Little Hearts
Cardiac Assessment in Children
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Objectives

• Assessment and management of:
  – Chest pain
  – Systemic hypertension
  – Arrhythmias
  – Syncope / POTS
• Discuss Congenital Heart Disease: School issues
• Describe special considerations for children with heart disease in the school/outpatient setting and emergency treatment

Normal Anatomy
Pediatric Vital Sign Normative Ranges

<table>
<thead>
<tr>
<th>Age Group</th>
<th>Respiratory Rate</th>
<th>Heart Rate</th>
<th>Systolic Blood Pressure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infant</td>
<td>20 - 30</td>
<td>80 - 140</td>
<td>70 - 100</td>
</tr>
<tr>
<td>Toddler (1-3 yrs.)</td>
<td>20 - 30</td>
<td>80 - 130</td>
<td>80 - 110</td>
</tr>
<tr>
<td>Preschooler (3-5 yrs.)</td>
<td>20 - 30</td>
<td>80 - 120</td>
<td>80 - 110</td>
</tr>
<tr>
<td>School Age (6-12 yrs.)</td>
<td>20 - 30</td>
<td>70 - 110</td>
<td>80 - 120</td>
</tr>
<tr>
<td>Adolescent (13+ yrs.)</td>
<td>12 - 20</td>
<td>55 - 105</td>
<td>110 - 120</td>
</tr>
</tbody>
</table>

Innocent vs. Pathologic Murmurs

Clinical Findings

- **Innocent/functional murmur:**
  - Healthy well-appearing child with otherwise normal physical examination
  - Normal S1 and S2
  - Systolic murmur
  - Not associated with a thrill
  - 50% of children have murmur at some point

- **Pathological murmur:**
  - Abnormal physical examination (failure to thrive, cyanosis, adventitious breath sounds, abnormal pulses)
  - Abnormal S1 or S2
  - Diastolic murmur
  - Associated with a thrill

Caring for the Child with Heart Disease in the School Setting
Congenital Heart Disease

- Congenital heart defects are the most common birth defect in the US, heart structure is abnormal
  - affecting nearly 1% of all infants or 40,000 births per year
- Improved diagnosis and management of heart defects have increased survival
  - More than 1 million adults in the US with congenital heart disease
  - More infants surviving with complex defects, have residual heart disease and complex management plans
  - More interventions can be done by catheterization, avoiding surgery

Common Congenital Heart Diseases

<table>
<thead>
<tr>
<th>Increased Pulmonary Blood Flow</th>
<th>Obstruction to Blood Flow</th>
<th>Decreased Pulmonary Blood Flow</th>
<th>Mixed</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASD</td>
<td>Coarctation of the Aorta</td>
<td>Tetralogy of Fallot</td>
<td>Transposition of the Great Arteries</td>
</tr>
<tr>
<td>PDA</td>
<td>Aortic Stenosis</td>
<td>Tricuspid Atresia</td>
<td>Total anomalous Pulmonary Venous Return</td>
</tr>
<tr>
<td>VSD</td>
<td>Pulmonary Stenosis</td>
<td></td>
<td>Truncus Arteriosus</td>
</tr>
<tr>
<td>CAVC</td>
<td></td>
<td></td>
<td>HLHS</td>
</tr>
</tbody>
</table>

Acquired Heart Disease

- Structurally normal heart with acquired cardiac disease at some point during a patient’s life
- Two most common acquired conditions:
  - **Rheumatic heart disease** - most likely to occur in children between 5 and 15 years of age, caused by streptococcal bacteria
  - **Kawasaki disease** - most likely to occur in children under the age of 5 years, inflammation of blood vessels that can result in coronary artery aneurysm
Cardiac Medications

- Most pediatric patients do not require lifelong management with medications
- Common medications include:
  - Diuretics (Lasix or Aldactone)
  - Afterload reducers (Enalapril)
  - Beta blockers (Atenolol)
  - Antiarrhythmics (Amiodarone, sotolol)
  - Anticoagulants (Coumadin), antiplatelet (ASA)
- Rule of thumb - 2 or more cardiac medications is associated with more severe disease

Neurodevelopmental Outcomes

- The majority of children with heart disease have a normal IQ
- Neurodevelopmental disabilities are more prevalent in school aged children with complex congenital heart disease treated in neonatal period
  - May be related to cardiopulmonary bypass, decreased cardiac output, and prolonged hospital stays
- Common problems include visual-motor and visual-spatial deficits and ADHD

Boston Circulatory Arrest Trial: 8 year olds
School Re-entry

- Develop 504 IEP plan as appropriate
- Some children may require tutoring / home schooling
- Some children will require abbreviated days and/or frequent absences due to doctors appointments

Exercise and Sports

- Exercise and physical activity are important for all children
- CHD kids are not immune to the obesity epidemic
  - A recent study suggests 25% of children with heart disease are obese
- Nearly all kids with heart disease can participate in some form of exercise

Common Recommendations for Physical Activity in School

1. May participate in the entire physical education program without restriction including varsity competitive sports
2. May participate in the entire physical education program except for varsity competitive sports where there is strenuous training and prolonged physical exertion
   - E.g. Football, hockey, wrestling, lacrosse, soccer, basketball
   - Less strenuous sports such as baseball & golf are acceptable at the varsity level
3. May participate in the physical education program except for restrictions from all varsity sports and from excessively stressful activities such as rope climbing, weight lifting, sustained running (e.g. laps) & fitness training. Must be allowed to rest when tired.
4. May participate only in mild physical education activities such as circle games, golf, badminton
5. Restricted from entire physical education program

When in doubt, contact the cardiologist!
Causes of Chest Pain

- Very Common Complaint
- Musculoskeletal
- Respiratory
- Gastrointestinal
- Idiopathic
- Psychogenic
- Cardiac

Musculoskeletal

- Muscle strain
- Trauma
- Growing pains
- Costochondritis
  - Most common musculoskeletal etiology of chest pain
  - Characterized by inflammation of cartilage at costochondral junction
  - Reproducible tenderness upon palpitation
  - + Pleuritic component to pain
  - Treatment with NSAIDS, rest, reassurance

Respiratory

- Respiratory etiology more common in children under the age of 12 years
- Cough
- Pneumonia
- Reactive airway disease
- Foreign body aspiration
- Pleural effusion
- Pneumothorax
Gastrointestinal

- Gastroesophageal reflux
- Esophagitis
- Gastritis
- Foreign body
- Caustic ingestion

Idiopathic / Psychogenic

- Most common etiology in adolescents (21-30%) and Females
- Idiopathic - Characterized by no organic or psychogenic etiology
- Psychogenic - Characterized by psychological etiology

Cardiac

- Less than 5% of chest pain in children has a cardiac cause
- Congenital & acquired anomalies
  - Coronary artery disease
    - anomalies, atherosclerosis, thrombosis or spasm
  - Left ventricular outflow tract obstruction
  - Cardiomyopathy (restrictive/dilated/hypertrophic)
  - Pulmonary hypertension
  - Pulmonary embolism
  - Pericarditis/Myocarditis/Arteritis
  - Aneurysm
- Arrhythmias
Assessment of Chest Pain
OLDCART

- Onset
- Location
- Duration
- Continuous
- Aggravating Factors
- Relieving Factors
- Treatments (effective and ineffective)

Case Study

- 10 year old male presents to your office accompanied by a friend because he complained of chest pain in the middle of his math test.
- What are you thinking?

Assessment of Chest pain

- How do they appear?
- Check Vital Signs
- Is the pain reproducible?
- Listen to heart and lungs
History Suggestive of Possible Cardiac Etiology

- Exertional chest pain
- Chest pain that radiates to the left shoulder or is exacerbated when supine
- History of prolonged immobilization, obesity, oral contraceptive pills, etc.
- History of recent intercurrent illness (Lyme disease, recent fever >38.5)
- Tachycardia in the absence of fever
- Palpitations
- Concerning family history
- Toxin exposure (cocaine, methamphetamine)

Chest Pain: When to worry

- Chest pain with exertion
  - Associated with dizziness or fainting
  - Occurs with palpitations or irregular rhythm
  - Radiates to back, jaw or left arm
  - Hemodynamic or respiratory instability
- Child with known heart disease or family history of heart disease or cardiomyopathy
- Chest pain that occurs only at rest, with a normal electrocardiogram, physical examination and without known risk factors, does not have a cardiac etiology

Systemic Hypertension

- Characterized by 3 or more elevated pressures on separate occasions
- Sphygmomanometer is the accepted gold standard
  - Automated Dinamap results in SBP increase by 10mmHg and DBP increase by 5 mmHg on average
  - Repeat all automated data indicating blood pressure >90th percentile

Definitions

- **Normotensive**
  - SBP & DBP < 90th percentile
- **Prehypertension**
  - SBP or DBP 90 - 95th percentile
  - BP > 120/70 in any patient under 18 years of age
- **Stage 1 hypertension**
  - SBP or DBP 95 - 99th percentile
- **Stage 2 hypertension**
  - >99th percentile

Information needed...

Normal Blood Pressure Values

Hypertension Screening

- All children 3 years of age and older should have BP checked at all health care encounters
  - Coarctation of the Aorta can present late in childhood,
  - Hypertensive arms, low BP in legs, faint or absent leg pulses
- If murmur is present on auscultation, consider obtaining 4 extremity blood pressures
- Screen children under the age of 3 years if risk factors for hypertension are present
  - Renal disease, prematurity, obesity, chronic steroid use or use of other medications which may result in hypertension

Management

- Children with evidence of systemic hypertension on 3+ occasions warrant cardiology referral
- For most children/adolescents the degree of vascular involvement is minor and the appropriate therapeutic approach is preventive- healthy lifestyle and behavior modification
  - Education
  - Optimizing nutrition
  - Promoting activity/physical education classes
Hypertension

• 8 year old chubby male presents to your office with a letter from her pediatrician asking to check his blood pressure weekly for the next 4 weeks and send back results.
• What would you expect his blood pressure to be?
• What would you begin counseling him on?

Arrhythmias

Normal Sinus Rhythm
Normal Sinus Rhythm

Palpitations

- An awareness or perception of the heart beat
  - “Skipped beats”
  - “Heart racing”

- Differential diagnosis is vast and includes several non-cardiac causes
  - Fever, anemia, drug use, thyroid disorders, anxiety, hyperventilation, arrhythmias, dehydration

Arrhythmias

- Too Fast: Tachycardias
  - Sinus Tachycardia
  - Supraventricular Tachycardia (SVT)
  - Ventricular Tachycardia (VT)

- Too Slow: Bradycardias
  - Sinus Bradycardia
  - Atrioventricular Block (AV Block)
  - Sick Sinus Syndrome
Tachycardias

- **Sinus tachycardia**
  - Most common by far, can be hardest to treat

- **Supraventricular tachycardia**
  - Most common arrhythmia of childhood

- **Ventricular tachycardia**
  - Rare, often associated with underlying heart disease or channelopathy

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Tachycardia

<table>
<thead>
<tr>
<th>SVT</th>
<th>Sinus</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Too fast to count</td>
<td>• Hard and fast</td>
</tr>
<tr>
<td>• Abrupt onset and termination</td>
<td>• Frequent symptoms</td>
</tr>
<tr>
<td>• Often felt in the neck</td>
<td>• May be situational</td>
</tr>
<tr>
<td>• May terminate with rest/squatting</td>
<td>• May terminate with rest/squatting</td>
</tr>
<tr>
<td></td>
<td>• Rates tend to be lower</td>
</tr>
</tbody>
</table>

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SVT

- **History**
  - Episodes are paroxysmal with abrupt onset and offset
  - Patients may complain of dizziness, palpitations, fatigue, chest pain or shortness of breath during an episode

- **Exam**
  - Heart rate is frequently >180 BPM
  - May appear pale or diaphoretic

- **Acute Management**
  - Calm observation
    - Count the heart rate, make the child comfortable (have them lie down with legs elevated)
  - Vagal Maneuvers
    - Ice to the bridge of the nose and forehead, Valsalva maneuver, stand on their head
    - Carotid massage NOT recommended
  - If an episode persists for >15-20 minutes despite vagal maneuvers
    - Notify parents: may need to be treated in the ER with IV medication.
SVT

- Chronic treatment options
  - Determined by age of the child, frequency and severity of symptoms; family preference
  - Observation and vagal maneuvers
    - Can be a good option for mild, infrequent symptoms
  - Medications
    - Beta-blockers, calcium channel blockers (CCB are contraindicated in WPW) are most common choices
  - Catheter ablation
    - Permanent cure rate of >90%
- Most patients with SVT have no restrictions on their activities

Cardiac Ablation

- Uses radiofrequency energy to destroy a small area of heart tissue that is causing rapid and irregular heartbeats.
- Low-risk procedure - generally successful.
- Takes place in an (EP) lab or a cardiac catheterization lab. It takes 2 to 4 hours. Discharged day of procedure or spend one night in hospital
- Back to normal activities in 3 days
WPW Definition

- A form of SVT:
  - intermittent tachycardia
  - a short PR interval
  - widened QRS complex on their ECG
- WPW results from an accessory pathway that directly connects the atria and ventricles and bypasses the AV node.
- Occurs in 1-1.5 / 1000 people

Normal Conduction and WPW Conduction

Findings on EKG

- PR shorter than 0.12 seconds
- Slurred upstroke of QRS “Delta Wave”
Management of WPW

• Needs to be evaluated by pediatric cardiologist
• Not everyone has tachycardia or is at high risk
• If WPW present on EKG:
  – further assess with an exercise test.
• If WPW pattern persists with exercise,
  – refer for EPS / Ablation to ablate pathway (risk of sudden death)
  – If no tachycardia and normal ETS, observe

Different Ambulatory Monitors

• Phones/ Aps
• Heart rate monitors for exercise
• http://www.alivecor.com/home

Case Study

• 13 year old female presents accompanied by a friend because her “heart is racing”. She is ambulatory and talking to you but looks pale. You attempt to take her pulse but it is “too fast to count”
• What are we going to assess?
• Are we worried?
• What do we think is going on?
Case Study

• This girl's heart rate persists and you are not successful with vagal maneuvers so you call her parents to come and get her. They take her to the emergency room where they are able to terminate the SVT with adenosine. She brings you a note that states she has WPW and is being referred to a pediatric cardiologist. She asks you what WPW is?

Bradycardias

• **Sinus node dysfunction**
  – Common in children with congenital heart disease who have had complex atrial surgery (ex. Fontan operation)
  – RARE in children with normal hearts
  – Symptoms include exercise intolerance and fatigue

• **Sinus bradycardia**
  – Largely benign
  – Can be seen in highly trained athletes, eating disorders, side effect of medication (ex. beta-blockers)
  – Unusual to be symptomatic

Bradycardias

– **AV block**
  • Can be congenital or acquired
    – (post surgical, Lyme disease, idiopathic)
  • Patients with complete heart block will likely need pacemaker placement at some point
  • Symptoms include dizziness, syncope, fatigue, increased sleep, exercise intolerance, history of rash (Lyme)
  • May be asymptomatic

  – Evaluate heart rate in the context of the physical exam
    • Perfusion, mental status, etc.
Pacemakers in Children

- Implant indications: complete heart block, sinus node dysfunction
- With or without congenital heart disease
- Pacing leads placed epicardially or transvenously
- Generators can be in either the chest or the abdomen

Pacemaker Care at School

- **Things to know**
  - Indication for placement
  - Patient’s underlying rhythm
  - Pacemaker settings (lower rate limit, upper rate limit)
  - Cell phones should be used 6 inches away from the device
  - Activity restrictions are usually due to the underlying heart disease, rather than the pacemaker itself
  - In general, patients should avoid collision sports
  - If necessary, an AED can be used. Avoid placing the pads directly over the pacemaker.

Ventricular Tachycardia

- Uncommon in children, can be life-threatening
- Often associated with underlying heart disease or channelopathy (ex. Long QT syndrome)
- May be triggered by adrenaline surges or myocardial ischemia associated with exercise or exertion
- Vagal maneuvers will not terminate VT, need AED
Implantable Cardiac Defibrillator (ICD)
Care at School

- **Things to know**
  - Indication for implantation
  - Patients underlying rhythm
  - ICD settings, especially the VT and VF zones
    - (ie. When patient will receive a shock)
  - AEDs can be used, avoid direct pad placement over ICD
  - Patients likely to have activity restrictions due to underlying cardiac disease or arrhythmia
  - ICDs have alarms that will beep if battery voltage is low or if the device or leads are malfunctioning
  - If patient receives one shock, but is stable, notify parents
  - If patient loses consciousness or receives multiple shocks, activate EMS

Syncope

- Characterized by sudden, brief, loss of consciousness with spontaneous recovery
- 25-50% of people will have an episode of syncope at some point
- Largely benign, but can represent pathology
- Differentiate using history (including family history), physical exam and ECG

Neurocardiogenic & Vasovagal Syncope

- **Most common cause of syncope**
  - 50% of cases presenting to medical attention
- **Cause**
  - Decrease in heart rate and blood pressure leading to inadequate cerebral perfusion, resulting in transient loss of consciousness
- **History**
  - Prodrome of nausea, lightheadedness, pallor, feeling of warmth, Symptoms more likely following position changes, after prolonged standing, warm environments
- **Exam**
  - Rapid return to consciousness, may appear pale, shallow and rapid breathing, sweaty, may feel unwell for several minutes after the episode.
Management of Neurocardiogenic & Vasovagal Syncope

- **Acute management**
  - Supine position and allow to awaken spontaneously- elevate feet
  - If hyperventilation, encourage to relax
  - Observe for several minutes

- **Chronic management**
  - Hydration, hydration, hydration: increase fluids 2-3 L per day
  - Salt – Increase sodium to 2-4 g / day
  - Aggressive hydration resolves symptoms in 90% of children
  - Anti-gravity maneuvers, avoidance of trigger situations, sodium supplementation
  - Prevention: recognize symptoms, steps to prevent syncope

- **Medications**
  - Reserved for those with persistent symptoms
  - Mineralocorticoids (fludrocortisone)
  - Alpha-agonists (Midodrine)
  - Beta-blockers

Different Types of Syncope

<table>
<thead>
<tr>
<th>Category</th>
<th>Abnormal +</th>
<th>Associated With Syncope</th>
<th>Postural Symptoms</th>
<th>Orthostatic Changes</th>
<th>Other Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>Typical</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Variable</td>
<td>1st arrest = male 2nd arrest = female</td>
</tr>
<tr>
<td>Ectopic</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Variable</td>
<td>Transient collapse</td>
</tr>
<tr>
<td>Catecholamine</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Variable</td>
<td>Very preluded spasms; usually micturition or vomitting after 2-3 seconds</td>
</tr>
<tr>
<td>POTS - Regional Palpitation</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Palpably pre-syncope sympotms</td>
</tr>
</tbody>
</table>

POTS – Postural Tachycardia Syndrome

- Most prevalent form of orthostatic intolerance
- An increase in the heart rate of at least 30 – 40 bpm when one changes from supine to an upright position within the first 10 minutes of standing
- Most commonly presents during late adolescence and in females
Symptoms of POTS

- In general these patients feel “bad”
- Lightheadedness / dizzy
- Tachycardia/ Palpitations
- Headache
- Fatigue
- Syncope/presyncope

Management of POTS

- No cure – Treatment focuses on treating underlying symptoms, lifestyle changes
- Similar to syncope – salt / fluids
- Fludrocortisone ( improves NA retention and increases BP)
- Midodrine – constrict peripheral blood vessels and aids in venous return
POTS/ Education

- Warning signs; dizziness, lightheadedness
- Slow position change
- Avoid prolonged standing/ keep moving

Hypertrophic Cardiomyopathy

- Hypertrophied, non-dilated left ventricle in the absence of another disease capable of producing the observed magnitude of hypertrophy

Presentation and Diagnostic Criteria

- Referral for family history, abnormal EKG, syncope, presence of a murmur
- Echocardiogram:
  - Septal thickness (typically >15mm), evidence of diastolic dysfunction, presence of left ventricular outflow tract obstruction
- EKG:
  - T-wave inversion or ST-segment depression, left atrial enlargement, left axis deviation, long QT interval, ventricular arrhythmias
### Management

- Annual evaluations: assessing for disease progression and risk stratification for sudden death
- Sports restriction
- Medication are used for symptoms
  - No available drug therapies have been shown to reduce the incidence of sudden death
  - Beta blockers, Verapamil, Lisinopril may help with chest pain and lowered outflow tract obstruction
- Surgical considerations for patients with disabling symptoms
  - Myectomy
  - Valve Repair or Replacement

### Sports Restrictions

- Restrictions
  - **Hockey, soccer, lacrosse, football, basketball**
  - Physical activity encouraged, avoid promoting a sedentary lifestyle!
  - Gym class and recreational sports are allowed
- Athletic Heart VS. Hypertrophic Cardiomyopathy
  - LVH on EKG prompting further evaluation, patient is referred to rule out HCM
  - Strict sports cessation for 6 months followed by re-evaluation with an echocardiogram

### Risk Stratification for Sudden Death

- Annual resting echocardiogram with EKG
- Cardiac MRI ~ Q3 years to detect for the presence of myocardial scarring
- Holter monitor
  - Every year for children < 8 years of age, then alternates with stress test every other year for children > 8 years of age
- Stress test with immediate post exercise echocardiogram
ICD Referral

• Referral made with combination of factors
  – **Major risk factors**: aborted sudden death, high risk genotype, family history of sudden death, and ventricular tachycardia
  – **Minor risk factors** (have significant but weaker association with sudden death): History of syncope, blunted blood pressure response to exercise, extreme wall thickness > 3cm and delayed enhancement on cardiac MRI (indicative of myocardial scarring and predictive of ventricular arrhythmias)

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Dilated Cardiomyopathy

Heart muscle becomes thin, the left ventricle becomes enlarged and unable to squeeze efficiently

Restrictive Cardiomyopathy

• Heart muscle is rigid, unable to relax and fill with blood

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Cardiomyopathy Genetics

- Pathogenic mutations are transmitted in an autosomal dominant pattern (offspring of affected person has a 50% chance of inheritance and risk of disease manifestation)
- Positive gene mutations in a first degree relative allow for familial testing
  - Genotype is unable to be determined 30-40% of the time
- All 1st degree relatives should have a screening EKG and echocardiogram
  - Children should have an evaluation every 3 years during 1st decade of life, then annually during the pubertal growth spurt from ages 10-14
  - After adolescence, can typically be followed ~ every 2 years

Secondary Cardiomyopathy

- Duchenne and Becker Muscular Dystrophy
- Mitochondrial and storage disorders
- Chemotherapy induced cardiomyopathy
- Friedreich’s Ataxia
- Noonan Syndrome

Causes of Sudden Cardiac Death

All are RARE events in childhood
- Hypertrophic Cardiomyopathy (33-50%)
- Long QT syndrome (15-25%)
- Arrhythmogenic Right Ventricular Dysplasia/Dilated Cardiomyopathy (10-20%)
- Coronary artery anomalies (10-20%)
- Primary VT/VF (10-15%)
- Wolff-Parkinson-White (3-5%)
- Aortic rupture (5%)
Tips for School Nurses

• Know your resources:
  – CPR, AED location, activate EMS
• Rest and Fluids are your friends
• When to worry:
  – Signs of poor perfusion
  – SVT symptoms that last longer than 15 minutes
  – Syncope associated with injury
  – Chest pain / syncope with exertion
• Never be afraid to pick up the phone and ask!

Additional Resources

www.nhlbi.nih.gov/guidelines/cvd_ped/summary
Includes information on nutrition, physical activity, tobacco exposure, high blood pressure, lipids and obesity

A Pocket Guide to Blood Pressure Measurements in Children
www.nhlbi.nih.gov/health/public/heart/hbp/bp_child_pocket


Thank you for your time!

Questions?