Objectives- At the end of this presentation you should be able to

- Identify emergent situations and place an APB*
- Identify urgent situations and “hack the system”
- Console any anxious parent and motivate those with a laid back approach
- Know the current appropriate use criteria
- Apply all of the above to “real-life” situations
  - CCHD screening
  - Adolescent with chest pain
  - Syncope
  - Bradycardia, tachycardia and other rhythm abnormalities

CCHD Screening- Friend or Foe?

- CCHD affects 1% of newborns, 25% of whom require intervention
- Fetal echo impact
  - Impact of fetal echocardiography
  - John M Simpson
- Fetal intervention
  - Change of course at time of delivery
- Consequences of not identifying
  - Mortality occurred in 28% of critical vs 8% of non-critical cases. Mortality was lower with an early diagnosis of critical CCHD (early diagnosis 16% vs late diagnosis 27%, p=0.04).
  - Eckersley L1, Sadler L2, Parry E3, Finucane K1, Gentles TL1.
  - Timing of diagnosis affects mortality in critical congenital heart disease.
- Cost of pulse oximetry CCHD
- Number of False Positives
  - 75% sensitive for critical cases, 49% for all cases
- Cost of pursuing False Positives

Hints there is CCHD- Hacking the system

- Family history of syndrome +/- CCHD
- Dysmorphic appearance or extracardiac anomalies- mother, father and child
- Ability to feed
- Abnormal 4 extremity blood pressures
- Abnormal exam
  - Comfortable respiration
  - Murmur
  - HSM
  - Pulses
- Now for the pulse Ox

Case 1

- 3.2 kg nondysmorphic FT male delivered NSVD, APGARs 9, 9
- Breastfeeding is OK, mother anxious
- Baby appears comfortable
- Exam remarkable for a fairly loud murmur, otherwise normal
- Pulse oximetry done at 24 hours of life 85%
- Surprising because baby does not look blue
  - Is it possible for the baby to NOT look blue?
  - Is it possible this is a false positive?

Case 1 –Next Steps

- Your next step:
  a. Ask for a different sat probe and/or machine, test probe on your finger
  b. Warm baby’s feet
  c. Call the cardiologist ASAP
  d. Obtain EKG and CXR
  e. Ask if there were any concerns on fetal sonogram
Case 1

- The cardiologist is covering hospital and office and cannot come right away, per office staff
- Your next step:
  a. Discharge the baby and send them to office as an outpatient ASAP
  b. Ask to speak to the cardiologist on the phone
  c. Obtain the EKG and Chest X-ray first, then call the cardiologist back
  d. Call 911 and have the baby transferred to a tertiary care children’s hospital
  e. Arrange for follow up with you in the office in 48 to 72 hours; after all, the mother is a critical care nurse

Risk: Benefit Ratio in CCHD Screening

- Objective:
  Prevention of discharge of infant with a ductal dependent lesion
  - If the ductus closes, infant is at risk of high mortality/morbidity
  - Left sided lesions: HLHS, Coarctation*, IAA, Aortic/mitral stenosis, Shone’s
  - Right sided lesions: Transposition (usually presents before screening) Pulmonary Atresia +/- VSD, Tetralogy of Fallot, Tricuspid Atresia, DORV

Prevention of discharge of infant with complex heart disease not dependent on the ductus, but with increased morbidity
- Truncus arteriosus
- TAPVR
- Single Ventricles
- AVC
- Heterotaxy

Our Friend, the Ductus

A Compendium of Ductal-Dependent Lesions

A Compendium of Non-ductal Dependent, but Complex Lesions
Answer

- Tricuspid Atresia
- HACK:
  - Always ask to speak to the cardiologist and NO ONE ELSE regarding the timing of evaluation of the patient
  - Trigger phrases: "more cyanotic than anticipated" + "blue" + "not feeding so well" + "I am not comfortable discharging" + "lives in a rural area" + "Primary relative with CHD" + "Fetal echo showed something"
  - Primary relative is mother, father, sibling, maybe grandparent, aunt, uncle cousin with STRUCTURAL heart disease

Case 1  Is it ever OK to discharge to outpatient for follow up?

Chest Pain and Appropriate Use Criteria

- Incidence and prevalence of chest pain in children in the US
  - 5.2% of all ER visits
  - Cardiac etiology 0 to 15%
- Current recommendations for evaluation
- Trigger words in history
- Physical Examination Points
- Usefulness of EKG
- Usefulness of CXR
- When to echo

From Appendix A- AUC

<table>
<thead>
<tr>
<th>#</th>
<th>Chest Pain</th>
<th>Appropriate Use Rule</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>Chest pain with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal EKG</td>
<td>Low</td>
</tr>
<tr>
<td>11</td>
<td>Chest pain with other symptoms or signs of cardiovascular disease, a benign family history, and a normal EKG</td>
<td>Moderate</td>
</tr>
<tr>
<td>12</td>
<td>Chest pain with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal EKG</td>
<td>High</td>
</tr>
<tr>
<td>13</td>
<td>Chest pain with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal EKG</td>
<td>Very high</td>
</tr>
</tbody>
</table>

From Appendix C

A Tale of 2 Chest Pains

- 15 year old competitive soccer player complains of chest pain at maximum exertion versus
- 15 year old self-professed “computer geek” with intensifying chest pain at rest and with activity, unremitting for the last month
- What questions do you want to ask these two?
- What tests do you want to order?
- When do you want to speak to the cardiologist?
Questions

Athlete
- 10/10 pain at peak exercise, forces him to stop exercise
- Alleviated by rest
- Recent respiratory illness and wheezing, new exercises
- No FHX SCD, deafness, seizures, syncope
- Pain is no worse with positional changes
- No change in activity
- Recently broke up with girlfriend
- Thinks there may be palpitations, but heart going so fast, uncertain
- No syncope

Geek
- Varies 6 to 10/10
- No exacerbating or relieving factors
- No recent illnesses, doesn’t exercise
- No FHX SCD, deafness, seizures, syncope
- Pain is no worse with positional changes
- More fatigued lately
- Doing well in school, same girlfriend for the last year
- No palpitations or syncope

What are the key questions with chest pain?
- Timing
- Intensity
- Exacerbating Factors
- Association with palpitations or syncope
- Associated illnesses (infectious disease, respiratory)
- Affect on Activities of Daily Living
- Family History

Which of these boys requires further testing and/or Cardiology Consultation?

- 15 yr old athlete- we need to exclude causes of SCD
- 15 yr old computer geek- pain is unremitting and intensifying
- Neither
- Both

15 year old athlete: AUC recommends further assessment for chest pain with or immediately following exercise

- EKG only; if normal allow to play
- EKG and echo only; if normal allow to play
- Consultation with pediatric cardiologist; restrict from play until evaluated
- EKG, Echo, Exercise stress test; if normal allow to play
- Tell him to suck it up. He is only 15 years old and fit. He does not have heart disease.

15 year old athlete
- P.E. Resting HR 45 BPM, BP 100/60, comfortable athletic male with no murmurs, no tenderness to palpation
- EKG: NSR, normal without LQTS, Brugada, pre-excitation, axis deviation
- Echo: Structurally normal heart with normal systolic function, coronaries difficult to visualize due to body habitus
- Do we need any further testing?
  a. MRI/MRA/CTA or coronary arteries
  b. Troponin levels with exercise the next time he has chest pain
  c. Exercise stress test
  d. Fitbit with exercise, have him send tracings
  e. Drug testing

15 year old Computer Geek
- P.E. HR 10 BPM, BP 110/70 This, mildly distressed, normal respirations, but preferred to sit up, leaning forward. Otherwise normal exam, although heart tones somewhat distant, normal pulses.
- EKG: NSR, lower voltage, but no LQTS, no Brugada, no pre-excitation of axis deviation, full pattern considered a normal variant
- Echo-poor resolution, not all structures well-visualized but function was normal
- Does he need further testing?
  a. MRI/MRA/CTA of coronary arteries
  b. Troponin, CRP, ESR
  c. CVR
  d. 24 hour Holter monitor
  e. Drug testing
Both patients have pathology. DDX of CP

- Pericardial effusion
- Myocardial diseases-
  - HCM
  - DCM
  - Myocarditis
- Coronary Disease
  - Structural
  - Atherosclerotic
- Dyshrhythmia
- Texidor’s Twinge
- POTS
- GERD
- Asthma/Pneumonia/Pleuritis
- Pneumothorax
- Costochondritis
- Other musculoskeletal
- Anxiety
- Extrinsic compression of cardiac structures
- Stimulant therapy to treat ADHD
- THC, meth, cocaine, others

Both Had Pathology

- Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)
  - Picked up on treadmill
- Hodgkin’s Lymphoma
  - Picked up on CXR then sent for CT

How far do we need to go in CP?

- AUC is strict regarding use of echo—however, if it is a competitive athlete, err on the side of over-testing
- AAOCAC—Anomalous aortic origin of the coronary artery
- Hypertrophic cardiomyopathy—sometimes difficult to distinguish
- Dysrhythmia, number one cause of sudden cardiac death, and can be difficult to identify
  - Baseline EKG
  - Holter and/or event monitor
  - Exercise stress testing

Hacking Chest Pain

- Personal History, including emotional history and school performance, sleep and use of caffeinated beverages and stimulants (especially ADHD meds)
- Recent history of febrile illnesses, rash, fever, respiratory illness, early satiety
- Associated palpitations +/- syncope
  - + Troponin, ESR, CRP, abnormal EXG
  
  **Trigger words to cardiologist**
  - + troponin, CP with max activity, associated syncope +/- palpitations, recent febrile illness, competitive athlete, sudden change in activity level

  Calming Parents
  - If it is the parental request to see the cardiologist, it may be easiest to acquiesce

Swooners and Other Tales

- Definitions*
  - Syncope: sudden loss of consciousness with loss of postural tone, not related to anesthesia with spontaneous recovery as reported by patient or observer. Patient may experience syncope when supine.
  - Dizziness: a sensation of light-headedness, unsteadiness, turning, spinning, or rocking.
  - Faceplant: a type of syncope in which there is no attempt to protect oneself from the effects of the fall.

  **Important questions:**
  - Where were you?
  - What were you doing?
  - Did you feel anything beforehand? Palpitations or racing heart, nausea, visual changes, tinnitus, hot then cold and clammy
  - Did you hurt yourself?
  - Did anyone witness this?

- Up to 15 percent of children experience a syncopal episode prior to the end of adolescence
- More common in athlete, particularly thin females
- There may be a family history
- True syncope may be dangerous and may be associated with trauma, but in and of itself is not life-threatening
- Life-threatening causes of syncope are to be excluded—ARRHYTHMIA, ANOMALOUS ORIGINS OF CORONARY ARTERIES, CARDIOMYOPATHIES

*Appendix B
A Cautionary Tale

- 16 yr, 6 feet tall, 130# basketball player – faints at least once per basketball game when he stops to shoot at free throw line
- Has never fainted in practice and keeps up with his team members
- Has no other associated complaints, no chest pain, shortness of breath or palpitations.
- Feels absolutely nothing prior to his LOC, and has nearly broken his jaw.
- Takes no medications or supplements, drinks water alternating with sports drinks, at least a gallon a day, and likes salty foods

Next steps

a. Ask for an echocardiogram to evaluate for Marfan syndrome
b. Obtain EKG; if normal, advise him to continue to hydrate and allow him to play
c. Test him for hypoglycemia
d. Obtain EKG and echo, and if normal, allow him to play
e. Obtain a stress test, and if normal allow him to play as long as he maintains hydration

An unusual case of syncope

- He continued to play basketball because his team relied on him as a shooting guard
- He continued to faint, but only when he stopped suddenly to shoot
- His EKG, echo, 24 hour Holter and exercise stress test were normal
- He was referred to an electrophysiologist, who performed a diagnostic test. Was it:
  a. Tilt table
  b. EP study
  c. Implantable event recorder
  d. Angiogram

Usual Coronary Anatomy

- This patient had an anomalous coronary, left, originating from right.
- He since has had surgery and continues to play basketball
- The incidence of AADCA is 1%
  - Higher incidence of coronary anomalies in young SCD victims (4 to 15%)
- High index of suspicion is necessary if patient continues to have syncope in spite of
  a. Normal baseline cardiac testing
  b. Adequate hydration and eating
  c. The story just does not make sense

Swooners

- This patient had an anomalous coronary, left, originating from right.
- He since has had surgery and continues to play basketball
- The incidence of AADCA is 1%
  - Higher incidence of coronary anomalies in young SCD victims (4 to 15%)
- High index of suspicion is necessary if patient continues to have syncope in spite of
  a. Normal baseline cardiac testing
  b. Adequate hydration and eating
  c. The story just does not make sense

Too Fast or Too Slow

- Arrhythmia constitute 55.1 per 100,000 pediatric emergency department (ED) visits
- Most common dysrhythmias
  - Sinus tachycardia (50%)
  - Supraventricular tachycardia (13%)
  - Bradycardia (6%)
  - Atrial fibrillation (4.6%) [Doniger & Sharieff, 2006]
- Because this is not a common pediatric clinical condition, it often can be a diagnostic challenge for clinicians.
Bradycardia

- Heart rate slower than the lower limit of normal for the patient’s age
- (See PALS)
- Mechanisms of bradycardia:
  - Depression of the pacemaker of the sinus node and conduction system blocks due to hypoxemia
  - Vagal stimulation (GERD)
  - Gastroesophageal reflux and other neurologic disorders
  - Acidosis
  - Acute intracranial hypertensive event (Cushing’s triad)
  - Athleticism
  - Anorexia nervosa
  - Underlying electrical disturbances in the heart (Doniger & Sharieff, 2006).

EKG - Normal or Abnormal?

EKG - Normal or Abnormal?

EKG - Normal or Abnormal?

EKG - Normal or Abnormal?

EKG - Newborn with heart rate varying between 60 and 180 BPM

EKG - Normal or Abnormal?

What kinds of questions do you want to ask?
Measuring the QT interval
Bazett's formula
Leads II and V5
Males >470 ms, Females >480 ms
Flat "funny" T waves
No change with exercise
Symptoms?

An Abnormal EKG
What is Brugada syndrome and why is it important to identify?

When to worry
• First- and second-degree heart block largely asymptomatic, may (rarely) progress
• Children/infants with complete heart block may present with CHF, fatigue/lethargy, poor feeding
• Older children with complete heart block
  • Dizziness
  • Fatigue
  • Decreased exercise tolerance
  • Confusion
  • Syncopal episodes

Causes of Third Degree Heart Block
• Structural heart disease
• Surgical heart block (if history of cardiac surgery)
• History of maternal connective tissue disease
• Thyroid disease
• Muscular dystrophy
• Cardiac tumor
• Electrolyte disturbance
• Infectious etiology (variety)
• Inflammatory disease (e.g., lupus erythematosus)
• Isolated anomaly

Hacking the System
• Cardiologist Trigger Words-
  • Abnormal EKG + syncope, fatigue, lethargy, change in activity level, unable to exercise, unable to feed (if infant) with associated pallor, diaphoresis, respiratory dysfunction
• Testing to Order –
  • In room jumping or jumping jacks
  • 24 hour Holter
  • Exercise stress test
• Comforting Parents
  • Remember, asymptomatic sinus bradycardia is not usually life-threatening

Thank you for the gift of your time!
Appendix A

Appropriate Use Criteria

<table>
<thead>
<tr>
<th>Table 1: Other Symptoms and Signs</th>
<th>Appropriate Use Rating</th>
</tr>
</thead>
<tbody>
<tr>
<td>40.</td>
<td>Symptoms and signs suggestive of malignant birth defect, including but not limited to: cyanotic or cyanotic state, poor or absent cardiorespiratory depression, and/or hypotension</td>
</tr>
<tr>
<td>41.</td>
<td>Clinical signs and symptoms of respiratory depression, and/or hypotension</td>
</tr>
<tr>
<td>42.</td>
<td>Signs and symptoms of increased venous return; increase of umbilical vein diameter, and/or abnormal venous pulsations</td>
</tr>
<tr>
<td>43.</td>
<td>Lymphedema and/or edema of the lower extremities</td>
</tr>
<tr>
<td>44.</td>
<td>Other clinical signs and symptoms suggestive of congenital heart disease</td>
</tr>
</tbody>
</table>

The rating in parentheses next to the rating reflects the median score for that indication.

Abbreviations: A. = Appropriate; B. = Below Appropriate; C. = Borderline Appropriate; D. = Derek Appropriate; E-IV = Extremely Inappropriate.
Appendix A

Appendix A

Appendix A

Appendix B  Pediatric Cardiology Thesaurus

Appendix C- Coming Attractions-
JACC August or September 2016

Appendix C Revised EKG Criteria
Appendix D  Known Causes of SCD

Appendix E Revised Bethesda Guidelines

Appendix F  14 point AHA Screening Guidelines

- **14-point AHA Screening Process**
  1. Obtain informed consent.
  2. Screen for personal history.
  3. Evaluate family history.
  4. Consider genetic testing.
  5. Perform cardiac structural assessment.
  6. Evaluate arrhythmia risk.
  7. Consider screening for non-cardiac conditions.
  8. Provide counseling and follow-up care.

- **Risk Stratification**
  - High-risk patients include:
    - Sudden cardiac death
    - Unexplained syncope
    - Family history of sudden cardiac death

- **Screening for Catheterization**
  - Consider catheterization in patients with positive screen results.

- **Treatment Options**
  - Medications
  - Device therapy
  - Surgical interventions

- **Follow-up and Counseling**
  - Regular follow-up appointments
  - Education about cardiac health
  - Psychosocial support

- **Resources**
  - American Heart Association
  - National Heart, Lung, and Blood Institute

- **References**
  - American Heart Association Guidelines
  - International Society for Heart Rhythm Disorders
  - European Society of Cardiology