OBJECTIVES

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

1. Describe the characteristics of innocent cardiac murmurs
2. List the causes of chest pain in children
3. Evaluate the cyanotic newborn
4. Identify causes, symptoms and treatment of congestive heart failure and cardiogenic shock in infants and children
5. Recognize and treat hypercyanotic spells

HEART MURMURS IN CHILDREN

Auscultation Basics

- Stethoscope
- Environment
- Examine all areas
- Changes with respiration and position
HEART MURMURS IN CHILDREN

Innocent Murmurs

- Vibratory (Still's) murmur
- Pulmonary flow murmur
- Peripheral pulmonary arterial stenosis murmur
- Aortic systolic murmur
- Venous hum

INNOCENT MURMURS IN INFANTS AND CHILDREN

<table>
<thead>
<tr>
<th>Murmur/PMS</th>
<th>Still's</th>
<th>Venous Hum</th>
<th>Pulmonary Flow</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pitch</td>
<td>Medium to high</td>
<td>Low</td>
<td>Low</td>
</tr>
<tr>
<td>Intensity</td>
<td>1/6</td>
<td>1/6</td>
<td>1/6</td>
</tr>
<tr>
<td>Heart Rate</td>
<td>Medium</td>
<td>Medium</td>
<td>Medium</td>
</tr>
<tr>
<td>Quality</td>
<td>Soft</td>
<td>Vibratory, Musical</td>
<td>Soft blowing</td>
</tr>
<tr>
<td>Location</td>
<td>1st &amp; 2nd ICS</td>
<td>RSB, LSB, back</td>
<td>2nd LICS</td>
</tr>
</tbody>
</table>

- Increased by:
  - Cardiac output
  - Supine, fever, exercise
  - Upright, Inspiration

- Decreased by:
  - Cardiac output
  - Standing, Valsalva
  - Supine, head turn, Jugular compression

Age Range:
- Birth to 2-3 months
- 1-10 years
- After age 10

- Confused with:
  - Pulmonary Branch stenosis
  - Hypertrophic Cardiomyopathy
  - PDA, ASD or Mild PS

HEART MURMURS IN CHILDREN

Pulmonary Branch Stenosis

- Systolic ejection murmur, grade 1-2/6
- Heard best in axillae and back
- Birth to 2-3 months
- Produced by relatively small size of pulm. arteries
- Confused with peripheral pulm stenosis or PDA
HEART MURMURS IN CHILDREN

Still's Murmur
• Systolic ejection murmur, grade 1-3/6
• “Musical”, “Buzzing”, vibratory
• Heard best at the mid to lower left sternal border
• One year to adolescence (also in infants)
• Increased by supine position, fever, or exercise
• Vibration in the left ventricular outflow tract
• Confused with hypertrophic cardiomyopathy

Venous Hum
• Continuous murmur, grade 1-3/6
• Heard best in the right or left infraclavicular area
• Two to ten years
• Increased in the upright position
• Abolished by jugular compression or head turn
• Produced by turbulent flow in the jugular veins
• Confused with patent ductus arteriosus

1. A loud systolic murmur is heard in a term newborn at 2 hours of age. Possibilities include all EXCEPT:
   A. Aortic stenosis
   B. Pulmonary stenosis
   C. Tricuspid regurgitation
   D. Large ventricular septal defect
   E. Mitral regurgitation
HEART MURMURS IN CHILDREN

“Innocent” Murmurs in the Newborn Infant

- Tricuspid regurgitation
- Closing patent ductus arteriosus
- Peripheral pulmonary stenosis
- Pulmonary flow
- Vibratory (Still’s)

HEART MURMURS IN CHILDREN

Six Cardinal Clinical Signs

- Pansystolic murmurs (VSD, TR, MR)
- Harsh murmurs (VSD, valve stenosis, outflow tract obstruction)
- Very loud murmurs (≥ grade 3)
- Murmurs heard at the upper left sternal border
- Systolic clicks (aortic or pulmonary stenosis, MVP)
- Abnormal S₂ (split-ASD, loud & single-pulm htn)

McCrindle, et al.
Arch Pediatr Adolesc Med 1996; 150:169
HEART MURMURS IN CHILDREN

When to Refer a Child with a Heart Murm

- All diastolic murmurs
- All pansystolic murmurs
- Late systolic murmurs
- Very loud murmurs (≥ grade 3)
- Continuous murmurs except venous hums
- Associated cardiac abnormalities

NEONATAL CYANOSIS
3. Arterial blood gas results from an infant with a cyanotic congenital heart defect should include:
A. Increased PO2 with crying
B. Increased PO2 with increased FiO2
C. Elevated PCO2
D. All of the above
E. None of the above

PULSE OXIMETRY NEWBORN SCREENING

- The screening is targeted toward healthy newborn infants in the newborn nursery.
- Screening should not be undertaken until 24 hours of life.
- Oxygen saturations should be obtained in the right hand and one foot.

- Pass: pulse oximetry reading of ≥95% in either extremity with a ≤3% absolute difference between the upper and lower extremity.
- Immediate evaluation: saturations <90%.
- In the event of a positive screening result, CCHD needs to be excluded with a diagnostic echocardiogram. Infectious and pulmonary causes of hypoxemia should also be excluded.
The proposed pulse-oximetry monitoring protocol based on results from the right hand (RH) and either foot (F).

SYNDROMES AND CHD

- Down (40-50%) AV Canal Defect, VSD, TOF
- Turner (35%) Coarctation, bicuspid aortic valve, AS
- Noonan (80-90%) Pulmonary stenosis, ASD, HCM
- Williams (60%) Supravalvar aortic stenosis, coarctation
  (del 7q11.23)
- DiGeorge (35%) Conotruncal malformations (Interrupted aortic arch, truncus arteriosus, TOF)
  (del 22q11)
- Alagille (95%) Pulm art stenosis, TOF, pulm stenosis

SYNDROMES AND CHD

- Alagille (arteriohepatic dysplasia) Peripheral pulmonary stenosis, PS, TOF
- Asplenia Complex cyanotic CHD
- Carpenter PDA, VSD
- CHARGE association VSD, ASD
- Cri-du-chat (5p-) Various congenital cardiac defects
- De Lange Tetralogy of Fallot, VSD
- Ellis van Creveld Common atrium (ASD)
- Fanconi PDA, VSD
- Fetal alcohol VSD, ASD, tetralogy of Fallot
- Fetal hydantoin ASD, VSD, coarctation
### Syndromes and CHD

<table>
<thead>
<tr>
<th>Syndrome/Condition</th>
<th>Associated Conditions</th>
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<tbody>
<tr>
<td>Goldenhar</td>
<td>Tetralogy of Fallot, ASD, VSD</td>
</tr>
<tr>
<td>Holt-Oram</td>
<td>Hypertrophic cardiomyopathy, VSD, TGA</td>
</tr>
<tr>
<td>Infant of a diabetic mother</td>
<td>Tetralogy of Fallot, VSD</td>
</tr>
<tr>
<td>Laurence-Moon-Biedl</td>
<td>Aortic root aneurysm, mitral prolapse</td>
</tr>
<tr>
<td>Marfan</td>
<td>Pulmonary stenosis</td>
</tr>
<tr>
<td>Multiple lentigines (leopard)</td>
<td>Complex CHD</td>
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<td>Polysyndrome</td>
<td>PDA, peripheral pulmonary stenosis</td>
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<tr>
<td>Rubinstein-Taybi</td>
<td>PDA</td>
</tr>
<tr>
<td>Smith-Lemli-Opitz</td>
<td>VSD, PDA</td>
</tr>
<tr>
<td>Thrombocytopenia-absent radius</td>
<td>ASD, tetralogy of Fallot</td>
</tr>
<tr>
<td>Trisomy D</td>
<td>VSD, PDA</td>
</tr>
<tr>
<td>Trisomy E</td>
<td>ASD, VSD</td>
</tr>
<tr>
<td>Wolf</td>
<td></td>
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</tbody>
</table>

### Family History and CV Disease

- Dilated Cardiomyopathy
- Hypertrophic Cardiomyopathy
- Marfan syndrome
- Muscular dystrophy
- Long QT syndrome (sudden death)
OBJECTIVE: Identification of “silent” cardiovascular abnormalities that can progress or cause sudden cardiac death.

- Personal history:
  - History of heart disease, including Kawasaki disease
  - Heart murmur
  - Systemic hypertension
  - Fatigue
  - Syncope/near-syncope
  - Excessive/unexplained exertional dyspnea
  - Exertional chest pain
  - Medication history
  - Illicit drug use (including performance enhancing drugs)

PHYSICAL EXAMINATION:
- Heart murmur (supine/standing)
- Femoral arterial pulses
- Stigmata of Marfan syndrome
- Brachial blood pressure measurement (sitting)

RED FLAGS:
- Syncope or near-syncope on exertion
- Chest pain on exertion
- Excessive dyspnea or fatigue with activity
- Family history: Marfan, cardiomyopathy, long QT syndrome, sudden death
- Irregular rhythm
- Weak or absent lower extremity pulses
- Hypertension
- Loud systolic murmur
- Any diastolic murmur
- Stigmata of genetic syndromes associated with CV disease

CONTRAINDICATIONS TO SPORTS PARTICIPATION

- Pulmonary vascular disease with cyanosis
- Severe pulmonary hypertension
- Excessive dyspnea or fatigue with activity
- Severe aortic stenosis or regurgitation
- Severe mitral stenosis or regurgitation
- Cardiomyopathy
- Acute pericarditis or myocarditis
- Vascular form of Ehlers-Danlos


2. All of the following may cause chest pain in children except:
A. Asthma
B. Costochondritis
C. Anxiety
D. Upper respiratory infection
E. Gastroesophageal reflux

CHEST PAIN IN CHILDREN

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>% of Children</th>
</tr>
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<tbody>
<tr>
<td>Idiopathic origin</td>
<td>21</td>
</tr>
<tr>
<td>Musculoskeletal origin</td>
<td>15</td>
</tr>
<tr>
<td>Cough</td>
<td>10</td>
</tr>
<tr>
<td>Costochondritis</td>
<td>9</td>
</tr>
<tr>
<td>Psychogenic origin</td>
<td>9</td>
</tr>
<tr>
<td>Asthma</td>
<td>7</td>
</tr>
<tr>
<td>Trauma</td>
<td>5</td>
</tr>
<tr>
<td>Pneumonia</td>
<td>4</td>
</tr>
<tr>
<td>Gastrointestinal disorders</td>
<td>4</td>
</tr>
<tr>
<td>Cardiac disease</td>
<td>4</td>
</tr>
<tr>
<td>Sickle cell disease</td>
<td>2</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>9</td>
</tr>
</tbody>
</table>

### CHEST PAIN IN CHILDREN

<table>
<thead>
<tr>
<th>Test</th>
<th>Total Patients</th>
<th>Normal</th>
<th>Abnormal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chest Xray</td>
<td>76</td>
<td>71</td>
<td>5</td>
</tr>
<tr>
<td>ECG</td>
<td>100</td>
<td>91</td>
<td>9</td>
</tr>
<tr>
<td>Urinalysis or HCT</td>
<td>28</td>
<td>28</td>
<td>0</td>
</tr>
<tr>
<td>SMAC</td>
<td>5</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Sed rate</td>
<td>7</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>T/T₄</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Echocardiogram</td>
<td>1</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Pregnancy test</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

#### When to Refer a Child with Chest Pain

- Acute distress present
- Significant trauma
- Pain associated with syncope, dizziness, palpitations, exertion
- History of cardiac or Kawasaki disease
- Pleural effusion or pneumothorax present
- Serious emotional problems
- Esophageal foreign body or caustic ingestion

Selbst, et al.
Pediatric Rev 1986; 8:56

#### Summary

- Generally benign and of noncardiac origin
- In adolescents, CP may be recurrent and may interfere with activities of daily life
- During evaluation of recurrent CP, a new etiology is frequently offered

Lam JC and Tobias JD
3. Arterial blood gas results from an infant with a cyanotic congenital heart defect should include:

A. Increased PO2 with crying
B. Increased PO2 with increased FiO2
C. Elevated PCO2
D. All of the above
E. None of the above
CONGESTIVE HEART FAILURE
ETIOLOGY

Congenital heart disease

- Birth – first week: Hypoplastic left heart, large AV fistula
  Critical AS or PS, TAPVR
  PDA (prematures)
- 1 – 4 weeks: Coarctation, critical AS,
  Large left to right shunts (prematures)
  Truncus arteriosus
- 6 weeks – 4 mo VSD, AV canal defect, large PDA
  Anomalous L coronary artery

4. All of the following cardiac conditions can present with heart failure in the first four months of life except:

A. Hypoplastic left heart syndrome
B. Coarctation of the aorta
C. Complete AV canal defect
D. Large ventricular septal defect
E. Moderately severe pulmonary stenosis

CONGESTIVE HEART FAILURE
ETIOLOGY

- Congenital heart disease
- Acquired heart disease (acute rheumatic carditis, myocarditis)
- Myocardial dysfunction (metabolic abnormalities, dilated cardiomyopathy)
- Miscellaneous: chronic tachycardia, complete AV block, severe anemia, acute hypertension
5. In infants, signs of congestive heart failure include all of the following except:
   A. Tachypnea
   B. Tachycardia
   C. Hepatomegaly
   D. Pedal edema
   E. Poor feeding

6. Which is most beneficial in treating CHF?
   A. K+ supplement
   B. Na+ supplement
   C. Furosemide
   D. Neosynephrine
   E. Morphine
CONGESTIVE HEART FAILURE
MANAGEMENT

• Oxygen (unless CHF is caused by excessive pulmonary flow)
• Diuretics – Lasix 1 mg/kg/dose
• Digoxin 8 – 10 mcg/kg/day (5 mcg/kg/day in prematures)
• ACE inhibitors
  ➢ captopril 0.5 – 6.0 mg/kg/day
  ➢ enalapril 0.1 mg/kg once or twice daily
• Misc: reduce energy expenditure, ± fluid restriction
• Beta-blockers: carvedilol (not yet standard therapy)

7. The following are true of furosemide EXCEPT:
   A. Should be given rapidly IV
   B. Usual dosage is 0.5-2.0 mg/kg/dose
   C. Can cause hypokalemia
   D. Can cause hyperostosis and nephrocalcinosis in newborns
   E. Can cause hypochloremic alkalosis

8. Digoxin levels are increased by all except:
   A. Quinidine
   B. Amiodarone
   C. Hypokalemia
   D. Carvedilol
   E. Erythromycin
9. An ACE inhibitor would be indicated for:
   A. Pulmonary stenosis
   B. Aortic stenosis
   C. Atrial septal defect
   D. Hypertrophic cardiomyopathy
   E. Dilated cardiomyopathy

CARDIOGENIC SHOCK
DEFINITION
Failure of the circulatory system to supply oxygen and nutrients to meet cellular metabolic demands

ETIOLOGY
- Newborns
  - Left heart obstructive lesions (HLHS, AS, COA)
  - Myocarditis
  - Tachyarrhythmia
  - Sepsis
- Infants and Older Children
  - Sepsis
  - Myocardial infarction
  - Myocarditis
CARDIOGENIC SHOCK

CLINICAL MANIFESTATIONS

- Pallor
- Tachycardia
- Tachypnea
- Hypotension, narrow pulse pressure
- Oliguria
- Metabolic acidosis

CARDIOGENIC SHOCK

EVALUATION

- Chest X-ray
- Electrocardiogram
- Echocardiogram
- Cardiology consultation

CARDIOGENIC SHOCK

MANAGEMENT

- Intubation and mechanical ventilation
- Positive inotropic agents (epinephrine, dopamine, dobutamine)
- Afterload reducing agents (milrinone, nitroprusside)
- Diuretics – Lasix 1 mg/kg/dose
- Judicious fluid replacement as indicated clinically
CARDIOGENIC SHOCK

Dopamine
1-5 mcg/kg/min: dopaminergic
5-15 mcg/kg/min: more beta-1
10-20 mcg/kg/min: more alpha-1

Dobutamine
2.5-20 mcg/kg/min: mostly beta-1, some beta-2

Epinephrine
0.05-0.1 mcg/kg/min: mostly beta-1, some beta-2
0.1 to 0.2 mcg/kg/min: alpha-1

Milrinone
50mcg/kg load then 0.375-0.75mcg/kg/min: phosphodiesterase inhibitor; increased inotropy and peripheral vasodilation

10. Long term complications of cyanotic congenital heart disease include:
A. Brain abscess
B. Stroke
C. Erythrocytosis
D. Bleeding disorders
E. All of the above

COMPLICATIONS OF CYANOTIC CONGENITAL HEART DISEASE

- Erythrocytosis: symptoms rare with Hct < 65%
- "Relative" anemia secondary to iron deficiency
- Stroke: associated with Fe" deficiency or paradoxical embolism
- Brain abscess – intracardiac shunting
- Bleeding disorders and thrombosis
- Hypercyanotic spells
11. A 3-month-old girl with known congenital heart disease has been having brief intermittent episodes of cyanosis. Today, a few minutes after feeding, she developed tachypnea, cyanosis, and persistent irritability. The MOST likely diagnosis is:
A. Breathholding
B. Colic
C. Congestive heart failure
D. Hypercyanotic spell
E. Seizure

CONGENITAL HEART DISEASE
HYPERCYANOTIC SPELLS
• Most frequent in tetralogy of Fallot
• Peak incidence, 2 to 4 months
• Early morning, after feeding, exercise, crying or defecation
• Cyanosis, increased respiration, fussy, syncope
• Treatment: knee-chest, O₂, sedation, fluids, phenylephrine
• Prevention: avoid dehydration, treat Fe²⁺ deficiency, β blocker

12. Hypercyanotic spells can be provoked by any of the following EXCEPT:
A. Pain
B. Induction of anesthesia
C. Dehydration
D. Iron deficiency
E. Squatting
13. **Treatment of hypercyanotic spells includes:**
   A. Isoproterenol
   B. Adenosine
   C. Digoxin
   D. Oxygen
   E. Furosemide

**True or False**

14. Nocturnal hypoxia can cause cor pulmonale

15. Cor pulmonale is irreversible

---

**COR PULMONALE**

- **Definition:** right ventricular hypertrophy or dilation secondary to pulmonary hypertension
- **Causes:** Congenital heart disease, alveolar hypoxia, pulmonary venous hypertension, primary pulmonary hypertension
- **Clinical manifestations:** dyspnea, fatigue, syncope, loud single S₂, hepatomegaly, venous distension
- **ECG:** right axis deviation, RVH
- **Management:** relieve airway obstruction, O₂, diuretics, ventilation, cardiac surgery, pulmonary vasodilators
ANSWERS FOR RECOGNITION OF CARDIOVASCULAR DISORDERS


SUGGESTED READINGS


SUGGESTED READINGS