, narrow QRS complexes unless aberrant conduction is present	Increase vagal tone (bag of ice water to face, Valsalva maneuver); adenosine; digoxin; sotalol; electrical cardioversion if acutely ill; catheter ablation
Atrial flutter	Atrial rate usually 300 beats/min, with varying degrees of block; sawtooth flutter waves	Digoxin, sotalol, cardioversion
Premature ventricular contraction	Premature, wide, unusually shaped QRS complex, with large inverted T wave	None if normal heart and if premature ventricular contractions disappear on exercise; lidocaine, procainamide
Ventricular tachycardia	>3 Premature ventricular beats; AV dissociation; fusion beats, blocked retrograde AV conduction; sustained if >30 sec; rate 120-240 beats/min	Lidocaine, amiodarone, procainamide, propranolol, cardioversion
Ventricular fibrillation	No distinct QRS complex or T waves; irregular undulations with varied amplitude and contour, no conducted pulse	Nonsynchronized cardioversion
Complete heart block	Atria and ventricles have independent pacemakers; AV dissociation; escape-pacemaker is at atrioventricular junction if congenital	Awake rate <55 beats/min in neonate or <40 beats/min in adolescent, or hemodynamic instability requires permanent pacemaker
First-degree heart block	Prolonged PR interval for age	Observe, obtain digoxin level if on therapy
Mobitz type I (Wenckebach) second-degree heart block	Progressive lengthening of PR interval until P wave is not followed by conducted QRS complex	Observe, correct underlying electrolyte or other abnormalities
Mobitz type II second-degree heart block	Sudden nonconduction of P wave with loss of QRS complex without progressive PR interval lengthening	Consider pacemaker
Sinus tachycardia	Rate <240 beats/min	Treat cause (fever), remove sympathomimetic drugs

TABLE 88.3 Complications of Kawasaki Disease

Coronary artery thrombosis
Peripheral artery aneurysm
Coronary artery aneurysms
Myocardial infarction
Myopericarditis
Heart failure
Hydrops of gallbladder
Aseptic meningitis
Irritability
Arthritis
Sterile pyuria (urethritis)
Thrombocytosis (late)
Diarrhea
Pancreatitis
Peripheral gangrene

Questions

1- The leading causative agents for endocarditis in pediatric patients are
A. group D enterococci
B. viridans-type streptococci
C. Pseudomonas aeruginosa
D. fungal organisms
E. Serratia marcescens

2- The following is a major Duke criterion for the diagnosis of endocarditis
A. new valve regurgitant flow by echocardiography
B. Osler nodes
C. single positive blood culture
D. serologic evidence of infection
E. high erythrocyte sedimentation rate

3- In Asia, the following hepatitis virus appears to be a significant cause of viral myocarditis
A. hepatitis A virus
B. hepatitis B virus
C. hepatitis C virus
D. hepatitis D virus
E. hepatitis E virus

4- Despite the use of antibiotic agents, mortality remains high, in the range of 20-25%.
Serious morbidity occurs in 50-60% of children with documented infective endocarditis.
Of the following, the MOST common morbidity is
A. heart failure
B. pulmonary emboli
C. mycotic aneurysms
D. acquired ventricular septal defect

5- The following cardiac lesions are at increased risk for bacterial endocarditis EXCEPT
A. mitral insufficiency
B. aortic stenosis
C. atrial septal defect secundum
D. coarctation of the aorta
E. patent ductus arteriosus

Answers

1- **(B).** Viridans-type streptococci (α-hemolytic streptococci) and Staphylococcus
aureus remain the leading causative agents for endocarditis in pediatric patients.

2- **(A).** The Duke criteria help in the diagnosis of endocarditis. Major criteria include
(1) positive blood cultures (2 separate cultures for a usual pathogen, 2 or more for less
typical pathogens), and (2) evidence of endocarditis on echocardiography (intracardiac
mass on a valve or other site, regurgitant flow near a prosthesis, abscess, partial
dehiscence of prosthetic valves, or new valve regurgitant flow).

3- **(C).** Coxsackievirus and other enteroviruses, adenovirus, parvovirus, Epstein-Barr
virus, parechovirus, influenza virus, and cytomegalovirus are the most common causative agents in children, though most known viral agents have been reported. In Asia, hepatitis C virus appears to be significant as well.

4-**(A).** The most common is heart failure caused by vegetations involving the aortic or mitral valve. Myocardial abscesses and toxic myocarditis may also lead to heart failure without characteristic changes in auscultatory findings and, occasionally, to life- threatening arrhythmias.

5- **(C).**