CHEST PAIN
IN THE PEDIATRIC PATIENT

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LINEE DI COMPORTAMENTO IN CARDIOLOGIA PEDIATRICA
Roma, 22 Ottobre 2016
Prevalence

- Chest pain is a common complaint in the office and emergency room
- Often can be a chronic complaint
- Male:Female ratio 1:1
- Prospective studies of chest pain in children have shown that pain is not as ominous as in adults
- Pain has cardiac cause in only 1-5% of cases (cardiac cause is least likely)
Causes of chest pain

- Idiopathic 21-45%
- Musculoskeletal 15-31%
  - Trauma
  - Costochondritis
  - Precordial catch
  - Slipping rib
- Respiratory 2-11%
  - Asthma
  - Pneumonia/effusion
  - Pneumothorax
  - Pleurisy
  - Pulmonary embolus
  - Malignancy
- Gastroesophageal 2-8%
  - Reflux
  - Foreign body
- Psychogenic 5-20%
- Miscellaneous < 10%
  - Skin infection
  - Breast disease
- Cardiac 1-6%

Friedman KG, Alexander ME. Chest pain and syncope in children: a practical approach to the diagnosis of cardiac disease.

J Pediatr 2013; 163:896-901
Musculoskeletal

- COSTOCHONDRITE
- MUSCLE STRAIN / OVERUSE
- PRECORDIAL CATCH
- BREAST MASSES
- SLIPPING RIB SYNDROME
- TRAUMA
Musculoskeletal

**COSTOCHONDRITIS**

- Tietze Syndrome
- Most common musculoskeletal cause
- All ages, more common in girls
- Inflammation of cartilage at costochondral margin

- **Cause**
  - Overuse
  - Preceding URTI with cough
  - Idiopathic

- Tenderness at junction of ribs and sternum, aggravated by deep breathing
- Treatment with NSAIDS, rest, reassurance
Musculoskeletal

- **SLIPPING RIB SYNDROME**
  - Displacement of the tips of the inferior ribs
  - Aggravated by deep breathing
  - Pain in one of the upper abdominal quadrants, epigastric area or inferior costal margins
  - Perception of slipping movement of ribs
  - Hooking Maneuver
Musculoskeletal

«SLIPPING RIB SYNDROME»

The Thoracic Cage: Anterior view

- True ribs (1–7)
  - Note that ribs run anteriorinferiorly
  - Note how Rib 1 is flattened

- False ribs (8–12)

- Floating ribs (11, 12)

- Manubrium
- Sternal angle-fibrocartilage joint
- Body
- Xiphisternal joint
- Xiphoid process
- Jugular notch
- Clavicular notch
- Note that the 2 Costal Margins form the Infrasternal Angle
- Intercostal spaces
- Costal cartilage
- Costal margin
Precordial Catch Syndrome

- Also known as “Texidor’s twinge”
- Brief, sharp, shooting pain
- Occurs at rest
- Localized at LSB or cardiac apex
- Sudden onset with deep breath
- Pain subsides with shallow breathing after hesitation
- No associated symptoms
- No physical findings
- Benign condition
Musculoskeletal

...take home message...

- No fever, no associated symptoms
- Characteristically sharp, fleeting, non exertional, positional
- Exacerbated by deep inspiration
- May be reproducible on palpation
Chest pain of respiratory etiology is more common under the age of 12.

- COUGH
- PNEUMONIA
- ASTHMA (AS HIGH AS 73%)
- PLEURAL EFFUSION
- PNEUMOTHORAX
- PULMONARY EMBOLISM
- FOREIGN BODY ASPIRATION
Respiratory

➢ ASTHMA

• May account for 10-20% chest pain in kids
• Personal or family history atopic conditions
• Associated with cough
• May be worse at night or with exercise
• Wheezing not always detectable
• Trial of bronchodilator
• Consider PFT for pain with exercise
Respiratory

- PNEUMOTHORAX/PNEUMOMEDIASTINUM
  - Children at risk
    - Asthma, bronchiolitis
    - Barotrauma
    - Cough, choking, vomiting
    - Crack, cannabis
    - Cystic fibrosis
    - Marfan syndrome
    - Tall male teenagers
Pulmonary embolism

- RISK FACTORS FOR PULMONARY EMBOLUS

- Immobility or recent surgery
- Neoplasm
- Hypercoagulability
- Central venous catheter
- Pleuritic pain
- Haemoptysis
- Hypoxia
- Consider chest radiogram, ECG, V/Q scan
Respiratory
...take home message...

- Associated with cough, shortness of breath, wheezing
- Pain elicited by respiration
- Associated with fever
Evangelista et al (2000) reported a prevalence of GI causes of chest pain as high as 8%

- **ESOPHAGITIS**
- **GERD**
- **ESOPHAGEAL FOREIGN BODY**
- **CAUSTIC INGESTION**

Gastrointestinal

- GASTROESOPHAGEAL REFLUX

  - Accounts for 5-10% of PED chest pain visits
  - Classic pain is temporally associated with meals
  - Burning, retrosternal
  - Trial of antacid, H2RA, PPI is appropriate
  - Consider pH probe if diagnostic testing needed
Gastrointestinal

- ESOPHAGEAL FOREIGN BODY
Gastrointestinal
...take home message...

- Burning, retrosternal
- Related to meals
- Associated with vomiting, dysphagia
- Loss of weight
Idiopathic / Psychogenic

- Most common cause in most studies of adolescents (21-30%)
- Idiopathic: no organic, psychogenic cause
- Psychogenic: pain of emotional origin
- Primarily affects girls
- Recent or current stressful situation
- Family illness, especially cardiovascular
- Family history of chest pain
- Other somatic and sleep complaints
- Depression
Miscellaneous

- THORACIC TUMOR
- SICKLE CELL DISEASE
- HERPES ZOSTER
- SCOLIOSIS
- BREAST DISEASE
- ANOREXIA NERVOSA
- HYPERVENTILATION SYNDROME
### Prevalence of cardiac chest pain

<table>
<thead>
<tr>
<th>Study</th>
<th>Location</th>
<th>Total patients</th>
<th>Patients with cardiac chest pain, n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Driscoll et al (1976)</td>
<td>Ped</td>
<td>43</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Pantell and Goodman</td>
<td>PC</td>
<td>100</td>
<td>1 (1)</td>
</tr>
<tr>
<td>Gastesi et al (2003)</td>
<td>Ped ED</td>
<td>161</td>
<td>1 (0.5)</td>
</tr>
<tr>
<td>Danduran et al (2008)</td>
<td>PC</td>
<td>263</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Friedman et al (2011)</td>
<td>PC</td>
<td>406</td>
<td>5 (1.2)</td>
</tr>
<tr>
<td>Saleeb et al (2011)</td>
<td>PC</td>
<td>3700</td>
<td>37 (1)</td>
</tr>
</tbody>
</table>

*PC, pediatric cardiology office; Ped, pediatrician’s office; Ped ED, pediatric ED.*

Prevalence of cardiac chest pain

- Records of children over 6 years old with chest pain seen at CHB from 2000 to 2009 were reviewed for demographic features, clinical characteristics, resource utilization, and presumed diagnosis.

- 3,700 children (7 to 22 yo, m 13 yo) with chest pain (33% exertional) were evaluated.

- 37 (1%) had heart issue, 0 cardiac deaths in nearly 18,000 patient years of follow-up.

Prevalence of cardiac chest pain

- Unknown: 52%
- Musculoskeletal: 36%
- Pulmonary: 7%
- Gastrointestinal: 3%
- Anxiety: 5-20%
- Cardiac: 1%

Cardiac causes of chest pain

- **MYOCARDIAL ISCHEMIA**
  - Congenital coronary abnormalities
  - Acquired coronary abnormalities (KD)
  - Hypercoagulable state
  - Cocaine

- **LEFT VENTRICULAR OUTFLOW OBSTRUCTION**
  - Valvular, subvalvular, supravalvular aortic stenosis
  - Coarctation of the aorta
  - Hypertrophic cardiomyopathy

- **MITRAL VALVE PROLAPSE**
- **CONNECTIVE TISSUE DISORDERS**
- **PERICARDIAL EFFUSION/PERICARDITIS**
- **MYOCARDITIS**
- **ARRHYTHMIAS**
- **PULMONARY HYPERTENSION**
- **CARDIAC DEVICE OR STENT COMPLICATION**
Myocardial ischemia

- **Congenital coronary abnormalities**
  - Prevalence 2:1000
  - Anomalous origin of left coronary from pulmonary artery
    - Presents in first months of life
    - Irritability, heart failure, cardiac enlargement
  - Anomalous origin from incorrect sinus of Valsalva
    - Presents later in childhood
    - Compression between aorta and pulmonary artery
  - Hypoplastic coronary arteries
Myocardial ischemia

- Congenital coronary abnormalities
  - Anomalous origin from incorrect sinus of Valsalva
    - Presents later in childhood
    - Compression between aorta and pulmonary artery
    - Sudden death
Acquired coronary abnormalities: Kawasaki disease

- Acute febrile vasculitis of childhood
- Features
  - Fever (> 39 degrees for 5 days)
  - Non-exudative conjunctivitis
  - Erythema of oral mucosa and tongue
  - Erythema and swelling of hands and feet
  - Cervical adenitis >1.5 cm
  - Rash
- Leading cause of acquired heart disease in kids
Kawasaki disease with coronary artery sequelae

- Acute and subacute
  - Myocarditis (50% of patients)
  - Pericarditis
  - Mitral, aortic insufficiency
  - Arrhythmias
  - Coronary aneurysms
    - 20-25% if untreated
    - 5% if treated with IVIG
    - Appear 7 days to 4 weeks after onset of fever

**Myocardial ischemia**

- **Kawasaki disease with coronary artery sequelae**

- Coronary aneurysms
- Coronary stenosis
- Thrombosis
- Myocardial ischemia
- Myocardial infarction
Myocardial ischemia

- **EKG**

- ST segment elevation in leads II, III, aVF, V6
- ST depression in leads aVL, V1-V4
Myocardial ischemia

- Hypercoagulable state
  - Sickle cell disease
    - Myocardial infarction uncommon but described
    - Perfusion defects in 5% children studied in a Paris sickle cell clinic (*Arch Dis Child* 2004;89:359-62)
    - Microvascular occlusion of small vessels
    - Exchange transfusion may be helpful for acute ischemia (*Pediatrics* 2003;111:e183-7)
  - Nephrotic syndrome
    - Thrombotic occlusion of coronary arteries

- Cardiac transplant

- Cocaine abuse
Aortic stenosis

- Valvar AS is a common congenital defect, though it is rare in infancy
- Tends to be progressive, associated with bicuspid aortic valve
- Can have chest pain with exercise
- Systolic ejection click may precede systolic murmur best heard over the aortic valve region
- May have thrill (check suprasternal notch)
- Can see hypertrophy or strain patterns on EKG
LV outflow obstruction

- EKG in aortic stenosis

*Left ventricular hypertrophy with strain pattern*
LV outflow obstruction

- Hypertrophic cardiomyopathy
  - Autosomal dominant
  - Symptoms in 2nd decade
  - May present with angina-like pain or syncope
    - impaired coronary perfusion
    - increased O2 demand
  - Risk of sudden death 6% in children
Hypertrophic cardiomyopathy

Diagnosis

- Systolic ejection murmur
  - Increases with decreased LV volume (Valsalva, squatting, standing)
- Normal or increased heart size on CXR
- ECG with LVH, LAD, conduction abnormalities
- Echocardiography diagnostic
LV outflow obstruction

- Hypertrophic cardiomyopathy

*Left ventricular hypertrophy with strain pattern*

*ST and T wave abnormalities*
Anginal pain
...take home message...

- Chest pain is located on the precordial or substernal area
- Chest pain radiating to the neck, jaw, arms, back or abdomen
- Crushing chest pain, deep heavy pressure
- Feeling of choking or squeezing sensation
- Pain at high level of exertion
- History of drug use, KD, cardiac surgery, hypercoagulable state
Arrhythmias

- **Supraventricular Tachycardia (SVT)**
  - Most common arrhythmia in children
  - In children >1 year
    - 82% present with palpitations (rate “too fast to count”)
    - 14% with pain
    - 14% perspiration
    - 14% dizzy
    - 4% pallor
  - 1-3% of chest pain complaints in ED
  - 6% of chest pain referred to cardiologist
  - Median time from symptoms to diagnosis 138d
  - Can sometimes convert to sinus rhythm with vagal stimulation
Arrhythmias

- Supraventricular Tachycardia (SVT)
Ventricular Tachycardia (VT)

- Wide complex tachycardia
- Sensation of palpitations
- 120-200 beats per minute
- Very rare
- Patients can have chest pain and syncope
- Causes: viral myocarditis, Long QT syndrome
Arrhythmias

- Ventricular Tachycardia (VT)
Arrhythmias
...take home message...

Mostly associated to
- Palpitations
- Irregular or rapid heart beat
- Syncope
- Exertion intolerance
Acquired cardiac conditions of chest pain

- PERICARDITIS

- Infectious etiology common in children
- Pain
  - More common in older children and adolescents
  - Worse when supine, relieved by leaning forward
- Physical findings
  - Friction rub if effusion small
  - Muffled heart sounds, pulsus paradoxus if large
Acquired cardiac conditions of chest pain

➢ PERICARDITIS

ST elevation, usually leads I, II, aVF, V4-V6
Pericarditis
...take home message...

- Acute onset symptoms in the setting of fever
- Chest pain associated with friction rub
- Increased pain while supine
Acquired cardiac conditions of chest pain

- **MYOCARDITIS**
  - Usually viral etiology
    - Enterovirus (coxsackie B), adenovirus
  - Presentation
    - Heart failure
    - Chest pain
      - More likely in older kids and adults
      - Ischemia or concurrent pericarditis
Acquired cardiac conditions of chest pain

- **MYOCARDITIS**
  - Physical findings
    - Tachycardia, tachypnea
    - Poor perfusion
    - Muffled heart sounds, S3, murmur
    - Hepatomegaly
  - CXR
    - Cardiomegaly
    - Pulmonary edema
Acquired cardiac conditions of chest pain

- **MYOCARDITIS**
  - ECG
    - Sinus tachycardia
    - Decreased voltages (<5 mm) limb leads
    - LVH
    - Prolonged PR interval, prolonged QT interval
  - Echocardiogram
    - Hypokinesis, impaired function
Acquired cardiac conditions of chest pain

- Sinus tachycardia
- Decreased voltages
- Diffuse ST and T abnormalities

MYOCARDITIS
Myocarditis
...take home message...

- Acute onset symptoms
- Associated with pathologic murmur
- Significant tachycardia, tachypnea, gallop
- Abnormal vital signs, ill appearance (low cardiac output)
Connective tissue disorders

- RISK OF AORTIC DISSECTION

- Children at risk
  - Marfan syndrome
  - Ehlers-Danlos
  - Turner syndrome
  - Coarctation
  - Aortic stenosis
  - Endocarditis
  - Cocaine use
Connective tissue disorders

- **MARFAN SYNDROME**

  - Caused by fibrillin gene mutation
  - Manifestations
    - Musculoskeletal: tall, long limbs and fingers, pectus
    - Ocular: lens dislocation
    - Cardiovascular: aortic root dilation, MVP
    - Pulmonary: spontaneous pneumothorax
Differential diagnosis
A thorough history is critical
History

- Description of the pain
  - Not as reliable in children as in adults
- Precipitating factors
  - Exertion
  - Eating
  - Deep breathing
  - Muscle use
  - Trauma
  - Emotional stress
History

- Frequency and chronicity
- Associated symptoms
  - Fever
  - Cough
  - Shortness of breath
  - Syncope
  - Dizziness
  - Palpitations
History

- Past medical history
  - Known heart disease
  - Asthma or atopic conditions
  - Prothrombotic conditions
    - Cancer
    - SLE
    - Nephrotic syndrome
  - Medications and drugs

- Family history
When to consider cardiac causes based on history

- Pt/FHx of seizures / syncope / sudden death
- Pt/FHx of congenital/acquired heart disease
- Pain with palpitations
- Pain with exertion, pain with dizziness
- Patient complaint of crushing chest pain
- History of prolonged fever
- History of abnormal EKG / CXR
The Physical Exam
Physical Exam

• Observation
  • Level of distress / anxiety
  • Breathing pattern / diaphoresis
  • Bruising / trauma / scarring of the chest
  • Posture / gait
  • Chest asymmetry
  • Syndromic appearance
Physical Exam

• Palpation
  • Tenderness at costochondral junction, xiphoid process, breasts, ribs
  • RV heave / thrill both over the PMI and at the suprasternal notch
  • Subcutaneous emphysema
  • Hooking maneuver
  • Abdominal exam
  • Pulses
Physical Exam

• Auscultation

  • Murmur, friction rub, extra heart sounds
  • Listen to heart in standing and squatting position, particularly over the aortic valve region
  • Tachycardia, Arrhythmias
  • Breath sounds (rales, wheeze, asymmetrical breath sounds)
What testing is used in the diagnosis of chest pain?
Red flags

- Chest pain/discomfort/tightness/pressure related to exertion
- Pain associated with palpitations, syncope or near-syncope
- Shortness of breath, acute distress
- Abnormal vital signs or physical findings
- Pain limits daily activities or disturbs sleep
- PMH consistent with Kawasaki disease
- Family history of sudden cardiac death, CMP, familial arrhythmias
- Physical stigmata of Marfan syndrome
- Substance abuse
- Presence of prothrombotic conditions
Cardiac Diagnostic Studies

- EKG
- Chest X-ray
- Echocardiogram
- Troponin testing
- 24 Holter Monitor
- Event Monitor
- Exercise Stress Test
- Cardiac MRI
# Cardiac Diagnostic Studies

## Table III. Utility and indications for cardiac testing in pediatric chest pain and syncope

<table>
<thead>
<tr>
<th>Symptom/test</th>
<th>Indication for test</th>
<th>Usefulness</th>
<th>Conditions diagnosed</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chest pain</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ECG</td>
<td>Abnormal physical exam, ECG, exertional chest pain, or palpitations</td>
<td>Selected patients</td>
<td>Cardiomyopathy, myocarditis, pericarditis, with or without pulmonary HTN</td>
</tr>
<tr>
<td>Echocardiography</td>
<td>Abnormal physical exam, ECG, family history, or exertional chest pain</td>
<td>Selected patients</td>
<td>Anomalous coronary artery origins, cardiomyopathy, myocarditis, pericarditis, pulmonary HTN, left ventricular outflow obstruction</td>
</tr>
<tr>
<td>Troponin testing</td>
<td>Suspected myocarditis or pericarditis</td>
<td>Selected patients</td>
<td>Myocarditis, pericarditis, coronary ischemia</td>
</tr>
<tr>
<td>Ambulatory ECG</td>
<td>Chest pain and palpitations</td>
<td>Rarely useful for chest pain</td>
<td>Atrial or ventricular arrhythmia, intermittent heart block</td>
</tr>
<tr>
<td>EST</td>
<td>Exertional chest pain and exertional syncope or palpitations</td>
<td>Rarely useful</td>
<td>Coronary ischemia, exercise-induced asthma (if spirometry included)</td>
</tr>
<tr>
<td>Syncope</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ECG</td>
<td>All patients with syncope</td>
<td>First-line diagnostic test</td>
<td>Long QT interval, short QT interval, Wolff-Parkinson-White syndrome, cardiomyopathy, pulmonary HTN, Brugada, heart block, with or without ARVD</td>
</tr>
<tr>
<td>Echocardiography</td>
<td>Abnormal physical exam, ECG, family history, or alarming event history</td>
<td>Selected patients</td>
<td>Cardiomyopathy, pulmonary HTN, with or without ARVD, coronary anomalies</td>
</tr>
<tr>
<td>Exercise ECG/EST</td>
<td>Exertional syncope or exertional symptoms</td>
<td>Selected patients</td>
<td>Catecholaminergic polymorphic VT, long QT interval</td>
</tr>
<tr>
<td>Ambulatory ECG</td>
<td>Syncope plus palpitations, frequent symptoms, exertional syncope</td>
<td>Selected patients</td>
<td>Atrial or ventricular arrhythmia, intermittent heart block, reflex bradycardia</td>
</tr>
<tr>
<td>Tilt-table test</td>
<td>Very frequent, recurrent syncope or atypical syncope</td>
<td>Rarely useful</td>
<td>Confirmation of NCS (low specificity), postural orthostatic tachycardia syndrome</td>
</tr>
</tbody>
</table>

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Risk factors for cardiac disease?
First episode of pain
Pain radiating to arm or back
Associated dizziness or collapse
History of cardiac, clotting, connective tissue or Kawasaki's disease
Long standing diabetes mellitus
Cocaine or other stimulant use
Abnormal pulse or blood pressure

Risk factors for pulmonary embolus?
Immobility or recent surgery
Neoplasm
Hypercoagulability
Central venous catheter
Pneumonia
Hemoptysis
Hypoxia

ECG, CXR
If high risk, consider troponin +/- echo

Is there still a possibility of cardiac disease?
Yes
Discuss with Cardiology

Yes
Consider PE
ECG
V/Q scan
Discuss with Respiratory unit

CXR clear or minor abnormality?
Yes

Respiratory symptoms or signs?
Yes
CXR
Abnormal CXR
Yes
Pneumothorax
Pneumonia
Foreign body

No

Exercise induced asthma
Radiolucent foreign body
Hyperventilation

No

Pain related to eating?
Abdominal tenderness?
Yes
Consider gastrointestinal cause

No

Risk factors for serious psychiatric disease?
Lowered affect or lack of motivation
Hypervigilance
Hyperventilation
Social withdrawal
Impairment of function at school
Drug and alcohol use

Yes
Consider serious underlying psychological cause

No

Pain reproducible with movement or palpation?
Yes
Consider musculoskeletal cause

No

Psychosomatic or undifferentiated chest pain?
Yes
Reassure and then refer to LMD, general pediatrician, adolescent medicine

No
Summary

- Chest pain in pediatrics usually due to benign, identifiable etiology

- Cardiac and other life-threatening causes of chest pain rare but do exist
  - Often can be ruled out by history and physical exam
  - Diagnostic tests appropriate in presence of red flags
Thanks!