UAE 5th Latifa International Pediatrics & Neonatology Conference

Outpatient Pediatric Cardiology: Evaluation and Management of Common Conditions

Paul E. Farrell, Jr, MD, MBA

Pediatric Cardiologist

The Children's Hospital of Philadelphia

Clinical Professor of Pediatrics

University of Pennsylvania School of Medicine

March 30, 2019

Objectives

- Review clinical evaluation of cardiac conditions
  - Vital signs, Pulse oximetry, BP (upper and lower extremities), Murmurs (Innocent versus pathologic), Pulses
- Brief comment on palpitations
- Review the evaluation and management of chest pain
  - functional, non-cardiac, or cardiac
- Review the evaluation and management of Syncope
  - Vasovagal (neuro-cardiogenic)
  - Cardiac- structural or arrhythmia induced

Cardiac Clinical Evaluation

- History
  - Infants
    - Prenatal history: Maternal diabetes, medication use, nuchal thickening, amniocentesis results, other organ system abnormalities, prematurity
    - History of respiratory distress, poor growth, feeding problems, or cyanosis
  - Children and adolescents
    - Frequent respiratory problems, exercise intolerance, chest pain, dizziness, syncope
  - Family history, social history

Cardiac Clinical Evaluation

- Physical exam
  - Growth: height, weight, HC in infants
  - Vital signs: HR, RR and work of breathing, BP (sitting with feet on the floor), 4 extremity BP in infants
  - Pulse oximetry:
    - Cyanosis not seen until saturation <88%
    - Lower extremity screening in infants (abnormal < 94%)
    - **Differential cyanosis**- leg lower than arm, seen with right to left shunting through PDA in critical left heart lesions
    - **Reverse differential cyanosis**- post ductal sat higher than preductal (TGA with PDA and elevated PVR, TGA with preductal CoA/interrupted aortic arch)

Cardiac Clinical Evaluation

- Palpitation
  - Pectus abnormalities, active precordium, thrills
- Auscultation
  - Heart sounds (S1, S2, S3, S4)
    - Narrow or single loud S2- pulmonary hypertension
    - Widely split S2- ASD, RBBB, PAPVC
  - Clicks
    - Ejection clicks in early systole, pulmonary clicks at ULSB, aortic clicks at LLSB
    - Mid-late systolic clicks-MVP
    - PDA and Ebstein anomaly- multiple clicks
  - Rubs- seen in pericarditis, sitting, LLSB or apex

Characteristics of Murmurs

- **Age at appearance**
  - Outflow tract murmurs can be heard in the first hours of life
  - As PVR drops in in newborns, shunt murmurs (PDA, VSD, AVC) can be heard
- **Timing during cardiac cycle**
  - Systolic
    - Ejection-crescendo/decrescendo, AS or PS
    - Holosystolic- VSD, mitral valve regurgitation
  - Diastolic- pathologic, AI/PI, flow rumbles in ASD/VSD
  - Continuous- PDA, AV malformations, coronary fistula
  - To-and Fro- TOF with absent pulmonary valve

Characteristics of Murmurs

- **Location, radiation, postural maneuvers**
  - Note where murmur is loudest on chest
  - Standing increases the murmur of hypertrophic cardiomyopathy with outflow tract obstruction (decreases venous return to smaller LV, increasing gradient)
  - Squatting increases venous return and LV volume, moves click and murmur to a later part of systole in MVP
- **Loudness, pitch, and quality** (harsh, blowing, to and fro, musical, vibratory)
  - Systolic murmurs Grade 1-6, diastolic murmurs 1-4
  - Grade 4, palpable thrill
  - Murmurs diminished with decreased cardiac output (critical aortic stenosis in a newborn)

Innocent murmurs

- Typically soft systolic murmurs < grade 4, not associated with heart disease
  - Still's murmur-loudest in supine position, decreases with sitting
  - PPS (newborn)
  - Venous hum-best heard sitting, decreases in supine position and with neck vein compression

Other important physical findings

- Lung exam- rales, wheezing
- Abdominal exam- hepatosplenomegaly, ascites
- Distal pulses- Femoral artery pulses, diminished, delayed, or absent
- Extremities- clubbing, increased arm span, upper to lower segment ratio (Marfan), edema, capillary refill

Palpitations and Irregular rhythms

- Premature atrial contractions
  - Common in fetus and neonates, usually benign and resolve over a few months if infrequent
- Premature ventricular contractions
  - Usually require cardiac evaluation
- Evaluation
  - History and physical exam
  - EKG with rhythm strip, Holter monitor, looping event monitors
- Treatment depends on type and frequency of events
- Abnormal EKG, rapid/irregular rhythms (SVT, atrial flutter, frequent PVC's)- recommend cardiac consultation

Chest Pain in Children and Adolescents

- Etiology
  - Idiopathic 40%
  - Musculoskeletal 30%
  - Hyperventilation/Emotional 20%
  - Breast related 5%
  - Respiratory/Gastrointestinal 4%
  - Cardiac 1%

Chest Pain in Children and Adolescents

- History most important in evaluation
- Cardiac etiology of chest pain not likely if:
  - Unrelated to exercise
  - Patient has systemic illness
  - Negative cardiac exam and EKG
- Chest pain with exertion and not at rest, or worse in supine position requires further workup

Chest Pain in Children and Adolescents

- Costochondritis
  - Usually unilateral, 2-3 continuous upper costochondral or sternal joints
  - Sharp pain, lasting seconds-minutes
  - Worse with deep breathing
  - Pain reproduced by pushing on joints
  - Treatment- anti-inflammatory medications, heating pad, physical therapy, localized steroid injections

Chest Pain in Children and Adolescents

- Respiratory Causes:
  - Asthma
  - Infection (pneumonia, bronchitis, pleurodynia)
  - Pneumothorax (abrupt onset, severe pain, SOB)
- Pulmonary vascular tree causes
  - Primary pulmonary hypertension
  - Pulmonary embolism (acute onset, severe SOB, pleuritic CP). Order d-dimer, oxygen saturation, CXR, CT angiogram
- GI causes
  - Esophageal reflux or spasm, dysmotility, foreign body
  - Ulcer

Chest Pain in Children and Adolescents

- Pericarditis
  - Etiology: idiopathic, infectious, systemic illness (SLE), post- pericardiotomy, trauma, uremia, neoplastic
  - Symptoms: substernal or left sided chest pain, pain worse in supine position and less in sitting position
  - Symptoms: Fever (2/3), friction rub (1/4), pulsus paradoxus (abnormally large decrease in pulse amplitude and systolic BP during inspiration)

Chest Pain in Children and Adolescents

- Pericarditis
  - Evaluation: CBC, Inflammatory markers, troponin (associated myocarditis), EKG, echocardiogram
  - Treatment: anti-inflammatory medications, steroids, pericardiocentesis for significant effusions with tamponade

Chest Pain in Children and Adolescents

- Cardiac causes:
  - Aortic stenosis
  - Hypertrophic cardiomyopathy
  - Coronary artery anomalies
    - Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA)
    - Anomalous origin of the LCA or RCA from the opposite sinus of Valsalva
- Evaluation: exam, EKG, echocardiogram, exercise testing, CT or MRI, cardiac catheterization

Etiology of Syncope

- Syncope is caused by an interruption of blood flow to the brain
- The single most common reason for syncope in children and adolescents is an abrupt change in vasomotor tone known as vasovagal syncope
- Generally the conditions are grouped into cardiac and non-cardiac causes

Vasovagal Syncope

- Exaggeration of these responses leads to a drop in blood pressure through several mechanisms
  - Hypotension (independent of heart rate) – vasodepressor type
  - Sinus bradycardia – cardio-inhibitory type
- Usually specific triggers can be identified
  - Postural changes, hair brushing, coughing, micturition, shaving, emotions, pain, swallowing, etc.
- Accompanied by a prodrome / pre-syncopal symptoms

Vasovagal syncope and exercise

- The benign exercise related syncope...
  - Must exclude cardiac etiologies carefully
- Commonly occurs **after** the **termination of activity**
  - Collapses after walking to sideline, not during activity
- Usually has the same prodrome as common fainting

Cardiac Causes of Syncope

- Structural
  - Aortic stenosis
  - Cardiomyopathy
    - HCM, DCM, acute
  - Coronary artery anomalies
  - ARVD
  - Pulmonary HTN
  - CHD
- Primary arrhythmia
  - LQTS
  - SQTS
  - Brugada
  - CPVT
  - Idiopathic VF
  - WPW with rapid ventricular response
  - Bradyarrhythmias

These are the etiologies that need to be referred due to an increased risk of sudden cardiac death (SCD)

Structural

- Aortic stenosis
  - Limited cardiac output due to physical obstruction
  - Sudden death likely related to arrhythmias, not obstruction, due to ischemia
- Coronary anomalies
  - Anomalous origins of coronary arteries
    - Particularly left coronary arteries from the right sinus
    - Again, SCD due to arrhythmias secondary to ischemia
  - Muscular bridge

Structural

- Arrhythmogenic right ventricular dysplasia
  - Typically an etiology in older children/adolescents
    - Mean age of presentation is 30 years
  - Fatty infiltration of the RV leading to ventricular arrhythmias
- Cardiomyopathy
  - Due to arrhythmias in dilated cardiomyopathy and acute myocarditis in the setting of poor output
  - Syncope in hypertrophic cardiomyopathy also likely due to ventricular arrhythmias but may also have some element of obstruction to outflow contributing to symptoms
    - Hypertrophic cardiomyopathy is the most common cause of sudden cardiac death in children and adolescents

Structural

- Pulmonary hypertension
  - Due to any number of etiologies
  - Can have RV arrhythmias if RV function is poor
  - Usually due to acute increases in pulmonary pressures with subsequent decrease in cardiac output
- CHD
  - Usually later in life related to poor function plus arrhythmias in complex CHD
    - TOF, TGA, AS are particularly at risk for sudden cardiac death
  - Some may present due to obstruction

Arrhythmia

- Long QT syndrome
  - Prolongation of the QT interval due to abnormal repolarization
  - Predisposes to polymorphic ventricular tachycardia
    - Torsades de pointes
  - Can be inherited or acquired
    - Inherited – types 1, 2 and 3 are most common
    - Acquired – drugs, electrolyte abnormalities (low K, low Ca, low Mg), hypothyroidism, bradycardia

Arrhythmia

- Brugada
  - Channelopathy resulting in ventricular tachycardia
  - Usually male dominant, SE Asian, and older population (older adolescents)
  - More common during fever to have arrhythmia
- Catecholaminergic polymorphic ventricular tachycardia (CPVT)
  - Channelopathy resulting in polymorphic ventricular tachycardia
  - Related to catecholamine stimulation
    - Emotion, exertion

Arrhythmia

- Wolff-Parkinson-White syndrome with rapid ventricular response (WPW with RVR)
  - An accessory pathway provides an alternative route for atrial signals to reach the ventricle
  - If the atrial rhythm is very fast it can conduct rapidly and lead to ventricular fibrillation
  - Not all accessory pathways can do this
- Short QT syndrome
  - Channelopathy leading to abnormal repolarization predisposing to ventricular arrhythmias
  - Very rare

History

- HPI
  - What they were doing when they passed out?
    - Exertional syncope is ALWAYS a cause for concern
    - Syncope AFTER exertion is usually vasovagal but still needs to be investigated
  - Where there any prodromal symptoms?
  - How long were they out?
  - Any symptoms while unconscious?
    - Was anyone around to witness the event?
  - How often does it occur?
  - Is it always in the same circumstances?

History

- PMH
  - Any significant cardiac, psychiatric or neurologic history
- FH
  - This is absolutely key – BE VERY SPECIFIC
  - History of sudden unexplained deaths, especially in those younger than 55 years
    - Unexplained drowning, car accidents, SIDS
  - History of syncope
  - History of arrhythmia disorders (e.g., pacer or ICD?)
  - History of cardiomyopathy

Physical Exam

- Make sure orthostatics are part of the vital signs
  - Laying, sitting, standing, standing 3 minutes
  - HR increase > 30 BPM- Postural Orthostatic Tachycardia Syndrome (POTS)
- A thorough neurologic exam can be useful to document focal lesions
- A good cardiac examination to evaluate for structural disease
  - Most cardiac etiologies will not be picked up this way

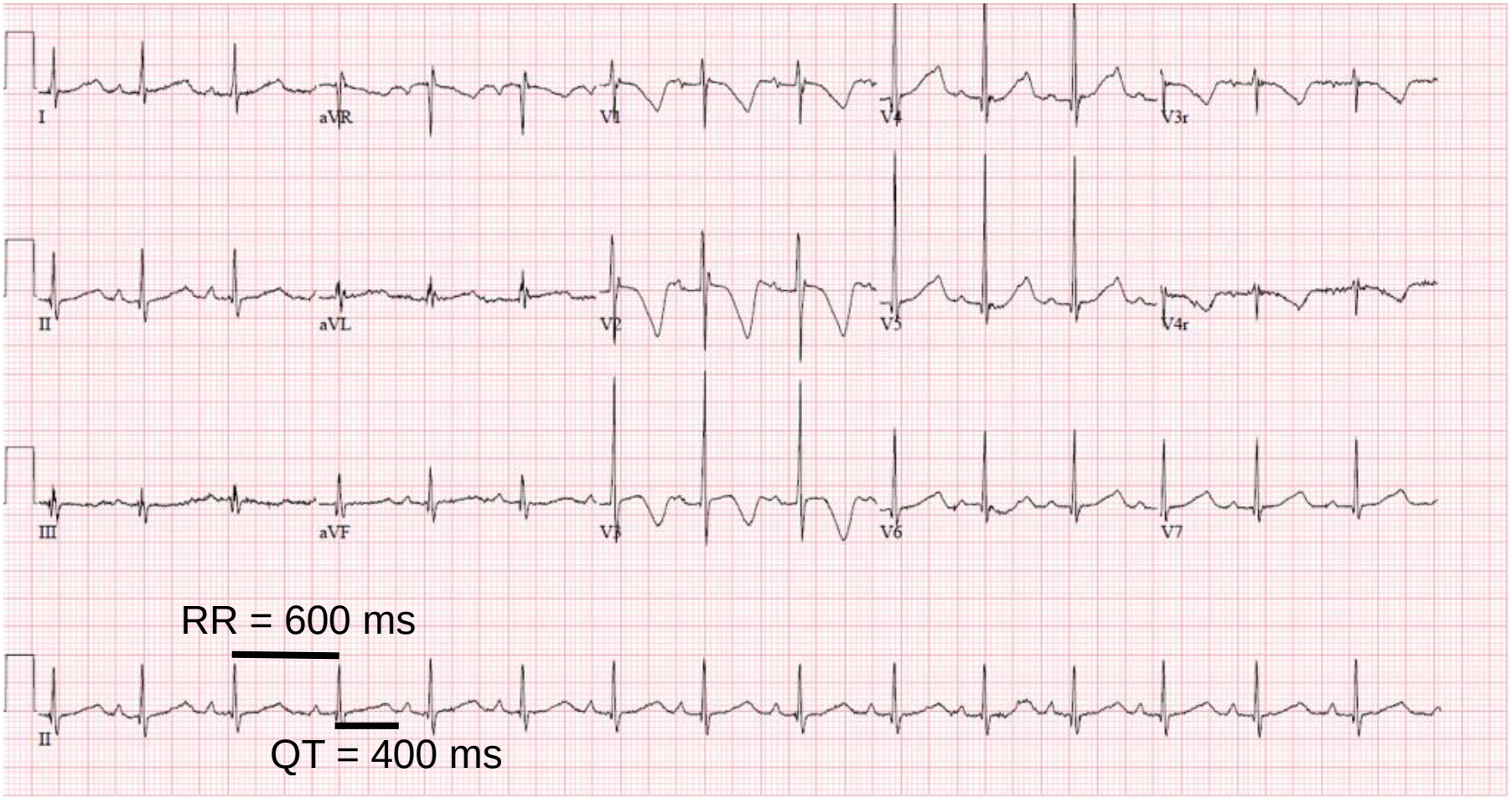
Case 1

- 15 y/o female presents to primary care clinic for recurrent syncope
- Occurs most often after standing long periods of time and when she is having blood tests
- Collapses, convulses, awakens
- PE normal
- ECG normal
- FH benign
- Diagnosis: Vasovagal (also known as neuro-cardiogenic syncope)

Case 2

- 13 y/o previously healthy girl with no history of syncope or pre-syncope collapses while playing soccer
  - Stays unresponsive
  - Bystanders “feel” a pulse
  - AED applied and shocks patient
  - Patient wakes up, but feels bad
  - Reports no prodrome prior to the collapse
  - Never had more extensive workup than a sports physical
  - Sibling died of SIDS (unknown to patient)
- Historical red flags?

Case 2 – Long QT syndrome

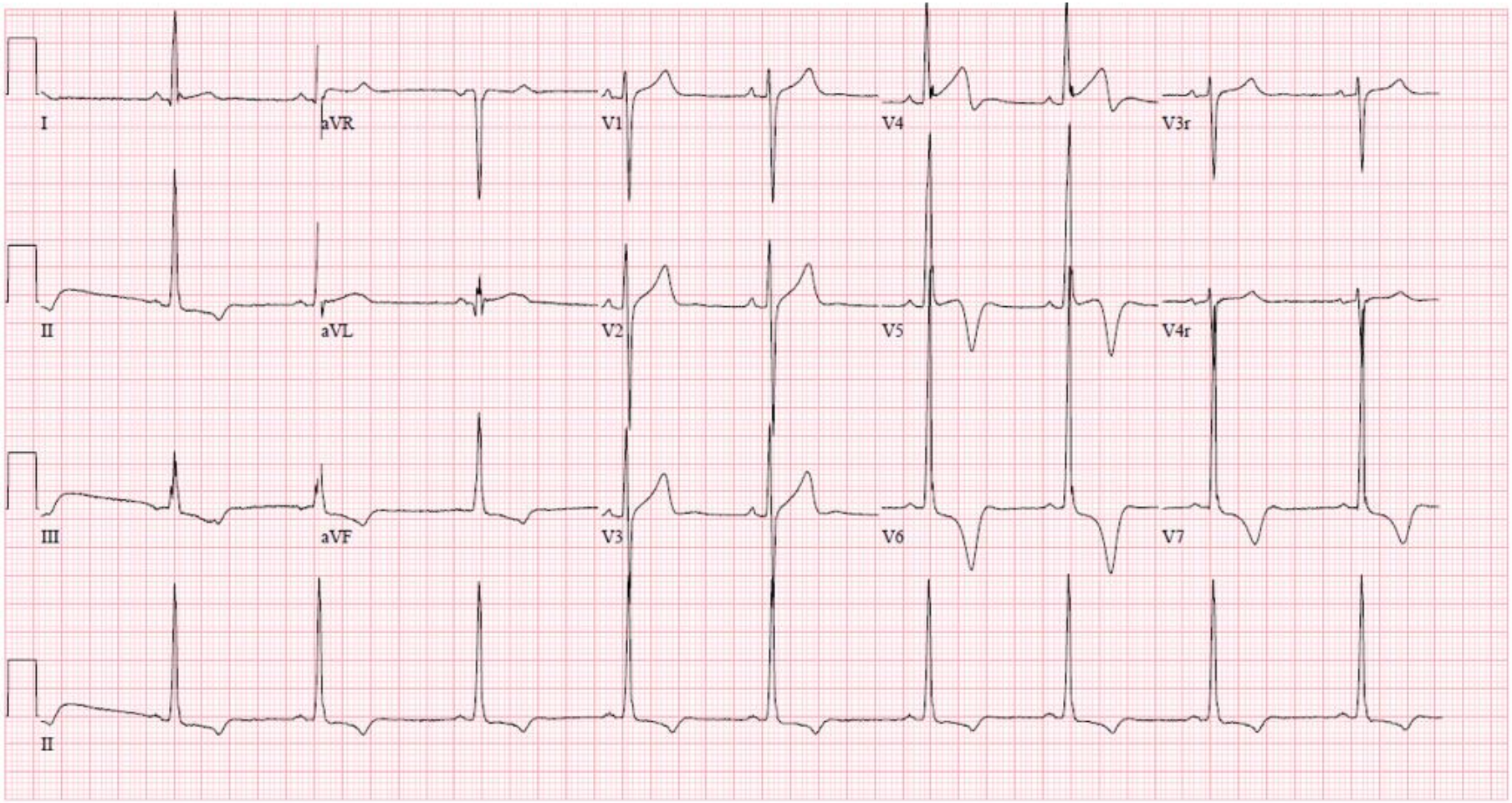


QTc = QT/√RR = 0.4/√0.6 = 515 ms

Case 3

- 16 y/o previously healthy boy collapses while playing basketball
- No prodrome, he does not recall falling
- Collapses two more times during time out
- Positive FH of sudden early cardiac death
- Exam in hospital unremarkable
- EKG and echocardiogram performed

Case 3 – Hypertrophic Cardiomyopathy



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- Thompson, W.R., Reinisch, A.J., Unterberger, M.J. et al. Pediatr Cardiol (2018). **Artificial Intelligence-Assisted Auscultation of Heart Murmurs: Validation by Virtual Clinical Trial.** Sensitivity and specificity for detection of pathologic cases were 93% (CI 90–95%) and 81% (CI 75–85%), respectively, with accuracy 88% (CI 85–91%).
- Lu et al, Congenital Heart Disease, November/December **Development of quality metrics for ambulatory pediatric cardiology: Chest pain.** The 3 approved quality metrics included: (1) documentation of family history of cardiomyopathy, early coronary artery disease or sudden death, (2) performance of electrocardiogram in all patients, and (3) performance of an echocardiogram to evaluate coronary arteries in patients with exertional chest pain
- Stewart, et al Pediatrics, January 2018, Vol 141 **Pediatric Disorders of Orthostatic Intolerance** (review article)