Problems in Pediatrics: Pediatric Cardiology Cases

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Case 1

• 12 day old ex-full term male presents to PMD for WCC
• Doing well at home, has regained birth weight
• Feeding 2 oz every 3 hours without difficulty
• No cyanosis, resp distress, extreme irritability
• No medications

• Vitals: 4.1 kg, HR 240 bpm, RR 44, BP 63/32, Sat 100% RA
• Exam: Well-appearing, alert, no wheezing, tachycardic, hyperdynamic precordium, no murmurs, no HSM, femoral pulses 2+ b/l
What test do you want?

A. Echocardiogram
B. CXR
C. ECG
D. Arterial blood gas
E. Electrolyte panel
Case 1

- To the ED...
Diagnosis?
Neonatal arrhythmias

Atrial arrhythmias
Supraventricular tachycardia
Ventricular tachycardia
Atrial arrhythmias

Most commonly seen in children with CHD who have undergone cardiac surgery

Types:
- Atrial ectopic tachycardia (AET or EAT)
- Atrial flutter
- Atrial fibrillation
Supraventricular tachycardia

Electrical impulse cycles repetitively through circuit from atria to ventricles and back again → results in rapid, regular ventricular contractions

Common manifestations in infants:

• Pallor
• Fussiness
• Irritability
• Poor feeding
Supraventricular tachycardia

SVT more likely to resolve if:
- Less than 1 year of age at onset
- Structurally normal heart
- Normal baseline ECG (no pre-excitation)

SVT less likely to resolve if:
- Older than 1 year of age at onset
- Structural heart disease
- Pre-excitation on ECG (WPW)
WPW
Emergency treatment of SVT

Vagal maneuvers:
- Ice to face, IV placement, Valsalva

Pharmacologic:
- Adenosine
  - Initial dose 0.05-0.1 mg/kg
  - Rapid IV bolus followed by rapid flush
  - May be repeated with escalating doses to 0.25 mg/kg
- Verapamil
  - Contraindicated in children less than 12 months

Synchronized electrical cardioversion
- 0.5 J/kg

Obtain 12 lead ECG in tachycardia and after conversion!!
SVT pre-Adenosine
SVT during Adenosine
Case 2

• 2 month old female with history of moderate-large perimembranous VSD presents for WCC

• Mom reports baby has not been feeding well for past week
  • Used to take 3 oz Enfamil every 3 hours; now will only take ½-1 oz at a time
  • Falls asleep during the feeds
  • Overall seems to be sleeping more than usual throughout day

• Vitals: 4.5 kg (was 4.6 kg at 1 month visit); HR 144; RR 60; sat 98%

• Exam: tachypneic with mild subcostal retractions; 4/6 holosystolic murmur over LSB, +thrill; liver edge felt 2-3cm BCM; 2+ femoral pulses
Characteristic signs and physical findings of impaired cardiac output in neonates typically include all of the following except:

A. Feeding intolerance
B. Tachycardia
C. Tachypnea
D. Poor perfusion
E. Swelling of extremities
Causes of CHF in neonates

Ventricular dysfunction

- Decreased “squeeze” of ventricles → impaired ejection of blood from ventricles (dilated cardiomyopathy, myocarditis)

Volume overload

- Typically due to significant L→R shunts (VSD, PDA)

Pressure overload

- Due to severe ventricular outflow obstruction that impedes ejection of blood from the heart (aortic stenosis, pulmonary stenosis) → results in inadequate cardiac output
Management goals in CHF

- Depends on etiology and severity
- Therapeutic goals are to relieve symptoms, decrease morbidity, slow progression of heart failure, and improve patient survival/quality of life
- Mainstay of treatment for volume overload or pressure overload due to structural defects → catheter-based interventions or surgical repair
  - Medical therapy (diuretics) as a temporizing measure
- Treatment for ventricular dysfunction → ACE inhibitor, aldosterone antagonist, +/- beta-blocker, digoxin
Case 3

A 15 year old healthy male collapses during soccer practice, while running laps around the field. Witnesses report some mild respiratory depression requiring CPR.
Syncope during all of the following situations is consistent with “benign” vagal syncope except:

A. While brushing hair in the morning
B. During the middle of a 5K road race
C. While standing in line at an amusement park
D. 3 minutes after finishing the time mile run at school
E. Immediately after micturition
Vasovagal (neurocardiogenic, reflex, benign) syncope

Most common cause of syncope among children

Typical clinical features are a precipitating event and a prodrome

- Standing or stress (physical/emotional)
- Reflex precipitants (hair grooming, micturition)
- Prodrome may include dizziness, visual changes, nausea, pallor, diaphoresis

Underlying mechanism: exaggeration of reflex-mediated alterations in vasomotor tone and heart rate normally responsible for maintaining blood pressure
“Red flag” syncope

Syncope during exercise
- Hypertrophic cardiomyopathy, Catecholaminergic polymorphic ventricular tachycardia (CPVT), severe aortic stenosis
- Associated with palpitations
  - SVT or other atrial arrhythmia leading to syncope
- Requiring CPR
  - Ventricular tachycardia or fibrillation, Torsades\n
Family history of sudden death (SIDS, drownings)
- LQTS
  - LQT1: arrhythmic events related to exercise, swimming
  - LQT2: events triggered by auditory stimuli (alarm clock, phone)
Evaluation of child with syncope

Situation in which it occurred
Symptoms before/after event
Family history
Physical exam/Vitals
ECG
+/- Echocardiogram
Case 4

• 13 year old female with history of being tall (height >99%), having long fingers and toes, scoliosis, and frequent changes in eyeglass prescription who presents for first well-child exam (recently moved to RI)

• No murmur on exam

• Mother is also very tall and had replacement of her aortic root 5 years ago
To whom should she be referred for concern for connective tissue disorder?

A. Cardiology  
B. Vascular surgery  
C. Genetics  
D. Ophthalmology  
E. A & C
Marfan syndrome

- Autosomal dominant connective tissue disorder affecting the ocular, skeletal, and cardiovascular systems
- Physical features and clinical presentation can vary between individuals, even within same family
- If family history of MFS, presence of ONE of the following is diagnostic:
  - Ectopic lentis
  - Ghent ≥7
  - Aortic root dilation (z-score ≥2 if older than 20, ≥3 if younger than 2)
- Absence of family history of MFS, presence of one of the following criteria is diagnostic:
  - Aortic root dilation (z-score ≥2) AND ectopic lentis
  - Aortic root dilation (z-score ≥2) AND FBN1 mutation
  - Aortic root dilation (z-score ≥2) AND Ghent score ≥7
Marfan syndrome

- Significant phenotypic overlap with other CTD
  - Loeys-Dietz syndrome (associated with risk of dissection from brain to pelvis)
  - Familial thoracic aortic aneurysm and dissection syndrome (FTAA)
  - Ehlers-Danlos syndrome (many sub-types)
Marfan syndrome

• Aortic root disease (leading to aneurysmal dilatation, aortic regurgitation, and dissection) is main cause of morbidity and mortality

• Dilatation of aorta is found in approximately 50% of young children with MFS, and progresses with time; 60-80% of adults have dilatation of root

• Dilatation can also involve other segments of thoracic and abdominal aorta

• Mitral valve prolapse also frequently identified in patients with MFS (40-55% of patients)
  • Frequency increases with age and greater in women

• **Normal aortic root size does not rule out Marfan Syndrome or other connective tissue disorder**
Question #1

SVT is most likely to resolve if:

A. Child is less than 1 year of age at onset
B. Child has had previous cardiac surgery
C. Baseline ECG shows pre-excitation (WPW)
D. Child has Ebstein’s anomaly of the tricuspid valve
16 year old female presents after 2 episodes of vasovagal syncope. These symptoms never occur during exercise. They usually occur in the mid-morning, especially after not eating breakfast. There is no family history of sudden death. She has had no prior evaluation for syncope. What is the next appropriate test to order?

A. 24-hour Holter monitor
B. ECG
C. 30-day event monitor
D. Tilt-table test
Question #3

17 year old male is referred to pediatric cardiology clinic after aortic root dilatation was found on echocardiogram performed secondary to chest pain. Remainder of echo is normal. Physical examination reveals patient has scoliosis and a pectus carinatum. Family history is significant for maternal grandfather with ascending aortic dissection. Which of the following genes should be tested for maximum yield?

A. Transforming growth factor, β receptor 1 (TGF-β)
B. Collagen, type III
C. Actin smooth muscle
D. Fibrillin 1 (FBN1)
Thank you!