At the end of this presentation the participant will be able to:

1. Describe the clinical findings, prevention and treatment of infective endocarditis
2. Explain the diagnosis and management of children with acute rheumatic fever and Kawasaki disease.
3. Diagnose and treat pericarditis
4. List the causes of dilated cardiomyopathy
5. Understand the role of lipid management in preventive cardiology

1. A 9-year-old boy with a bicuspid aortic valve develops subacute bacterial endocarditis after extraction of a tooth. The organism MOST likely causing this patient’s endocarditis is:
   A. Candida albicans
   B. Escherichia coli
   C. Haemophilus influenzae
   D. Staphylococcus aureus
   E. Streptococcus viridans

INFECTIVE ENDOCARDITIS

MICROBIOLOGY

- Streptococcus viridans 32 - 43%
- Staphylococcus aureus 27 - 33%
- Strep. Faecalis (enterococcus) 4 - 7%
- Pneumococci 3 - 7%
- Staphylococcus epidermidis 2 - 12%
- Gram negative (HACEK) 4 - 5%
- Fungi 0 - 12%
- Culture negative 5 - 7%
INFECTIVE ENDOCARDITIS

CLINICAL MANIFESTATIONS

• History: heart defect; recent dental procedure
• Fever 56 – 100%
• Anorexia, weight loss, malaise 40 – 83%
• Splenomegaly 36 – 67%
• New / changing murmur 9 – 44%
• Skin manifestations, embolic events 15 – 50%

INFECTIVE ENDOCARDITIS

TESTING

• Positive blood culture 68 – 98%
• Elevated ESR 71 – 94%
• Anemia 19 – 79%
• Microscopic hematuria 28 – 47%
• Echocardiography
• Serum bactericidal concentrations

INFECTIVE ENDOCARDITIS

Modified Duke Criteria for Diagnosis of IE

Major Criteria

• Positive blood culture (growth on 2 occasions of microorganisms “typical for” IE OR persistently positive cultures for an organism “consistent with” IE)
• Positive echocardiogram (vegetation, paravalvular abscess, or valve dehiscence after surgery)
• New valvular regurgitation (by auscultation)

Clinical criteria:
• two major, or one major and three minor, or five minor criteria
INFECTIVE ENDOCARDITIS

Modified Duke Criteria for Diagnosis of IE

Minor Criteria

- Predisposing heart condition
- IV drug use
- Fever (>38º C)
- Major arterial emboli
- Septic pulmonary infarcts
- Mycotic aneurysm
- Intracranial hemorrhage
- Conjunctival hemorrhage

Clinical criteria:
- two major, or one major and three minor, or five minor criteria

INFECTIVE ENDOCARDITIS

Modified Duke Criteria for Diagnosis of IE

Definite

- Pathologic evidence of intracardiac or embolized vegetation or intracardiac abscess OR
- Clinical criteria: two major, or one major and three minor, or five minor criteria

Possible

- One major and one minor or three minor criteria

INFECTIVE ENDOCARDITIS

Modified Duke Criteria for Diagnosis of IE

Rejected

- Firm alternate diagnosis
- “IE syndrome” resolved within 4 days of antibiotic therapy
- No pathologic evidence of IE at surgery or autopsy within 4 days of antibiotic therapy
- Case does not meet “possible IE” criteria
INFECTIVE ENDOCARDITIS

PROCEDURES FOR WHICH PROPHYLAXIS IS RECOMMENDED

• All dental procedures that involve manipulation of gingival tissue or the periapical region of teeth or perforation of the oral mucosa
• Tonsillectomy and/or adenoidectomy
• Surgical procedures or biopsy of respiratory mucosa
• Incision and drainage of infected tissues

2. Patients with which of the following diagnoses should receive antibiotic prophylaxis for dental procedures:
   A. Coarctation of the aorta
   B. Secundum atrial septal defect
   C. Post-op ventricular septal defect with patch leak
   D. Aortic stenosis
   E. Patent ductus arteriosus

INFECTIVE ENDOCARDITIS

PROPHYLAXIS FOR DENTAL PROCEDURES

• Prosthetic cardiac valve
• Previous infective endocarditis
INFECTIVE ENDOCARDITIS
PROPHYLAXIS IN CONGENITAL HEART DISEASE

- Unrepaired cyanotic CHD, including shunts and conduits
- Completely repaired CHD with prosthetic material within 6 months
- Repaired CHD with residual defects at the site or adjacent to the site of prosthetic patch or device.
- Cardiac transplant recipients with valvulopathy
- Establish an accurate cardiac diagnosis
- Follow the guidelines of the American Heart Association.

*Except for the conditions listed above, antibiotic prophylaxis is no longer recommended for any other form of CHD.*

3. A 10-year-old boy with a mechanical prosthetic aortic valve is scheduled for routine dental care, including examination and cleaning.

Among the following, the agent of CHOICE for standard prophylaxis against bacterial endocarditis is:

A. Amoxicillin  
B. Cefaclor  
C. Cloxacillin  
D. Erythromycin  
E. Penicillin

4. A 4-year-old is seen 10 days following an upper respiratory infection. She has swollen painful knees and ankles and temperature of 104°F (40°C). The cardiac examination is significant for a grade IV/VI systolic murmur at the apex.

These findings are consistent with:

A. Acute rheumatic fever  
B. Septic arthritis  
C. Juvenile rheumatoid arthritis  
D. Pericarditis  
E. Kawasaki disease
ACUTE RHEUMATIC FEVER

ETIOLOGY

• Immunologic lesion
• Delayed sequela of group A type 18 (M-18) β-hemolytic streptococcal infection of the pharynx (not skin)

ACUTE RHEUMATIC FEVER

REVISED JONES CRITERIA

<table>
<thead>
<tr>
<th>MAJOR</th>
<th>MINOR</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Joints (60-85%)</td>
<td>Arthralgia</td>
</tr>
<tr>
<td>• Carditis (40-50%)</td>
<td>Prolonged PR int.</td>
</tr>
<tr>
<td>• Nodules (2-10%)</td>
<td>Fever</td>
</tr>
<tr>
<td>• Erythema marginatum (10%)</td>
<td>Previous ARF</td>
</tr>
<tr>
<td>• Sydenham’s chorea (15%)</td>
<td>↑ acute phase reactants</td>
</tr>
</tbody>
</table>

2 major \ OR \ 1 major and 2 minor \ PLUS \ recent strep.

ACUTE RHEUMATIC FEVER

TREATMENT

• Benzathine penicillin G 0.6 – 1.2 million Units IM
• Withhold anti-inflammatory meds until diagnosis established
• Aspirin 100 mg/kg/day in 4 – 6 divided doses
• For severe carditis: prednisone 2 mg/kg/day
• Bedrest during the inflammatory process
### ACUTE RHEUMATIC FEVER

#### SECONDARY PREVENTION

- Benzathine penicillin G 1.2 million Units IM q 21-28 d
- Penicillin V 250 mg PO bid
- Sulfadiazine 500 – 1000 mg PO daily
- Allergic to penicillin: Macrolide or azalide
- Continue prophylaxis through childhood (longer with carditis)

### ACUTE RHEUMATIC FEVER

#### Duration of Secondary Rheumatic Fever Prophylaxis

<table>
<thead>
<tr>
<th>Category</th>
<th>Duration After Last Attack</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rheumatic fever with carditis and residual heart disease</td>
<td>10 years or until 40 years of age (whichever is longer)</td>
</tr>
<tr>
<td>Rheumatic fever with carditis but no residual heart disease</td>
<td>10 years or until 21 years of age (whichever is longer)</td>
</tr>
<tr>
<td>Rheumatic fever without carditis</td>
<td>5 years or until 21 years of age (whichever is longer)</td>
</tr>
</tbody>
</table>

*Clinical or echocardiographic evidence.


5. **The MOST appropriate treatment for the above patient would be:**

   - A. Intravenous steroids
   - B. Oral aspirin (ASA) and prednisone
   - C. Oral aspirin and IM penicillin
   - D. Intravenous digoxin
   - E. Intravenous furosemide
6. Regarding acute rheumatic fever arthralgia is a major criterion.
7. Patients with Sydenham’s chorea should receive penicillin.
8. Erythema multiforme is a major criterion.
9. A systolic ejection murmur is evidence of carditis.

10. A 2-year-old boy presents with 5 days of fever, swollen hands and feet, strawberry tongue, maculopapular rash and conjunctivitis. The MOST likely diagnosis is:
   A. Lyme disease
   B. Kawasaki disease
   C. Stevens-Johnson syndrome
   D. Mononucleosis
   E. Acute rheumatic fever

**KAWASAKI DISEASE**

**DIAGNOSTIC CRITERIA**
Fever for five or more days and at least 4 of 5 features

- Skin changes (rash)
- Extremity changes - erythema, induration, desquamation
- Oropharyngeal changes
- Cervical lymphadenopathy (>1.5 cm)
- Bilateral nonpurulent conjunctivitis
KAWASAKI DISEASE
CARDIAC SYMPTOMS AND FINDINGS

Early
• Myocarditis and/or pericarditis
• Conduction abnormalities or arrhythmias

Late
• Coronary artery aneurysms (15-20%)
• Aneurysms of other arteries

KAWASAKI DISEASE
ASSOCIATED NON-CARDIAC SYMPTOMS OR FINDINGS

• GI: diarrhea, hydrops of gallbladder, hepatitis
• Ocular: uveitis
• Genitourinary: Urethritis
• CNS: aseptic meningitis
• Musculoskeletal: arthralgia, arthritis, myositis
• Laboratory: leukocytosis, anemia, thrombocytosis

KAWASAKI DISEASE
LABORATORY FINDINGS

• Leukocytosis
• Anemia
• Thrombocytosis
• Elevated acute phase reactants
• Elevated liver enzymes and bilirubin
• Hyponatremia
11. Treatment should include:
   A. Hospitalization
   B. Aspirin
   C. Intravenous immune globulin
   D. All of the above
   E. None of the above

KAWASAKI DISEASE
TREATMENT RECOMMENDATIONS
• IVIG 2 G/kg – one dose
• High dose aspirin: 75-100 mg/kg/day
• Reduce aspirin to 3-5 mg/kg/day once afebrile; continue for 6-8 weeks
• No firm evidence for late treatment, treatment in the presence of established aneurysms, or recurrent symptoms

12. A 12-year-old boy presents with a 24 hour history of sharp, pleuritic chest pain worse in the supine position. His temperature is 38.5° and a pericardial friction rub is heard. The MOST likely diagnosis is:
   A. Musculoskeletal chest pain
   B. Pericarditis
   C. Bacterial endocarditis
   D. Mycoplasma pneumonia
   E. Pulmonary embolus
PERICARDITIS

CLINICAL FEATURES

• Chest pain
• Fever
• Pericardial friction rub
• Tamponade: JVD, ↑HR, ↓BP, muffled heart tones
• Sepsis: purulent pericarditis

PERICARDITIS

ETIOLOGY

• Idiopathic
• Viral
• Acute rheumatic fever
• Bacterial, including tuberculosis
• Collagen vascular diseases (SLE)
• Post-op: post cardiotomy syndrome
• Malignancy
• Uremia

13. The most useful diagnostic test is:
A. Chest x-ray
B. Sed rate
C. ECG
D. Arterial blood gas
E. Complete blood count
14. Treatment should include:
   A. Nonsteroidal anti-inflammatory medication
   B. Corticosteroids
   C. Pericardiocentesis
   D. Hospitalization
   E. All of the above
DILATED CARDIOMYOPATHY

DEFINITION
Weakened myocardium with reduced systolic function. Pathological examination generally demonstrates myocardial fibrosis and necrosis.

DILATED CARDIOMYOPATHY

ETIOLOGY
• Neuromuscular disorders
• Viruses - Coxsackie B, adenovirus, parvovirus
• Familial
• Toxic, or metabolic agents: adriamycin >500 mg/M2 total dose, especially with adjunctive radiation therapy
• Coronary abnormalities
• Metabolic, nutritional, endocrine disorders

DILATED CARDIOMYOPATHY

DIAGNOSIS
• History: prior viral illness, family history
• Clinical Exam: congestive heart failure, weakness, and fatigue.
• ECG: left ventricular hypertrophy and ST-T wave changes, LAE
• Chest x-ray: cardiomegaly with increased pulmonary venous markings.
• Echocardiogram: dilated left ventricle with diminished systolic ventricular function.
DILATED CARDIOMYOPATHY

MANAGEMENT

• ACE inhibitors
• Beta blockers: carvedilol
• Diuretics ?
• Digoxin ?
• Anticoagulation
• Cardiac transplantation

HYPERTROPHIC CARDIOMYOPATHY

• Asymmetric hypertrophy, especially the septum
• LVOT obstruction can occur
• Normal systolic LV function; impaired diastolic function
• Genetically transmitted as an autosomal dominant trait

HYPERTROPHIC CARDIOMYOPATHY

CLINICAL MANIFESTATIONS

• Usually undetected until adolescence or young adulthood
• Positive family history in 30%
• Symptoms: exercise intolerance, syncope, arrhythmia
• Murmurs: LVOT ejection murmur (louder upright), MR
• ECG: LVH, ST-T wave changes, arrhythmia
• Echocardiogram: diagnostic, asymmetric hypertrophy
HYPERTROPHIC CARDIOMYOPATHY

MANAGEMENT

• Activity restrictions
• Beta blockers, calcium channel blockers
• Avoid digoxin and positive inotropic agents
• Avoid tachycardia, hypovolemia and hypotension
• Septal myomectomy for obstruction
• Defibrillator for high risk patients

PREVENTIVE CARDIOLOGY

RISK FACTORS FOR ATHEROSCLEROSIS

<table>
<thead>
<tr>
<th>Unmodifiable</th>
<th>Modifiable</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male sex</td>
<td>Diet</td>
</tr>
<tr>
<td>Advancing age</td>
<td>Sedentary lifestyle</td>
</tr>
<tr>
<td>Coronary anatomy</td>
<td>Smoking</td>
</tr>
<tr>
<td>Positive family history</td>
<td>Obesity</td>
</tr>
<tr>
<td>Dyslipidemia</td>
<td>Insulin resistance</td>
</tr>
<tr>
<td>Hypertension</td>
<td>Hypertension</td>
</tr>
<tr>
<td>Diabetes</td>
<td>Dyslipidemia</td>
</tr>
<tr>
<td>Atherosclerosis</td>
<td></td>
</tr>
</tbody>
</table>
HYPERLIPIDEMIA IN CHILDREN

<table>
<thead>
<tr>
<th>Total Cholesterol</th>
<th>LDL Cholesterol</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Desirable</td>
<td>&lt;170 mg/dL</td>
<td>&lt;110 mg/dL</td>
</tr>
<tr>
<td>Borderline</td>
<td>170-200 mg/dL</td>
<td>110-130 mg/dL</td>
</tr>
<tr>
<td>High</td>
<td>≥200 mg/dL</td>
<td>≥130 mg/dL</td>
</tr>
<tr>
<td></td>
<td></td>
<td>≥190 mg/dL</td>
</tr>
</tbody>
</table>

Other concerns: triglycerides > 150 mg/dl, HDL-C < 35 mg/dl


PREVENTIVE CARDIOLOGY

PEDIATRIC DISEASES WITH CV RISK FACTORS

- Familial hypercholesterolemia
- Diabetes mellitus, type 1 and type 2
- Chronic kidney disease
- Post-heart transplantation
- Kawasaki disease
- Chronic inflammatory disease
- Congenital heart disease
- Childhood cancer survivors

FIGURE 1 Directions: Step 1: risk stratification by disease process (Table 1)
HYPERLIPIDEMIA: THERAPY

- Primary: lifestyle changes (diet, weight, activity)
- Medication (>8 years old or onset menses):
  - Consider if LDL
    - $\geq 130$ diabetes
    - $\geq 160$ family history or $\geq 2$ add’l risk factors
    - $\geq 190$ all patients
  - Statins (contraindicated during pregnancy)
  - (Not for isolated elevated triglycerides)


MYOCARDITIS

Definition: inflammation of the myocardium

Etiology:
- Viral: Coxsackie, rhinovirus, adenovirus, influenza, EB virus, CMV, parvovirus, HSV type 6.
- Other infections: rickettsia, bacteria, fungi, and parasites.
- Acute rheumatic fever
- Inflammatory and collagen vascular diseases
- Toxins

Clinical Manifestations:
- History of preceding upper respiratory or other viral prodrome
- Nonspecific respiratory or gastrointestinal symptoms
- Signs of congestive heart failure, specifically an S3 gallop.

ECG changes: low QRS voltage, ST-T wave changes, conduction disturbances and pseudoinfarction.

Cardiac MRI: regional or global myocardial signal increase (T2), early gadolinium enhancement, at least one focal lesion with nonischemic regional distribution on late gadolinium enhancement.
MYOCARDITIS

Treatment:
Supportive and symptomatic.
Activity restrictions until signs of inflammation resolve.
Diuretics (furosemide 1 mg/kg) may be given.
Oxygen
Intravenous inotropic agents (e.g. dopamine, dobutamine).
Immunosuppressive agents, corticosteroids, or IVIG have not been reproducibly demonstrated to be efficacious and their use remains controversial.

SUGGESTED READINGS


ANSWERS FOR ACQUIRED HEART DISEASE