Evaluation of Chest Pain in the Pediatric Patient

Jennifer Thull-Freedman, MD, MSc

Many causes of chest pain in children are benign and self-limited. Nonetheless, serious and life-threatening etiologies exist, and the challenge to the practitioner is to be able to identify the few patients who have a serious cause for their pain. Furthermore, chest pain is a worrisome symptom for families who often fear a cardiac cause, and the symptom may lead to school absence and limitation of activities. The differential diagnosis for pediatric chest pain is extensive (Box 1), and physicians caring for children must be familiar with the possible causes for chest pain and attempt to identify an etiology. In many cases a thorough history and physical examination are sufficient to identify the source of the pain, and diagnostic testing can be performed on a selective basis to address concerns identified. Only after a serious cause has been excluded should reassurance and symptomatic care be offered.

EPIDEMIOLOGY

Chest pain accounts for approximately 0.3% to 0.6% of pediatric emergency department (ED) visits.1-3 The frequency of visits is fairly constant throughout the year,4 with a slight excess in summer months reported in one study.1

In EDs treating children up to 18 years of age, the median age for presentation with chest pain was 12 to 13 years.1,5,6 The reported male to female ratio is fairly even, ranging from 1:1 to 1.6:1.1,5,6 In adolescents, relatively more girls present with chest pain.5 Many ED studies report that most children present with acute pain of less than 1 day in duration.2,6 In contrast, a study done in Turkey reported that 59% of patients described pain greater than 1 month in duration.7

KEYWORDS

- Cardiac
- Electrocardiogram
- Pediatric chest pain
- Pulmonary embolism

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<table>
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<tr>
<th>Box 1</th>
<th>Differential diagnosis of pediatric chest pain</th>
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<td><strong>Cardiovascular</strong></td>
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<td>• Arrhythmia</td>
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<td>• Coronary artery disease (anomalous coronary arteries, acute Kawasaki disease [coronary arteritis], premature atherosclerosis [eg, dyslipidemia])</td>
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<td>• Coronary artery vasospasm (toxicologic ingestion [cocaine, marijuana])</td>
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<td>• Structural (hypertrophic cardiomyopathy, valvular stenosis [pulmonary, aortic], mitral valve prolapse)</td>
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<td>• Myocarditis</td>
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<td>• Pericarditis</td>
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<td>• Endocarditis</td>
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<td>• Congenital absence of pericardium</td>
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<td>• Aortic aneurysm or dissection (Marfan, Turner, and Noonan syndromes)</td>
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<td><strong>Respiratory</strong></td>
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<td>• Asthma</td>
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<td>• Pneumothorax/pneumomediastinum</td>
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<td>• Pulmonary embolism</td>
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<td>• Pleuritis/pleural effusion (eg, systemic lupus erythematosus)</td>
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<td>• Pleurodynia (coxsackievirus)</td>
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<td>• Chronic cough</td>
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<td>• Airway foreign body</td>
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<td><strong>Abdominal and gastrointestinal</strong></td>
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<td>• Esophagitis (gastroesophageal reflux disease, eosinophilic esophagitis, bulimia, pill esophagitis)</td>
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<td>• Esophageal foreign body</td>
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<td>• Esophageal spasm/dysmotility</td>
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<td>• Gastritis</td>
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<td>• Hiatal hernia</td>
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<td>• Referred pain from abdominal trauma (Kehr sign)</td>
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<td>• Cholecystitis</td>
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<td><strong>Musculoskeletal and chest wall</strong></td>
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<td>• Chest wall strain (exercise, overuse injury, forceful coughing)</td>
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<td>• Skeletal (chest wall or thoracic spine) anomaly</td>
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<td>• Trauma (contusion/rib fracture)</td>
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<td>• Costochondritis/Tietze syndrome</td>
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<td>• Slipping rib</td>
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<td>• Precordial catch (Texidor twinge)</td>
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<td>• Breast tenderness</td>
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<td>• Cutaneous (eg, herpes zoster)</td>
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CAUSES OF CHEST PAIN IN CHILDREN

Most of what is known about frequency of various causes of pediatric chest pain comes from studies performed in pediatric EDs and cardiology clinics. Table 1 provides a list of frequencies of causes according to organ system. In general, the most frequent cause reported is musculoskeletal pain, including costochondritis. These conditions represent between 7% and 69% of cases presenting to an ED, with the reported frequency dependent somewhat on how strictly musculoskeletal pain is defined and whether it is used as a diagnosis of exclusion or reported in combination with idiopathic causes. Respiratory causes including asthma are the second most common organic etiology identified, representing 13% to 24% of cases. Gastrointestinal and psychogenic causes are identified in less than 10% of cases, and a cardiac cause is found infrequently, representing not more than 5% of cases. An idiopathic etiology was frequently assigned in several studies, accounting for 20% to 61% of diagnoses made.\(^5\) Children who were given a diagnosis of nonorganic chest pain were more likely to have pain greater than 6 months in duration and more likely to have a family history of chest pain or heart disease.\(^5\) Children who were given a diagnosis of organic disease were more likely to have pain of acute origin, pain awakening them from sleep, fever, or abnormal examination findings. Children less

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<th>Table 1</th>
<th>Frequency of causes in children complaining of chest pain</th>
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<tr>
<td>Cause</td>
<td>Emergency Department or Pediatric Clinic (%)(^1,4-6,8,9)</td>
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<tr>
<td>Idiopathic/cause unknown</td>
<td>12–61</td>
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<tr>
<td>Musculoskeletal/costochondritis</td>
<td>7–69</td>
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<tr>
<td>Respiratory/asthma</td>
<td>13–24</td>
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<tr>
<td>Gastrointestinal/gastroesophageal reflux disease</td>
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<tr>
<td>Psychogenic</td>
<td>5–9</td>
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<tr>
<td>Cardiac</td>
<td>2–5</td>
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than 12 years of age were two times more likely to have a cardiac or respiratory cause for their pain, whereas adolescents were 2.5 times more likely to have a psychogenic cause.\(^5\) Rowe and colleagues\(^1\) demonstrated an increased frequency of traumatic causes in boys.

**Chest Wall**

Direct trauma has been reported as the cause for chest pain in approximately 5% of cases.\(^4,5\) Frequent or severe cough can cause chest wall pain because of muscle strain. Children who engage in a new or intense physical activity may experience delayed-onset muscle soreness of the pectoralis or shoulder muscles. Delayed-onset muscle soreness typically peaks within 2 days following activity. Because of the lag in development of soreness, children and parents may not recognize the association of the pain with the preceding activity before seeking medical attention. The pain of delayed-onset muscle soreness can generally be reproduced by palpation or engagement of the involved muscle group. Treatment is with nonsteroidal anti-inflammatory agents and rest.

Chest wall deformities, such as pectus excavatum or pectus carinatum (Fig. 1), can be associated with musculoskeletal chest pain. Patients with chest wall deformities should be examined carefully for findings consistent with Marfan syndrome, which is associated with an increased risk of aortic root dilation and dissection and spontaneous pneumothorax (Fig. 2). Pectus excavatum in isolation may also be associated with aortic root dilation, even when other stigmata of Marfan syndrome are absent.\(^13\)

Costochondritis is defined as a pain localized to a costal cartilage that is reproducible on palpation. Many patients complaining of chest pain are found to have areas of tenderness at the costochondral or costosternal junctions (26%–41%).\(^1,2\) A diagnosis of costochondritis is assigned when pain reproducible by palpation is not attributed to

another specific diagnosis. The causes and natural history of this condition are not well understood. Postulated etiologies include minor trauma, cough, and postviral reaction. In adults, costochondritis can be associated with fibromyalgia and other rheumatologic conditions in a minority of individuals; however, in children this association has not been described. In a study that was limited by a large loss to follow-up, 62% of adolescents (15% of the original population) who had been diagnosed with costochondritis still reported pain after 1 year.

Tietze syndrome is a specific form of costochondritis characterized by localized, painful, nonsuppurative costochondral swelling. Mukamel and colleagues described
a series of eight Israeli children between 10 months and 12 years of age with a clinical diagnosis of Tietze syndrome. Masses were usually tender; varied in size from 1 to 4 cm; and were located in lower, middle, and upper costochondral junctions. None had fever or systemic symptoms, although elevated erythrocyte sedimentation rate was reported in some. All cases resolved within 2 months. The etiology of this condition is unknown.

Slipping rib syndrome is an unusual cause of lower chest pain that results when the medial fibrous attachments of the 8th, 9th, or 10th ribs are inadequate or ruptured, allowing the costal cartilage tips to sublux and possibly impinge on intercostals nerves. In some cases there may be a preceding trauma. Patients may be aware of a popping sensation at the onset of pain. The diagnosis is supported by a positive “hooking maneuver,” which consists of hooking the fingers under the lowest costal cartilages and drawing them anteriorly and superiorly, reproducing the symptoms. Saltzman and colleagues described a case series of 12 patients diagnosed with slipping rib syndrome who experienced temporary relief of pain with intercostal nerve block (nine of nine) and complete relief of pain following excision of the offending rib tip (nine of nine).

Precordial catch syndrome is a clinical diagnosis applied to a characteristic pattern of benign chest pain. The pattern was first described in 1955 by Miller and Texidor and came to be known as “Texidor twinge.” The pathophysiology of the syndrome is not known, and although it has been described anecdotally as a frequent cause of pediatric cardiology referral, it has not been specifically studied in the pediatric population. It is described, however, as a sharp pain of sudden onset localized to the anterior chest wall that occurs mostly at rest. It tends to last from a few seconds to 3 minutes and may be exacerbated by taking a deep breath. There are no associated symptoms, and physical examination is negative.

Breast tenderness can also be the source of chest wall pain. This can be physiologic during thelarche, or caused by infectious or inflammatory conditions, such as mastitis. Cutaneous chest wall pain may occur during an episode of herpes zoster. Pain occurs in a unilateral dermatome distribution and may precede the development of characteristic skin findings by several days.

**Pulmonary**

Approximately 13% to 24% of children with chest pain seen in an ED or ambulatory setting are found to have a pulmonary origin for their pain. The most frequently implicated respiratory cause is asthma. Selbst and colleagues identified asthma as the diagnosis in 7% of patients presenting to the ED with chest pain. Exercise-induced asthma may be an underrecognized cause of pediatric chest pain. A study of children with chest pain who performed exercise stress testing found significant improvement in symptoms and pulmonary function after bronchodilator use. In a study of pediatric patients with chest pain referred to a cardiac stress laboratory, 26% had abnormal pulmonary function testing, despite only 19% having a known history of asthma.

Pneumonia has been reported in 2% to 5% of ED patients with chest pain. In patients with sickle cell disease, chest pain accompanied by lower respiratory tract symptoms and an infiltrate on radiograph should be managed as an acute chest crisis (Fig. 3). Pleurodynia (historically “devil’s grip”) is characterized by fever and pleuritic chest pain. It may occur in localized epidemics and is often associated with coxsackievirus B1, although other enteroviruses have also been implicated. Pleuritis and pleural effusions are uncommon causes of pleuritic chest pain but can be seen in children with infections and such conditions as collagen vascular disease, malignancy, and familial Mediterranean fever.
Pneumothorax and pneumomediastinum may account for up to 3% of cases of chest pain presenting to a pediatric ED (Fig. 4).\(^1,6\) Chest pain is present in nearly all children with pneumothorax.\(^2,3\) In a study by Lee and colleagues,\(^2,4\) however, only 68% of children with pneumomediastinum had chest pain; neck pain (44%) and sore throat (33%) were also present. These entities are typically found in children with asthma; bronchiolitis; or other lower airway diseases, such as cystic fibrosis. Marfan syndrome is also a risk factor. Previously healthy children may present with a spontaneous pneumothorax from a ruptured bleb. Pneumothorax or pneumomediastinum may also occur after an episode of choking or aspiration or after inhalation of cocaine or marijuana.\(^2,5\) Pneumothorax should be suspected in a child with risk factors or unexplained dyspnea, tachypnea, or decreased breath sounds. Pneumomediastinum should be suspected if subcutaneous emphysema or Hamman sign (precordial crackles that correlate with the heartbeat) are present.

Pulmonary embolism (PE) is rare in healthy children but may be seen in the presence of risk factors, such as a central venous catheter; malignancy; coagulopathy; nephrotic syndrome; major surgery (especially cardiac); trauma; or sepsis.\(^2,6,2,7\) Nearly all children identified as having a PE are symptomatic. In a study of pediatric trauma patients with PE, 73% had dyspnea, 70% had tachypnea, 51% had rales, and 66% had pleuritic chest pain.\(^2,8\) Typical electrocardiograms (ECGs) findings in PE are presented in Table 2.

**Gastrointestinal**

Gastrointestinal causes for chest pain have been identified in up to 8% of patients presenting to pediatric EDs.\(^9\) Gastroesophageal reflux disease is the most frequently
diagnosed gastrointestinal cause for chest pain in children, occurring in 3% of patients in one study.\textsuperscript{9} Because gastroesophageal reflux disease is usually a clinical diagnosis, however, it is difficult to estimate the true contribution of gastroesophageal reflux disease in chest pain. In an uncontrolled study by Berezin and colleagues,\textsuperscript{29} 27 children with idiopathic chest pain and no symptoms of reflux underwent endoscopy. Sixteen were found to have esophagitis, four had gastritis, and one had abnormal

\textbf{Fig. 4.} Pneumothorax accentuated with expiration. (A) In this patient with chest pain, on a typical inspiration chest radiograph, no pneumothorax is identified. (B) With expiration, the superior lung margin (arrows) becomes smaller, but the pneumothorax stays the same size; relatively it appears bigger and can be easier to see. (From Mettler FA. Chest. In: Essentials of radiology. 2nd edition. Philadelphia: Saunders/Elsevier; 2005. p. 102; with permission.)
manometry. In another study, patients who presented to a cardiology clinic with both chest pain and epigastric tenderness had a high prevalence of esophagitis or gastritis on endoscopy (41 of 44 patients). Most of those who received treatment had resolution of symptoms.  

Structural abnormalities, inflammatory or motility disorders, and foreign bodies involving the esophagus or stomach may also produce chest pain in children. Eosinophilic esophagitis is an increasingly recognized disorder in children, which may cause chest pain because of esophageal inflammation, dysmotility, and reflux. Bulimia nervosa is another cause of esophagitis in the pediatric population, particularly in adolescent girls. Coins and other objects lodged in the esophagus typically present with chest pain that is often accompanied by drooling and dysphagia. Pill esophagitis is chemical irritation of the esophageal mucosa from certain medications, particularly iron preparations, tetracyclines, and nonsteroidal anti-inflammatory agents. The classic pediatric presentation of pill esophagitis is the adolescent patient who ingests a tetracycline antibiotic capsule with too little water, especially before going to bed. Chest pain, dysphagia, and occasionally hemoptysis are present.

Esophageal rupture from nontraumatic causes (Boerhaave syndrome) has also been described as a cause of chest pain in children. The perforation of the esophagus is believed to be caused by increased pressure transmitted from retching or vomiting but has also been associated with coughing, asthma, defecation, seizures, childbirth, nose-blowing, and immunosuppression. Patients may present with chest pain, vomiting, and subcutaneous emphysema (Mackler triad) and hematemesis, respiratory distress, and hemorrhagic or septic shock. Chest radiography may reveal pneumomediastinum, pneumothorax, mediastinal widening, or pleural effusion, and diagnosis is confirmed by contrast esophagram or CT scan.

**Cardiac**

Cardiovascular disease is identified in only 2% to 5% of patients seen in pediatric EDs for chest pain; however, it is often the leading concern of patients and families.
seeking care. In one study 56% of adolescents feared chest pain was cardiac. The presence of fever, dyspnea, palpitations, pallor, or abnormal cardiac auscultation has been found to be statistically significantly related to a cardiac etiology. Outside of North America, the frequency of cardiovascular etiologies for pediatric chest pain may be higher. A Turkish study of pediatric patients with chest pain referred for cardiology evaluation, of whom 9% had a known cardiac diagnosis, determined that 42.5% had cardiac disease, including 14% with rheumatic heart disease and 12% with evidence of dysrhythmia.

In children, acute myocardial infarction (AMI) has been described in association with coronary artery anomalies; congenital heart disease; Kawasaki disease; familial hypercholesterolemia; previous heart transplant; sickle cell disease; cardiac myxoma; hypercoagulable states; substance abuse; and certain metabolic conditions, such as homocystinuria and mucopolysaccharidosis. Most information on AMI in the pediatric population comes from case reports. A population database study, however, estimated the risk in adolescents as 6.6 per 1 million patient-years. Of the 123 patients between 13 and 18 years who had suffered AMI, 23% had a history of substance abuse (cocaine 41%, amphetamines 31%, cannabis 10%, multiple substances 10%). AMI was more common in males (OR = 3) and smokers (OR = 4.1) compared with patients of the same age admitted for other reasons. Pre-existing conditions (systemic lupus erythematosus and hyperlipidemia) were found in 2.5% of patients. The most common location of infarction was subendocardial, accounting for 40% of infarcts. A study of all myocardial infarction cases in a single pediatric institution in an 11-year period identified nine cases. The average age was 15.5 years; eight were boys. The one female patient was 4 months postpartum. All were previously healthy except for migraine headaches in two and attention deficit disorder in one patient who was taking methylphenidate. Drug screens, hypercoagulability studies (performed in seven of nine patients), and lipid profiles were negative in all patients. All presented with chest pain and responded to nitroglycerin. Abnormal ECGs were found in eight patients, and all had elevated cardiac enzymes. Of interest, all nine patients had normal coronary angiograms, and all had normal exercise stress testing after hospitalization. The authors concluded that coronary spasm was the most likely cause for the ischemia. Intracoronary thrombus can also be a cause of AMI in children. Risk factors include hypercoagulability and emboli from endocarditis or prosthetic valves.

Congenital coronary artery abnormalities have also been associated with an increased risk of myocardial infarction in children. Anomalous origin of the left coronary artery from the pulmonary artery is often identified in early infancy when the pulmonary artery pressure declines, usually around 2 to 3 months of age. Patients may present with crying, poor feeding, and signs of congestive heart failure. Anomalous origin of the left coronary artery from the pulmonary artery may also present in later childhood and may present with anginal pain. The typical ECG pattern is that of an anterolateral infarction with large and wide Q waves, ST changes, and T wave inversion in leads I, aVL, V5, and V6. Other coronary artery abnormalities, including anomalous origin of the left main coronary artery or right coronary artery from the contralateral sinus of Valsalva and hypoplastic coronary arteries, may also present in childhood. Numerous case reports have been published of children, especially young athletes, who experience chest pain, myocardial ischemia, or sudden cardiac death because of the presence of coronary artery anomalies.

Kawasaki disease has been associated with myocardial infarction both in the acute and subacute phases and as a long-term consequence. Acute cardiac complications of Kawasaki disease include coronary artery aneurysms, myocarditis, pericarditis, and
arrhythmias. Aneurysms, which generally occur 10 days to 4 weeks after the onset of symptoms, are the most frequent complication and occur in 20% to 25% of untreated patients\textsuperscript{42} and 5% of patients treated with intravenous immunoglobulin. Infarction can occur during the acute phase caused by intimal proliferative inflammation or during resolution caused by obstruction, stenosis, or irregularities of the arterial wall. Successful use of intravenous immunoglobulin to treat Kawasaki disease was first reported in 1983.\textsuperscript{43} In a cohort of patients with acute Kawasaki disease in the preintravenous immunoglobulin era, 25% had coronary aneurysms. Forty-nine percent experienced regression of their aneurysms within 1 to 2 years after diagnosis; however, 10 to 21 years after diagnosis, 19% were found to have coronary stenosis (5% of the original cohort), and 8% had experienced myocardial infarction (2% of original cohort).\textsuperscript{44} Presumably children with a missed diagnosis of Kawasaki disease who do not receive intravenous immunoglobulin would have a similar prognosis for stenosis or infarction.

Myocardial ischemia has also been reported in children with sickle cell disease. In a study of pediatric sickle cell patients with a history of chest pain, heart failure, abnormal ECG, left ventricular dilation, or hypokinetic left ventricle, 64% had perfusion defects on thallium-201 single-photon emission CT. Three children who were started on hydroxyurea therapy underwent repeat single-photon emission CT, and all showed improvement. No occlusion of coronary arteries was found, suggesting that pathology of the microcirculation is responsible for the defects.\textsuperscript{45}

Myocarditis is a rare but serious cause of chest pain in children. In one study, 56% of children with myocarditis ages 10 to 17 years presented with symptoms of chest pain or palpitations, and 25% presented with lightheadedness, syncope, or seizure.\textsuperscript{46} In children less than 10 years old respiratory presentations were more common, accounting for 47% of cases. Common physical findings include tachypnea (60%–68%); hepatomegaly (36%–50%); and tachycardia (32%–58%).\textsuperscript{46,47} ECG abnormalities are detected in 93% to 100%, and chest radiography is abnormal in 55% to 90%. Laboratory abnormalities include elevated troponin (54%); elevated creatine kinase (73%); elevated erythrocyte sedimentation rate (38%–57%); and elevated aspartate aminotransferase (85%). Pericarditis and endocarditis can also be associated with chest pain. Typical ECG findings in myocarditis and pericarditis are presented in Table 2, and representative ECG examples are presented in Figs. 5 and 6. Partial or complete congenital absence of the pericardium is rare but may produce chest pain in children. In a series of 10 patients, the median age of presentation was 21 years (the youngest was 2 years of age). All except the youngest child presented with chest pain. The ECG findings include sinus tachycardia, ST-segment changes (elevation and depression), T-wave changes, and incomplete left bundle-branch block. (From Brady WJ, Ferguson JD, Ullman EA. Myocarditis: emergency department recognition and management. Emerg Med Clin North Am 2004;22:865–85; with permission.)
pain, which was typically stabbing and nonexertional, and often could be induced or relieved by postural changes. Chest radiographs of 7 of the 10 showed displacement of the cardiac silhouette to the left with loss of the right heart border.48

Arrhythmias are one of the more common causes of cardiac-related chest pain in children. ED studies by Massin and coworkers4 and Lin and coworkers6 reported arrhythmias in approximately 2% of children presenting with chest pain. Tachyarrhythmias are associated with decrease in duration of diastole and may cause chest pain because of a reduction in myocardial blood flow. In one study, 14% of children over the age of 1 year presenting with supraventricular tachycardia reported having chest pain.49 Structural heart disease may also cause chest pain. Left ventricular outflow obstruction caused by aortic stenosis or hypertrophic cardiomyopathy causes pain that is typically exertional and is caused by subendocardial ischemia. In hypertrophic cardiomyopathy, the physical examination is characterized by a harsh systolic ejection murmur that is heard best at the apex and lower left sternal border. An increase in the intensity of the murmur is seen when the patient assumes an upright posture from a squatting, sitting, or supine position, and with the Valsalva maneuver. A decrease in intensity is heard after going from a standing to a sitting or squatting position, or with passive elevation of the legs. The decrease in intensity occurs when increased ventricular filling increases the size of the outflow tract and decreases the gradient across the obstruction. Mitral valve prolapse has been reported in pediatric patients with chest pain. The role of mitral valve prolapse, however, in causing chest pain is unclear. In adults, the prevalence of chest pain in patients with mitral valve prolapse is similar to that in the general population.50 It has been hypothesized, however, that severe mitral valve prolapse could cause pain because of papillary muscle dysfunction or ischemia.

Children with severe pulmonary stenosis or pulmonary hypertension are at risk for myocardial ischemia. Pain often occurs with exercise. The murmur of pulmonary stenosis is audible at the upper left sternal border and may radiate to the ipsilateral axilla and back. Pulmonary arterial hypertension is a serious and often fatal condition that may be initially difficult to diagnose and rarely may present with chest pain. It may be idiopathic or secondary to congenital heart disease; pulmonary disease; or systemic disease, such as collagen vascular disease. In a study of 63 pediatric patients with an average age at presentation of 5.8 years, the most common symptoms were exercise-induced dyspnea (98%); dyspnea at rest (25%); chest pain (3%); and syncope (13%).51
Aortic aneurysm and dissection have been described both in healthy pediatric patients and in those with known risk factors. Approximately 3.5% of aortic dissections occur in children under 19 years of age,\(^5\) and aortic dissection is responsible for approximately 1 in 3000 pediatric deaths.\(^5\) Risk factors include congenital anomalies, such as coarctation of the aorta and aortic valvular stenosis. Other causes include Marfan syndrome (see Fig. 2), Ehler-Danlos syndrome, Turner syndrome (Fig. 7), trauma, cocaine use, and weight lifting. A study of patients under 28 years of age with Marfan syndrome found a prevalence of aortic root dilation of 83%. Half of the patients studied began to develop aortic root dilation by 10 years of age.\(^5\) Before the development of preventative medical and surgical therapy, life expectancy for Marfan patients was greatly reduced and aortic dissection and other

![Fig. 7. A patient with Turner syndrome and typical features including webbed neck (A), cubitus valgus (B), ankle edema (C), and short stature and widely spaced nipples. (From Gawlik A, Malecka-Tendera E. Hormonal therapy in a patient with delayed diagnosis of Turner's syndrome. Nat Clin Pract Endocrinol Metab 2008;4(3):173–7; with permission.)](image)
cardiovascular complications were responsible for over 90% of deaths. Turner syndrome is another cause of aortic dissection in children. Approximately half of girls with Turner syndrome have underlying cardiac defects, most commonly bicuspid aortic valve and aortic coarctation. A study describing 85 cases of aortic dissection in Turner syndrome reported an average age of 30.7 (range, 4–64) years. Fifteen percent had underlying hypertension, 30% had congenital heart disease, and 34% had both. In 11% of the cases, however, no risk factors were identified. The prevalence of aortic root dilation in Turner syndrome is approximately 6% of patients. Pectus excavatum may be associated with aortic root dilation, even when other stigmata of Marfan syndrome are absent. In one study, patients with isolated pectus excavatum without a suspected connective tissue disorder were evaluated with echocardiograms. The patients with pectus excavatum had a significantly higher prevalence of aortic root dilation than controls. Several patients underwent genetic testing and were diagnosed with Marfan syndrome despite lacking the usual phenotypic appearance. The pain of aortic dissection is often described as severe and knifelike or tearing. It tends to be located in the anterior or posterior chest, neck, jaw, or shoulder. The chest radiograph is likely to show mediastinal widening, pleural effusion, abnormal aortic contour, or cardiomegaly. Diagnosis can be confirmed by echocardiography.

Psychiatric

In studies based in a pediatric ED, chest pain has been attributed to a psychiatric cause in approximately 5% to 9% of cases. One study found adolescents to be 2.5 times as likely to have a psychogenic cause for their chest pain compared with younger children. A history of a stressful event, such as death or hospitalization in the family, family separation, or school changes, has been reported in 31% of adolescents with chest pain. Other reported psychiatric causes include anxiety disorders and depression. A study of 27 children referred to a cardiology clinic for chest pain found that 56% of children who did not have a cardiac etiology were given a Diagnostic and Statistical Manual-IV diagnosis of an anxiety disorder, including panic disorder (33%), generalized anxiety disorder (26%), social phobia (19%), and other specific phobia (19%). One child had major depression. A study of 36 children diagnosed with psychogenic chest pain found that 55% had other somatic complaints and 30% had sleep disturbances.

APPROACH TO THE PEDIATRIC PATIENT WITH CHEST PAIN

The primary goals in evaluation of a child with chest pain are to rule out cardiac and other serious causes and to classify the origin of the pain. A thorough history and physical examination are often sufficient to accomplish these goals. In cases in which the cause remains unclear or if concerning features are identified, further evaluation and sometimes referral are warranted.

History

A complete history is perhaps the most important part of the assessment of a child with chest pain. The history should begin with the onset of pain, with the knowledge that acute pain is more likely to be caused by an identifiable organic cause. One study reported that 31% of children stated that the pain had awakened them from sleep; this was shown to be associated with a higher likelihood of an organic cause. The family should be asked about events that may have precipitated the pain, such as exercise, trauma, eating, potential foreign body ingestion, or psychologic stressors. In many
cases the child’s description of the pain does not help in identifying the etiology, with most children indicating an anterior location to their pain, and 90% of children characterizing their pain as moderate to severe. Descriptions of pain being sharp, constant, or radiating can be nonspecific and have not been found to be significantly associated with cardiorespiratory causes. Most studies of pediatric chest pain are small, however, and include few patients with serious organic causes, so the studies may not be powered to demonstrate such an association. Characteristic pain patterns have been described with certain conditions. Chest wall pain is often localized and sharp, and exacerbated by moving or taking a deep breath. Pleural or pulmonary pain may also be accentuated with inspiration or cough, although pain is less likely to be well-localized than musculoskeletal pain, and less likely to be reproduced with palpation. Pleuritic pain is often sharp and superficial, whereas pulmonary pain, such as that associated with asthma, is more likely to be diffuse and deep. A description of midsternal or precordial pain that worsens after eating or when lying down may be esophageal. The classic description of cardiac pain is that of pressure, crushing, or a squeezing sensation that may radiate to the neck or arm. There is little information on whether this classic description is typical in pediatric cases. Pain that is mitigated by sitting up and leaning forward may be caused by pericarditis. The presence of blood or other irritants in the peritoneal cavity may cause referred chest or shoulder pain (Kehr sign). Psychogenic pain is expected to be vague, poorly localized, varying in location, and possibly associated with other somatic complaints.

Pain associated with palpitations or syncope should be considered a possible indicator of cardiac disease, and pain associated with exertion could be either cardiac or related to a respiratory cause, such as exercise-induced asthma. A history of fever is likely to be reported with pneumonia, but may also be present with myocarditis, pericarditis, or pleural effusion. A history of drooling or reluctance to swallow may be present in a child with an esophageal foreign body. The presence of joint pain or rash may suggest collagen vascular disease. The patient and family should be asked about emotional stressors or presence of anxiety or depression. Adolescents should be asked about use of medications, especially oral contraceptives and pills that have been associated with esophagitis, such as tetracycline. They should also be interviewed privately and asked about use of illicit substances, such as cocaine or marijuana. A complete review of systems is beneficial in identifying relevant information that may not be volunteered by the patient.

In taking the past medical history, certain illnesses should be asked about directly, such as Kawasaki disease, asthma, sickle cell disease, diabetes, or connective tissue disorders, such as Marfan syndrome. The family history should focus on history of unexplained or sudden death, serious underlying conditions, and whether family members have a history of chest pain or heart disease. Although a family history of heart disease may help to identify a child at risk of the same, it has actually been demonstrated that a family history of heart disease or chest pain is associated with a higher likelihood of nonorganic disease.

It should be recognized that the symptom of chest pain is often very worrisome for children and their families. In a study of adolescents seen in a pediatric chest pain, 61% reported that they did not know what was causing their pain, but 56% were afraid of heart disease or a heart attack, and 12% were worried they had cancer. It is important to recognize this fear and address patients’ and families’ concerns during the assessment. Children who present to EDs with chest pain are likely to have missed school, with estimates ranging from 30% to 41%. Families should be specifically asked about school absenteeism so that recommendations for returning to school can be given.
Physical Examination

Physical examination abnormalities have been reported in 39% to 49% of pediatric ED patients with chest pain.2,6 The examination should include a full set of vital signs and an assessment of the general appearance, noting level of alertness, color, and presence of distress or anxiety. Fever may suggest the presence of pneumonia or another infectious or inflammatory condition, and tachycardia or tachypnea suggests the possibility of cardiac, respiratory, or other serious organic etiology. The chest wall should be inspected for signs of trauma, asymmetry, pectus carinatum or excavatum, or costosternal swelling. Tenderness of the chest wall or costochondral and costosternal junctions has been reported in 24% to 54%2,6 of pediatric ED patients with chest pain and suggests a musculoskeletal etiology.

After chest wall tenderness, abnormal lung auscultation is the second most commonly identified abnormality in the examination of pediatric chest pain patients, occurring in approximately 13% of patients seen in an ED setting.6 Auscultation of the lungs for crackles, wheezes, and decreased breath sounds may suggest pneumonia, asthma, or pneumothorax. Pneumomediastinum may cause subcutaneous emphysema, which can be detected by crepitus on palpation of the supraclavicular area or neck. The heart should be auscultated to identify the presence of an irregular rhythm, murmur, rub, gallop, or muffled heart sounds. The rub of pericardial effusion is best appreciated when the patient is leaning forward. If a large effusion is present, the patient may have distant heart sounds, jugular venous distention, narrow pulse pressure, and increased pulsus paradoxus. Patients with myocarditis may have tachycardia, gallop rhythm, displaced point of maximal impulse, or a murmur of mitral regurgitation. If coarctation or aortic dissection is suspected, four-limb blood pressures should be obtained.

Palpation of the abdomen may reveal epigastric tenderness in patients with a gastrointestinal cause for their pain. In a study of children referred to a pediatric cardiology clinic in Iran for evaluation of their chest pain, 33% had epigastric tenderness, and of these, 93% had positive findings on endoscopy.30 If a history of trauma is present, the abdomen should be assessed from tenderness and peritoneal signs. Hepatomegaly may be a sign of heart failure. The skin and extremities should be examined for evidence of trauma, chronic disease, or dysmorphology. Xanthomas on the hands, elbows, knees, and buttocks are characteristic of familial dyslipidemia. Range of motion and resistance testing of the upper extremities may reveal a musculoskeletal source for pain, such as muscle strain or delayed-onset muscle soreness. Special attention should be given to identifying findings associated with Marfan syndrome or other connective tissue disorders, because these conditions carry an increased likelihood of serious pathology.

Investigations

If concern for serious etiology is raised by the history or physical examination, or if pain is severe or disruptive to usual activities, further investigation is warranted (Table 3). Although it may be difficult to identify a precise cause for the pain, it is important to exclude life-threatening pathology. A chest radiograph should be obtained if there is unexplained pain of acute onset, respiratory distress, abnormal pulmonary or cardiac auscultation, fever, significant cough, history of drooling or foreign body ingestion, or significant underlying medical conditions. In a study by Rowe and colleagues,1 chest radiography was obtained in 50% of patients presenting to an ED with chest pain. Of 18 positive results, 15 were infiltrates, and there were two cases of pneumomediastinum and one pneumothorax. Lin and colleagues6 described 103 children who visited
an ED in Taiwan for chest pain; chest radiographs were obtained in 98%. Abnormal-
ities were found in 28% and were reported as pulmonary infiltrates (13%); hyperinfla-
tion (7%); pneumonia (5%); and pneumothorax (3%). A 12-lead ECG should be
obtained if there is pain or syncope with exertion, abnormal cardiac auscultation,
a clinical suspicion for myocarditis or pericarditis, or serious underlying medical condi-
tions that carry an increased risk of cardiac disease. The ECG should be evaluated
with age-appropriate criteria for evidence of arrhythmia, conduction delay, preexita-
tion, hypertrophy, or ischemia. In a study by Selbst and colleagues,5 191 children
with ill-defined or potentially cardiac-related chest pain received ECGs. Of these,
16% were abnormal. All abnormalities were minor or were previously known, however,
except in four patients. Of these, three had arrhythmias identified by physical exami-
nation, and one child with systemic lupus erythematosus who was febrile had changes
consistent with pericarditis. In a study by Rowe and colleagues,1 ECG was performed
in 18% of patients presenting with chest pain. Of ECGs obtained, six (10%) were posi-
tive, with four being previously known or unrelated findings, one patient with ST
segment and T-wave changes found to have myocarditis, and one with a history of
palpitations found to have Wolff-Parkinson-White syndrome.1 Lin and colleagues6
described 103 children who visited an ED in Taiwan for chest pain; ECGs were
obtained in 85%. Four (4.6%) showed abnormalities, including first-degree AV block,
second-degree AV block, premature ventricular contraction, and Wolff-Parkinson-
White syndrome.

Laboratory investigations are rarely necessary in the evaluation of children with
chest pain, but may be useful when certain conditions are suspected. A complete
blood count may be obtained for suspected infectious causes or in a patient with
an underlying condition, such as sickle cell disease. In a patient with suspected
cardiac ischemia or myocarditis, cardiac enzymes and aspartate aminotransferase

<table>
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<tr>
<th>Workup</th>
<th>History/Symptom</th>
<th>Sign</th>
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<tbody>
<tr>
<td>Chest radiograph</td>
<td>Fever</td>
<td>Fever, tachypnea, rales, distress</td>
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<tr>
<td></td>
<td>Cough</td>
<td>Ill-appearing</td>
</tr>
<tr>
<td></td>
<td>Shortness of breath</td>
<td>Evidence of significant trauma</td>
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<td></td>
<td>History of trauma</td>
<td>Unexplained tachycardia</td>
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<td></td>
<td>Pain awakening from sleep</td>
<td>Pathologic heart auscultitation</td>
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<td></td>
<td>History of drug use (eg, cocaine)</td>
<td>Decreased breath sounds</td>
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<td>Association with exercise</td>
<td>Subcutaneous air/crepitus</td>
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<td></td>
<td>Acute onset of severe pain</td>
<td>Tall, thin habitus, pes excavatum or carinatum</td>
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<td>Underlying medical problems (eg, Marfan syndrome, lupus, Kawasaki disease)</td>
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<td></td>
<td>Foreign body ingestion</td>
<td>Drooling</td>
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<tr>
<td>Electrocardiogram</td>
<td>Shortness of breath</td>
<td>Pathologic heart auscultitation</td>
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<td>Association with exercise</td>
<td>Unexplained tachycardia</td>
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<td>Association with syncope</td>
<td>Unexplained respiratory distress</td>
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<td></td>
<td>Palpitations</td>
<td>Diminished perfusion</td>
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<td></td>
<td>History of drug use</td>
<td>Decreased pulses</td>
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<td></td>
<td>Precardial trauma</td>
<td>Evidence of trauma</td>
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<tr>
<td></td>
<td>Personal or family history of heart disease</td>
<td>Ill-appearing</td>
</tr>
</tbody>
</table>

may be useful. Troponin is elevated in 54% of pediatric patients with myocarditis and may also be elevated with pericarditis. D-dimer may be obtained if PE is suspected, although there are limited data about D-dimer test performance in pediatrics. One study found that children with PE were as likely as control patients with similar risk factors to have a D-dimer value within the normal range, and in another study D-dimer was normal in 40% of pediatric patients with PE. Other tests that are rarely necessary but may be useful include a drug screen when there is a concern about possible substance abuse, Holter monitor if arrhythmia is suspected, exercise stress test or pulmonary function test for unexplained exertional pain, and endoscopy for possible gastrointestinal sources of pain.

**Treatment and Referral**

If musculoskeletal pain is identified, analgesics (ibuprofen or acetaminophen) should be offered. Patients with infectious, respiratory, or cardiac sources for their pain need treatment directed at their underlying condition. If esophagitis or gastritis is suspected, a therapeutic trial of an H2 blocker or proton pump inhibitor can be initiated. Patients with possible exercise-induced asthma may be offered a trial of a β-agonist if more serious respiratory or cardiac disorders are not suspected. For patients with idiopathic or undiagnosed respiratory pain, analgesics and close follow-up are appropriate.

Patients being seen in a nonhospital setting should be referred to an ED if experiencing significant distress, if trauma has occurred, or if serious pathology cannot be ruled out. Consultation or referral to a cardiologist may be appropriate if there is exertional pain; history of palpitations, syncope, or presyncope; abnormal cardiac auscultation, chest radiograph, or ECG; concerning family history; or significant recurrent pain of unknown etiology. Referral to a gastroenterologist or pulmonologist may be considered for specific concerns. If significant anxiety, depression, or emotional stress is present, the patient should be referred to a psychiatrist, psychologist, or primary care provider with experience in mental health issues.

**SUMMARY**

Chest pain is common in children seen in EDs, ambulatory clinics, and cardiology clinics. Although most children have a benign cause for their pain, some have serious and life-threatening conditions. The symptom must be carefully evaluated before reassurance and supportive care are offered. Because serious causes of chest pain are uncommon and not many prospective studies are available, it is difficult to develop evidence-based guidelines for evaluation. The clinician evaluating a child with chest pain should keep in mind the broad differential diagnosis and pursue further investigation when the history and physical examination suggest the possibility of serious causes.

**REFERENCES**


