Cardiovascular Diseases: Case Presentations

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Objectives

• Recognize and manage atrial and ventricular arrhythmias.

• Distinguish cardiac etiologies for chest pain and syncope.

• Develop an appreciation for the impact that childhood obesity has had on the incidence of childhood hypertension and hyperlipidemia.
Case #1

• 2 week old infant comes to your office for well child check. Benign history, feeding well with good weight gain. On exam, occasional irregular heart beats are noted.
• What is the differential diagnosis?
• Important question: what is the heart rate?
Atrial Premature Beats**
(APB, PAC)

Normal

Very common in newborns
Aberrant Conduction of PAC

Look for some variation of a P wave preceding the beat.
Aberrant Conduction of PAC
Ventricular Premature Beats**
(VPB, PVC)

Typically normal

Should suppress with exercise

Check family history for syncope, sudden death or drowning
Same Baby, but...

• On auscultation the heart rate is noted to be slow.....
Blocked PACs
What is the Rhythm?

Can be seen in infants of mothers with anti-SSA and antiRo antibodies
Can be associated with structural heart disease
Often results in this.....
Another 2 week old

• This time noted to be feeding poorly, fussy, sleeping more. Exam with tachycardia and tachypnea, hepatomegaly, pulse too rapid to count......
Narrow Complex Tachycardias
Differential Diagnosis of Tachycardia**

• Look for the P wave
  – Narrow complex, regular with 1:1 P:QRS
    • Prior to QRS: sinus, ectopic atrial, PJRT
    • Within QRS: AVNRT
    • Behind QRS: Accessory pathway (WPW or other)
  – Narrow complex, irregular, more Ps than QRS
    • Atrial flutter or fibrillation
  – Wide complex, regular
    • No relationship of P to QRS
      – Ventricular tachycardia
    • With P waves
      – SVT with aberrancy
Rhythm Snapshots: Sinus Tachycardia

- ST can be a hard diagnosis to confirm.
- P-waves buried in t-waves at faster HR’s.
- Max HR = 220 – age in years
- Diagnosis: P-wave before each QRS w/ NL p-wave axis (+) 2,3,aVF, (-) avR, +/ - in V1.
- Hard to differentiate from “regular” ectopic atrial tachycardia from right atrium.
- Extreme sinus tach ominous. Usually indicates poor cardiac function and impending trouble. (r/o tamponade, bad ventricular function, hypovolemia, shock)
Orthodromic Recipricating Tachycardia

Narrow complex, regular rapid tachycardia.
Retrograde p-wave behind each QRS in 1:1 relationship
Starts and stops suddenly
Treatment: vagal maneuvers, adenosine, medication in infants, typically beta blocker
Rhythm Snapshots: SVT acute termination

- SVT due to pathway. Tiny retrograde p-waves behind QRS’s and on top of t-waves.
- Terminates to NSR with adenosine with obvious sinus p-waves in front of QRS’s.
- T-waves now smooth in NSR without notches (superimposed p-waves).

- If funny notches without QRS’s persist and tachycardia restarts after few seconds, consider AFL with transient AV node block due to adenosine rather than SVT.
- If SVT clearly breaks to NSR then restarts after few beats/seconds/minutes, try adenosine a few more times. If able to stop SVT but with frequent spontaneous restarts, add another drug then retry adenosine once other drug on board (BB or amiodarone).
Adenosine

• Intravenous, fast push
• Dosing based on patient weight
  – First dose: 100mcg/kg (max dose 6mg)
  – Second dose: 200mcg/kg (max dose 12mg) PALS protocol
• Transiently blocks conduction through AV node. If you are not seeing a pause in ECG tracing, you are not getting the drug to the patient or not enough drug
• Will terminate reentrant SVT or will slow ventricular response in atrial flutter
Rhythm Snapshots: Fast VT

- Treatment: Hemodynamically stable; meds +/- elective cardioversion.
  - Hemo unstable; Immediate cardioversion (synchronized preferred).
  - Meds: Amiodarone 5-15 mg/kg bolus, same as JET.
  - Others acceptable but significantly less effective.
  - Trial bolus of adenosine acceptable since SVT with aberrancy more common than VT in pediatrics.
- Common in ventricular dysfunction (IDCM) or in surgeries with ventriculotomy or potential for coronary insufficiency (TOF, ALCAPA, TGA-ASO).
Bibliography for Rhythm

- How to Read Pediatric ECG’s by Myung K. Park
- Pediatric Advanced Life Support Circulation 2010; 122[Suppl3]S876-S908
Case #2

• 14 year old female presents for school physical. Has gained quite a bit of weight since evaluation last year. Not participating in organized sports. BP 150/90, HR 80. No murmur on exam.

• What additional historical information would help?
• What other exam features?
• What is your differential?
AAP HTN Guidelines

• Hypertension is more common in preschoolers who are overweight and obese and who had low activity

• 60 min/day of moderate to vigorous activity
Measure BP & HT & calculate BMI
Determine BP category for age, Ht, gender

Stage 2 Hypertension
- Diagnostic Work-up
  Include Evaluation for Target Organ Damage
  - Secondary HTN
  - Or Primary HTN
  - Consider Referral to Provider with Expertise in Pediatric HTN
- Drug RX
- Weight Reduction & Drug RX

Stage 1 Hypertension
- Repeat BP
  Over 3 visits
  - >95%
  - Diagnostic Work-up
    Include Evaluation for Target Organ Damage
  - Secondary HTN
  - Primary HTN
- RX specific for cause
- Drug RX
- Weight Reduction
- Weight Reduction

Pre-Hypertensive
- Therapeutic Life Style Changes
- Repeat BP in 6 months
  - 90–<95% or 120/80mmHg

Normotensive
- Educate on Heart Healthy Lifestyle for the Family
- Consider Diagnostic Work up & Evaluation for Target Organ Damage if Obesity or co-morbidity exists

- Monitor Q 6 Mo
- Weight Reduction
## Plasma Lipid Concentrations in Children and Adolescents

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<th>Category</th>
<th>Low, mg/dL&lt;sup&gt;a&lt;/sup&gt;</th>
<th>Acceptable, mg/dL</th>
<th>Borderline-High, mg/dL</th>
<th>High, mg/dL&lt;sup&gt;a&lt;/sup&gt;</th>
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<td>&gt;120</td>
<td>115–120</td>
<td>—</td>
</tr>
</tbody>
</table>

<sup>a</sup> TC: Total Cholesterol; LDL: Low-density lipoprotein; Non-HDL: Non-high-density lipoprotein; HDL: High-density lipoprotein.
Bibliography

• Pediatrics 2004; 114(2):555-575, National High Blood Pressure Education Program (NHBPEP)
• http://www.nhlbi.nih.gov/guidelines/hypertension/child_tbl.htm
Case #3 - Child with Chest Pain

• 6 year old male presents with episodes of chest pain. Parent reports ‘I can see his heart beating through his shirt when he complains about this pain’.

• What additional history is important?
• What physical findings?
• What testing if any is indicated?
Causes of Chest Pain

• Most of pediatric and adolescent chest pain in noncardiac
• 45% of cases of chest pain the etiology will not be determined
• Supratentorial component cannot be overlooked
  – Referrals increase dramatically when an athlete dies suddenly
Cardiac Causes of Chest Pain

Arrhythmia
  Nonsustained ectopy
  Supraventricular tachycardia (SVT)
  Ventricular tachycardia (VT)
Mitral Valve Prolapse (MVP)
Coronary artery anomalies or ischemia
Myocarditis
Arrhythmia

• Most often supraventricular
• ECG most often normal
  • If WPW, then ECG was useful
  • If long QTc, then ECG may or may not be useful!
    • How long does the QTc have to be to be long???
• Before you order another test, have the family or patient count the pulse and teach them how to do a Valsalva maneuver
Case #4 – Adolescent Chest Pain

• 14 year old female presents with episodes of chest pain which is left sided.

• What additional history is important?
• What physical findings?
• What testing if any is needed?
Evaluating Chest Pain

• History
  • With or without activity?
  • Awakens them from sleep?
  • Only at school or with one parent?
  • Dietary balance and fluid intake

• Family History
  • Sudden death?
  • Marfan or aortic dissection?

• Social History
Evaluating Chest Pain - Exam

• Vital signs
• BMI
• Reproducible chest wall tenderness
• Presence of murmur
• Dysmorphic characteristics
Noncardiac Causes of Chest Pain

• Idiopathic
  • Some 25-45% of cases of chest pain

• Musculoskeletal
  • Trauma
  • Over exertion
  • Costochondritis
  • Precordial catch
Noncardiac Causes Of Chest Pain

• Pulmonary
  • Asthma
  • Exercise Induced Bronchospasm
  • Vocal Cord Dysfunction
  • Pneumothorax

• Rheumatologic
  • Pleural or pericardial effusion
    • SLE (lupus)
    • Juvenile Rheumatoid Arthritis
Noncardiac Causes of Chest Pain

• GI
  • Reflux
  • Esophagitis secondary to medication
    • Minocycline
    • Nonsteroidal anti-inflammatory agents
Case #5 – Chest Pain

• 14 year old female presents with episodes of chest pain which is left sided. The pain occurs only with activity.

• What additional history is important?
• What physical findings?
• What testing if any is needed?
Chest Pain with Exertion

- Overuse injury
- Exercise induced bronchospasm or asthma
- Vocal cord dysfunction
- Deconditioning
- Coronary artery abnormality
- Hypertrophic cardiomyopathy
- Aortic stenosis

- Unless a pathologic etiology is suspected, do not remove child from gym/sports
So do you need any testing???

- Pain that interferes with sleep,
- is precipitated by exercise, or
- is associated with
  - dizziness,
  - palpitations,
  - syncope, or
  - shortness of breath
- Electrocardiogram and/or chest radiograph
Myocarditis

- Inflammation of the myocytes resulting from viral illness
- Laboratory testing
  - ECG
  - Troponin
  - BNP
- Diagnosis by
  - MRI
  - Endomyocardial biopsy
Pathogens Known to Cause Myocarditis

• Varicella
• Parvovirus B-19
• Adenovirus
• Streptococcus
• Influenza
• Epstein-Barr virus
• Coxsackievirus and echovirus
• Chlamydia
• HIV
Coronary Ischemia

• Coronary Vasospasm
  • Substance use
    • Cocaine
    • Methamphetamine
  • Prinzmetal angina

• Coronary artery anomaly
  • Aberrant origin of the right or left coronary artery
  • ECG likely typically normal in these conditions, even with exercise stress test
Case #6 – The Weak and Dizzy

• 16 year old female complains of dizziness and ‘curtain like’ visual change when standing up too fast after watching a movie or sitting in class.

• What additional history is important?
• What physical findings?
• What testing if any is needed?
**Evaluation**

- **History**
  - Prodrome: Visual changes, ringing in ears, sweating, nausea
  - Dietary history – ask what they are drinking!
- **Family History**
  - Sudden death, arrhythmia, medications in house
- **Physical Exam**
  - Includes orthostatics and assessment for murmur
  - +/- ECG; beta HCG, CBC
What about the Dizzy?

• Orthostasis
• Autonomic Instability

• How to treat?
• How to monitor hydration?
  How to find time in the day to drink and use the bathroom!!!!
“It’s all about the letter P”

- **Dehydration – What color is your urine?**
  - Assess caffeine intake
  - Assess beverage consumption
  - Urine color
  - Assess use of bathroom at school

- **Treatment – Hydration and increase salt intake**
  - Pretzels, popcorn, baked potato chips, pickles, pepperoni
  - Water bottle – may need a note for school
POTS

- Postural Orthostatic Tachycardia Syndrome (POTS)
- Chronic day to day symptoms of orthostatic incompetence associated with excessive tachycardia
- Defined by meeting criteria for Orthostatic Incompetence
- In the average adolescent, an increase in heart rate of >35 beats/min or heart rate >120-130 bpm after two minutes standing
Case #7 - The Fainter

- 14 year old male presents after a syncopal episode which occurred in the early morning after his alarm went off.

- What additional history is important?
- What physical findings?
- What testing if any is needed?
Calculating the QTc

- Use the LONGEST value over 3-5 cardiac cycles
- Leads: II, V5, V6
- Bazett Formula:
  - measured QT
  - divided by square root of RR (preceding)

- Bazett overestimates during tachycardia and underestimates during bradycardia
- Computer measurements of QTc are wrong ~25% of the time
Long QT syndrome

**Family history** of premature sudden death
- Drowning, SIDS, single car accident

**Personal history** of palpitations with syncope
- Syncope with fright
- Alarm clock
Neurally-mediated syncope
(Simple faint)

- Peak incidence 15 yrs
- Female:Male 2:1
- Seen in 15-25% of all normal adolescents
- Triggered by:
  - Prolonged standing
  - Dehydration, hot days
  - Intercurrent illness
  - Disgust, sight of blood

- Micturation syncope
  - Or in shower, while washing or combing hair
- May occur post-exercise, NOT during exercise
- May occur during or after a meal
Classification of Syncope

- **Neurally mediated**
  - Vasovagal
  - Carotid
  - Situational
  - Atypical

- **Orthostatic hypotension**
  - Primary autonomic failure
  - Secondary autonomic failure
  - Drug-induced
  - Volume depletion

- **The good news**, treatment is pretty much the same regardless of the exact diagnosis
  - Volume repletion
  - Fludrocortisone 0.1mg daily
  - beta blockers (Atenolol)

- **Unless, Cardiovascular**
  - Arrhythmia
  - Structural cardiac disease
Non-cardiovascular causes:

• Breath-holding spells
• Hypoglycemia
• Hyperventilation
• Vertigo
• Seizures
• Basilar artery migraine
• Hysterical syncope
Cardiac Causes of Syncope: What can you die from?**

- Hereditary long QT syndrome
- Cardiomyopathy
  - Hypertrophic
  - Dilated
  - Arrhythmogenic RV dysplasia (ARVD)
- Wolff-Parkinson-White syndrome
- Coronary anomalies
- Late post-operative arrhythmias
- Congenital complete AV block
- Valvar aortic stenosis
Cardiac Causes of Syncope: And How to Screen for Them**

- Hereditary long QT syndrome
  - Cardiomyopathy
    - Hypertrophic
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    - Arrhythmogenic RV dysplasia (ARVD)
- Wolff-Parkinson-White syndrome
- Coronary anomalies
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- Congenital complete AV block
- Valvar aortic stenosis

- Family history and ECG
- Family history, ECG and echo
- And possible murmur
- Family history and MRI
- ECG
- Echo
- ECG or event recorder
- ECG
- Murmur on exam
Hypertrophic Cardiomyopathy

- Clinical Signs & Symptoms
  - Dyspnea on exertion
  - Chest pain
  - Syncope or presyncope
  - Palpitations
  - Sudden death
  - Midsystolic murmur that increases in intensity with standing (dynamic auscultation)

- None of the symptoms are specific for HCM!
Hypertrophic Cardiomyopathy

- Autosomal dominant, but many affected relatives have much milder disease
- Clear age-related progression
  - Repeated screening of family members is indicated, more often in adolescence
- +/- Genetic testing
Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA)
Conclusion

• The history of the event(s), the family history and the physical exam are the three legged stool of pediatric cardiology.
Bibliography


• Guidelines on management (diagnosis and treatment) of syncope – Update 2004 The task force on Syncope, European Society of Cardiology. European Heart Journal DOI: http://dx.doi.org/10.1016/j.ehj.2004.09.004
Potentially Useful Classification:

What can you die from?

- Wolff-Parkinson-White syndrome
- Hereditary long QT syndrome
- Cardiomyopathy
  - Hypertrophic
  - Dilated
  - Arrhythmogenic RV dysplasia (ARVD)
- Coronary anomalies
- Valvar aortic stenosis
- Aortic dissection
Practice Change

• Evaluation of the patient with CHEST PAIN should include
  • A careful history including family history
  • A careful physical examination
  • Testing only in those cases where a specific pathologic diagnosis is suspected

• Restriction from activity is rarely warranted, and usually only in those with chest pain with exertion or with associated signs/symptoms