Chest pain in children and adolescents

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Cardiovascular disease is the most common contributor to morbidity and mortality in North America. It is not surprising that when a person experiences chest pain, one’s first thought is the heart. Indeed, in the adult population, unexplained chest pain requires exploring cardiac causes early in the medical evaluation. Conventional wisdom, however, suggests that chest pain in the pediatric age group would unlikely be contributed to a cardiac etiology. Despite this “common sense,” chest pain in children provokes considerable anxiety for patients and parents. Commonly in clinics, phrases like “I don’t want him/her to be the one who dies on the playing field” can be heard. Rare but well publicized incidences of an adolescent experiencing sudden death has fueled these fears. It is fortunate that sudden death in the pediatric age group in general, and specifically, sudden cardiac death, is rare \cite{1,2}.

This article explores chest pain in the pediatric population. The common causes of pediatric chest pain and the approach to its evaluation are reviewed. Special attention is given to the cardiac causes of pediatric chest pain and how it potentially can be differentiated from more common benign etiologies. The aim is to reassure the reader of his or her clinical instincts so that this anxiety-provoking condition can be best managed and that appropriate referral to the subspecialist can be made.
General considerations

Chest pain in children and adolescents is a commonly encountered problem. The extent of the complaint of chest pain is unclear and probably under-appreciated because many cases are dealt with in the primary care office and unreported. Driscoll et al [3] found that presentation for chest pain occurred at a rate of 0.29% in a prospective study of pediatric outpatient visits. A similar rate of occurrence (0.25%) was noted by Selbst [4] in pediatric patients presenting to the emergency department.

Several prospective studies have shown strikingly similar characteristics of chest pain in children and adolescents, although disparities regarding the etiology were present [3,5–7]. It is apparent that chest pain in the pediatric age group overwhelmingly is benign. It is unfortunate that the pain is difficult to treat and, in many instances, recurrent [8]. Interestingly, despite its benign nature, chest pain had significant life-altering effects on those children experiencing this condition. Approximately one half of children with chest pain had missed school at some point during their ailment [6,7]. A surprising number of adolescents in one study (69%) had restricted their own activity secondary to the chest pain [7]. In many instances, activities are either curtailed or completely stopped by parents, coaches, or primary care physicians until further testing is completed or the child is evaluated by a subspecialist.

When considering all pediatric-range patients, the mean age of patients experiencing chest pain was reported to be between 11 and 13 years of age by two independent studies [3,5]. Selbst et al [5] noted that younger age patients tended to have a higher percentage of organic disease, whereas adolescents made up the majority of patients with unknown or psychogenic etiologies of their chest pain. Taking into account all pediatric patients experiencing chest pain, there was no sex predilection; however, some differences in sex were seen with specific etiologies and are discussed later in this article [4–6,8,9].

Although there is much concern that chest pain in the pediatric age group may have an underlying cardiac origin, the percentage of pain attributed to the heart or great vessels is low. Two reports evaluated pediatric patients with chest pain referred to the cardiology clinic [10,11]. Theoretically, it is assumed that these patients would be biased toward cardiac disease by the nature of their referral. Kaden et al [10] found 12% with underlying cardiac conditions attributable to the chest pain. In the study by Fyfe and Moodie [11], a cardiac etiology was found in only 6% of those suffering from chest pain. Overall, the incidence of chest pain attributable to a cardiac etiology is less than 5% [9].

Evaluation of chest pain

Several organ systems have the potential to cause or contribute to pain localizing to the thorax (Fig. 1), including the musculoskeletal system and its
array of muscles with numerous insertions throughout the thoracic cage (Fig. 2). Pain can originate from muscle bodies, tendons, ligaments, cartilage, or bone. Other organ systems include the respiratory, cardiovascular, gastrointestinal, and nervous systems. Therefore, it is not surprising that the most important aspect in the evaluation of chest pain is a detailed history that includes a complete review of these systems. More often than not, the diagnosis or a suspected diagnosis can be ascertained from the history alone.

A detailed description of the chest pain should include information about quality, intensity, and location. In addition, timing of the pain can be helpful; this would include temporal relationships to exercise, eating, or trauma. Temporal relationships to the onset of pain can provide clues as to the etiology. Information about the frequency and duration of chest pain can aid in determining the seriousness of the episode.

Although all complaints of chest pain must be taken seriously despite etiology, pain that is constant or frequently occurring without completely subsiding typically is more worrisome than brief, infrequently occurring episodes of pain.
The length of time that the patient has been experiencing chest discomfort is helpful in elucidating the presence or absence of serious organic disease. Chest pain persisting longer than several months is unlikely to be related to serious organic etiology.

Finally, information about what makes the pain better or worse can pinpoint seriousness and etiology. For example, in an otherwise healthy individual with no fevers or respiratory symptoms, pain that gets worse with inspiration is generally reassuring, implicating the musculoskeletal system, and decreases the possibility of potentially life-threatening conditions. This having been said, it is therefore important to determine whether there are any associated complaints. Constitutional symptoms such as fever, weight loss, or fatigue; respiratory complaints such as cough, shortness of breath, or wheezing; cardiac findings of palpitations, racing heart, dizziness, or syncope; and gastrointestinal issues such as vomiting or dysphagia are all important pieces to the puzzle and why a routine, complete review of systems can be beneficial in identifying any valuable information that was missed when discussing the chief complaint. Some of the pertinent history
and associated symptoms are explored when discussing the specific etiologies for pediatric chest pain later in this article.

To completely understand the complaint of chest pain, it is important to know how the patient and the parents perceive the pain. The simple, direct question that asks the adolescent what they think the pain is from may be all that is required to discover the etiology. Many times, just the reassurance that their heart is fine allows the patient to deal more effectively with their pain, even if a direct cause is not found. Parents are especially grateful to learn that the pain is not life threatening. Not knowing what the cause may be but knowing what it is not may make a big difference. On the other hand, elucidating the patient’s and parents’ understanding of the chest pain may provide clues that lead the physician to suspect a more psychogenic cause to the pain or an atmosphere that may perpetrate the chest pain. It is important to understand how the chest pain may be interrupting the patient’s life and, for that matter, affecting the family dynamics. Chest pain is not always a concern for serious organic disease but can be an entity (organic or not) that is extremely disruptive for the patient and family. Therefore, an extension to the history obtained about perceived notions is a good understanding of the patient’s and family’s social history. Recent stressors at home and at school should be elucidated. Pain and incapacitation secondary to a recent life stressor is real and needs medical attention. Family dynamics can be evaluated during the interview process by noting the interaction between child and parent. Are there signs of stress or anxiety? Does the child speak for him/herself or does a parent speak for him/her? Does the parent take over the consultation? All of these observations can influence further evaluation and potential treatment. Finally, a thorough family history may provide clues to help establish a diagnosis.

Just like the evaluation of any illness with unknown etiology, a complete general physical examination is required. This examination must include accurate measurements of the vital signs. It is unfortunate that blood pressure readings commonly are omitted in the pediatric setting. There is a misconception that accurate blood pressures are difficult to obtain in the pediatric patient or that alterations in blood pressure do not happen frequently enough in pediatrics to make measurements necessary. Accurate pediatric blood pressures are obtainable and are an important part of every physical examination for an unknown illness or even during routine well-child examinations. Special attention needs to be made when examining the chest. On general inspection, signs of injury such as bruising or swelling need to be looked for and investigated. Symmetry needs to be assessed, including the breasts in girls and in boys. Frequently, the source of the chest pain is determined during palpation. The entire thoracic cage should be palpated for signs of tenderness; palpation of the precordium for signs of cardiac disease can be performed at the same time. Complete auscultation of the lung fields, evaluating for evidence of infection, bronchospasm, or effusion, is necessary. When examining the heart, the clinician should pay close attention to the second heart sound. Is splitting normal? Is the second heart sound loud or accentuated? Heart murmurs are expected to be common in this age group. Pathologic versus benign murmurs need to be differentiated; this topic is discussed
elsewhere in this issue. Heart murmurs per se do not cause chest pain; however, a few cardiac conditions that are associated with murmurs can lead to chest pain, as is discussed later. Finally, in many instances, patients can be put through several maneuvers in an attempt to illicit the pain. These include having patients twist at the torso, raise their arms over their head, push or pull against resistance, and inhale deeply. More often than not, the examination is normal, which is reassuring because serious organic disease seldom results in a normal examination. When the history is negative for serious pathology and there is a normal physical examination, further testing is not necessary. Several studies have shown that without a specific indication, routine tests were of no benefit in determining the cause of the chest pain [4–6,11]. Included in these tests were echocardiography and exercise stress testing [11]. When a normal history and physical is obtained, the physician may be tempted to order routine tests to reassure the families and, possibly, themselves. Ordering these tests, however, may have the opposite effect, raising anxiety that there is, in fact, something wrong.

**Etiologies of chest pain**

There are numerous causes of chest pain (Box 1). In this section, the causes are classified as noncardiac and cardiac etiologies. A brief description of those more likely to be encountered is presented in this section.

**Noncardiac causes**

**Idiopathic causes**

Idiopathic is the most frequently encountered diagnosis for chest pain in pediatrics. The typical patient presents with a few weeks–to–1-month history of occasional episodes of chest pain. The pain is described as sharp, occurring with or without exercise, and usually is short in duration. There are usually no specific associated symptoms, although many patients admit to feeling anxious about the pain. A few may temporarily stop their activity but most patients can continue what they were doing despite the pain. Recurrence is common, but long-term occurrences are not typical. The physical examination always is normal, with the pain not being reproducible.

**Musculoskeletal causes**

Musculoskeletal or chest wall pain also is a common etiology in pediatrics. The most common types are chest wall strain or costochondritis and arise from overuse. Frequently, there is a history of recent participation in sports or weight lifting. The pain is described as sharp and can be radiating. At times, there is increased pain with certain positions or activity. The pain can occur with or without activity and tends to increase with activity, particularly running, which likely is secondary to jarring of the affected area. Deep inspiration makes the pain worse. Chest wall strain typically involves one of the major muscle groups of the
Box 1. Causes of chest pain in children and adolescents

Noncardiac causes

Idiopathic
Musculoskeletal
Chest wall strain
  Trauma
  Costochondritis
  Precordial catch syndrome
  Slipping rib syndrome
Psychogenic
Respiratory
Asthma
Pneumonia
Pneumothorax
Pneumomediastinum
Chronic cough
Gastrointestinal
Gastroesophageal reflux
Esophagitis
Gastritis
Miscellaneous
Breast mass
Cigarette smoking
Sickle cell disease
Thoracic tumors
Herpes zoster
Pleurodynia

Cardiac causes

Coronary artery disease
Vasospasm
Anomalous origin
Kawasaki disease
Hyperlipidemia/early coronary artery disease
Cocaine abuse
Dysrhythmias
Supraventricular tachycardia
Ventricular tachycardia
Inflammatory
Pericarditis
Costochondritis affects one or more of the rib cartilages and, in one study, was found more frequently in girls [12]. Costochondritis can be preceded by respiratory illnesses and probably is related to frequent or excessive coughing. Pain is reproducible with palpation or maneuvers.

Precordial catch syndrome has features consistent with chest wall pain and is described in this section, although it would fit just as easily under idiopathic chest pain because an exact etiology is unknown [13]. Precordial catch syndrome, or Texidor’s twinge, is described as a sharp pain that is well localized. The pain usually occurs at rest and has a split second onset taking the patient by surprise. Typically the pain lasts only seconds to minutes with deep breathing making the pain worse. Patients tend to breathe shallowly and may even hyperventilate. Patients also may sit straight up to help relieve the pain, suggesting that posture may have a role in pathophysiology. Physical examination is normal, without reproducible pain.

A less-known etiology in the pediatric population involves the eighth, ninth, and tenth ribs and is known as the slipping rib syndrome [14]. These ribs do not attach directly to the sternum but join together by fibrous tissue. There can be some laxity and the ribs can slip over one another. Patients describe a popping or clicking sensation followed by a dull ache in either upper abdominal quadrant. The symptoms are related to activity. Trauma is the suspected inciting event. On physical examination, a positive hooking maneuver in which the rib is lifted anteriorly with curved fingers reproduces the popping sensation, and pain is pathognomonic. In some cases, persistent, life-changing pain can be addressed with surgical removal of the affected rib.

**Psychogenic causes**

Psychogenic chest pain in adolescents accounts for about 9% to 20% of patients seen, which is a significant proportion [15]. The pain described is vague, sometimes changing, and long standing. Various associated symptoms are reported, which may include headaches and abdominal pains [15]. Symptoms consistent with hyperventilation and panic attacks, such as dizziness, also are common. Asnes et al [16] reported that a stressful event preceding the onset of chest pain almost always can be uncovered. Interestingly, adolescent girls are the typical patients who suffer psychogenic chest pain, although boys also can be affected. Sometimes, the interaction between patient and parent can
lead the clinician to suspect a psychogenic origin to the pain. The authors often note parents who are overly anxious about the situation, believing something is seriously wrong with their child. The physical examination is completely normal.

Respiratory causes

Asthma is a common malady seen in pediatrics and it has been attributed to cause relatively frequent respiratory-induced episodes of chest pain [4–6,17]. Wiens et al [17] reported a staggeringly high incidence of exercise-induced asthma in pediatric patients with chest pain. Chest pain usually occurs with exercise and, in many cases, is described as chest tightness. Associated findings are shortness of breath and, occasionally, wheezing. Many patients have a history of bronchospasm or allergies or, frequently, there is a family history of asthma. Pain and respiratory symptoms subside relatively quickly with rest. Because symptoms are usually associated with activity, these patients frequently are referred to the pediatric cardiologist. Exercise testing with pulmonary function studies confirms suspected exercise-induced asthma.

Pneumonia can present with chest pain; however, associated respiratory symptoms and fever are typical. Pneumothorax and pneumomediastinum present with chest pain. With pneumothorax, the pain is unrelenting and there are the typical respiratory findings. Pneumomediastinum chest pain is ill defined. There often is dysphagia and subcutaneous emphysema [18–20].

Gastrointestinal causes

In the category of gastrointestinal disease as a cause of chest pain in pediatrics, only gastroesophageal reflux disease has any significant contribution [21]. Chest pain as a result of “heart burn” is relatively uncommon in pediatrics [15]. The diagnosis is made by the history. Like adults, children describe a burning pain that at times may cause them to double over. Pain usually is worse in the recumbent position. Temporal relationships to eating or to eating specific foods typically are uncovered. Symptoms improve with empiric antacid therapy. Physical examination typically is normal, although epigastric tenderness sometimes may be elicited when disease is severe and esophagitis is present.

Miscellaneous causes

Of the numerous, unusual etiologies of chest pain in children, one deserves some discussion. Breast masses can occur in girls and in boys. They usually are seen during puberty and can be anxiety provoking, especially in boys. Specific questions about the breasts in the history usually uncovers the concerns. The diagnosis is made easily with the breast examination and, in most cases, reassurance is all that is required. Concern for more serious pathology requires appropriate referral.
Cardiac causes

The most feared cause of chest pain in children is that of cardiac origin. Several cardiac problems have the potential to cause chest pain. Thankfully, those that would be considered life threatening are rare.

Coronary vasospasm

Vasospasm of the coronary arteries in association with atherosclerotic coronary artery disease is a well-known phenomenon in adults. Case reports describing myocardial ischemia without predisposing risk factors in adolescents have been sporadically presented in the literature [22–25]. These ischemic episodes have been attributed to vasospasm. At the author’s institution over the past 2 years, three adolescent boys have suffered myocardial ischemia and damage as demonstrated by elevated cardiac enzymes and ischemic changes on ECG. Fig. 3A shows an ECG from one of these patients and is compared with an ECG showing early repolarization (Fig. 3B), a pattern that is a normal variant but sometimes confused with ischemic changes. The pain described with these episodes is crushing, diffuse, and unrelenting. Associated features that can be seen with ischemic pain are diaphoresis, nausea, dyspnea, and syncope. Physical examination reveals a patient in distress and, possibly, pale and diaphoretic. A gallop rhythm and a heart murmur of mitral insufficiency may be noted on auscultation. Signs of decreased perfusion and evidence for elevated catecholamines may be apparent. Vasospasm is presumed when the workup reveals no other risk factors and includes a negative toxicology screen. Acetylcholine provocation testing has been used to diagnose vasospasm; however, this potentially dangerous test is not routinely done [25]. Cocaine abuse has a similar presentation to vasospasm, with evidence of cardiac ischemia [26,27]. Pathophysiology is believed to be related to high cardiac oxygen consumption, with coronary artery vasospasm. Examination may reveal an anxious patient with confusion or combativeness. Toxicology screening will confirm recent cocaine use.

Anomalous coronary arteries

Origin of the coronary arteries from the opposite sinus of Valsalva can result in myocardial ischemia and sudden death [1,28]. The predominant presenting symptom is sudden death; however, chest pain can be a presenting symptom. Frommelt et al [29] reported a series of 10 patients, 2 of whom had chest pain consistent with ischemia. Five of the 27 patients in a study by Basso et al [30] experienced chest pain up to 24 months before the episode of sudden death. The chest pain usually occurs with intense physical activity. Pathophysiology is related to inadequate coronary perfusion with intense exercise, either by compression between the great vessels, relative ostial stenosis of the anomalous coronary artery, or both. This rare entity is one in which significant pathology exists in light of a normal physical examination. The key to diagnosis is a careful history that reveals symptoms only during intense exercise. ECG and exercise
Fig. 3. Comparison of ECGs showing ischemic changes versus normal variant. (A) Acute myocardial infarction in an adolescent boy. Note the S-T elevations in leads I, V4, V5, and V6. The lateral precordial leads V4, V5, and V6 also have an abnormal inversion of the T wave. (B) Early repolarization pattern in a teenage patient. S-T segments exhibit typical sloping pattern. T waves are normal.
stress testing were not helpful in making a diagnosis before the sudden-death event [30].

**Kawasaki disease**

Healing of the coronary artery aneurysms seen following the inflammatory phase of Kawasaki disease results in areas of stenosis. Like any coronary artery obstruction, reserve for myocardial perfusion is decreased. In addition, thrombosis from insufficient laminar flow within the aneurysms can decrease coronary perfusion further. Chest pain in a child with a previous history of Kawasaki disease and coronary artery changes should be considered ischemia until proved otherwise.

**Left ventricular outflow obstruction**

Hypertrophic cardiomyopathy is the most common cardiac cause of sudden death; however, chest pain is an unusual feature [2,28]. Like anomalous origin of the coronaries, sudden death is a common presenting feature likely secondary to ventricular dysrhythmias. Syncope is more common than chest pain, although both symptoms can be considered associated features. In many cases, a family history positive for the disease or for unexplained sudden death is uncovered due to the inherited nature of the phenotype. Spontaneous mutations occur, however, and the history may be normal. Physical examination may reveal a systolic ejection murmur of subaortic stenosis. The murmur changes with maneuvers, becoming more intense with standing or the Valsalva maneuver.

Chest pain can occur with aortic stenosis but, typically, only with severe obstruction [28]. Due to the impressive nature of any significant valvar aortic stenosis murmur, it is not likely to go undiagnosed. Any pathologic murmur found in the evaluation of chest pain needs referral.

**Tachyarrhythmias**

Supraventricular tachycardia has been perceived as chest pain, especially in younger children. Theoretically, prolonged episodes can result in endocardial ischemia with subsequent chest pain or ischemia, but this has not been the authors’ general experience. In patients presenting to the pediatric cardiology clinic for evaluation of chest pain, Kaden et al [10] found supraventricular tachycardia to be the most frequent reason for chest pain. On questioning, the patient or parents may state that the heart feels like it is pounding, with abrupt initiation and termination. Episodes occur with and without activity. The patient may experience nausea or fatigue during episodes. Parents relate how the child just stops his/her activity and lies down. Physical examination usually is normal in the absence of supraventricular tachycardia. Long-term supraventricular tachycardia leads to tachycardia-induced dilated cardiomyopathy, resulting in a gallop rhythm and possible mitral insufficiency murmur on examination.

Premature ventricular contraction and ventricular tachycardias rarely produce isolated chest pain [28]. Other factors such as palpitations, racing heart, syncope, or exercise intolerance almost always are present.
**Inflammatory**

Pericarditis and myocarditis can be associated with chest pain; however, associated findings almost always are found. Pericarditis presents with fevers and a friction rub in most cases and, in one report, was a relatively common cardiac cause of chest pain in the cardiac clinic setting [11]. Myocarditis always has an associated tachycardia, and a gallop rhythm usually is auscultated. In many cases, pericarditis and myocarditis are associated with a preceding viral illness. A recent history of trauma may exist in some patients with pericarditis.

**Connective tissue disorders**

The physician must be cognizant of chest pain that occurs in patients with a diagnosed or suspected connective tissue disorder. Aortic dilatation with the potential for aortic dissection and rupture exists [31,32]. The chest pain is unrelenting and becomes severe. Frequently, there are associated symptoms such as dyspnea and abdominal pain. The patient exhibits significant distress; signs of diminished cardiac output may be present. Physical examination may reveal the stigmata of Marfan syndrome or other connective tissue disease.

**Mitral valve prolapse**

Chest pain caused by mitral valve prolapse is vague and controversial [33]. Clinicians should be leery of attributing chest pain to mitral valve prolapse in pediatric patients. It should be considered only when no other etiology can be found and the classic findings of a midsystolic click and murmur that moves with maneuvers such as standing or the Valsalva maneuver are present. Other entities associated with mitral valve prolapse, such as connective tissue disorders and panic disorders, need to be considered as the cause of the chest pain [28,33].

**Treatment of chest pain in pediatrics**

The lack of significant pathology in most children and adolescents suffering from chest pain makes reassurance the mainstay of therapy. Patients and their parents want to hear that the heart is normal. Interestingly, following initial evaluation by their primary care physicians, many patients exhibited heightened anxiety concerning the heart and thought something was wrong with their heart based on a study questionnaire [10]. A patient with a normal history and physical examination rarely has pathology, and routine tests or unnecessary referrals may be detrimental. Follow-up studies on pediatric chest pain were reassuring because no significant pathology was found [3,4,7,11]. A medical provider who exhibits confidence and takes an active role in counseling patients and families is instrumental in resolving most chest-pain issues. Due to the recurrent nature of the chest pain, follow-up visits are important. It is advisable to let patients and parents know that the condition can be frustrating and recurrent pain is expected. This information helps families to cope without allowing their fears to alter lifestyle. Of course, chest pain related to organic causes needs appropriate treat-
ment or referral. Chest pain that is associated with syncope; diaphoresis, pallor, or nausea; palpitations or sensation of a racing heart; or that occurs only with strenuous activity requires prompt referral to the pediatric cardiologist.

Summary

Chest pain in the pediatric population is common but generally benign. Cardiac causes of chest pain are uncommon and the overwhelming majority should be suspected from the history and physical examination. Routine testing has not been shown to be helpful and should be used only when indicated by the history and physical examination. A strong caregiver–family relationship is the most beneficial aspect of care to ensure optimal outcomes.

References