Acquired Heart Disease

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Acquired Heart disease

- Disease affecting cardiac tissue and function which does not have its inception at birth and usually is secondary to an extraneous agent.
Types of Acquired Heart Disease

- Ischemic / hypoxic
- Hypertensive
- Infectious
- Inflammatory
- Metabolic
- Nutritional
- Traumatic
Ischemic / Hypoxic

- Coronary occlusion
  - Atherosclerosis
  - Kawasaki Disease
  - Sickle cell anemia
- Hypoperfusion
  - Surgical ischemic arrest
  - Severe hypotension
- Asphyxia
Hypertensive

- Systemic hypertension
- Pulmonary hypertension
Infectious

- Pericarditis
- Myocarditis
- Endocarditis
Inflammatory

- Post-pericardiotomy Syndrome
- Rheumatic fever
- Collagen vascular Diseases
Metabolic

- Endocrine adenopathy
  - Adrenal
  - Pituitary
  - Pancreatic
  - Thyroid
  - Parathyroid

- Storage diseases
  - Glycogen
  - Mucopolysaccharides
Nutritional

- Nutritional deficiencies
  - Starvation
  - Vitamin
  - Mineral
  - Carnitine

- Nutritional Excesses
  - Obesity
  - Vitamin
  - Mineral
Traumatic

- Penetrating
- Blunt
• Kawasaki Disease
• Pericarditis and post-pericardiotomy syndrome
• Myocarditis / congestive Cardiomyopathy
• Infectious Endocarditis
• Rheumatic heart disease
Kawasaki Disease

- Recognized in 1970’s
- Inflammatory disease of unknown etiology
- 9.2/100,000 cases per year; usually <4 y/o
- Winter and spring; 3yr”epidemics”
- Asiatics and blacks > white: 9&1.5/1
Kawasaki’s Disease Pathophysiology

- Immunoregulatory anomalies
  - Activation of T and B lymphocytes
  - Production of immunoglobulins and cytokines
  - Wide spread immune reaction
- Generalized microvasculitis
- Myocardial and pericardial inflammation
- Coronary vasculitis
Kawasaki’s Disease Clinical Manifestations

- Fever of 5+ days duration

- Physical findings
  - Polymorphous rash
  - Non-purulent conjunctivitis
  - Erythema of oral membranes including tongue
  - Indurative edema of hands and feet
  - Cervical lymphadenopathy

- Acute and often severe toxic presentation

- Multi-organ involvement
Kawasaki’s Disease Laboratory Findings

- Elevated acute phase reactants
- Elevated ESR
- Elevated Platelets
- Myocardial dysfunction
- Pericardial effusion
- Coronary thickening ---> coronary dilatation and aneurysm
Kawasaki's Disease- Cardiovascular Stages

• 20% of untreated; 2-4% with treatment
• Stage 1: Week 1-2
  – Microvasculitis
  – Peri, myo, and endocarditis
  – Endocarditis and perivasculitis of coronaries
• Stage 2: Week 1.5-3
  – Vasculitis of coronaries with aneurysms and thrombi
  – Intimal proliferation of coronaries
  – Peri, myo-, and endocarditis
• Stage 3: Week 4-5
  – Scarring and intimal thickening of Coronaries
  – Myocardial infarction
• Stage 4: >2m0
  – Advanced coronary artery disease
  – Myocardial Fibrosis
Echocardiographic Findings

- Acute phase:
  - Pericardial effusion
  - LV dysfunction
  - Diffuse coronary artery wall thickening and dilatation in 30-50%
- Coronary dilatation
  - $<5$y/o, lumen $>3$ mm
  - Sacular or fusiform
Treatment

- IVGG: 2g/kg over 24 hrs
- ASA:
  - 20-25 mg/kg/dose, q 6 hrs
    - until afebrile 2-3 days
  - 3-5 mg/kg/day
    - 6-8 weeks, until ESR and plt count normal
    - Indefinitely if coronary artery anomalies
Pericarditis / post-pericardiotomy Syndrome

- Inflammation(infection) of pericardial space
- Chest pain
- Friction rub
- Pericardial effusion
- Fever
- Elevated ESR
Pericarditis

- Viral
- Purulent
- Tuberculous
- Rheumatic
- Kawasaki
- Uremic
**Post - pericardiotomy Syndrome**

- 30%, if pericardium opened
- 1-2 weeks post surgery
- Etiology??
  - Viral Autoimmune
- Symptoms:
  - Fever
  - Chest pain
  - Friction rub
  - Pericardial effusion
Post - pericardiotomy syndrome

- Treatment:
  - ASA: 50-75 mg/kg/day; 4-6 weeks
  - Steroids: 2mg/kg/day; taper over 3-4 weeks
  - Diuretics (cautiously)
Cardiac tamponade

• Pathophysiology
  – Increase in pericardial fluid which elevates filling pressures, impedes ventricular filling and decreases cardiac output
  – Rapid small volume increase versus large chronic volume
**Cardiac Tamponade**

- Physical findings
  - Decreased heart sounds
  - Distended jugular veins
  - Pulsus paradoxus
    - >10 mmHg decrease in SBP with inspiration
    - Increased pooling of blood in pulmonary bed due to decreased LV filling
Cardiac tamponade

- **ECG:**
  - Low voltage
  - ST - T wave changes
  - Electrical alternans

- **CXR**
  - “Water - bottle” heart, if large volume
  - Normal, if acute

- **ECHO**
  - space between heart and pericardium
  - Swinging heart
  - Inspiratory variation in Doppler flows
Myocarditis / Congestive Cardiomyopathy

- Infection of myocardium with lymphocytic infiltration
- Degenerative process affecting myocytes
- Impairment of myocardial function
Myocarditis - Etiology

- Viral - Coxackievirus, ECHO, adeno, etc.
- Bacterial - Tuberculosis, strep, etc.
- Fungal - unusual
- Protozoan - Chaga’s disease (T.cruzi), malaria, toxoplasmosis
- Rickettsial
- Spirochetal
- Metazoal - trichinosis, echinococcosis, etc.
Congestive Cardiomyopathy - Etiology

- Infectious - viral
- Familial - duchenne’s
- Metabolic - glycogen storage
- Ischemic - Kawasaki
- Toxic - anthracyclines
- Nutritional - carnitine
Clinical Manifestation

- General malaise or viral syndrome
- Low cardiac output state (Shock)
- Gallop rhythm (mitral insufficiency)
- ST - T wave changes
- ECHO:
  - Reduced shortening fraction
  - Segmental wall motion anomalies
  - Valvar insufficiency
Course

Myocarditis

70-80% Mild-Mod CHF
20-30% Severe CHF

60-70% Recovery 10-20% Dil Cardiomyo 10% Death/Trans
Diagnosis

- Clinical findings
- Identification of etiologic agent
- Endomyocardial biopsy
  - Lymphocytic infiltrate
  - Etiologic agent
  - Necrosis
  - “Staging”
Treatment

• Symptomatic
  – inotropes
  – Diuretics
  – afterload reduction

• Correction of etiology

• Immunosuppression
  – Steroids
  – Anti-virals
  – Cyclosporin
  – Interferon

• Transplantation
Infective Endocarditis

- Microbial infection of endocardial surface of heart - valves or wall
- “Acute” (virulent) / “subacute” (prolonged)
- 1:1800 to 1:4500 ped cases: admissions
- Any age; greater in 5th decade
- Pre-v. post-antibiotic era - no change
- Factors:
  - Better diagnosis
  - Drug abuse
  - Treatment modalities
Etiology

- Alpha hemolytic strep: most common (>60%); prolonged
- Staph aureus: 2nd most common (20%); virulent
- Beta hemolytic strep: uncommon
- Coagulase negative Staph: increasing
- Candida
Risk Factors

• High Risk
  – Prosthetic valves
  – Surgical shunts
  – Indwelling catheters
  – Previous SBE

• Moderate Risk
  – PDA
  – VSD
  – ASD (not seconundum)
  – Bicuspid aortic valve
  – RHD
  – MVP with MR
Clinical Manifestations

- Fever
  - High (Staph)
  - Low (Strep)
- “Viral syndrome”
- New murmur
- CHF
- Petechiae
- Inc ESR, anemia, hematuria
Diagnosis

- Blood Culture
  - Positive off antibiotics
  - 5-8% negative cultures
  - 2-3 sets over 24 hrs; (as much as possible)

- ECHO:
  - Vegetations
  - Valve insufficiency
Treatment

- Specific anti-microbial
- 4-6 weeks IV
- 2 weeks w/wo P.O.
- Surgery, esp. prosthetic valves
Prophylaxis

- AHA guidelines
  - Amoxicillin - oral, upper resp procedures
  - Clindamycin (penicillin allergic)
  - Amp and gent or vancomycin - GU or GI
This patient has a heart lesion ( ) that require bacterial endocarditis prophylaxis.
For your convenience a schedule has been given below.

Cardiac Conditions Associated With Endocarditis

Endocarditis Prophylaxis Recommended

High-risk category
- Complex cyanotic congenital heart diseases (e.g., single ventricle defects, transposition of the great arteries, tetralogy of Fallot)
- Surgically constructed systemic-pulmonary shunts of conduits
- Mechanical cardiac valves, including bioprosthetic and homograft valves
- Previous bacterial endocarditis

Moderate-risk category
- Most other congenital cardiac malformations (other than above and below)
- Acquired valve dysfunction (e.g., rheumatic heart disease)
- Hypertrophic cardiomyopathy
- Initial valve replacement with valve prosthesis on native valves
- Prosthetic heart valves

Negligible-risk category (no greater risk than the general population)
- Initial recurrence after valvular defect (ACE I)
- Surgery repair or valve replacement defect, ventriculotomy defect, or patent ductus arteriosus (without residual shunt beyond 6 months)
- Mitral valve prolapse without valve regurgitation
- Muscle, pericardial, or connective tissue defects
- Previous Kawasaki disease without valve dysfunction
- Previous rheumatic fever without valve dysfunction
- Cardiac pacemakers, cardioverter and defibrillators
- Intracavitary catheter or pacemaker wires

Endocarditis Prophylaxis Not Recommended

Prophylactic Regimens for Dental, Oral, Respiratory Tract, or Esophageal Procedures

<table>
<thead>
<tr>
<th>Situation</th>
<th>Agent</th>
<th>Regimen</th>
</tr>
</thead>
<tbody>
<tr>
<td>Standard general prophylaxis</td>
<td>Amoxicillin</td>
<td>Adult: 2 g orally 1 hr before procedure Children: 50 mg/kg orally 1 hr before procedure</td>
</tr>
<tr>
<td>Unusual or late cardiac malformations</td>
<td>Ampicillin</td>
<td>Adult: 2 g IM or IV, within 30 min before procedure Children: 50 mg/kg IM or IV, within 30 min before procedure</td>
</tr>
<tr>
<td>Angina or to Percutaneous</td>
<td>Clarithromycin</td>
<td>Adult: 500 mg, orally 1 hr before procedure Children: 25 mg/kg, orally 1 hr before procedure</td>
</tr>
<tr>
<td>Cardiac surgery or Catheterization</td>
<td>Clarithromycin</td>
<td>Adult: 500 mg, orally 1 hr before procedure Children: 25 mg/kg, orally 1 hr before procedure</td>
</tr>
<tr>
<td>Allergy to penicillin and cannot take alternative medications</td>
<td>Clarithromycin</td>
<td>Adult: 500 mg IM or IV within 30 min before procedure Children: 25 mg/kg IM or IV within 30 min before procedure</td>
</tr>
<tr>
<td>Cardiac #</td>
<td>Clindamycin</td>
<td>Adult: 1 g IM or IV within 30 min before procedure Children: 50 mg/kg IM or IV within 30 min before procedure</td>
</tr>
</tbody>
</table>

Prophylactic Regimens for Genitourinary Gastrointestinal (Excluding Esophageal) Procedures

<table>
<thead>
<tr>
<th>Situation</th>
<th>Agent</th>
<th>Regimen</th>
</tr>
</thead>
<tbody>
<tr>
<td>High-risk patients</td>
<td>Ampicillin or Gentamicin</td>
<td>Adults: ampicillin 3 g, intramuscularly, IM or intravenously, IV (p.o. dose: 1.5 mg/kg, not to exceed 3 g) plus gentamicin 1.5 mg/kg within 30 min of starting the procedure. Children: ampicillin 1 g, IM or intravenously, within 30 min of starting the procedure.</td>
</tr>
<tr>
<td>Moderate-risk patients</td>
<td>Amoxicillin</td>
<td>Adult: amoxicillin 1.5 g orally 1 hr before procedure, or ampicillin 1.5 g IM within 30 min of starting the procedure. Children: amoxicillin 0.5 mg/kg IM or IV given 1.5 mg/kg within 30 min of starting the procedure.</td>
</tr>
<tr>
<td>Moderate-risk patients</td>
<td>Carbenicillin</td>
<td>Adult: Carbenicillin 50 mg/kg IM or IV within 30 min of starting the procedure Children: Carbenicillin 20 mg/kg IM or IV within 30 min of starting the procedure.</td>
</tr>
</tbody>
</table>

* Data and comments should be noted on dental chart
* Penicillin should not be used in individuals with immediate-type hypersensitivity reaction (urticaria, angioedema, anaphylaxis) to penicillin

Adapted from: Prevention of Bacterial Endocarditis - Recommendations by the American Heart Association. JAMA June 11, 1997 - Vol 277, No. 22
Rheumatic Fever

- Most common cause of acquired heart disease in children (5-15 y peak of 8 y)
- USA: 0.5-3.0/100,000 (1900: 100-200/100,000)
- Post- infectious connective tissue response in susceptible host
- Group A beta- hemolytic streptococcus infection of the pharynx
- F/H of RHD and low socioeconomic status.
Pathophysiology

- 1960 Kaplan and coworkers show an antigenic “similarity” between strep cell walls and myocardium.
- An autoimmune response to strep group A with cross reaction to myocardium.
Inflammation of brain causing involuntary limb movements

Fever

Initial stage: tonsillitis or sore throat

Skin rashes

Inflammation of heart valves

Inflammation of joints

Nervous system attacked

Nodules over joints
Jones Criteria

- **Major**
  - Carditis: 40-50%
  - Arthritis: 60-85%
  - Chorea; 15%
  - Erythema marginatum: 10%
  - Subcutaneous nodules; 2-10%

- **Minor**
  - Clinical: Arthralgia, fever and H/O RF or RHD
  - Laboratory: Elevated ESR, C-reactive protein and Prolonged PR interval

- **Must have evidence for strep infection** (Inc ASO, +ve culture or recent scarlet fever).
Arthritis

- Most common manifestation
- Monoarticular, usually large joints
- Migratory or Fleeting
- Good response to ASA
- No residual effect
Carditis

- 1-2 weeks after Strep; may be delayed
- Inflammation of:
  - Endocardium: Valves
  - Myocardium (Tachycardia, cardiomegally and Heart failure).
  - Pericardium: Rub or PE (rare)
- Prior attack predisposes to recurrence
- The only feature which cause permanent damage.
Valvular Involvement

- Mitral
  - Insufficiency; mild to severe (Carey-Coombs)
  - Congestive heart failure
  - Stenosis, late

- Aortic
  - Insufficiency
  - Less common but more severe
Chorea

- Sydenham’s chorea or St. Vitus’ dance
- Prepubertal girls (8-12y)
- First emotional lability and personality changes
- Followed by loss of motor coordination - characteristic spontaneous, purposeless movement and motor weakness
- It is often an isolated manifestation.
Erythema Marginatum

- Nonpruritic serpiginous or annular erythematous rashes.
- Most prominent on the trunk and inner proximal portions of the extremities.
Subcutaneous Nodules

- Hard, painless, nonpruritic, freely movable, swelling, 0.2-2.0 cm in diameter.
- Symmetrical, single or clusters
- On the extensor surfaces of both large and small joints, scalp or along the spine.
Investigations

- Elevated ESR
- ASO > 333 Todd units
- Throat culture
- Leukocytosis
- Hypochromic microcytic anemia
- ECG: first degree heart block, arrhythmia.
- CXR: progressive cardiac enlargement.
- ECHO.
Treatment

- Cardiac supportive
  - Bed rest 1-2 W
  - Immobilise inflamed joints
  - ASA 100 mg/kg/d (level 20 mg/100 ml) - side effects (after diagnosis of RF is made)
  - Benz Penicillin G 0.6-1.2 million U for eradication
  - Steroids Prednisone (severe carditis) 2 mg/kg /d 2-4 W ???
  - Treatment of CHF- Digoxin toxicity
Prevention

- Any pt with documented H/O RF
- Prophylaxis after attack: until 21-25y of age
- Benzathine Penicillin 1.2 million U IM q 28 d. or Erythromycin 250 mg BID for penicillin allergies