Pediatric Cardiology for the Primary Care Provider

Mary Anne Milbert, RN, MSN, CPNP-PC
DISCLOSURES

• There has been no commercial support or sponsorship for this program.
• The planners and presenters have declared that no conflicts of interest exist.
• The program co-sponsors do not endorse any products in conjunction with any educational activity.
ACCREDITATION

Boston College Connell School of Nursing Continuing Education Program is accredited as a provider of continuing nursing education by the American Nurses Association Massachusetts, an accredited approver by the American Nurses Credentialing Center’s Commission on Accreditation.
SESSION OBJECTIVES

• Name four most common congenital heart defects.
• Identify when to refer for chest pain, syncope and murmur.
Primary Care Cardiology and Case Studies

Mary Anne Milbert RN, MSN, CPNP-PC
Pediatric Nurse Practitioner
Cardiovascular Program
Boston Children’s Hospital
Lecture Overview

- Assessment of Pediatric Heart Sounds
- Congenital Heart Disease
- Chest Pain
- Syncope
- Case Studies
Assessment of Pediatric Heart Sounds
Heart Murmurs

- Key Points
  - Heart murmurs in children are quite common and are the most frequent referral to a pediatric cardiologist.
  - Studies have shown anywhere from 32%-75% of children have been diagnosed with an innocent murmur between the ages of 1-14 yrs.
  - Structural congenital heart disease is uncommon, affecting less than 1% of children or 9 out of every 1000 infants or 36,000 infants are born with a heart defect.
Assessment of Pediatric Heart Sounds

- **Regions to Auscultate**

- **APT-M**
  - Aortic area
    - Is centered at the second right intercostal space (ICS), includes the suprasternal notch and neck
  - Pulmonic Area
    - Is centered at the second left ICS
  - Tricuspid Area
    - Extends from the fourth or fifth intercostal space to the subxiphisternal region
  - Mitral Area
    - Includes the apex, extends to the left sternal edge and laterally to the axillary region
Listening Posts for Pediatric Heart Sounds

- Aortic area
- Pulmonary area
- Tricuspid area
- Mitral area
- Midclavicular line
Heart Sounds

- The first heart sound (S1) is associated with the closure of the mitral Valve (M1) and the tricuspid valve (T1)
  - Best heard the beginning of systole and is synchronous with the apical impulse
  - Best heard at the mid to left lower sternal border and apex
- The second heart sound (S2) is produced by closure of the aortic valve (A2) and the pulmonic valve (P2)
  - Marks the beginning of diastole
  - Best heard at the left upper sternal border
**Physiologic Splitting of S2**

- Most important to recognize in infants and children
- Normal S2 splitting varies with respiration, increasing with inspiration and decreasing or single with expiration
- During inspiration, a greater negative pressure occurs in the thoracic cavity, which increases systemic venous return to the right side of the heart.
- This increased volume of blood in the right ventricle prolongs the duration of the RV ejection time, delaying closure of the pulmonic valve, which widens the splitting of the S2
- Splitting of S2 during inspiration is a NORMAL finding
Schematic of the Splitting of S2

- **Expiration**
  - S1
  - A2P2
  - S1

- **Inspiration**
  - S1
  - A2P2
  - S1
Third (S3) and Fourth (S4) Heart Sounds

- **S3** is commonly heard in children and elite athletes
  - Low frequency, best heard at the apex, represents early diastole and is related to the rapid filling of the ventricle
  - A LOUD S3 is abnormal and is audible in conditions characterized by dilated ventricles
- **S4** almost always indicates pathology
  - Low frequency, best heard at the apex but can be widely transmitted, late diastole
  - Characterized by decreased ventricular compliance or CHF
Schematic of the Timing of the Third Heart Sound
Schematic of the Timing of the Fourth Heart Sound
Ejection Clicks and Mid-Systolic Clicks

- Ejection clicks are high frequency snappy sounds that follow S1 very closely and occur in early systole
  - Associated with semi-lunar valve obstruction such as Pulmonic Stenosis or Aortic Stenosis

- Mid-Systolic clicks occur mid-systole
  - Associated with Mitral Valve Prolapse
  - Exaggerated by standing, lying on left side and with the Valsalva maneuver
Schematic of Ejection and Mid-systolic Clicks in Relation to S1 and S2

- Expiration
  - S1
  - EC
  - S2
  - S1

- Inspiration
  - S1
  - MC
  - S2
  - S1
Grading the Intensity of Heart Murmurs

1. Barely Audible
2. Soft but easily audible
3. Moderately loud but no thrill
4. Louder with a thrill
5. Audible with the stethoscope barely on the chest; thrill detectable
6. Audible with the stethoscope off the chest; thrill detectable
Systolic Murmurs

- Most murmurs are systolic, occurs between S1 and S2
- Two types: Ejection or Regurgitant
  - **Systolic Ejection Murmur (SEM)**
    - Interval occurs between S1 and the onset of the murmur, can be blowing or harsh, ends with the onset of S2
    - Best heard upper left/right sternal border
      - Ex: Aortic Stenosis (AS), Pulmonic Stenosis (PS), Atrial Septal Defect (ASD), Innocent Murmur (IM)
  - **Systolic Regurgitant Murmur (SRM)**
    - Begins with S1 and usually heard throughout systole (pansystolic and holosystolic)
    - Best heard at the left lower sternal border and apex
      - Ex: Ventricular Septal Defect (VSD), Tricuspid Regurgitation (TR) and Mitral Regurgitation (MR)
Diastolic Murmurs

- Much less common in children than adults
- Presence is highly suggestive of a structural problem
- Four types: Early diastolic, Mid-diastolic, Late diastolic and Continuous
  - Early Diastolic Murmur
    - occur immediately following S2
      - Ex: Aortic regurgitation (AR), high pitched, best heard at the left mid-sternal border and radiates to the apex
      - Ex: Pulmonary regurgitation (PR), best heard at the upper left sternal border and radiates along the sternal border
Diastolic Murmurs Cont.

- **Mid-Diastolic Murmur**
  - Start with a loud S₃
    - Ex: Mitral Stenosis (MS) or LARGE left to right shunts ie VSD or PDA which produce MS secondary to the large blood flow across the normal-sized mitral valve

- **Late-Diastolic Murmur**
  - Pre-systolic
    - Ex: MS or Tricuspid Stenosis (TS)

- **Continuous Murmur**
  - Begin with S₁ and continue without interruption through S₂ into all parts of diastole
    - Ex: Patent Ductus Arteriosus (PDA), Atrio-Ventricular Fistula and (AVF), Truncus Arteriosus (TA), Venous Hum
Congenital Heart Disease
Congenital Heart Disease

- Aortic Stenosis
- Pulmonic Stenosis
- Bicuspid Aortic Valve
- Coarctation
- Mitral Valve Prolapse
- Patent Ductus Arteriosus
- Atrial Septal Defect
- Ventricular Septal Defect
- Tetralogy of Fallot
Normal Intracardiac Anatomy and Blood Flow

- Superior Vena Cava
- to Lungs
- Pulmonary Veins from Lungs
- Lungs
- Atrial Septum
- Tricuspid Valve
- Inferior Vena Cava
- Pulmonary Valve
- RA (Right Atrium)
- PA (Pulmonary Artery)
- LA (Left Atrium)
- LV (Left Ventricle)
- RV (Right Ventricle)
- Mitral Valve
- Aortic Valve
- Ventricular Septum
Valvar Stenosis (AS or PS)

- **Valvar** - the valve leaflets are thickened and or narrowed
- **Supravalvar** - above the valve is thickened or narrowed
- **Subvalvar (infundibular)** - the muscle under the valve area is thickened, narrowing the outflow tract
Aortic Stenosis/Bicuspid Aortic Valve

- Overall incidence is 0.2-0.5 per 1000 children. Most cases of aortic valve stenosis in young people are due to a **bicuspid aortic valve**, the aortic valve has only 2 leaflets instead of the normal 3.

- Familial incidence with left ventricular outflow obstruction (LVOO) 3-5%, recommend screening for first degree relatives.

- The murmur of aortic stenosis is typically a mid-systolic ejection murmur, heard best over the “aortic area” or right second intercostal space, with radiation into the right neck.

- Grading of AS is mild, moderate and severe and is dependent on the degree of narrowing.
Bicuspid Aortic Valve
Treatment Options for AS

- Wide spectrum of treatment options
- Most children with mild aortic valve stenosis require bi-annual follow-up (with or without aortic regurgitation), the degree of stenosis often gradually worsens over time
- Severe or moderate to severe aortic valve stenosis require intervention to prevent long term damage to the heart, initial intervention is a balloon valvuloplasty through a cardiac catheterization
- Valvotomy is surgical release of scar tissue within the aortic valve leaflets that is preventing the valve leaflets from opening properly
Pulmonic Stenosis

- Overall incidence is about 8-10% of all cases of congenital heart defects, one of the most common defects
- Thickened leaflets with commisural fusion, post-stenotic dilatation common
- Classic systolic ejection murmur in pulmonic area, with an ejection click present, diastolic murmur is related to the “leaking” PR,
- Connective tissue disorders i.e. Marfan’s Syndrome, Ehlers-Danolos have a high incidence of MVP, more than 50%
Pulmonic Stenosis

Normal heart

Pulmonary valve stenosis

Pulmonary artery
Treatment for PS

- Mild pulmonary stenosis often does not require treatment
- Moderate or severe stenosis is treated with repair of the pulmonic valve, balloon valvuloplasty most common
- Valvotomy is surgical release of scar tissue within the pulmonary valve leaflets that is preventing the valve leaflets from opening properly
- Patch enlargement:
  - Subvalvar PS, an incision is made into the right ventricle and a patch is sewn into the cut edges of the right ventricle to enlarge the area below the pulmonary valve where the narrowing was
  - Supravalvar PS, the narrowing is in the artery just beyond the pulmonary valve, patches are sewn into this artery to enlarge its diameter and relieve the narrowing
Coarctation of the Aorta

- Overall incidence is 5 in 10,000 births or 5% of all congenital heart defects
- Cause is unknown, certain chromosomal abnormalities such as Turner's syndrome high incidence, more than 50% have a Bicuspid Aortic Valve
- The murmur of a Co-Arc is typically a systolic ejection murmur, heard best over the upper sternal border with intrascapular murmur, high blood pressure in upper extremities and decreased femoral pulses
- Elastic tissue from the ductus arteriosus may encircle the aorta and cause a "lasso effect" on it as the ductus closes, resulting in a coarctation of the aorta
Coarctation of the Aorta
Treatment options for Co-Arc

- In a critically ill newborn, prostaglandin (PGE-1), is used to open the ductus arteriosus allowing blood to flow to areas beyond the coarctation.
- Severity and length of the coarctation segment will dictate the most appropriate technique, surgical vs balloon dilation.
- Recurrence of coarctation at the area of the repair is possible, even years following treatment, re-stenosis is highest among newborns, and decreases in older children.
- The majority of cases can be managed with cardiac catheterization and balloon dilation.
Mitral Valve Prolapse

- Mitral valve prolapse is the most prevalent cardiac valve disorder.
- Overall incidence is about 3-5% of pediatric patients likely have clinically significant mitral valve prolapse, recent article in CA however stated 0.6%
- The prolapse occurs from various underlying causes affecting one or more portions of the mitral valve leaflets, chordae tendineae, papillary muscle, and/or valve annulus.
- Classic mid-systolic click heard at the apex, regurgitant systolic murmur is related to the “leaking” MR,
- Connective tissue disorders i.e. Marfan’s Syndrome, Ehlers-Danolos have a high incidence of MVP, more than 50%
Mitral Valve Prolapse

- Normal closed mitral valve
- Prolapsed mitral valve
- Left atrium
- Chordae tendineae
- Left ventricle
- Papillary muscles
- LV
- LA
- PML

APEX LLD
Treatment for MVP/MR

- Asymptomatic patients require no specific treatment and they should be reassured of their excellent prognosis
- MVP patients without mitral regurgitation should be evaluated every 3 to 5 years
- Mild-mild+ should be evaluated every 2 years
- Moderate to severe mitral regurgitation or high-risk features should be reviewed with an echocardiogram yearly or more often
- When surgery is required, mitral valve repair is usually feasible, repair is characterized by low mortality and long-lasting durability; the 10-year reoperation-free survival rate ranges between 93% and 96%
Patent Ductus Arteriosus

- Abnormal blood flow between the aorta and pulmonary artery, typically closes in a couple of days after birth.
- Occurs in about 8 out of every 1,000 premature babies, compared with 2 out of every 1,000 full-term babies, more girls than boys.
- Infants with genetic disorders, such as Down syndrome, and whose mothers had rubella during pregnancy are at higher risk for PDA.
- Classic continuous murmur, machinery sounding, in premature infants, a heart murmur may not be heard.
Patent Ductus Arteriosus
Treatment for PDA

- If treatment is needed, medications such as indomethacin or a special form of ibuprofen are often the first choice, works well with little side effects, the earlier treatment is given, the more likely it is to succeed
- Transcatheter device closure; few different types small metal coil or other blocking device
- Surgery may be needed if the catheter procedure does not work or it cannot be used, surgery involves making a small cut between the ribs to repair the PDA (Trans-thoracic)
- If other heart problems or defects, keeping the ductus arteriosus open may be lifesaving. Medicine may be used to stop it from closing
SCAMP Criteria for PDA

Isolated PDA on any ECHO

If unexplained poor wgt gain then cath for HD/PDA closure

ECHO reveals: LVEDV z score >2.5 or PDA >2mm (<2 y/o) OR > 3mm (>2 y/o) or RVp > ½ systemic

Yes

No  LVEDV z score > 1.8 Yes

No  RTC < 2 y/o at 3 yrs of age

2-4 y/o in 18 mons

>4 y/o in 3 yrs, then 5 yrs then exit
Atrial Septal Defect

- Overall incidence is 2-3 per 1000 children, second most common congenital heart defect
- Cause is unknown, certain genetic disorders have a high incidence ex. Down syndrome, trisomy 13, and trisomy 18
- The murmur of an ASD is typically a systolic ejection murmur, heard best over the upper sternal border with radiation, fixed or widely split $S_2$
- Size of defect determines treatment options small, moderate and large
Atrial Septal Defect
Treatment options for ASD

- Wide spectrum of treatment options, small no treatment generally close spontaneously, typically surgery vs cath closure between 2-5 yrs of age
- Surgical closure is quite rare, done for large defects or defects with not enough septum to attach device
- Device closure if small to moderate, new the last 10-15 years, cath procedure which is minimally invasive
  - Advantages: less discomfort and shorter stay
  - Disadvantages: lack of long term follow-up on the safety and efficacy of device closure compared with surgery ie: Amplatzer device recently issues with erosion of walls
Ventricular Septal Defects

- A VSD is the most common type of congenital heart defect; 5-7 per 1,000 infants born have a VSD.
- Two types: Muscular and Membranous.
  - Muscular: small-medium sized defects rarely cause problems, close spontaneously up to 70%, never get bigger only get smaller.
  - Membranous: same as above, close spontaneously only 30-40% of the time.
- Large Defects: cause CHF, generally need medications, Lasix, Aldactone and Digoxin and increased caloric feeds; defects will become smaller over time, will electively close at 4-6 mons if not controlled with meds.
Ventricular Septal Defect
Treatment for VSD

- Surgical closure of isolated ventricular septal defects is uncomplicated in 99 percent or more of cases
- Some ventricular septal defects may be closed using an FDA approved closure device
- Since the 2007 SBE recommendations at BCH:
  - Small hemodynamically insignificant muscular defects are D/C at 3 y/o
  - Small membranous defects are followed every 2-3 yrs
    - Can develop LVOT obstruction, Sub Ao membrane
    - Increase in RV/LV muscle bundle
Tetralogy of Fallot

- Clinical picture of this defect is wide, from asymptomatic forms to extreme forms in which the anatomical and functional communication between right ventricle and pulmonary artery is absent (pulmonary atresia with Tetralogy of Fallot), with severe clinical expression
- Pulmonary artery stenosis is constantly present and represents the central element of Tetralogy of Fallot
- It may be a pulmonary valve stenosis in the right ventricular outflow tract or a hypoplasia of the pulmonary artery trunk
- May be associated with chromosomal abnormalities, such as 22q11 deletion syndrome otherwise known as DiGeorge Syndrome
- Cyanotic TOF
Tetralogy of Fallot

Four abnormalities that result in insufficiently oxygenated blood pumped to the body:

1. Narrowing of the pulmonary valve
2. Thickening of wall of right ventricle
3. Displacement of aorta over ventricular septal defect
4. Ventricular septal defect - opening between the left and right ventricles
Treatment for TOF

- Clinical picture of this defect is wide, acyanotic infants to cyanotic
- Depending on O2 levels determines timing of surgery, if critically low surgical repair in the neonatal period
- Infants with normal oxygen levels or only mild cyanosis are usually able to go home in the first week of life and elective surgery at 6 mons of life
- Closure of the ventricular septal defect with a synthetic Dacron patch so that the blood can flow normally from the left ventricle to the aorta
- The narrowing of the pulmonary valve and right ventricular outflow tract is then augmented (enlarged) by a combination of cutting away (resecting) obstructive muscle tissue in the right ventricle and by enlarging the outflow pathway with a patch
SBE Guidelines

- New guidelines in 2007 the American Heart Association simplified its recommendations
- Dental procedure prophylaxis:
  - Artificial valve or a valve repaired with artificial material
  - History of endocarditis
  - Heart transplant with abnormal heart valve function
  - Congenital heart defects including:
    - Cyanotic heart disease, not fully repaired, has a conduit or shunt
    - S/P repair with artificial material or a device for 6 mons
    - Residual defects, leaks or abnormal flow at/or adjacent to a prosthetic patch or device
Chest Pain
Chest Pain

- Common symptom in children and adolescents
- CP produces much anxiety and concern in parents and patients and can lead to school absences and unnecessary restrictions from sports
- Careful history and physical exam can help determine the cause without the need for multiple diagnostic tests
- If no fever, negative cardiac or pulmonary exam, chest pain does not occur with exertion or exertional syncope, if the pain does not radiates to back, jaw, left arm or shoulder or increases with supine position then **cardiac basis for the chest pain is unlikely**
- Literature: low incidence of cardiac pathology ranging from 0%-5%
- There is a large dichotomy between a family’s understanding of pediatric chest pain and the medical cause of the symptom as is outlined in table below
## Causes of Pediatric Chest Pain

**Family’s concern vs Medical Dx**

<table>
<thead>
<tr>
<th>Cause</th>
<th>Family’s Estimate (%)</th>
<th>Medical Diagnosis (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiac</td>
<td>52-56%</td>
<td>1-6%</td>
</tr>
<tr>
<td>Musculoskeletal</td>
<td>13%</td>
<td>15-31%</td>
</tr>
<tr>
<td>Resp Tract</td>
<td>10%</td>
<td>2-11%</td>
</tr>
<tr>
<td>Cancer</td>
<td>0-12%</td>
<td>0</td>
</tr>
<tr>
<td>Idiopathic</td>
<td>10-19%</td>
<td>21-45%</td>
</tr>
</tbody>
</table>
### Conditions associated with chest pain

- Analyzed charts of patients aged 7-21 yrs from Jan 00’-Dec 09’, identified nine cardiac conditions known to cause chest pain (Total of 484)
- Excluded patients with known CHD or acquired heart disease

<table>
<thead>
<tr>
<th>Condition</th>
<th>Included Pts</th>
<th>Pts with CP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic Dissection</td>
<td>1</td>
<td>0%</td>
</tr>
<tr>
<td>Coronary anomalies</td>
<td>131</td>
<td>34 (26%)</td>
</tr>
<tr>
<td>Dilated cardiomyopathy</td>
<td>61</td>
<td>5 (8%)</td>
</tr>
<tr>
<td>Hypertrophic</td>
<td>100</td>
<td>5 (5%)</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>62</td>
<td>46 (74%)</td>
</tr>
<tr>
<td>Pericarditis</td>
<td>65</td>
<td>62 (95%)</td>
</tr>
<tr>
<td>Pulm Embolus</td>
<td>19</td>
<td>13 (68%)</td>
</tr>
<tr>
<td>Pulm hypertension</td>
<td>37</td>
<td>6 (16%)</td>
</tr>
<tr>
<td>Takayasu arteritis</td>
<td>8</td>
<td>0%</td>
</tr>
</tbody>
</table>
Chest Pain SCAMPS (Standardized clinical action and management plans) summary

- July 2010-December 2011 at BCH and NECCA sites
  - 109 providers
  - 1,018 patients
  - Average age 13 yrs

- Results
  - 99.1% had single visit; 0.9% had a return

- ?? Cardiac Cause
  - Pericarditis
  - Anomalous origin of the right coronary

- Musculoskeletal and pulmonary most common

- 9 return pts had no disease
SCAMP Data thru Jan 2015

- Total pts enrolled 3175
- Total Chest Pain Encounters 3390 (Included f/u encounters)

Chest Pain Overview

- Pts with exertional CP 39.7%
- Pts with and w/o exertional CP that followed ECHO recommendations 60.3%
- Pts Identified with a Cardiac Dx for CP ??Guess??
### Cardiac Chest Pain:

<table>
<thead>
<tr>
<th>Cardiac Diagnosis</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myocarditis</td>
<td>1 (0.83%)</td>
</tr>
<tr>
<td>Anomalous Coronary Artery</td>
<td>2 (1.67%)</td>
</tr>
<tr>
<td>Pericarditis</td>
<td>4 (3.33%)</td>
</tr>
<tr>
<td>Hypertrophic Cardiomyopathy</td>
<td>1 (0.83%)</td>
</tr>
<tr>
<td>Other Cardiac Diagnosis</td>
<td>11 (9.17%)</td>
</tr>
<tr>
<td>Diagnosis Pending</td>
<td>101 (84.17%)</td>
</tr>
<tr>
<td><strong>Total Patients with cardiac dx</strong></td>
<td><strong>19</strong></td>
</tr>
</tbody>
</table>

### Non-Cardiac Chest Pain:

<table>
<thead>
<tr>
<th>Final Diagnosis (Non-Cardiac)</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-Cordial Catch Syndrome</td>
<td>53 (2.10%)</td>
</tr>
<tr>
<td>Not Recorded</td>
<td>428 (16.99%)</td>
</tr>
<tr>
<td>Psychological</td>
<td>88 (3.49%)</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>111 (4.41%)</td>
</tr>
<tr>
<td>Palpitations / Arrhythmia</td>
<td>2 (0.08%)</td>
</tr>
<tr>
<td>Deconditioning</td>
<td>26 (1.03%)</td>
</tr>
<tr>
<td>Neurological</td>
<td>7 (0.28%)</td>
</tr>
<tr>
<td>Pulmonary</td>
<td>172 (6.83%)</td>
</tr>
<tr>
<td>Syncope</td>
<td>3 (0.12%)</td>
</tr>
<tr>
<td>Musculoskeletal</td>
<td>1446 (57.40%)</td>
</tr>
<tr>
<td>Other</td>
<td>183 (7.26%)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>2519</strong>*</td>
</tr>
</tbody>
</table>
Syncope
Syncope

- Common disorder in pediatrics
  - High incidence in 6 months to 5 years, declining until adolescents, 20-40% will have at least one faint
  - Typical syncope is not associated with an increased incidence in mortality however the symptoms are frightening and often trigger extensive unnecessary testing
- Therapy usually depends of frequency, physiology and patient/family perceptions
Classification of Syncope

- Breath Holding Spells
  - Onset 6 months-3 yrs
  - Triggering event
  - Infrequent
  - Aborted cry
  - Incidence 5-10%

- Typical Syncope
  - Onset 3-18 yrs
  - Early adolescent or older highest incidence
  - Positional changes with prodrome
  - Females >> Males
  - Injury and incontinence very rare
Classifications of Syncope Cont

- Postural Orthostatic Tachycardia Syndrome (POTS)
  - Frequent symptoms
    - Frequent palpitations
    - SOB
    - Dizziness with out syncope
    - Chest pain
    - Headaches and other somatic symptoms
  - Females>>> Males
  - May have a connective tissue disorder
Classification of Syncope Cont

- Convulsive Syncope
  - Prolonged standing
  - Identifiable trigger
    - Stretch
    - Hair brushing
    - Micturition
    - Cough
    - Needle
    - Minor Injury

- Exertional Syncope
  - Two Patterns
    - Abrupt syncope at peak exercise
    - Post-Exertional dizziness/collapse
## SCAMP History and Scoring

### Severity Scores

<table>
<thead>
<tr>
<th>Subjective Syncope Score (done by patient)</th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
<th>8</th>
<th>9</th>
<th>10</th>
</tr>
</thead>
<tbody>
<tr>
<td>How dizzy are you on a daily basis?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10 being every time you stand, 0 being never</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>How well do you feel?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10 being &quot;perfect&quot;, 0 being &quot;dead&quot;</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Objective Syncope Score

*Please circle or check most applicable in each individual section*

<table>
<thead>
<tr>
<th>Number of overall syncopal events</th>
<th>0-1</th>
<th>2-4</th>
<th>5-10</th>
<th>&gt; 10</th>
</tr>
</thead>
<tbody>
<tr>
<td>Never</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt; Weekly</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Multiple events weekly</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Daily</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Lightheadedness/dizziness without syncope</th>
<th>Never</th>
<th>&lt; Weekly</th>
<th>Multiple events weekly</th>
<th>Daily</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clustered and/or &gt; 1 ER visit</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Injury</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Severity of Spells</th>
<th>&lt;1 minute, minimal residual</th>
<th>Brief, modest residual</th>
<th>Clustered and/or &gt; 1 ER visit</th>
<th>Injury</th>
</tr>
</thead>
<tbody>
<tr>
<td>Disability/ School Attendance</td>
<td>Normal School Attendance</td>
<td>Early pickup and/or &gt; 2 school nurse visits</td>
<td>Missed 3-5 days</td>
<td>Missed &gt; 5 days</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Total Score</th>
<th></th>
</tr>
</thead>
</table>
Initial Evaluation

Flowchart:
- Obtain History, Physical Exam, Orthostatic Blood Pressures, and ECG
  - Normal ECG?
    - YES: Normal Echo?
      - YES: Does the patient have M&V or prolonged QTc > 0.45 seconds?
        - NO: Choose the box that appropriately fits the patient and follow the categorical decision tree
        - YES: Exit SCAMP
    - NO: Obtain Echocardiogram
  - NO: Clear diagnosis of cardiac disease or suspected seizure?
    - YES: Exit SCAMP
    - NO: Does the patient have M&V or prolonged QTc > 0.45 seconds?
      - NO: Refer to Neurology
      - YES: Exit SCAMP

Decision Points:
- Normal ECG
- Normal Echo
- Does the patient have M&V or prolonged QTc > 0.45 seconds?

Conditions:
- Transient episode of loss of consciousness with the total duration of unconsciousness and loss of tone lasting ≤ 1 min. Includes patients who present with clearly triggered prodromal symptoms or situational syncope.
- Transient collapse with or without significant prodrome, while actively involved in significant exercise.
- Abrupt progression of prodromal symptoms and transient collapse with severe and limited opisthotonic posturing or myoclonic jerks that resolve within seconds.
- Does not fit into the other 3 categories of syncope or has the combination of a ≥ 35 bpm increase in heart rate with orthostatic vital signs, and a 3 minute standing heart rate ≥ 110 bpm and at least plausible pre-syncope symptoms.

Categories:
- Typical Syncope
- Exertional Syncope
- Convulsive Syncope
- POTS/Atypical Syncope
Typical Syncope Decision Tree

*Non-Pharmacologic Management
- Increase sodium (2-4g/day)
- Increase fluids (2-3L/day)
- Antigravity maneuvers
- Recognition prophylaxis

Provide patient/family with teaching sheet and non-pharmacologic counseling*

Syncope Severity?

LOW 0-4 OBJECTIVE SCORE

IS THE SYNCPE SITUATIONAL?
- e.g. blood draws, noxious stimuli

YES NO

Provide reassurance

No Follow-Up Recommended

Follow-Up: Best Clinical Judgment

Observe symptoms

FOLLOW-UP 2 MONTHS

Moderate 5-8 OBJECTIVE SCORE

Obtain labs through PCP prior to next visit

Normal?

YES NO

Observe symptoms without drugs

Follow-Up 2 Months

FOLLOW-UP 2 MONTHS

Disable 9-12 OBJECTIVE SCORE

Follow POTS/Atypical decision tree on page 8

**Ferritin <50ng/mL Ferrous Sulfate 3mg/kg/day with a max dose of 325mg
Exertional Syncope Decision Tree

- **Exertion?**
  - Yes: Provide non-pharmacologic treatment
  - No: Obtain Echocardiogram
    - Normal? (Yes) Exit SCAMP
    - Anomaly? (Yes) Refer to EP and Exit SCAMP (Anomaly? (No) Continue)

- **Syncope during Exertion**
  - Low (0-4) Objective Score: Observe symptoms with no treatment recommended
  - Moderate (5-8) Objective Score: Order Holter or loop monitor based on frequency of symptoms
  - Severe (9-12) Objective Score: Follow FDOTS/Topical decision tree on page 8

- **Follow-Up Best Clinical Judgment**

- **Syncope at Rest**
  - Provide NPPV and/or prescribe Fludrocortisone
  - Follow-up 2 Months

- **Syncope with Exercise**
  - Provide NPPV and/or prescribe Midodrine with exercise
  - Follow-up 2 Months

- **Testing Recommendations**
  - Order Holter monitor if multiple symptoms each day
  - Order loop monitor if reasonable expectation of symptoms within 2 weeks
  - If less frequent symptoms, use best clinical judgment

- **Non-Pharmacologic Management**
  - Increase sodium (2-4g/day)
  - Increase fluids (2-3L/day)
  - Antigravity maneuvers
  - Recognition prophylaxis
Treatment of Syncope

- Fluid recommendation
  - 2-4 L per day
  - 1-2 gms NA per day
  - Nuns Na Tabs (REI or EMS)
  - Include an electrolyte solution (G2 or Power-ade Zero)
  - Salty Snacks
    - Pretzels, wheat thins, gold fish, pickles, canned soups

- Medical management
  - Low ferritin (normal 10-180) however if less than 50 tx with Ferrous gluconate
  - Florinef 0.1 mg QD or BID
  - If not successful, refer to EP
Case Studies
Importance of Vital Signs

Clinical History

- At 4 y/o WCC to have a “slow heart rate”
  - Referred for outpt chest x-ray and EKG
    - Complete heart block on EKG (AP 57) with mild cardiomegaly
    - History 5/18/12 tick noted, no bull's-eye just redness followed at PCP next day, (not engorged, no rash, no symptoms)
    - 6/6/12 c/o H/A’s, sm bump on back of head (occipital node), Lyme titers drawn
    - 7/11/12 c/o fatigue, no other symptoms, results of Lyme titers
      - Lyme Elisa pos, Western Blot (IgG 1/3, IgM 2/10) neg
      - Tx with 14 oral Amox
    - 8/12 c/o of fatigue, napped on vacation, NO VSS on visit
      - Tx with oral hydration and resolved over 2 weeks
Admitted to BCH

- ECHO revealed mild LV dilation with LVEDV z 3.68 and LVESV z 2.41 with an EF 68%
- Tx with 21 day IV course of Ceftriaxone (PICC line) and then 10 day po Amox
- ETT tests CHB throughout, increase AP 100, 50% predicted
- Expect Pacemaker in adolescence

- Recurrent tick exposure 5/14 with bull's-eye treated with 30 days Amox per ID

- F/U q 6mons with ECHO/Holter and ETT, no change

- Question Congenital Heart Block vs Lyme Carditis
  - No Maternal hx Lupus or any other Rheumatoid disease
  - Monitoring during delivery reportedly no arrhythmia or decrease fetal heart rate
Murmur

- 16 y/o murmur noted at WCC
- Junior HS, recent immigrant from Uganda
  - Plays pick up basketball, gym class c/o SOB and increase HR, rests and is able to participate. No c/o CP, palps, syncope, No Family hx, no recent illness or fever
    - P/E unremarkable with 1-2/6 SEM
    - EKG T-wave inversion (LLL T-wave neg), no LVH
    - ECHO revealed anomalous right coronary artery arising from the Lcusp, normal LV size and fxn
    - Cardiac MRI revealed single coronary ostium from asc aorta above the L sinus of Valsalva, short inter-arterial course between aorta/pulm artery
    - SestaMibi Stress revealed 85% predictated for age/gender, no myocardial ischemia, normal LV size and fxn
- F/U annually with SestaMibi stress and ECHO
Chest Pain

- 17 y/o with c/o CP at PE, No other concerns, Tx with 600mg Advil TID for 2 wks, no improvement
  - Normal EKG/Chest x-ray
- Junior in HS daily c/o CP since December, episodes lasts few hours to the entire day
- Missed 100 days of school (no feeling well c/o migraines, backaches and overall pain), no organized or rec sports, Gym class describes a squeezing CP no other symptoms, sits out of gym and does not return
- PE: Wgt 93%, Hgt 23% and BMI 96%, No Murmur, EKG nrml
- EHCO revealed a RACA arising from the L SoV, looks to have a longer inter-arterial course, nrml LV size and fxn
- Scheduled Cardiac MRI and SestaMibi Stress in April
Syncope

- 12 y/o with 4-5 episodes of syncope
  - c/o of tunnel vision, ringing in ears, all with postural changes, no c/o cp or palps, no loss b/b, no injury, awake and alert within a min
  - Family history.....Dad with an LVAD, Dx with Cardiomyopathy in 2008 when ICD placed and on LVAD in 2012. No genetic testing done
    - ECHO normal
    - F/U based on Dad’s genetic testing, however if no testing or Dad negative then f/u 2 years

- 15 y/o with 3-4 episodes of syncope
  - Most recent episodes occurred a few wks ago with blood draw, another during health class, c/o tunnel vision and ringing in ears, no c/o cp or palps, no loss b/b, no injury, awake and alert within a min , first one in 9/14 while playing hockey, felt knees get weak, tunnel vision and went down face first.
    - ECHO normal inter-cardiac anatomy, nrml LV size and fxn
    - ETT scheduled
Easiest Heart Patient

Thank You!