

A Teen with Chest Pain

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KEYWORDS

• Chest pain • Cardiac • Noncardiac • Teen • Adolescent

KEY POINTS

- Chest pain in teens is most commonly noncardiac and musculoskeletal in nature.
- Chest pain from a cardiac cause is rare in teenagers.
- A thorough history and physical examination are typically the only evaluation required for assessment of chest pain.
- Patients with a history of repaired congenital heart disease, Kawasaki disease with coronary artery aneurysms, certain connective tissue disorders, symptoms of exertional chest pain, or association with syncope should be referred to a pediatric cardiologist for further evaluation.

INTRODUCTION: NATURE OF CHEST PAIN

Chest pain is a commonly encountered symptom in the outpatient setting as well as the emergency room. The onset of chest pain in adults usually necessitates prompt cardiac evaluation because coronary vascular disease, while potentially life-threatening, can be managed and treated successfully if detected in a timely fashion. When this symptom is seen in the teenage population, teenagers and their parents are typically concerned about cardiac causes for the pain. Fortunately, chest pain in the teenage population is most commonly benign.¹⁻⁸ Media coverage of rare and unfortunate events of sudden cardiac death only contributes to teen and parental anxiety about chest pain complaints. Chest pain has been associated with higher health care utilization in children with noncardiac chest pain particularly if the parent or child has increased psychological stress.⁹ These factors all add to the challenge for the medical professional who is evaluating the teenage patient with chest pain.

Chest pain has previously been reported to account for 0.29% of patient chief complaints to the emergency room in a prospective trial by Driscoll and colleagues.¹ In more recent studies, chest pain accounted for 5.2% of all cardiology consultations¹⁰

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and 15% of all outpatient visits at a large, tertiary center pediatric cardiology practice.¹¹ However, less than 5% of chest pain complaints are associated with a cardiac condition.^{12,13} Although media attention to these episodes in the community gives the sense that these episodes occur on a more frequent basis, several studies have shown that the incidence of sudden cardiac death in teens is fortunately rare.¹²⁻¹⁴ Most recently, Roberts and Stovitz¹⁵ reported an incidence of sudden cardiac death in adolescents as 0.24 per 100,000 athlete-years over the last 19 years in Minnesota. Chest pain in teens is most commonly divided into noncardiac and cardiac causes.

Key message

- Chest pain is a common complaint seen in primary care office, emergency room, and pediatric cardiology practice
- Chest pain in adolescents is most commonly benign
- Chest pain in this age group is perceived as cardiac by parents and patients and associated with tremendous anxiety
- Less than 5% of the chest pain is cardiac in origin

NONCARDIAC CHEST PAIN

Musculoskeletal chest pain is a very common type of noncardiac chest pain with reported prevalence anywhere from 15% to 31%.^{12,13} Several types of musculoskeletal chest pain are seen in teenagers (**Box 1**).

COSTOCHONDRITIS OR COSTOSTERNAL SYNDROME

Costochondritis or costosternal syndrome typically presents as a sharp, stabbing pain along 2 or more contiguous costochondral joints. Deep breathing usually exacerbates the pain and this pain usually lasts just a few seconds to a few minutes. Signs of joint inflammation are absent, but palpation of the chest over the area reproduces the pain.

TIETZE SYNDROME

Tietze syndrome is most often seen in teens and adults. There is frequently a history of recent upper respiratory infection. Excessive coughing is thought to be a possible mechanism. There is localized inflammation of a single costochondral joint with the second and third ribs most often involved. Signs of inflammation, such as warmth, swelling, and tenderness, are found at the specific costochondral, costosternal, or sternoclavicular joint involved. Signs of inflammation are what helps differentiate this from costochondritis.

TRAUMA AND MUSCLE STRAIN

Trauma and muscle strain are particularly common in teenagers who are active in sports and are prone to chest wall trauma or muscle strain. Skeletal trauma in one series was the cause of chest pain in 2% of teenagers and children.¹² If trauma is the underlying cause, there may be inflammation or signs of injury at the site of pain. In cases where the history of trauma is significant, signs or symptoms of hemo-pericardium and myocardial contusion should be evaluated. Weight training or history of heavy lifting is often underreported unless specifically asked in cases where muscle strain is suspected to be the source of the chest pain.

Box 1**Common causes of chest pain in teenagers**

- Noncardiac causes
 - Musculoskeletal
 - Costochondritis
 - Idiopathic
 - Tietze syndrome
 - Trauma and muscle strain
 - Slipping rib syndrome
 - Scoliosis
 - Chronic cough
 - Asthma
 - Pneumonia
 - Pneumothorax/pneumomediastinum
 - Pulmonary embolism
 - Gastroesophageal reflux disease
 - Gastritis
 - Esophagitis
 - Psychogenic
 - Breast disease, gynecomastia
 - Herpes zoster
 - Sickle cell disease (acute chest syndrome or vaso-occlusive crisis)
- Cardiac causes
 - Arrhythmia—supraventricular tachycardia, ventricular tachycardia
 - Pericarditis—infectious, noninfectious or autoimmune, postpericardiotomy syndrome
 - Left ventricular outflow tract obstruction—aortic stenosis, subaortic stenosis, supraaortic stenosis
 - Anomalous origin of the coronary artery
 - Kawasaki disease
 - Coronary artery vasospasm
 - Hyperlipidemia or family history of early coronary artery disease
 - Cocaine use
 - Other—cardiac device or stent complications, aortic dissection, ruptured aortic aneurysm, pulmonary hypertension

IDIOPATHIC CHEST-WALL PAIN

Idiopathic chest pain typically is located over the midsternum or inframammary area and is sharp, lasting only a few seconds to minutes, and is exacerbated by deep inspiration. Palpation over the sternum or rib cage may elicit pain.

Slipping rib syndrome is seen infrequently and involves intense pain in the lower chest or upper abdominal area. The 8th, 9th, and 10th ribs are attached

to each other and not directly to the sternum. Trauma or dislocation in this area can lead to this condition and can be diagnosed by a positive “hooking maneuver” whereby the examiner pulls on the inferior rib margin, which pulls the lower rib cage out.

PULMONARY

A pulmonary cause is also a common cause of chest pain. Asthma represents the most common pulmonary cause of chest pain. Seventy-three percent of children with chest pain were found to have evidence of asthma in a study by Weins and colleagues.¹⁶ Infections of the bronchial tree or lungs, such as pneumonia, bronchitis, empyema, pleural effusion, or pleurisy, can cause acute chest pain. Pulmonary embolism can also be associated with chest pain and may be suspected in teens presenting with hypoxia and a predisposition to thrombosis.

GASTROINTESTINAL

Gastrointestinal causes can also account for a certain percentage of teenagers with chest pain. Evangelista and colleagues¹⁷ reported a prevalence of gastrointestinal causes for chest pain as high as 8%. Gastroesophageal reflux disease, peptic ulcer disease, esophagitis, gastritis, and cholecystitis may present as chest pain. More rarely, ingestion of caustic substances, foreign body, or an esophageal stricture may present as chest pain.

PSYCHOGENIC

Psychogenic chest pain is typically caused by anxiety or a history of a recent stressful event. Often, psychogenic chest pain is associated with other somatic complaints such as abdominal pain or headache. Psychosocial factors have been attributed to the development and maintenance of chest pain as defined by Gilleland and colleagues.¹⁸ Psychogenic chest pain has also been demonstrated in other somatic complaints such as abdominal pain and headache.^{19–21}

MISCELLANEOUS

There are various other causes of chest pain that can be placed in this category. Chest pain from breast-related causes can be seen in postmenarche teen girls in association with mastitis, fibrocystic disease, or pregnancy. In teen boys, gynecomastia may occasionally cause unilateral or bilateral chest pain. Herpes zoster infection may initially present with pain or paresthesia in a dermatomal pattern, be extremely painful and uncomfortable, and precede the rash by several days. Scoliosis or other deformities can cause chest pain because of nerve compression or abnormal posture and positioning or stretching of the chest wall.

Key message

- Musculoskeletal chest pain is extremely common in this age group
- Associated symptoms may suggest a pulmonary, gastrointestinal cause for the chest pain
- Psychogenic chest pain is a diagnosis of exclusion and may be seen with other somatic symptoms such as headaches and abdominal pain

CARDIAC CHEST PAIN

Actual cardiac causes of chest pain are rare. However, there are several cardiac diagnoses for which chest pain can be one of several presenting symptoms.

Inflammatory

Pericarditis, whether present with or without a pericardial effusion, is typically infectious in origin. Pain can be associated with pericarditis, but there are often other symptoms present as well. Patients usually have fever and may have a friction rub on examination. The pain is typically retrosternal and can radiate to the left shoulder. The pain tends to be more constant and unrelenting than the noncardiac chest pain. It worsens with deep breathing and lying in the supine position. Sitting up and leaning forward tends to alleviate the pain.

Tachyarrhythmia

Supraventricular tachycardia, particularly in younger patients, may be reported as chest pain. However, there are other symptoms such as a feeling of the heart “pounding,” nausea, dizziness, or fatigue. There should be no symptoms in between these episodes. In studies focusing on patients presenting to a pediatric cardiologist, Kadun and colleagues²² found that supraventricular tachycardia or palpitations interpreted as chest pain was the most common presentation in young, school-aged children and some teenagers presenting for palpitations. The chest pain, however, was usually also associated with other symptoms such as nausea and/or vomiting along with the episodes. Isolated premature ventricular contractions and ventricular tachycardia have a rare association with chest pain. Ventricular tachycardia usually has symptoms of palpitations, exercise intolerance, and/or syncope. Patients with a history of cardiac disease and surgical repair such as d-transposition of the great arteries with a Mustard or Senning repair are at risk of intra-atrial reentry arrhythmias. Teenagers with single-ventricle physiology who have undergone Fontan palliation are also at risk for intra-atrial reentry arrhythmias, which may present with chest pain but more likely with palpitations.⁸

Left Ventricular Outflow Tract Obstruction

Left ventricular outflow tract obstruction (aortic valve, subaortic area, or supra-aortic area) and coarctation of the aorta may have complaints of chest pain, but will usually also have other symptoms, such as dizziness, fatigue, and/or syncope with exertion. Severe aortic obstruction increases the likelihood of chest pain. The physical examination will have a harsh, systolic ejection murmur, which radiates into the neck. The most common cause of sudden cardiac death in this age group is hypertrophic cardiomyopathy. However, chest pain is actually an unusual symptom.^{8,22} Sudden death, likely from ventricular arrhythmia, is a frequent initial presentation in teenagers with hypertrophic cardiomyopathy. Syncope is more often the presenting complaint compared with chest pain, and a family history for unexplained sudden death should raise suspicion for this diagnosis. However, many cases are de novo so the family history may not always be helpful. A systolic murmur may be heard on examination over the left ventricular outflow tract, which should increase in intensity with standing or Valsalva maneuver.

Kawasaki Disease

Coronary artery stenosis is a well-known long-term complication for those who had a history of coronary artery aneurysms due to Kawasaki disease. After the inflammatory

phase, the healing coronary aneurysms can lead to areas of stenosis. It is also known that, due to these areas of stenosis, there may be decreased reserve for myocardial perfusion. The aneurysms themselves can cause decreased flow and thrombosis, which affects myocardial perfusion even further. Chest pain in a teenager with a history of coronary artery complications from Kawasaki disease usually is exertional in nature and should prompt a thorough evaluation for myocardial ischemia. Known risk factors that contribute to the likelihood of developing coronary aneurysms, coronary stenosis, or obstruction include male gender and onset at an early age (<6 months) or at an older age (>5 years).²³

Coronary Artery Anomalies

Congenital anomalies of the coronary arteries can also lead to an increased risk of myocardial ischemia, which may present with anginal chest pain. After hypertrophic cardiomyopathy, they are the second most common reason for sudden cardiac death in teenagers. Chest pain is not the typical presenting symptom. Teenagers with these coronary anomalies may have sudden death as the first symptom. For those that do present with chest pain, it is most often associated with exertion. Chest pain related to ischemia is usually described by patients as a crushing tightness, pressure, burning, or fullness in the chest. These patients often are diaphoretic, nauseous, and/or dyspneic. They may also present with syncope. A typical example in this category is the left main coronary artery or left anterior descending coronary artery arising from the right sinus of Valsalva or from the right coronary artery. The left main coronary artery or left anterior descending coronary artery then typically courses between the aorta and pulmonary artery, which leads to compression of the coronary artery during exertion, resulting in myocardial ischemia and/or ventricular arrhythmia and sudden death in a teen.

Coronary vasospasm is known to be associated with atherosclerotic coronary artery disease in adults. There are case reports^{24–26} of ischemia presumed to be secondary to coronary vasospasm in the literature in teenagers with no known risk factors. Cocaine abuse during adolescence can also present with chest pain due to coronary vasospasm and evidence of myocardial ischemia. Additional symptoms may include combativeness or confusion and the drug screen will be positive.

Teenagers with a family history significant for hyperlipidemia and early coronary artery disease are also at an increased risk for lipid abnormalities and myocardial ischemia.

Some teenagers with repaired congenital heart disease may experience chest pain. Patients at an increased risk for coronary complications are those who have undergone manipulation of their coronary arteries during their initial surgical repair as seen in the arterial switch operation for d-transposition of the great arteries and the Ross procedure. Both of these surgical procedures require reimplantation of the coronary arteries, which puts these patients at an increased risk for developing coronary artery ostial stenoses.

Connective Tissue Disorders

Although rare in this age group, teenagers with Marfan syndrome may present with sudden onset of severe chest pain along with back pain due to a dissecting aortic aneurysm. Teenage girls with Turner syndrome are also at risk of aortic root dissection, which is associated with sudden, intense chest pain. The other associated connective tissue disease with cardiac implications for chest pain is Ehlers-Danlos syndrome type IV. These patients may experience acute aortic dissection and may complain of chest pain that is mid sternal and radiates to the back. Pain associated

with dissection is typically severe and may be described as a “tearing” quality. Patients with any one of these syndromes complaining of chest pain with these qualities should have a workup (cross-sectional imaging, CT or MRI) to rule out aortic dissection immediately because of the life-threatening implications.

Mitral Valve Prolapse

Chest pain associated with palpitations, dizziness, and panic attacks have been seen in patients with mitral valve prolapse. However, Bisset and colleagues²⁷ reported only 18% of patients with mitral valve prolapse in their study group presented with complaints of atypical chest pain. Mitral valve prolapse can also be seen in individuals with connective tissue disorders.

Key message

- Cardiac cause for chest pain in this age group is extremely rare but clinical implications can be serious
- A thorough history and physical examination can be helpful in ruling out a cardiac cause

PATIENT HISTORY

Even though cardiac causes of chest pain are rare, it is important to perform a thorough history and physical examination to rule out the possibility of underlying disease. A comprehensive history and physical examination are the most helpful elements in evaluation of chest pain (**Box 2**).

Within the patient history, a description of the pain in the patient’s own words, severity of the pain, associated symptoms, aggravating, precipitating, and alleviating factors are important to describe. The acuteness or chronicity of the pain is also helpful. Acute pain can be associated with trauma, asthma, pulmonary embolism, and cardiac causes such as aortic dissection or ischemia due to coronary anomalies. Chronic, recurrent pain is most often likely due to noncardiac causes, such as pain from musculoskeletal, gastrointestinal, or psychogenic causes.

Pain location may also aid the practitioner in assessing the underlying cause. Pain that is localized is typically chest wall or pleuritic pain. Pain that is diffuse can be from underlying diseases of the lungs or heart. Whether and where the pain radiates may also be helpful to pinpoint a specific cause. An example would be pain that radiates to the neck, jaw, upper extremity, or shoulder is commonly associated with myocardial ischemia. Chest pain that radiates to the left shoulder can be seen in patients with pericarditis. Pain due to aortic dissection may radiate to the back or interscapular space.

Aggravating and/or precipitating factors are also very useful in aiding diagnosis. Pain exacerbated by deep breathing, change in body position, or with specific movements is typically musculoskeletal. Pain brought on by exertion or associated with dyspnea should prompt a search for a cardiac or respiratory cause. A recent history of respiratory illness and fever makes the diagnosis of an infectious cause leading to pericarditis or pneumonia likely. Patients with arrhythmia often complain of chest discomfort, but usually in association with palpitations. If complaints of vomiting or regurgitation are present with the chest pain and the pain worsens with eating or swallowing, the patient may have a gastrointestinal cause. Chest pain in association with various other symptoms like abdominal pain, headache, or limb pain makes a psychogenic cause most likely. Lightheadedness and paresthesias are often seen during panic attacks and/or hyperventilation episodes.

Box 2
History

- Chest pain description
 - Onset, duration, location, quality, severity, and radiation
 - Precipitating, aggravating, or alleviating factors
- Other elements of history
 - Associated symptoms
 - Trauma
 - Drug abuse
 - Presence of psychological stressors
 - Recent weight training or heavy lifting
- Past medical history
 - Cardiac disease, Kawasaki disease, hypercholesterolemia
 - Asthma, recent respiratory illness
 - Autoimmune disease
 - Sickle cell disease
 - Other chronic diseases
- Surgical history
 - Any previous chest or abdominal surgery
- Family history
 - Cardiomyopathy
 - Sudden cardiac death
 - Arrhythmia
 - Dyslipidemia
 - Premature coronary artery disease
- Genetic diseases
 - Marfan syndrome
 - Turner syndrome
 - Ehlers-Danlos syndrome, type IV

A history of asthma may suggest chest pain due to the underlying condition. Also, association of chest pain with difficulty in breathing, and cough temporally related to exercise, raises concern for exercise-induced asthma. A recent history of trauma, weight lifting, or a muscular trauma is the likely cause for the pain in teens involved in sports. Illicit drugs such as cocaine and other sympathomimetics are strong vasoconstrictors and can cause coronary vasospasm and vasoconstriction, which leads to pain from myocardial ischemia. It is crucial for health care providers to elicit a drug history and this might require a private interview with the adolescent.

It is also important to know about any history of cardiac diseases in the teen's past such as Kawasaki disease or d-transposition of the great arteries with an arterial switch operation. These patients could have coronary ostial stenosis leading to myocardial ischemia and pain. Patients who have undergone a recent cardiac catheterization

and device placement for an atrial septal defect or stent placements may have pain if there is device embolization leading to impingement on other cardiac structures. Patients with coronary vasculopathy from cardiac transplantation, primary coronary anomalies, or coronary artery stenosis in association with familial hypercholesterolemia may have anginal chest pain. History of sickle cell disease or other chronic conditions may be important as patients may experience chest pain caused by complications of their underlying chronic disease. Sickle cell patients may experience chest pain during acute chest syndrome and patients with autoimmune disorders such as Crohn disease or systemic lupus erythematosus may develop pericarditis with or without pericardial effusions and may complain of chest pain.

Familial genetic or connective tissue disorders are also important to elucidate because these can be associated with conditions that cause chest pain (Marfan, Turner, or Ehlers-Danlos syndrome type IV).

Key message

- The description of the pain, exact location, and associated activity is key to the diagnosis
- Recent initiation of weight training or heavy lifting is underreported by teenagers
- Chronic pain ongoing for several years suggests a benign nature
- Private interview with a teen may be warranted in cases of suspected drug abuse

PHYSICAL EXAMINATION

Vital signs and anthropometric measurements should be obtained during the comprehensive examination. Certain vital signs, anthropometric measurements, and/or clinical examination abnormalities may help direct the provider to further evaluations or testing (**Box 3**).

A teen with a very tall stature may need evaluation for Marfan syndrome. Tachypnea and fever may raise concern for a respiratory illness. Tachypnea and fever may also be associated with pericarditis (with or without effusion) if accompanied with tachycardia. In this situation, chest pain worsens with lying down and is relieved by sitting and leaning forward. Hypotension with evidence of jugular venous distention may indicate systolic or diastolic ventricular dysfunction, which would raise concerns for myocarditis or cardiomyopathy. Evidence of dysmorphism raises concern for a genetic disorder. The chest needs to be inspected thoroughly, looking for any chest wall or bony abnormalities, pectus excavatum or carinatum, scoliosis, or surgical scars. Thelarche in teen girls or gynecomastia in teen boys is not an uncommon source of chest pain.

On palpation of the chest, reproducible chest tenderness similar to the experienced pain is a very important and reassuring sign, effectively ruling out a primary cardiac cause. Rales or wheezing on chest auscultation might raise concerns for a pneumonic process or a hyperreactive bronchial airway (asthma, exercise-induced asthma).

A detailed cardiac examination is very important to look for potential cardiac causes of chest pain. Palpation of the chest is performed for thrills (obstructive lesions) or heaves (increased right ventricular impulse seen in pulmonary hypertension). Attention should also be paid to the examination of the heart sounds. Distant or muffled heart sounds are suspicious for a pericardial effusion. When the fluid accumulation in the pericardial cavity is in the mild to moderate range, a pericardial rub may be heard. Myocarditis often has evidence of a gallop rhythm and may have a mitral regurgitation murmur. Fixed left ventricular outflow tract obstruction from supravalvar, valvar, or subvalvar aortic stenosis and obstructive hypertrophic cardiomyopathy usually

Box 3**Physical examination**

- Vital signs
 - Height, weight, body mass index, heart rate, blood pressure (lower extremity blood pressure with hypertension), respiratory rate
- General inspection
 - Dysmorphic features
- Chest inspection
 - Chest wall abnormalities (pectus excavatum or carinatum)
 - Scoliosis
 - Signs of trauma, healed surgical scars
 - Reproducible chest wall tenderness on palpation
- Cardiac
 - Hyperdynamic precordium
 - Distant heart sounds, abnormally loud second heart sound
 - Murmur (+/-), systolic click (+/-), gallop (+/-)
 - Upper extremity (right arm) hypertension (greater than 20 mm Hg difference between upper and lower limb blood pressure)
 - Decreased femoral pulse

presents with a harsh, mid-systolic ejection type of murmur. An ejection systolic click may be heard in aortic stenosis. Patients with mitral valve prolapse typically have a mid-systolic click and an apical mid to late systolic honking murmur. If a continuous murmur is heard, a coronary fistula may be present. Patients with coarctation of the aorta will have weak femoral pulses and upper limb hypertension (blood pressure ideally obtained from the right arm). In some coarctation patients, a continuous murmur over the back in the lower scapular margin may suggest the presence of extensive collateral arteries. Congestive heart failure should be suspected if hepatomegaly, ascites, and peripheral edema are present.

Key message

- Reproducible chest wall tenderness is the single most important examination finding in the evaluation of a teenager with chest pain, yet is commonly missed

IMAGING AND ADDITIONAL TESTING

Many teenagers who present with chest pain have a normal history and physical examination, and in that case, no further evaluation or testing is required. If there are any positive elements in their history or physical examination as described above, further testing should be performed accordingly. In patients with benign causes of chest pain, reassurance to the both the teenager and the parents that this pain is not cardiac and that the cardiac examination is normal is usually all that is required. Families should also be educated that these chest pain episodes may be recurrent but it does not imply heart disease. Several studies^{5,7,8} have shown that additional or routine testing is not required. Follow-up in these patients²⁸ has shown that those

who had previously been evaluated for chest pain do not present later with concerning pathologic abnormality.

Patients with chest pain related to other disease processes like asthma or gastroesophageal reflux disease should be referred to the appropriate specialists as deemed necessary. Most cases of teenage chest pain do not require cardiology referral and can be managed by the primary providers. However, in patients with chest pain that occurs with exertion, palpitations, exertional syncope, or in those with known cardiac disease, the chest pain episodes should be evaluated by a pediatric cardiologist.

Key message

- Most adolescents with chest pain do not require any additional testing
- Only those with chest pain associated with activity, syncope, palpitations, abnormal cardiac examination, or those with known cardiac disease require referral to cardiology

SUMMARY

Chest pain in the teenage population is seen frequently, but is typically benign. A thorough history and physical examination should provide enough information to confirm or rule out suspicions for cardiac disease. Time spent in counseling and reassurance of the teenager and parents in regards to the benign nature of their chest pain is most important. In those individuals who have symptoms with exertion, additional associated symptoms, or an abnormal cardiac examination require referral to a pediatric cardiologist.

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