Office Cardiology: *From Heart Murmurs to Chest Pain*

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Heart Murmurs... How Common?

“Studies have suggested that virtually all children demonstrate a heart murmur at some time during childhood”

What is a Heart Murmur?

- A heart murmur is a series of audible sounds whose duration is sufficiently long to exceed that which one could call a sound “Extra noise”

- Turbulence of blood flow through the heart structures

- Gradient (pressure difference)... Blood flows downhill
Before You Listen!

- Height & Weight
  - Failure to thrive
  - Falling off the normal curve
- Vital Signs
  - General appearance
  - Associated signs and symptoms
  - Cardiac examination
History... Heart Murmur

- When was the murmur detected?
- Was the patient sick at that time?
- Has the murmur changed over time?
- Do the parents have any concerns about the child’s general health?
Concerning Symptoms from History

- Cyanosis
- Chest Pain
- Dizziness/Syncope
- Failure to Thrive
- Recurrent respiratory infections, wheezing
- Disorders Associated with Heart Disease
- Family History of Congenital/Acquired Heart Disease
Concerning Signs from Exam

- Cyanosis/clubbing/edema
  - Dysmorphic features
  - Chest deformities
- Hyperactive precordium
- Palpable thrill or heave
- Increased cardiac impulses
- Diminished lower limb pulses (also if weak or bounding)
  - Skin lesions
  - Hypatosplenomegaly
  - Joints abnormalities
Don’t Forget the Pulses

- **Normal**: Equal full upper/lower extremity pulses with no delay
- **Coarctation**: Diminished and delayed femoral pulses
- **Aortic Stenosis**: Diminished pulses - decreased stroke volume
- **Bounding pulses**: Diastolic runoff as in moderate-severe AI, PDA or BT shunt
Tips for Better Auscultation!

- Keep the child calm (listen while feeding/asleep)
- Quiet room
- Distraction with a toy might help
- Don’t give up until they give up
- Stick to one stethoscope
- Be systematic (S1, S2, added sounds, murmurs)… *Listen to the murmur last*
- Note effect of positional changes on the murmur
- Learn maneuvers that affect the intensity of the murmur
S2... The Good, the Bad and the Ugly

- **Physiologically split S2.. GOOD**
  (Inspiration increases systemic venous return to RV → ↑ RV ejection time)

- **Wide & fixed split S2.. (delayed P2) Sort of BAD**
  - Atrial septal defect
  - Partial anomalous pulmonary venous return
  - Right bundle branch block

- **Reversed (paradoxically) split S2.. Just BAD**
  (Closes with inspiration, opens with expiration)
  - Left bundle branch block
  - Severe valvar aortic stenosis
Single S2... UGLY

Always ABNORMAL

- Severe pulmonary hypertension
- Severe pulmonary stenosis
  - Truncus arteriosus
  - Tetralogy of Fallot
- D-Transposition of the great arteries
  - Pulmonary atresia
- Single ventricle lesions (HLHS, HRHS)
Clicks

**Constant**
- Aortic valve stenosis

**Variable**
- (more in expiration)
  - Pulmonary valve stenosis

**Mid-late systolic**
- Mitral valve prolapse
Murmurs Locations & Radiation

AS (Valvular, supra/sub valvular)

VSD, AVC, Still’s, IHSS, TR

PS, ASD, Pulm. Flow, PPS, PDA, COA

Still’s, MR, MVP, AS, IHSS
Murmur Type
(In relation to S1)

- Systolic Ejection (possibly benign)
  - Crescendo-decrescendo (diamond) pattern
  - Ejection of blood across open semilunar valve
- Systolic Regurgitant (always pathologic)
  - Heard throughout systole (including isovolumetric period)
  - Blood flow across AV valve or VSD
- Continuous (sometimes benign)
  - Murmur continues through systole and diastole
- Diastolic Regurgitant (AI, PI)
- Diastolic Rumble (MS, TS)
Classification of Heart Murmurs

- Systolic Ejection
- Holosystolic (Regurgitant)
- Early diastolic
- Mid diastolic (Rumble)
- Continuous
Murmur Intensity- The Fun Part

- **Respiratory cycle**
  - Inspiration $\rightarrow$ increase blood flow to right heart
  - Expiration $\rightarrow$ increase blood flow to left heart

- **Supine $\rightarrow$↑ preload**
  - Exaggerates flow murmurs

- **Sitting $\rightarrow$↓ preload (↓ LV volume)**
  - Diminishes flow murmurs

- **Exercise & hyperdynamic circulation**
  (fever, anemia, pregnancy) $\rightarrow$↑ contractility
  - Exaggerates IHSS and flow murmurs
Murmur Intensity- The Fun Part

- **Standing** → ↓ afterload (↓ LV volume)
  - Increases intensity of IHSS & MVP (click earlier in systole)
- **Squatting & hand grip** → ↑ afterload & venous return → ↑ LV filling
  - Decrease intensity of IHSS murmur (↓ LVOT obst.)
  - Improve MVP (click & MR later in systole)
- **Valsalva maneuver** → ↓ preload (↓ LV filling)
  - Increases intensity of MVP and IHSS
  - Decreases intensity of innocent murmurs
Innocent Murmurs: Six Characteristics

1- Systolic ejection murmurs *(Never diastolic)*

{All innocent systolic murmurs are caused by blood flow in response to ventricular contraction and therefore must be mid-systolic in timing, as they cannot occupy isovolumetric contraction}

2- Grade 2 or less

3- No associated other pathologic sounds

4- No associated concerning symptoms/signs

5- Come and go

6- Localized with minimal radiation
What to Expect?

First Week of Life
- Ductal-Dependent Lesions
  - HLHS/HRHS
  - Critical aortic/pulmonic stenosis
  - Severe CoA/Interrupted aortic arch

At 6-8 weeks
- Shunt Lesions
  - “Pulmonary overcirculation”
  - VSD, ASD, AVSD, PDA
  - May be PPS

Preschool and School Age
- Mostly innocent
- ? ASD
- ? Acquired
Pulmonary Vascular Resistance Changes

- Pulmonary Arterial Mean Pressure (mm Hg)
- Pulmonary Blood Flow (ml/min/kg)
- Pulmonary Vascular Resistance (mm Hg/ml/min/kg)

Weeks: -7 to 7
BIRTH
When to Refer

“PATHOLOGIC MURMURS”

- **L → R Shunt lesions**
  - VSD, ASD, PDA
- **Obstructive lesions**
  - Valve stenosis
  - Outflow tract stenosis
  - Supravalvar stenosis
- **Regurgitant lesions**
When to Refer
“$A$ Concerning Systolic Murmur”

- Signs of CHF or cyanosis?
- Significant failure to thrive
- Associated syndrome/anomaly strongly associated with CHD
- ? Age < 1 month
- Loud (Grade III/VI or higher)
- Associated thrill
- Heard while standing
- Hyperactive precordium
- Single or widely fixed split S2
- Additional sound(s): click, gallop, rub
- Abnormal pulses, absent or weak femoral pulses
When to Refer

"A Concerning Murmur"

- A continuous murmur that persists in the supine position
- A regurgitant (holosystolic) murmur
- All diastolic murmurs
- New onset of a murmur (+/- stigmata of Rheumatic fever, Infective endocarditis)
- Louder murmur than before
- Unusual radiation
- Unclassified murmurs
- Associated abnormal CXR and ECG
Heart Songs: Test Your Skills

- Normal S2
- Still’s murmur
- Venous hum
- Mitral valve prolapse
Heart Songs: Test Your Skills

- Ventricular Septal Defect
- Atrial Septal Defect
- Mitral regurgitation
- Aortic regurgitation
- Aortic/pulmonary stenosis
- Patent ductus arteriosus
Take Home Message

- Innocent murmurs are common (~50%-85% of healthy children)
- Practice makes perfect
  "The more you listen, the better you get"
- Diagnosis can be reached by auscultation only without the need for any further tools
- Referring all children with murmurs for "echo first" is cost-ineffective in many centers
- **Always refer when in doubt**
Chest Pain in Children

- Very common referral complaint
  - Accounts for 650,000 physician visits/year in patients 10-21 years of age (0.25% - 0.29% of all physician visits)
- Requires some degree of body awareness
  - Starts in school age children, peaks in adolescence
  - Can be source of great child/parental anxiety
    - Chest pain equals heart pain
    - Sudden cardiac death stories
    - Improved heart attack awareness
- Usually non-cardiac in origin
- History and physical exam usually sufficient to exclude significant heart disease
Chest Pain in Children

- Very rarely life-threatening
- Male = Female
- Mean age of presentation 9 to 14 years
  - Children less than 12 more likely to have cardiorespiratory cause
  - Children over 12 likely to have psychogenic cause
- Chronic condition (~ 20% for > 3 years)
- Reassurance is all that is needed in most cases
Chest Pain in Children… Etiologies

- Idiopathic (12%-85%)
- Chest wall syndrome “Musculoskeletal” (15%-31%)
- Pulmonary (12% - 21%)
- Other (4% - 21%)
- Psychogenic (5% - 17%)
- Gastrointestinal (4% - 7%)
- Cardiac (4% - 6%)

Chest Pain... History Taking

- Location
- Quality
- Severity (scale of 1 to 10)
- Duration and frequency
- Radiation
- Relation to physical activity and body position
- Relation to respiration/food
- Aggravating/relieving factors
- Associating symptoms
- History of trauma to the chest/heavy backpack
Chest Pain- Cardiac or Not

**Cardiac**
- Pressure/crushing quality
- Substernal
- Worse with activity
- Stops with rest
- Radiates to neck, arms
- Associated nausea, diaphoresis, dyspnea, palpitations, dizziness
- No association with food, breathing

**Non-Cardiac**
- Sharp/burning quality
- Occurs anywhere
- No change with activity
- Lasts seconds to minutes
- No radiation or associated symptoms
- Reproducible on exam
- ? Worse with food
- Pleuritic component
Chest Pain- Evaluation

- Family History
  - Sudden death, cardiac disease
  - Long QT syndrome
  - Hypertrophic cardiomyopathy (CM), Dilated CM

- Social History
  - Alcohol, Tobacco, cocaine, drugs
  - Family disruption/school difficulties (psychogenic)

- Review of Symptoms
  - Fever, malaise, weight loss, fatigue

- Medical history- Sickle cell disease, asthma, shingles, etc.
Chest Pain- Evaluation

- Usually normal physical examination
- General appearance
- Vital Signs: Vital signs (hypertension) and Ht./Wt.
- Chest: deformity, **wall tenderness**, breath sounds, adventitious sounds
- Heart: PMI, S2, clicks, murmur, gallop, rubs, femoral pulses
- Abdomen: Hepatosplenomegaly
- Skin: rashes, lesions
Evaluation of Chest Pain

- Additional tests are rarely helpful
- Electrocardiogram
- Chest X-Ray if clinically indicated
- For concerning cardiac related CP:
  - Echocardiogram
    - HCM, origin of coronary arteries, ventricular size & function
  - Treadmill stress test
  - Cardiac enzymes
  - Angiography
  - Radionuclide study “Cardiolite stress test”
Evaluation of Chest Pain

- 191/235 patients had **EKG**
  - 16% were abnormal
    - All minor changes
    - Arrhythmias (3 pts, noted on PE)
    - Pericarditis (1) (SLE)
- 137/235 patients had **CXR**
  - 27% were abnormal
    - Pulmonary infiltrates
    - Atelectasis
    - Hyperinflation
    - Pneumothorax (1)
    - Pleural effusion (1)

Chest Wall Chest Pain

**Musculoskeletal CP**
- Very common
- Costochondritis
- Trauma or muscle overuse/strain
- Chest deformity
- Tietze syndrome
  - Inflammation of one costochondral junction (the area is tender, warm and swollen)
- Slipping rib syndrome
  - Usually related to 8th -10th ribs excess mobility
  - Positive “hooking maneuver”

**Precordial Catch Syndrome “Texidor’s Twinge”**
- Common in adolescents
- Brief (few seconds)
- Sharp in quality and may be pleuritic
- Located inferior to the left nipple/LLSB without radiation
- Unrelated to physical activities
- Accentuated by bending forward
- It forces the patient to breathe shallowly
- Self-limiting natural history
## Other Causes of Chest Pain

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<td>Eating disorder: Bulimia</td>
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Chest Pain- Cardiac Causes

- **Myocardial**- imbalance between myocardial demand and CO
  - Myocarditis, hypertrophic/dilated cardiomyopathy
- **Valvular**- supply-demand mismatch
  - Severe aortic stenosis, pulmonary stenosis
  - ? MVP
- **Pericardial**- pericarditis
- **Great vessels “Aorta”**- dissection of the aorta
- **Coronary arteries abnormalities**
  - LAD from RCA, RCA from left sinus, ALCAPA, coronary fistulae
  - Kawasaki disease (20% to 25%)
  - Coronary vasospasm (cocaine abuse)
- **Rhythm disturbances**- may cause coronary ischemia due to diminished ventricular diastolic filling and low cardiac output
Myocarditis

An inflammation of the myocardium associated with myocardial necrosis

- Microbial etiologies
  - Viral: Enteroviruses (Coxsackie Group B, Echovirus, Poliovirus, Adenovirus, Mumps, EBV, CMV, HIV)
  - Others: Mycoplasma, bacterial, rickettsial, fungal or parasitic organism.
- Other causes: Autoimmune
  - ARF, Kawasaki’s Disease, JRA, and SLE
Dilated (Congestive) Cardiomyopathy

CP is related to diminished stroke volume leading to impaired coronary perfusion

- Mostly idiopathic
- Genetic: Carnitine deficiency, Freidriech’s ataxia
- Ischemic: ALCAPA, KD, DM, Cocaine
- Toxins: Anthracyclines, alcohol, Methamphetamine
- Infections: Myocarditis (viral), HIV, Severe sepsis
- Nutritional: Severe iron def., Thiamine def
- Arrhythmias: SVT, VT, CAVB
Hypertrophic Cardiomyopathy
Hypertrophic Cardiomyopathy (IHSS, HOCM)

- Mechanisms of chest pain (angina):
  - Imbalance between myocardial demand and cardiac output
  - Marked increase in myocardial O2 demand exceeds coronary flow during exercise
  - LVOT obstruction exacerbates the imbalance, leading to increased myocardial work and myocardial O2 consumption
  - Coronary artery compression by myocardial bridging
- Very variable presentation ranging from chest pain, exercise intolerance to sudden death
- May have positive family history
- Echocardiogram is diagnostic
Pericarditis

Acute inflammation of the pericardium

- Idiopathic, presumed viral
- Purulent (bacterial)
- Autoimmune (JRA, SLE)
- Uremia
- Neoplastic disease
- Post pericardiotomy syndrome
Pericarditis

- Severe pain, worse while supine
- Pain improves when leaning forward
- Constitutional symptoms (fever, etc.)
- PE: Distant heart sounds and pericardial friction rub
- Labs: Abnormal ECG and cardiomegaly on CXR
Congenital Coronary Arteries Anomalies

RCA from left sinus

Normal

LCA from right sinus
Coronary from Pulmonary Artery

Anomalous Left Coronary Artery from Pulmonary Artery

“ALCAPA”
ALCAPA

- **Symptoms**: irritability, especially while feeding
- **Signs**: CHF
- **EKG findings**: Q waves in I, aVL, V4-6 (anterolateral MI)
- **Pathophysiology**:
  - After birth, the LV is perfused by the desaturated PA blood.
  - Ischemia occurs during exertion (crying, feeding), which further increases myocardial oxygen demand resulting in LV free wall (anterolateral) MI.
  - Dilation of the ventricle occurs due to chronic hypoxia
- **Treatment**: Surgery
Acute lateral infarction. ECG 1 hr after the onset of chest pain. Typical changes in the lateral chest leads (V4–V6) and the high lateral limb leads (I and aVL). Note the reciprocal changes in leads II, III and AVF. This pattern usually reflects occlusion of the circumflex coronary artery or, perhaps, a large diagonal branch of the left anterior descending coronary artery.
Myocardial Infarction.
EKG Changes & Cardiac Enzymes

Fig 1. This electrocardiogram (ECG) demonstrates the diagnostic criteria for childhood myocardial infarction (MI). Note the wide Q wave (>35 msec), ST-segment elevation (>2 mm), and prolonged QT interval.

(Troponin I level. >2 ng/ml abnormal, >8 ng/ml poor prognosis)

Myocardial Infarction in Healthy Adolescents with Normal Coronary Anatomy*

- Nine patients presented to ED over a period of 11 years (June 1995 to May 2006). {8 boys; age range: 12-20 yrs; mean 15.5 yrs} and 1 female met the criteria of acute myocardial ischemia and infarction:
  - Typical anginal CP
  - ST elevation or non-specific ST-T wave changes on EKG
  - Abnormal cardiac wall motion by echo or angiographic imaging
  - Elevated cardiac enzymes (CK, CK-MB and Troponin I)

*John R. Lane and Giora Ben-Shachar: Pediatrics 2007;120;e938-e943
Cocaine Induced Coronary Vasospasm

- May cause angina, myocardial infarction, arrhythmias, or sudden death
- Mechanism:
  - It blocks the reuptake of norepinephrine in the CNS and peripheral sympathetic nerves → increased circulating catecholamines → coronary vasoconstriction
  - It induces the activation of platelets → increased endothelin production and decreased nitric oxide production
Kawasaki Disease

**KD- Diagnostic Criteria**

**Fever for 5+ days and 4 of the 5 criteria:**

1. Bilateral non-purulent limbic sparing conjunctival injection
2. Mucous membrane changes: Injected pharynx, fissured lips, strawberry tongue
3. Extremity changes: Edema, induration, erythema, desquamation
4. Polymorphous rash
5. Cervical adenopathy (>1.5 cm), unilateral
Kawasaki Disease
Acute Rheumatic Fever - Revised Jones Criteria

MAJOR
Carditis

MINOR
Arthralgia
Dissecting Aortic Aneurysm

- Associated with connective tissue disorders, e.g. Marfan syndrome
- Children with Turner’s, Noonan’s and Williams syndromes are also at risk
- Severe chest pain radiates to the back
- Diagnostic tools- Stat Echo, TEE, chest CT scan
Thumb sign

Ectopia lentis
Aortic Root Aneurysm
Mitral Valve Prolapse (MVP)

- Vague and controversial
  - No direct link between MVP and CP, dizziness, syncope, rhythm disturbances, panic attacks
- Prevalence: 4-21% (overdiagnosed)
- Echocardiogram is diagnostic
- Often idiopathic with/without regurgitation
- May be associated with connective tissue disorders
- Physical examination:
  - Marfanoid body habitus
  - Mid-systolic click and murmur change with standing and Valsalva maneuver
Chest Pain- Management

- Reassurance

- Treat underlying cardiac cause if present
Screening Echo

- May be helpful (R/O HCM, PS, AS)
- Does not exclude all causes of CP
  - Technical difficulties
- May give false sense of security
Chest Pain - When to Worry?

- Chest pain on exertion
- Associated cardiac symptoms - presyncope, syncope, dyspnea, or palpitations
- Family history of sudden death
- Abnormal physical findings
- Family anxiety level (persistent CP)
Presyncope/Syncope
Syncope

- A sudden transient loss of consciousness and postural tone as a result of decreased cerebral perfusion
- Common in children and adolescents
- About 15% of children have a syncopal episode between the ages of 8-18 years

Causes:
- Autonomic (Neurocardiogenic)
- Non-cardiac
- Cardiac
Syncope... Differential Diagnosis

- **Neurocardiogenic/vasovagal syncope** "Simple faint":
  - Due to loss of resistance in peripheral circulation
  - Provoked by emotional factors
  - Most common form of syncope (~80%)
  - Positive family history (33%)

- **Orthostatic**:
  - Excessive and prolonged fall in BP and HR with sudden positional change
- **Cardiac**: Due to loss of cardiac output from cardiac disease
- **Respiratory**: Cough, hyperventilation, or breath holding
- **Neurologic**: Seizure
- **Metabolic**: Anemia and hypoglycemia
Syncope: Proposed Mechanisms

- Decrease or loss of cerebral circulation
- Cardiac problem with diminished cardiac output
- Decreased systemic BP inadequate to perfuse the brain
- Inadequate supply of energy substrates to supply the brain (glucose, O₂)
“The causes of syncope are multifactorial, and the possibility that syncope may be a premonitory sign of sudden death has resulted in extensive evaluations which are not usually cost effective.”

Syncope... When to Worry?

- Exercise-induced syncope
- Associated symptoms: palpitations/CP
- Atypical heart murmur on examination
- Abnormal EKG
- Recurrent episodes
- Younger children (< 6 years of age)
- Family history of:
  - Sudden death < 40 years
  - QT prolongation
  - Cardiomyopathy
Cardiac Causes of Syncope

- **Obstructed Outflows**
  - Significant aortic Stenosis
  - Hypertrophic Cardiomyopathy
  - Severe pulmonic stenosis

- **Myocardial Dysfunction**
  - Dilated cardiomyopathy
  - Coronary Artery anomalies

- **Others:**
  - Pulmonary hypertension
  - Tetralogy spell

- **Arrhythmias**
  - Ventricular Tachycardia
    - Brugada syndrome
    - Arrhythmogenic RV dysplasia
  - Supraventricular Tachycardia
    - Wolff- Parkinson- White Syndrome (WPW)
  - Long QT Syndrome
  - High-grade AV Block
  - Atrial fibrillation/flutter
Diagnostic Tools
Diagnostic Tools
Reveal implanted.

Fainting episode occurs.

Patient uses Activator after waking.

Heart rhythm related cause ruled in or out. Next steps defined.

Physician analyzes data from Reveal.
Narrow QRS Complex tachycardia

Wide QRS complex tachycardia
Wolff-Parkinson-White Syndrome
Bradyarrhythmias
Bradyarrhythmias
Third Degree Heart Block
Normal Values of QTc:
Infants up to 6 months old <0.45 seconds, Children <0.44 seconds, Adolescents and Adults <0.42 seconds
LQTS with Torsades de Pointes

![ECG Tracings]

Diagram showing ECG tracings labeled as follows:

- I: aVR
- II: aVL
- III: aVF
- V1
- V2
- V3
- V4
- V5
- V6

Tracings labeled as:

A

B
Syncope During Exercise is a Grave Sign