Clinical Pearls of Pediatric Cardiac Assessment: A Workshop for APNs

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Disclosures

- I have no disclosures
Learning Objectives

- Describe key features of the history and cardiac exam that assist in discerning pathologic from innocent murmurs and when to refer
- Understand basic components of pediatric EKG interpretation and describe key features of common pediatric arrhythmias
- Describe newborn assessment and history taking skills when congenital heart disease is suspected
- Review several of the more common congenital and acquired heart disease cases that the primary care provider may encounter in the clinic setting from newborn through adolescent visits
True or False:

Heart murmurs are common in the pediatric population.
True or False:

All patients with a murmur need an echocardiogram
Patients with atrial septal defects may have all of the following on physical examination except:

- A. A widely split second heart sound
- B. Diminished femoral artery pulses
- C. A systolic murmur at the ULSB
- D. A diastolic rumble in the tricuspid area
- E. Increased precordial activity
Joint Policy Statement

2017: American Academy of Pediatrics and American College of Cardiology

-Organized resource for primary care providers who manage patients and families with CHD through life stages
Murmurs and Heart Sounds
Normal Anatomy
Normal Pressures
Temporary increases in cardiac output can increase turbulence and therefore cause murmurs to be temporarily present.

- Pregnancy
- Fever
- Exercise
- Excitement or fear
Heart Sounds and Murmurs

What characteristics are consistent with innocent murmurs and which are associated with pathologic disease?
Assessment: History

- Maternal Health History
- Family History
- Birth and present history
Assessment: History

Maternal History

Infections: Rubella
- PDA
- PPS

Maternal Chronic Illness
- Diabetes
- Maternal CHD
- Lupus

Drugs during pregnancy
- Thalidomide
- Coumadin
- Tegretol
- Lithium
- Cocaine
- Alcohol
Assessment: History

Family History

- Syncope
- Sudden unexplained death
- Arrhythmias
- History of CHD
- History of Acquired Heart disease
Assessment: History

Birth History

- Birth weight
- GA, Apgars
- Length of Stay
- Cyanosis Apnea
- Murmur at birth
Assessment: History

Present History

- Rate of weight gain
- Feeding: -diaphoresis -Length of feeds
- Exercise intolerance
- Cyanosis -Tachypnea -Infections -Hospital admissions
- Symptoms: -chest pain -palpitations -syncope -dizziness
Assessment: Physical Exam

Exam

- Inspection
- Palpation
- Auscultation
Physical Exam

Inspection

Clubbing
  Refer

Cyanosis
  Refer

Left Parasternal Bulge
  Refer
Clubbing
Inspection

Left parasternal bulge
CXR: Anatomic Correlation

Courtesy of Maureen Madden
Palpation

- Increased Precordial Activity or Heave
  - Refer (Increased cardiac output) heart disease
    - Anemia, left to Right shunts
    - Hyperthyroid

- Palpable Thrill

- Decreased Femoral Pulses
  - Refer (Increased cardiac output) heart disease
    - Anemia, left to Right shunts
    - Hyperthyroid

- Bounding Pulses
  - Refer
    - Anemia
    - PDA
    - Aortic Valve Insufficiency
Auscultation Pearls

- Listen in both supine and sitting positions
- Listen over the entire chest wall
- Listen anteriorly and posteriorly
- If rhythm sounds abnormal, consider EKG
- Take your time
- Make sure you listen at the RUSB
Heart Sounds

First Heart Sound (S1)
- Represents closure of mitral and tricuspid valves
- Best heard at LLSB

Second Heart Sound (S2)
- Represents closure of pulmonary and aortic valves
- Best heard at mid sternal border
- Splitting should be intermittent and vary with respirations
Auscultation – Second Heart Sound (S2)

Listen to S2

- Widely Split and Fixed
  - Refer
    - ASD
    - CRBBB
- Loud and Single
  - Refer
    - Pulmonary Hypertension
- Normal
Describing Murmurs

- Intensity
- Character
- Timing
- Location
- Transmission
Intensity

Grade I - Very faint; barely audible
Grade II - Quiet, but heard immediately
Grade III - Moderately loud (S1, S2)
Grade IV - Loud, associated with thrill
Grade V - Very loud, assoc. with thrill (45 degree rule)
Grade VI - Loudest, heard without stethoscope (1 cm off chest rule), thrill
Character

- The “harsh” murmur -
  - more likely to be pathologic murmur
  - monotonous sound

- The “musical” murmur -
  - more likely to be innocent murmur
  - similar to plucking guitar string
Timing

In Systole
More information needed

In Diastole
Refer

Continuous
Refer

More information needed
Thinking More

Murmur Loudest at LLSB

Vibratory, Musical? Gets softer when the patient stands or sits up?
- Reassure Functional or “Stills Murmur”

Harsh? Gets louder when the patient stands?
- Refer HCM
True or False:

Only some diastolic murmurs are pathologic.

FALSE
1. Where is the murmur loudest?

- Upper Right Sternal Border: Refer Aortic stenosis
- Upper Left Sternal Border: Consider referral ASD PS PPS
- Lower Left Sternal Border: Think More
- Apex: Refer Mitral valve insufficiency

2. Widely radiating murmur?

Referral likely
Take Home Pearls

- **Innocent**
  - Localized
  - Short in duration
  - Changes with position
  - Normal heart sounds
  - Asymptomatic patient
  - Musical
  - Systolic

- **Pathologic**
  - Palpable thrill
  - Widespread transmission
  - Abnormal S2
  - Harsh
  - Holosystolic, diastolic, or continuous
  - Murmur at RUSB
  - Abnormal pulses
  - Concerning signs and symptoms
  - Concerning family history
Putting the Puzzle Pieces Together

- Diastolic sound
- Palpitations
- Cyanosis
- Palpable thrill
- Dizzy
- History
- Family hx
- Syncopal
- Pulses
Pediatric ECG Interpretation
Assessing ECGs

- Rate:
  - Varies with age—the younger the child, the faster the HR
  - Varies with status at the time of the EKG (crying, fever, sleeping)

- Normal heart rates for age:
  - Newborn – 110-150
  - 2 years – 85-125
  - 4 years 75-115
  - 6 years 65-100
  - Older than 6 years – 60-100
Interpreting ECGs

- Rhythm (sinus or non-sinus) with consideration to the P axis
- Heart Rate
- QRS axis, T axis, and QRS-T angle
- Intervals: PR, QRS, QT
Review of Cardiac Electrophysiology

- One cardiac cycle = P wave + QRS complex + T wave
- Two important intervals: PR and QT
- Two segments: PQ and ST
The sinoatrial (SA) node is the pacemaker for the heart

SA node impulse depolarizes right and left atria-
Produces a P wave

Atrial impulse arrives at atrioventricular (AV) node and passes through node at slower force-produces PQ segment

Impulse reaches Bundle of His-faster velocity-passes to left and right bundle branches to ventricles through Purkinje fibers-produces narrow QRS

Repolarization of the ventricle produces T wave (repolarization of atria not visible on ECG)
The Conduction System
Lead Placement
Normal EKG
Normal EKG
We Will Focus On:

- Heart rate
- Rhythm
- Intervals: PR, QT
- Significance of a delta wave, QRS complexes
- Common arrhythmias
- ST segment changes
Arrhythmias From the AV node

- Nodal Premature Beat
- Sinus Pause and Nodal Escape Beat
- Nodal or Junctional Rhythm
- Nodal Tachycardia
What Happened To the P Waves?
Measurements of ECG
Heart Rate

- Why? Fast or Slow?
- Time is measured horizontally on ECG paper
  - 1 mm = 0.04 second
  - 5 mm = 0.20 second (one large division between heavy lines)
  - 30 mm = 1.2 seconds (6 large divisions)
ECG Paper

ECG Paper is divided into large squares and small squares

0.20 secs

0.04 secs

Large Squares are defined by dark lines and are 5 small squares high by 5 small squares long
What’s wrong?
Intervals PR

- PR: onset of P wave to beginning of QRS complex
- Tells provider normalcy of conduction through atrioventricular node
- Easiest to measure in Lead II
Important intervals and How to Measure Them

- PR interval is the time from activation of the atria (manifest as the P wave) and activation of the ventricles (manifest as the QRS complex).

- PR interval varies with age and heart rate
  - The older the person and the slower the heart rate, the longer is the PR interval
  - The lower limits of normal PR interval according to age:
    - Less than 3 years – 0.08 seconds
    - 3-16 years – 0.10 seconds
    - More than 16 years – 0.12 seconds
Prolongation of PR interval

- Indicates a delay in conduction through AV node
- Myocarditis
- Atrial Septal Defect (ASD)
- Toxicity: digitalis
- Hyperkalemia
- Normal heart with vagal stimulation
Short PR interval

- Wolff-Parkinson-White (WPW) pre-excitation
- Accelerated atrioventricular conduction to one ventricle through accessory pathway
- AV reentry tachycardia
- Criteria: Short PR interval for age; Delta wave (initial slurring of QRS complex), wide QRS duration
Wolff-Parkinson-White Syndrome
Supraventricular Tachycardia

Most common arrhythmia in children
- Narrow complex tachycardia
- Accessory electrical pathway
- Rate that is “too fast to count”
- Associated with chest pain, dizziness, syncope
- Can sometimes convert to sinus rhythm with vagal stimulation
QRS Duration - Normal Range

- Premature infants: 0.04 seconds
- Full term infants: 0.05 seconds
- Children: 1-3 years old: 0.06 seconds
- Children > 3 years old: 0.07 seconds
- Adults: 0.08 seconds
Wide QRS Complexes

- Bundle branch block (right or left)
- Intraventricular block
- Ventricular arrhythmias
- Implantable ventricular pacemaker
- Ventricular hypertrophy
Right Bundle Branch Block
Ventricular Tachycardia
Ventricular Tachycardia in a 16 yo Female with Syncopal Episode
ST Segment

- Normal ST segment is horizontal/isoelectric
- In limb leads: ST segment elevation or depression up to 1 mm is not abnormal in children and infants
- ST segment elevation in precordial leads (may represent pericarditis (sub epicardial myocardial damage or pericardial effusion))
ST Segment Changes Associated With Myocardial Infarction

- Wide Q waves (>0.035 seconds) with or without Q-wave notching
- ST segment elevation (>2 mm)
- Prolongation of QTc interval (>0.44 second) with abnormal Q waves
Vent. rate 99 bpm
PR interval 170 ms
QRS duration 88 ms
QT/QTc 374/479 ms
P-R-T axen -1 15 40

Normal sinus rhythm
RSI or Q3 pattern in V1 suggests right ventricular conduction delay
Voltage criteria for left ventricular hypertrophy
Nonspecific ST and T wave abnormality
Prolonged QT
Abnormal ECG

Unconfirmed
<table>
<thead>
<tr>
<th>Metric</th>
<th>Value</th>
<th>Notes</th>
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</thead>
<tbody>
<tr>
<td>Vent. rate</td>
<td>103 bpm</td>
<td>Sinus tachycardia</td>
</tr>
<tr>
<td>PR interval</td>
<td>160 ms</td>
<td>Possible Left atrial enlargement</td>
</tr>
<tr>
<td>QRS duration</td>
<td>102 ms</td>
<td>Incomplete right bundle branch block</td>
</tr>
<tr>
<td>QT/QTc</td>
<td>366/479 ms</td>
<td>Left ventricular hypertrophy</td>
</tr>
<tr>
<td>P-R-T axes</td>
<td>22 0 111</td>
<td>Marked ST abnormality, possible lateral subendocardial injury</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Abnormal ECG</td>
</tr>
</tbody>
</table>

Referred by:  
Unconfirmed
Intervals:

QT

- Measured from onset of Q wave to end of T wave
- Measure in leads with visible Q waves
- Usually expressed in rate-corrected interval
Calculating the QTc

Bazett’s Formula

- QTc = QT measured/square root of RR interval
- Normal QTc interval (mean +/- SD) is 0.40 +/- 0.014 second with upper limit of normal 0.44 seconds in children > 6 months
- QTC interval longer in newborn and small infants with upper limit of normal QTc 0.47 second (first week of life) and 0.45 second in first 6 months of life
\[ QT_C = \frac{QT}{\sqrt{RR}} \]
ECG of Prolonged QT Interval

15 year old with collapse, pulled from pool

QT = 0.51, QTc = 0.48
Long QT Syndrome

- Disorder of myocardial repolarization characterized by prolonged QT interval on ECG and ventricular arrhythmias
- May result in sudden death
- Patients may present with complaints of syncope, seizures, palpitations
- First manifestation may be cardiac arrest
QT Prolongation

- Congenital: mutations of cardiac ion channel genes
- Acquired: drugs, electrolyte disturbances
Take Home Pearls

- Normal EKG but concerning family history
- Abnormal EKG, especially in setting of chest pain or syncope
- Abnormal rate for age
- Abnormal QTc - have reviewed by a cardiologist
- Palpitations that turn on/off suddenly
- Share clinical information with cardiologist reviewing the EKG
Neonatal Cardiac Exam
What makes the neonatal exam unique?
- Transition from fetal circulation to extra-uterine circulation
- Identifying ductal dependent congenital heart disease is time sensitive
- Primary care provider has a great responsibility of examining the newborn, as they are the first safety net to finding cardiac defects that may otherwise be missed
- Prenatal history is important

Focus here will be detecting and screening for CHD in the cardiac exam of the neonate
The Fetal Circulation

4 shunts:
- Placenta
- Ductus venosus
- Foramen ovale
- Ductus arteriosus
History

- **Prenatal History**
  - Maternal history of diabetes, thyroid disorders, obesity, HTN, connective tissue disorders
  - Fewer than 50% of cases of CHD are identified on routine prenatal ultrasound
  - Often they have a benign birth history

- **Family History**
  - Congenital abnormalities
  - Inheritable diseases with known cardiac lesions
  - Childhood deaths and siblings or first-degree relative with structural heart disease
History

- Delivery
  - Premature?
  - PROM? Maternal Group B Strep (GBS) status?

- Feeding History
  - Frequency? Quantity? How long does it take to feed?
  - Symptoms during feeding? (tachypnea, diaphoresis, color change)

- Developmental milestones
  - Often delayed in infants with CHD
Differential Diagnoses in Cyanotic Neonate

- Lung disease
- Persistent pulmonary hypertension of the newborn (PPHN)
- Congenital heart disease
Newborns with congenital heart disease may be tachypneic, but are usually not in respiratory distress.

May appear completely normal (initially).

Any newborn presenting with clinical features of shock should be evaluated for congenital heart disease.

If CHD not detected during hospitalization, generally will see signs and symptoms of CHD by two weeks of age.

- Most common sign is difficulty feeding.
- Other features: color changes, excessive irritability, diaphoresis, poor weight gain, decreased activity/excessive sleep, delay in developmental milestones.
The Physical Exam

- Should be systematic approach
- Vital signs, including oxygen saturations (pre & post ductal)
- Observe warm, unclothed infant
  - Note respiratory status, perfusion, color
- Weight, length, and head circumference
  - Decrease in weight percentiles when compared to length and HC should raise question of CHD
The Physical Exam

- Observe for:
  - Central cyanosis
  - Respiratory status
    - Tachypnea, retractions, grunting, nasal flaring
  - Signs of decreased systemic perfusion
    - Temperature and color of skin, mottling
  - Dysmorphic features
  - Extracardiac abnormalities
Cardiac exam begins with palpating the precordium to assess right ventricular impulse

- Normally can easily palpate along the lower sternal border in the newborn
- Will be normal or increased in patients with right ventricular outflow obstruction (pulmonary atresia, pulmonary valve stenosis) or in PPHN
- RV impulse is decreased with inflow obstruction (tricuspid atresia, hypoplastic right heart syndrome)
The Physical Exam

- Pulses
  - Palpating upper and lower extremity simultaneously
  - Palpate the RIGHT upper and RIGHT lower extremities together
  - Note their presence or absence, bounding or weak, thready
The Physical Exam

- Capillary refill
  - ~ 3 seconds or less
- Abdominal exam
  - Hepatomegaly
The Physical Exam

- Auscultation
  - Note rate, rhythm
  - S1 and S2
    - widely split second heart sound suggests increased pulmonary blood flow (PDA)
    - Fixed second heart sound (no split) may indicate issue with pulmonary valve (atretic or diminutive) or PPHN
  - S2 can be difficult to appreciate in the newborn
  - Presence of clicks or gallops
  - Note any murmurs (innocent or pathologic?)
  - Note lung sounds – presence of crackles (rales) may indicate pulmonary edema (RVOTO, right sided failure)
Innocent vs Pathologic Murmurs In the Infant

- Pathologic
  - Patent Ductus Arteriosus
  - VSD, PFO/ASD
  - TR (in setting of PPHN)
- May not necessarily have a murmur with congenital heart disease
- Cannot rely on the presence of absence of murmur to indicate CHD
Innocent vs Pathologic Murmurs In the Infant

- **Innocent Murmurs** – produced by normal flow
  - PPS – Peripheral Pulmonary Stenosis
    - Pulmonary flow murmur due to rapid increase of pulmonary blood flow after birth
    - a soft systolic ejection murmur heard mostly along the upper left sternal border, radiates to axillae. PPS is common in newborn infants and usually disappears at 4-6 months of age
  - Venous hum
    - Produced by returning blood flow from the great veins to the heart
    - Quality of sound will change with head and neck position
Testing

- EKG has limited value in newborn, unless arrhythmia is present
- Chest x-ray
- Echo – mainstay in diagnosis of CHD in newborn, has chiefly replaced cardiac cath
- Genetics evaluation if indicated
- Pulse oximetry screening
Pulse Oximetry

- Screen all infants with pulse oximetry, pre and post ductal after 24 hours of age.
- Screen considered positive if the O2 saturation is:
  - <90% in any one extremity
  - <95% in both extremities on 3 separate measures taken 1 hour apart
  - >3% absolute difference in O2 saturation between upper and lower extremities on 3 separate measures taken 1 hour apart
Take Home Pearls

- Findings that warrant further evaluation
  - Pathologic murmurs
  - Suspicious history of poor growth, difficulty feeding
  - Positive pulse ox screening test
  - Reports of color changes (cyanosis) during feeds, crying, agitation
  - PPHN that does not resolve as expected
  - That “gut” feeling
Pediatric Cardiology: Toddler and School Age
Management of Innocent Murmurs

- You will play a pivotal role in reassuring parents and allaying their anxiety while educating them about their child’s innocent murmur
Common Innocent Murmurs

- Still’s Murmur
- Cervical Venous Hum
- Pulmonary ejection murmur
Still’s Murmur

- Most common innocent murmur
- Quality similar to plucked guitar string (very musical)
- Best heard along LLSB while supine
- May diminish/disappear in sitting position
- Can become louder with exercise, excitement, and fever
- Most commonly ages 2 - 7 years
Cervical Venous Hum

- Only “continuous” innocent murmur
- Typically located on right, just below mid-clavicular region
- Loudest in sitting position
- Disappears in supine position
- May be obliterated by pressing lightly over jugular vein on affected side
- Usually heard between 3 & 6 years of age
<table>
<thead>
<tr>
<th>Type (timing)</th>
<th>Description</th>
<th>Age Group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Still’s (systolic)</td>
<td>Most common, btw LLSB &amp; apex, grade 2-3/6, musical, turbulence in LVOT</td>
<td>3-6 yo, occasionally in infancy</td>
</tr>
<tr>
<td>Pulmonary ejection murmur (systolic)</td>
<td>ULSB, grade 1-3/6, blowing in quality</td>
<td>8-14 yo</td>
</tr>
<tr>
<td>Peripheral pulmonary stenosis (systolic)</td>
<td>ULSB, radiates to L/R chest, axillae, back, grade 1-2/6</td>
<td>Premature/full-term newborns, usually disappears 3-6 mos of age</td>
</tr>
<tr>
<td>Venous hum (continuous)</td>
<td>Continuous, L/R supra/infra clavicular areas, grade 1-3/6, inaudible in supine position</td>
<td>3-6 yo</td>
</tr>
<tr>
<td>Carotid bruit (systolic)</td>
<td>Right supraclavicular area &amp; over carotids, grade 2-3/6</td>
<td>Any age</td>
</tr>
</tbody>
</table>
Atrial Septal Defect

- Can be single or multiple ASDs
- EKG may be abnormal
- Murmur of relative pulmonary stenosis
- Shunting occurs from left to right
- Widely Split S2 - varies little with respirations
- RV heave
- Closure is necessary if echo reveals right ventricular volume overload, FTT, and children beyond age of 5 years - small ASDs may close spontaneously
Atrial Septal Defects

From the Multimedia Library of Congenital Heart Disease, Boston Children’s Hospital, editor Robert Geggel, MD, www.childrenshospital.org/mml/cvp with permission
Radiological Findings

From the Multimedia Library of Congenital Heart Disease, Boston Children's Hospital, editor Robert Geggel, MD, www.childrenshospital.org/mml/cvp with permission
Ventricular Septal Defect

- 50% of all children with CHD have VSD
- High incidence of spontaneous closure with small, muscular VSDs
- May not be diagnosed until several weeks or months after birth
- The larger the shunt, the more quiet the murmur (low frequency and low intensity)
- VSDs never become “larger”
- Smaller VSDs have louder murmurs
Radiological Findings

Fig. 1 - Chest X-ray showing increased pulmonary arterial vasculature. When the right atrium and left ventricle are enlarged, a communication between chambers must be considered.
Kawasaki Disease

- Peaks in toddler years (80% affected <4 years of age)
- At least 3-4% have coronary artery aneurysm sequelae
- Can be asymptomatic due to collateral growth supplying ventricle
- May present with fatigue / SOB / diaphoresis
Mucocutaneous Lymph Node Syndrome (Kawasaki, 1967)

Fever persisting at least 5 days, no other diagnosis AND 4 of the 5 criteria:

1. Bilateral conjunctival injection
2. Erythema & cracking of lips, strawberry tongue, erythema of pharynx
3. Erythema & edema of hands and feet; later peeling
4. Polymorphous exanthem/rash
5. Cervical lymphadenopathy (> 1.5 cm.), usually unilateral
Laboratory Findings

- Leukocytosis with neutrophilia
- Elevated inflammatory markers: ESR/CRP
- Anemia (normochromic, normocytic)
- Sterile pyuria
- Elevated transaminases
- Hypoalbuminemia
Kawasaki Disease in the 21st Century

- Use of a new Algorithm for Assessment and Treatment of Incomplete Kawasaki Disease

- Pts with fever and < 4 clinical criteria
Management

- Intravenous Gamma Globulin
  - 2 g/kg over 8 - 12 hours

- Plus Aspirin
  - 80-100 mg/kg/day until afebrile,
  - then 3 - 5 mg/kg QD
Importance of Diagnosis

- Immediate IVIG administration reduces the risk of coronary artery abnormalities—serum immunoglobulin G levels are inversely related to development of CA abnormalities.

- Aspirin used in therapy—reduces thrombus formation—does not reduce coronary artery aneurysm.
Risk of Not Diagnosing

- Coronary Artery Aneurysms
Risk Factors for Coronary Artery Aneurysms

- Male gender
- Young (< 1 yr) or old (> 5 yrs.) age
- Persistent fever despite IVIG
- Labs at presentation
  - Low Hct or Hgb
  - Low platelet count
  - Low Albumin
  - High CRP
  - Higher absolute band count
Typical locations of aneurysms

Kitamura, et al, 1994
Take Home Pearls

- History of innocent murmur now with characteristics of pathologic heart disease
- History of repaired CHD with concerning symptoms
- Chronic respiratory conditions that don’t improve with treatment
- History of treated Kawasaki
  - Who doesn’t have regular follow up
  - Concerning clinical picture, abnormal EKG
Pediatric Cardiology: Adolescence

COMMON CARDIAC ABNORMALITIES AND THE PRE-PARTICIPATION SPORTS PHYSICAL
Sudden Cardiac Death (SCD)
Primary Objective: screen for health risks that may place participants at higher risk of injury, illness, and/or life-threatening conditions.

No cost-effective practical guidelines for screening that have proven effective in identifying potential candidates for sudden death.

Medical clearance for sports doesn’t imply absence of heart disease or complete protection of sudden death.
CDC estimates 2000 deaths annually in athletes less than 25 years of age

Leading causes of SCD:
- HCM
- Coronary anomalies
- Arrhythmias
- Myocarditis
## AHA’s 14-Point Screening Tool

<table>
<thead>
<tr>
<th>Personal History</th>
<th>Family History</th>
<th>Physical Exam</th>
</tr>
</thead>
<tbody>
<tr>
<td>2. Unexplained syncope/near-syncope</td>
<td>9. Disability from heart disease in close relative &lt; 50yo</td>
<td>12. Femoral arterial pulses (to exclude CoA)</td>
</tr>
<tr>
<td>3. Excessive &amp; unexplained dyspnea/fatigue or palpitations, assoc. with exercise</td>
<td>10. HCM or DCM, long-QT syndrome, or other ion channelopathies, Marfan syndrome, or clinically significant arrhythmias: specific knowledge or genetic cardiac conditions in family members</td>
<td>13. Physical stigmata of Marfan syndrome</td>
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<tr>
<td>5. Elevated SBP</td>
<td></td>
<td></td>
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<tr>
<td>6. Prior restriction from participation in sports</td>
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<tr>
<td>7. Prior testing for the heart, ordered by MD/NP</td>
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Pre-Participation Sports Physical

Family History

- Syncope
- Sudden unexplained death
- Arrhythmias
- History of heart disease
- Seizures
Pre-Participation Sports Physical

Present History

- Chest Pain
- Palpitations
- Pre-syncope/Syncope
- Shortness of breath
- Easy fatigability
Pre-Participation Sports Physical

Exam

- Murmur
- Marfanoid body features
- Weak femoral pulses
- Increased blood pressure
- Abnormal heart sounds
Hypertrophic Cardiomyopathy

- Abnormal thickening of the myocardium
- Predisposition to both left ventricular outflow tract obstruction and cardiac arrhythmia
- Autosomal dominant inheritance
- Exam: Systolic ejection murmur at RUSB which becomes quieter with squatting
- Can be progressive
Normal heart

Hypertrophic cardiomyopathy
Coronary Anomalies

- Abnormal origin or course of coronary arteries
- Normal physical exam
- History is often notable for presyncope, syncope, or exertional chest pain
- EKG can be normal, especially at rest
Anomalous Left Coronary Artery
Marfan Syndrome

- Connective tissue disease involving the protein fibrillin-1
- Autosomal dominant with high penetrance
- Phenotypic variation
- Progressive
- Involvement of skeletal, ocular, cardiovascular, skin, and pulmonary systems
Marfan Syndrome

Dilation of the ascending aorta and subsequent dissection
Marfan Syndrome

- Disproportionate (tall) stature
- Long limbs/digits
- Arm span > height
- Upper to lower segment ration < 0.85
- Hyperextensibility, flat feet
- Other skeletal abnormalities (pectus, chest asymmetry, scoliosis)
Marfan Syndrome

- Family history
- Myopia
- High arched palate, dental crowding
- Striae of skin
- Spontaneous pneumothorax
Aortic Stenosis

- Diseased aortic valve (often bicuspid valve) that obstructs left ventricular outflow
- Can be progressive, more common in males
- Exam findings can include:
  - thrill at suprasternal notch
  - systolic click
  - decreased peripheral pulses
  - murmur at RUSB that can become louder with squatting
Position of aortic valve in the heart

Normal aortic valve
- Open
- Closed

Aortic valve stenosis
- Open
- Closed
Take Home Pearls

- Chest pain, palpitations and/or syncope especially with exercise
- Dysmorphic features especially Marfanoid
- Family history of sudden death
- New onset murmur, especially at RUSB
- RUSB murmur that changes with position
- No clearance until cardiology clearance occurs
Antibiotic prophylaxis with dental procedures is reasonable for patients with cardiac conditions associated with the highest risk of adverse outcomes from endocarditis, including:

- Prosthetic cardiac valves, including transcatheter-implanted prostheses and homografts
- Prosthetic material used for cardiac valve repair, such as annuloplasty rings and chords
- Previous endocarditis
Current SBE Prophylaxis Recommendations

Congenital heart disease (CHD) only in the following categories:

- Unrepaired cyanotic CHD, including those with palliative shunts and conduits

- Completely repaired congenital heart defect with prosthetic material or device, whether placed by surgery or catheter intervention, during the first six months after the procedure

- Repaired CHD with residual shunts or valvular regurgitation at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization)

  Cardiac transplantation recipients with valve regurgitation due to a structurally abnormal valve
Case Study #1

- Six year old boy referred to cardiology for new murmur
  - Seen for first time by new provider one month prior
- ROS: Otherwise healthy, no syncope, no dyspnea, no chest pain, Mild intermittent asthma
- Negative Family history for CHD
Vital Signs/Physical Exam

- BP - RA: 136/74, LA: 139/56, RL: 92/43
- Pulse-85
- RR-24
- O2 sat 99%
- Decreased femoral pulses
- Perfusion normal
Well-functioning bicuspid aortic valve (inter-coronary commissure fusion), left aortic arch with normal branching pattern, and severe distal coarctation
As the length of the obstruction as well as presence/absence of collaterals was not well seen- underwent cardiac MRI

- Demonstrated a very tight but discrete coarctation and multiple collateral vessels bypassing the obstruction. Left ventricular size and function were normal
Surgical Repair

- End to end anastomosis repair
- Thoracotomy
Case Study #2

- 14 year old athlete, otherwise healthy
- Murmur heard by PNP on pre-sports physical exam
- Harsh murmur heard at RMSB 2/6
- Widely split, fixed S2
- ROS otherwise negative
History

- No dyspnea
- No syncope
- Great growth, nutritional status
- No daily medications
- No palpitations
- No chest pain
- No cyanosis
Family/Social History

- Negative family history
- Very active soccer player
- Would you refer?
Atrial Septal Defect
Case Study #3

- 16 year old presents for sports physical
- Problem list includes history of sharp chest pain diagnosed as costochondritis, now resolved
- VSS; no concerns or worrisome signs or symptoms
- On exam: normal S1, physiologic splitting of S2
- Murmur heard RUSB but it sounds musical to you....you aren't sure.....
- What should you do next?
Case Study #3

- In the squatting position, murmur is quieter
- EKG and CXR are unavailable to you
- Is there anything else that you can do or ask that may help lead you in the right direction?
Case Study #3

- Family history is unusual: several people under the age of 50 have died of ‘heart attacks’
- Clear this patient?
- Refer for evaluation? What next?
Subaortic Stenosis
6 week old infant presents to primary care family practice with chief complaint of fussiness and not feeding well and diaphoresis present during feeds
Case Study #4

- What other questions do you want to ask?
  - Feeding? “Was breastfeeding well, but lately when she is this fussy, she doesn’t eat”
  - Growth? Only in the 39th percentile for weight, but is growing steadily
  - Delivery history? Term baby, C-section for fetal intolerance to labor, required brief CPAP in DR, no NICU stay
  - Family history? Mother’s brother had some “hole” in his heart, but she couldn’t remember what it was called. She only knows he needed surgery to fix it.
Case Study #4

- Physical exam
  - Fussy, crying infant, mildly tachypneic but breath sounds clear and equal
  - Saturations in the mid 90’s, no differential from pre and post
  - Pale, mottled appearance, weak peripheral pulses, cap refill sluggish, non-edematous
  - Abdominal exam benign, precordium is quiet
  - CV exam is notable for a significant tachycardia, difficult to appreciate any murmurs
Due to her poor perfusion, irritability, and significant tachycardia, baby is sent to the ER with suspected ...
Case Study # 4

Supraventricular tachycardia (SVT)
Case Study #4

- Vagal maneuver is done (ice to the forehead) and her SVT is quickly resolved
- However, now a murmur is appreciated on exam that was not noted before
Case Study #4

- Murmur best heard along the left upper sternal border, has a soft, blowing quality
- You also note it under her left axilla
- Normal S1, S2
- RRR, normally active precordium
- She now appears better perfused, brisk capillary refill
- Pulses are 2+, equal in all extremities
  - Lungs clear and equal bilaterally, non labored breathing, still mildly tachypneic
Parents say no one has ever told them that their baby has a murmur

What kind of murmur is this?

**Peripheral Pulmonary Stenosis**
What Questions Do You Have?
References

- Pictures from the Multimedia Library of Congenital Heart Disease, Boston Children’s Hospital, editor Robert Geggel, MD, www.childrenshospital.org/mml/cvp with permission


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