Pediatric Chest Pain

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Education Gaps

Lack of familiarity with a systems-based approach and differential diagnosis of chest pain in pediatric patients may lead to inappropriate use of electrocardiography and echocardiography. Clinicians should be aware of the various causes of chest pain in children and the current guidelines and algorithms being used to determine which children are at risk for cardiac chest pain and should have further testing performed.

Objectives After completing this article, readers should be able to:

- 1. Be familiar with a system-based differential diagnosis of pediatric chest pain and specific diagnosis within each system.
- Understand the initial approach to assessing chest pain in a pediatric patient.
- 3. Identify when echocardiography should be used to further assess chest pain.
- 4. Understand the appropriate use criteria for echocardiography as clinical practice guidelines as well as the use of standardized clinical assessment and management plans.
- 5. Be familiar with specific questions to identify patients at risk for sudden cardiac death.

INTRODUCTION

Chest pain is a common complaint in the pediatric population, encountered in both outpatient settings and emergency and urgent care settings. There are numerous causes of chest pain in children, with most studies ultimately not determining an etiology (I) or attributing the pain to musculoskeletal causes. The complaint may be anxiety provoking for patients, their families, and care providers alike because a cardiac cause is often feared (2)(3)(4); however, the incidence of a cardiac-related cause for chest pain in the pediatric population is exceedingly low, ranging from 0.2% to 1% of cases. (5)(6)(7) Most etiologies of chest pain in pediatric patients can be grouped into musculoskeletal, pulmonary, gastroenterologic, psychogenic, or cardiac causes. Despite the variety of causes, the initial evaluation for chest pain in pediatric patients is fairly uniform, relying

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ABBREVIATIONS

ECG	electrocardiogram
NSAID	nonsteroidal anti-inflammatory
	drug
SCAMP	standardized clinical assessment
	and management plan

heavily on a thorough history and physical examination, and occasionally necessitating an electrocardiogram (ECG) if cardiac etiologies remain on the differential diagnosis. Less frequently, when abnormalities are found on history and physical examination or on the ECG, obtaining an echocardiogram may be required, as well as cardiology consultation and/or follow-up. Several recently published guidelines and standardized clinical assessment and management plans have been proposed during the past decade that aim to reduce the overuse of unnecessary testing and evaluation while maximizing patient care delivery. (6)

ETIOLOGIES

Most chest pain is ultimately of unknown origin, with studies failing to find a clear cause of the patient's complaints. (2)(5) A large-scale systematic review of studies from 1950 to 2010 compiled the causes of chest pain in pediatric patients, finding idiopathic chest pain in 35.63%, followed by musculoskeletal causes in 19.75%, psychological causes in 16.31%, gastrointestinal causes in 10.36%, cardiac causes in 9.57%, and respiratory causes in 8.39%. (8) This review showed a higher-than-previouslydescribed proportion of chest pain due to cardiac causes; however, the authors note that this may be attributed to some studies taken from pediatric cardiology departments, in which cardiovascular disease is more likely. Table 1 shows a summary of the major causes of chest pain in children by system, specific diagnosis within each system, differentiating features of each, and general management, all of which are described in greater detail herein.

Musculoskeletal Causes

Costochondritis. Costochondritis is defined as a self-limited inflammation of the costochondral junctions of the ribs at the sternum. (9) The condition may also be referred to as costosternal syndrome, parasternal chondrodynia, or anterior chest wall syndrome. (9) The pain from costochondritis is reproduced by palpation of the affected joints and may have radiation to the chest wall. (9) Most often there is more than I costochondral junction affected, and those most frequently affected include the second to fifth junctions. (10) Usually diagnosis requires only a history or physical finding of reproducible pain on palpation of the costal cartilage. (9) Treatment is largely supportive and comprises nonsteroidal anti-inflammatory drugs (NSAIDs) and avoid-ance of activities that lead to chest musculature overuse. (9)(II)

Tietze Syndrome. Tietze syndrome may be thought of as a variant of costochondritis because the process of inflammation is similar; however, the differentiating factor is that in Tietze syndrome there is visible swelling at the costochondral junction. (9) In addition, Tietze syndrome usually presents with only I joint affected, and the joint in question is usually at the level of the second or, less commonly, third costochondral junction. (IO) Treatment is similar to that for costochondritis: using NSAIDs and promoting rest. (II)

Slipping Rib Syndrome. A less frequent cause of chest or upper abdominal pain, slipping rib syndrome occurs due to costal cartilage subluxation and resultant intercostal nerve impingement. (12) The disorder is generally caused by trauma of the costal cartilages of the eighth, ninth, and tenth ribs; ribs that are not directly attached to the sternum but rather to each other with a shared cartilaginous connection to the sternum. (13) The condition is thought to occur due to inadequacy or rupture of the interchondral fibrous attachment of the anterior ribs. (12) The symptoms experienced include pain and a perception of a slipping movement of the ribs, (13) giving this condition its name. Treatment ranges from minimal intervention, including avoidance of triggers such as certain postures or movements, to strapping the ribs or injecting local anesthetic nerve blocks. (12)

Precordial Catch Syndrome. Precordial catch syndrome presents as a well-localized, sharp, stabbing, or needlelike pain. (14) The pain usually occurs at rest without provocation and never during sleep. (14) The pain is characteristically worse when the patient takes a deep breath, which leads to deliberate shallow breathing during the episode of pain, which generally lasts 30 seconds to 3 minutes. (14) Rarely there may be underlying structural defects such as an atrial septal defect or pulmonary valve stenosis in affected children. (14) The pathophysiology causing the pain remains unclear, but the origin of the pain is reassuringly neither cardiac nor pericardial in origin. (14) Treatment involves reassurance and explaining the benign nature of the condition, despite the severe pain that may be experienced, without the requirement for medication. (11)

Muscle Strain. Another frequent cause of musculoskeletal chest pain occurs due to muscle strain, with common eliciting factors including trauma and overuse. (15) Commonly implicated muscles include the intercostal, pectoralis, internal and external oblique, and serratus anterior muscles. (15) A history of athletic endeavor is common, including weight lifting and rowing, but it may also occur in the context of coughing. (15) Younger children experience more injury due to trauma than do adolescents, who are

TABLE 1. Causes, Features, and Management of Chest Pain in Pediatric Patients

CAUSE	FEATURES	MANAGEMENT
Musculoskeletal system		
Costochondritis	Often >1 costochondral junction, typically involving second to fifth rib junctions Reproducible pain on palpation	Largely supportive management with NSAIDs and avoidance of pain-provoking activity
Tietze syndrome	Often only 1 costochondral junction affected at the level of the second or third rib junction Visible swelling with reproducible pain on palpation	Largely supportive management with NSAIDs and avoidance of pain-provoking activity
Slipping rib syndrome	Often involving costal cartilages of the eighth, ninth, or tenth ribs Perceived slipping sensation of the rib	Initially trigger avoidance; if necessary, strapping the affected ribs or local nerve blocks
Precordial catch syndrome	Well-localized and sharp pain occurs at rest Pain abruptly subsides	Reassurance with explanation of benign nature of the condition
Muscle strain	Due to trauma or overuse, especially in athletes or those with chronic cough	Rest, stretching as tolerated, NSAIDs
Trauma	May have chest wall bruising but less likely to have fractures compared with adults	Assess for associated injuries, with rib fracture in young children raising abuse concern Assess for intrathoracic trauma, especially if pneumothorax or hemothorax at presentation
Pulmonology system		
Asthma	Pain most often with exercise-induced variants or perception of "tightness" as pain	Bronchodilator treatment Encourage gradual warm-up to lessen severity
Respiratory infections	Suspect with focal findings on auscultation Often present with cough in the context of a febrile illness	Consider chest radiography if presenting with febrile illness and associated symptoms Treatment with antibiotic drugs when bacterial cause likely, often with pleural irritation
Pleurodynia	Sharp and spasmodic chest pain Presents with fever due to infectious etiology; coxsackie B most common	Self-resolving over weeks; analgesics to minimize pain when necessary
Gastrointestinal system		
Gastroesophageal reflux	Commonly presents epigastric pain and regurgitation May have associated burning sensation	Acid suppression using either histamine receptor blockade or proton pump inhibitors
Esophagitis	Due to reflux, allergic, infectious, or iatrogenic causes due to medications Treatment of underlying condition; controlling reflux above, treating infectious causes and changing medications if found to be causative Eosinophilic esophagitis treated with elimination dief and topical corticosteroids	
Foreign body ingestion	Common in children <5 y old presenting with sudden- onset symptoms May have drooling or reluctance to swallow	Chest radiography including posterior-anterior and lateral imaging Endoscopy may ultimately be needed both for diagnosis and treatment in removal of foreign body
Cardiac system		
Pericarditis	Often presents with fever and is typically retrosternal Patients often sit upright and lean forward to alleviate pain	Obtain an ECG to assess for described findings NSAIDs or aspirin in older children as analgesia and to control inflammation
	Friction rub on auscultation and widespread ST-segment elevation and PR depression on ECG Often viral or idiopathic cause	Corticosteroids or colchicine also used occasionally

Continued

TABLE 1. (Continued)

CAUSE	FEATURES	MANAGEMENT
Myocarditis	May present with nonspecific symptoms mimicking respiratory disease or sepsis Often viral causes, including group B coxsackie as well as parvovirus B19 and human herpesvirus 6	Low threshold to obtain ECG, especially when respiratory diagnosis is uncertain Treatment relies on supportive therapy for left ventricular function, including pharmacologic therapies for circulatory support or occasionally mechanical support
Mitral valve prolapse	Associated with a mid-to-late systolic click with a high- pitched late systolic murmur	No specific treatment Antibiotic prophylaxis not required Occasional symptomatic treatment with β -blockers Severe regurgitation may require surgical intervention
Arrhythmia	 Supraventricular tachycardia Most common dysrhythmia, tracing with narrow-complex QRS Ventricular tachycardia Many potential causes all requiring prompt treatment to avoid ensuing hypotension and degeneration into ventricular fibrillation 	 Hemodynamically stable supraventricular tachycardia Initial treatment with vagal maneuvers, rapid push adenosine if ineffective Hemodynamically stable ventricular tachycardia Identify and treat underlying abnormalities
		Hemodynamically unstable supraventricular or ventricular tachycardiaDirect current cardioversion
Ischemia	 Increased myocardial demand Left ventricular hypertrophy Right ventricular hypertrophy Decreased myocardial supply Acquired (Kawasaki disease, cocaine/stimulant use, thrombotic event) Congenital (aberrant coronary course, stenotic coronary orifice, coronary arising from pulmonary artery) 	 Various causes all requiring thorough history and physical examination ECG to be obtained on abnormal findings Echocardiography likely indicated (see Table 2) Cardiology consult and follow-up likely required
Psychogenic causes	 Most prevalent in older children Often with recent stressor May have specific fears regarding heart attack or heart disease 	Attempt to limit unnecessary testing with thorough history; provide reassurance if psychogenic cause most likely

more prone to injury due to overuse. (16) Treatment is similar to that for most other muscle strain injuries, with rest and stretching as tolerated being paramount in recovery as well as NSAIDs if necessary.

Trauma. Trauma to the thorax, although encountered more frequently in adults, can cause significant morbidity and mortality in children. (17) There are numerous additional injuries that may occur in children who have experienced thoracic trauma that differ from those in adults due to the relatively greater thoracic cage flexibility in children, resulting in fewer fractures but more pulmonary contusions. Pulmonary contusion from direct lung injury results in consolidation of nonanatomical areas usually with associated chest wall bruising but without rib fractures. (17) Rib fractures are uncommon in newborn to 3-year-olds and should raise concern for child abuse. (15)(17) In older children, rib fractures must also prompt the treating physician

to consider associated injuries, including pneumothorax and hemothorax. (17)

As children become older and the incidence of fractures increases, flail chest may occur when multiple ribs are broken in 2 or more sites, leaving a portion of the chest wall to move paradoxically during respiration.

Pneumothorax secondary to trauma may present with chest pain and should raise concern for additional intrathoracic injury, seen in one-third of patients, or a combination of intrathoracic and extrathoracic injury, also seen in another one-third of patients. (17)

Pulmonary Causes

Asthma. Asthma, especially exercise-induced asthma, may result in chest pain in children, although the pain itself is reported in only a minority of children with asthma. (I) Bronchodilator treatment leads to improvement in a

significant proportion of affected children. (I) Children with exercise-induced asthma present with more pronounced symptoms with exercise that demands short, intense burst of activity without sufficient warm-up, (I) which should be kept in mind by providers in advising patients and families on how to decrease the severity of the child's chest pain. Children with asthma exacerbations reporting chest pain are more likely to have chest radiographs ordered by providers that infrequently yield positive findings, (I8) suggesting that in an otherwise straightforward asthma exacerbation with chest pain, management should remain the same as for an asthma exaceerbation without chest pain.

Respiratory Infections. Respiratory infections, specifically pneumonias, may present with chest pain. Pneumonia may cause pleural irritation, which results in chest pain, a finding that is suggestive of a bacterial pneumonia in an otherwise healthy child. (19) In addition, children with complicated disease are more likely to experience chest pain than those with uncomplicated disease. (20) In conjunction with focal chest findings on auscultation during a febrile illness, chest pain acts as another predictor of pneumonia. (21) When a bacterial cause is believed to be the etiology of pneumonia, appropriate antibiotic agents should be instituted.

Pleurodynia. Pleurodynia, also known as Bornholm disease, occurs most frequently in children and results in sharp, spasmodic chest pain along with fever due to its infectious etiology. (22) Viral infection is the believed cause of the disease, with coxsackie B being the most commonly implicated pathogen. (23) Pleurodynia is basically a myositis that affects the intercostal muscles, leading to pleuritic pain and splinting of the chest. (22) The condition occurs epidemically and, despite initially severe pain, is self-resolving over the course of a few weeks with analgesics as needed used to minimize pain. (22)

Gastrointestinal Causes

Gastroesophageal Reflux. Gastroesophageal reflux represents another potential cause of chest pain, (24) and although less common than experiencing epigastric pain and regurgitation, chest pain is experienced in 1.8% to 5.0% of children with reflux, (25) with older children and adolescents more likely to describe chest pain. (26) Notably, it is uncommon for a child to complain of more than 1 gastroesophageal symptom, (25) meaning that although chest pain is a less frequent symptom, it may be the only complaint in some children. When pain from reflux is experienced, it is often described in the middle of the chest and may also involve a burning sensation. (25) The substernal pain

experienced is caused by esophageal acid exposure. (24) The severity of symptoms is not associated with the histologic grade of the disease. (27) The mainstay of treatment remains acid suppression in the form of histamine receptor blockade or proton pump inhibition.

Esophagitis. Esophagitis describes the inflammation that results from tissue damage to the esophagus. This damage primarily occurs due to reflux as described previously herein but may also be due to infections, certain medications, and allergies, the latter being increasingly described in the past 2 decades in the form of eosinophilic esophagitis. Eosinophilic esophagitis may present similarly to gastroesophageal reflux and include symptoms of chest pain, with I defining difference being a lack of response to acid suppression treatment. (28) Eosinophilic esophagitis is distinguished from other types of esophagitis by significant eosinophilia of the mucosa, (28) giving this condition its name and diagnostic criteria by biopsy. Dietary changes, including elimination diets due to the proposed allergic mechanism of eosinophilic esophagitis, have been recommended, as well as topical corticosteroids. (28)(29) Exclusive amino acidbased elemental formulae have been shown to offer relief to most children with eosinophilic esophagitis. (29) Esophagitis can also be caused by certain medications, including oral contraceptives and antibiotics such as tetracyclines, (30) as well as NSAIDs. (29) Diagnosis relies on endoscopy with biopsy, (29) and treatment involves cessation of offending medications.

Foreign Body Ingestion. Foreign body ingestion is most common in children younger than 5 years with a wide array of ingested items, including coins most frequently as well as magnets, small toys, buttons, bones, jewelry, and retained foods. (31) Although most children with esophageal foreign bodies are asymptomatic, (32) symptoms often include retrosternal pain or a pressure sensation along with cyanosis and dysphagia. (31)(33) A history of drooling or reluctance to swallow should prompt consideration of foreign body ingestion. (30) Management after physical examination includes both posterior-anterior and lateral chest radiographs, as well as considering neck and abdominal films for objects that are above the thoracic inlet or past the pylorus. (31) Ultimately, endoscopy may be used for both diagnosis and treatment in symptomatic children without radiographic findings if the object is radiolucent. (31)

Cardiac Causes

Cardiac conditions account for the lowest proportion of chest pain experienced in the pediatric population. Of the cardiac causes of pain, a division can be made between nonischemic and ischemic causes, with the former accounting for more causes of cardiac chest pain, including pericarditis, myocarditis, mitral valve prolapse, and supraventricular and ventricular tachycardia.

The ischemic causes of cardiac chest pain can be further subdivided into ischemia due to increased cardiac demand and decreased blood supply. Conditions that result in increased demand are ultimately attributed to either left ventricular hypertrophy or right ventricular hypertrophy. Causes for left ventricular hypertrophy in children include aortic stenosis, hypertrophic cardiomyopathy, and systemic hypertension. Right ventricular hypertrophy in children may be due to pulmonic stenosis or pulmonary hypertension. There are several potential causes for decreased blood supply leading to ischemia that can be acquired, including Kawasaki disease and cocaine use, as well as congenital causes, including an anomalous aortic origin of a coronary artery, a stenotic orifice from which the coronaries arise, and anomalous left or right coronary arteries from the pulmonary artery. In addition, familial hypercholesterolemia and other prothrombotic conditions can also lead to cardiac ischemia due to impaired blood supply.

Syncope or palpitations associated with chest pain are possible indicators of cardiac disease. (30) In comparing patients with vasovagal syncope and those with cardiac syncope, the latter significantly occurs surrounding activity, (34) a clue in differentiation.

Nonischemic Cardiac Causes.

Pericarditis. Inflammation of the fibrous pericardium around the heart commonly presents in children with chest pain and fever (35) and is most often due to viral or idiopathic causes. (36) The pain is typically retrosternal with sudden onset and worsened with inspiration. (36) The pathological process leads to worsening pain in the supine position and accounts for the classic presentation of a patient sitting upright and leaning forward. (36) A key physical examination finding is that of a friction rub on auscultation. (36) An ECG will generally demonstrate widespread ST-segment elevation and PR depression. The only other valuable test is a complete blood cell count; a markedly elevated white blood cell count suggests purulent pericarditis. NSAIDs and aspirin in older children remain the mainstay of treatment for analgesia and inflammation control, (36) with corticosteroids used less frequently. (37) Colchicine, used to treat adults with pericarditis, is used far less in children in current practice (37) but may be considered in recurrent cases.

Myocarditis. Myocarditis—inflammation to the cardiac muscle tissue itself—may present nonspecifically with a range of symptoms from flulike symptoms or chest pain to

overt heart failure and shock. (35)(38) Most cases are initially attributed to respiratory diseases, including asthma and pneumonia or sepsis, (39)(40) because complaints of chest pain may be minimal in children younger than 10 years in contrast to those older than 10 years. (39) Similar to pericarditis, viruses are responsible for most cases of myocarditis, (38) with group B coxsackie commonly considered but also including parvovirus B19 and human herpesvirus 6. (38) Screening tests should include chest radiographs for cardiomegaly and ECG. When pneumonia or asthma diagnosis is uncertain, a low threshold for ECG should be had by providers. (39) Nonspecific ECG findings include sinus tachycardia, ST-wave and T-wave abnormalities, low voltage readings, and, less frequently encountered, atrioventricular or bundle branch block. (38) Most pediatric patients require care in an ICU with pharmacologic therapies aimed at providing circulatory support but may require mechanical circulatory support if pharmacologic therapies are ineffective. (41)

Mitral Valve Prolapse. Mitral valve prolapse occurs when I or both of the mitral valve leaflets prolapses a defined amount into the left atrium. (42) The cause for this prolapse is attributed to valvular tissue abnormalities, including myxomatous degeneration, geometric disparities between the ventricle and valve, and connective tissue disorders. (42) The valvular abnormality is predominantly congenital but may not be recognized until adolescence. (43) The classic finding on auscultation is a mid-to-late systolic click often with a high-pitched late systolic murmur, (42) with the click occurring earlier when standing or sitting. (43) Symptoms of mitral valve prolapse syndrome include chest pain, exertional dyspnea, palpitations, syncope, and anxiety. (42) Specific therapy is not indicated, and antibiotic drug prophylaxis is no longer recommended for surgical or dental procedures (43); however, treatment for symptomatic patients may include β -blockers or may require surgical intervention if severe regurgitation is present. (42)

Arrhythmias.

Supraventricular Tachycardia. The most frequent cardiac dysrhythmia seen in children is supraventricular tachycardia, which represents several dysrhythmias, the most common being Wolff-Parkinson-White syndrome, which are characterized by their narrow-complex rhythms originating above the atrioventricular junction. (44) The 3 major causes of supraventricular tachycardia are reentrant tachycardias using an accessory pathway, reentrant tachycardias without an accessory pathway, and ectopic or automatic tachycardias. (45) Presentation in infants may include fussiness, irritability, poor feeding, tachypnea, diaphoresis with feeds, and hepatic congestion, whereas older children may

experience chest pain as well as palpitations, nausea, dizziness, and light-headedness. (44) Older children and adolescents who are able to verbalize chest pain with supraventricular tachycardia will have heart rates of 180 to 220 beats/min. (44) Treatment for patients without hemodynamic instability begins with increasing vagal tone, which can be done using various vagal maneuvers, (44) including the Valsalva maneuver in cooperative children or placing bagged ice over the nasal bridge in younger children to elicit the diving reflex. (44) Importantly, ocular pressure must never be implemented, and carotid sinus massage is very rarely effective. (45) Medical therapy using adenosine by rapid push may subsequently be used should vagal maneuvers not be effective. (44)(45) In situations of hemodynamic instability or with symptoms of severe heart failure, direct current cardioversion using 0.5 to 2 J/kg should be implemented. (45)

Ventricular Tachycardia. In pediatric patients, ventricular tachycardia is less common than supraventricular tachycardia (45) but may also present with chest pain (5) along with palpitations, which may be perceived as pain. (7) Ventricular tachycardia may be associated with a variety of conditions, some of which are described in this review, including anomalous origin of a coronary artery, mitral valve prolapse, dilated or hypertrophic cardiomyopathy, myocarditis, primary cardiac tumors, arrhythmogenic right ventricular dysplasia, Wolff-Parkinson-White syndrome, and drug use, including cocaine and amphetamines. (45) Treatment must be prompt because ventricular tachycardia may lead to hypotension and degeneration into ventricular fibrillation. (45) As with patients experiencing hemodynamically unstable supraventricular tachycardia, patients with unstable ventricular tachycardia should immediately be treated with direct current cardioversion using 2 J/kg initially, followed by searching for and correcting any underlying abnormalities. (45)

Cardiac Ischemia. There are several potential causes of myocardial ischemia, any of which may present with chest pain. As mentioned previously herein, relative to other causes, pain brought on by ischemia to the cardiac tissue accounts for a very small proportion of all causes. When ischemia is severe, myocardial infarction may result, which can occur in the context of long-standing diabetes mellitus, Kawasaki disease, chronic anemias, or cocaine use leading to coronary vaso-spasm. (30) Rarely, thrombotic events in children may cause cessation of blood flow to the cardiac tissue, leading to ischemia and subsequent infarct, both of which will cause pain. Anomalous coronary arteries may lead to ischemia due to their coursing between the atria and ventricles, leading to vessel compression during systole and subsequent decreased blood flow to the cardiac tissue. Similarly, a stenotic orifice at

the origin of the coronary artery can result in decreased blood flow to the cardiac tissue. Aberrant left or right coronaries arising from the pulmonary arteries represent another rare condition that may be seen during infancy, which can cause decreased blood flow to the coronaries. (30) Finally, any condition leading to either left or right ventricular hypertrophy may predispose to a scenario in which the increased demand of the cardiac tissue cannot be met by the available blood supply, leading to ischemia and pain. Most ischemic causes of chest pain occur during exercise, which is why specific questioning during history is crucial in leading to the diagnosis, as discussed in more detail later herein.

Psychogenic Causes

Psychogenic causes of chest pain are an important consideration, especially in older children, with I study finding increased incidence in children older than 12 years. (46) In adolescents presenting with chest pain, I study found that 31% had experienced a recent stressful event, such as a death in the family, major illness or accident, family separation, or school changes. (3) Questioning the adolescents' underlying concerns regarding their chest pain elucidated that heart attack or heart disease as well as cancer were fears in most adolescents, although few actually believed these reasons to be the cause of their pain. (3) Another study found a fear of heart disorder or cancer to be present in 39%. (4) Overall, the predominating psychiatric disorders seen in adolescents with chest pain are anxiety disorders and major depression. (47) In I study, once no medical cause was identified as the cause for chest pain in 8- to 17-year-olds, 81% were diagnosed as having an anxiety disorder and were retrospectively noted to commonly present other somatic symptoms of pain in other locations. (48) These findings are congruent with previous studies showing additional somatic symptoms in more than half and sleep disturbances in one-third of patients. (49) The significance in this adolescent population is 2-fold, both in that investigations seeking an organic cause may increase anxiety, the potential primary cause for the pain, but conversely that anchoring on psychogenic causes and missing an organic cause is of major concern to health-care providers.

EVALUATION

A thorough and complete history is of the utmost importance in arriving at a correct diagnosis (2) and minimizing unnecessary testing and evaluation. As shown by the wide variety of possible diagnoses, there are often key historical features that may differentiate the source of a child's chest pain, including the time course of symptom onset (acute onset more often leading to an identifiable cause), (30) the timing and duration and symptoms, the site of pain and its reproducibility, the quality and radiation of pain experienced, aggravating and relieving factors, and associated complaints. A history of syncope should be investigated because there are major differentiating features of having a cardiac versus vasovagal cause, (34) with cardiac causes more often implicated when syncope surrounds activity, with a family history of cardiac disease or sudden death, or with abnormalities found on physical examination and ECGs.

The medical and surgical history should be explored with specific questioning regarding previous cardiac history or surgery. A family history of cardiac disease should be explored, as should a history of sudden death in the family, with specific questions listed in the Complications section of this review. Additional questions should include inquiring about any history of Kawasaki disease, asthma, sickle cell disease, diabetes, or connective tissue disorders, for example, Marfan syndrome, which places the patient at increased risk for aortic dissection due to aortic aneurysm. (50) In the case of Turner syndrome, Ehlers-Danlos syndrome type IV, and homocysteinuria, patients may carry a higher risk of aortic root dissection. (50) After obtaining a detailed history, a thorough and targeted physical examination may yield insight into the cause of chest pain. Both tenderness to palpation and well-localized pain may lead to a possible musculoskeletal cause of chest pain. Bruising may act as an obvious finding indicating trauma and may necessitate further investigations to determine the extent of intrathoracic or extrathoracic injury. (17) Abnormal auscultatory findings on the lung examination may lead to a pulmonary source of chest pain, and of course any extracardiac sounds should lead the provider to assess for possible cardiac involvement. Pertinent negative findings are also useful when the physical examination findings are normal as psychogenic causes may be explored, although an organic etiology may not be excluded.

ASSESSMENT IF CARDIAC ETIOLOGY IS SUSPECTED

If either the history or physical examination, including the child's vital signs, lead to any indication that the cause of chest pain may be cardiac, the next step in evaluation is to obtain an ECG. (7) Abnormalities in any of these initial assessments may necessitate echocardiography. (6) Of note, in investigating a potential cardiac cause of chest pain,

exercise testing and rhythm monitoring using Holter monitors have been found to provide no clear diagnostic benefit in initial assessment despite their previous recommendation. (6) Many new algorithms and methods to determine benefits of echocardiography have been described in the past decade. In 2014 the first appropriate use criteria for initial transthoracic echocardiography in outpatient pediatric cardiology were established and made specific recommendations regarding the appropriateness of obtaining an echocardiogram in children experiencing chest pain without previously known heart disease. (34) The guideline has a total of 113 possible indications, 11 of which pertain to chest pain, and is useful for education and for improvement initiatives in health-care delivery, outcomes, and resource utilizations. (34) Although the appropriate use criteria were created for outpatient cardiology, the authors note the usefulness for general pediatricians and family practitioners, understanding that they are often the first clinicians to face the decision of whether to use echocardiography. (34) The recommendations made are shown in Table 2 and further enforce previous findings described in this review-that a thorough history and physical examination with ECG should be used to lead further investigation.

Historically, clinical practice guidelines have been used to guide practice; however, in the past decade, a new tool has arisen known as the standardized clinical assessment and management plan (SCAMP). (6) SCAMPs differ from clinical practice guidelines in that data acquisition is mandated, the analysis of set data is on a recurring basis, and the SCAMP itself is altered based on the recurring analysis. The aims of SCAMPs include reducing the diversity of assessment and management, improving patient care delivery through the use of SCAMP algorithms, and reducing ineffective or unnecessary resource utilization while assessing the effectiveness of testing and interventions. (51) One study implementing SCAMPs demonstrated a reduction in cost by 22% in assessing and managing pediatric chest pain despite a higher percentage of patients being recommended for echocardiography, (52) and another large study retrospectively found a theoretical reduction in referral to pediatric cardiology by more than 50%. (5) The use of SCAMPs has suggested that the diagnostic approach to chest pain in pediatric patients should use only the data gathered from the history and physical examination in conjunction with ECG to suggest when further testing is indicated, (6) as described throughout this review.

TABLE 2. Appropriateness of Initial Transthoracic Echocardiography in Outpatient Pediatric Cardiology in Children Seen with Chest Pain (34)

Appropriate use of echocardiography	 Exertional chest pain Nonexertional chest pain with abnormal ECG findings Chest pain with a family history of sudden unexplained death or cardiomyopathy
Echocardiography may be appropriate	 Chest pain with other symptoms or signs of cardiovascular disease, a benign family history, and normal ECG findings Chest pain with a family history of premature coronary artery disease Chest pain with recent onset of fever Chest pain with recent illicit drug use
Echocardiography is rarely appropriate	 Chest pain with no other symptoms or signs of cardiovascular disease, a benign family history, and normal ECG findings Nonexertional chest pain with no recent ECG Nonexertional chest pain with normal ECG findings Reproducible chest pain with palpation or deep inspiration
ECG—electrocardiography.	

Regardless of the method used to determine the necessity of echocardiography, the decision is made judiciously, understanding the low probability of having any significant findings. In I large-scale study, 38% of children with chest pain had echocardiography performed, with abnormalities found in 11.9%, although only 0.8% of these echocardiograms had abnormalities that were potentially related to the patient's chest pain. (50) Most patients with a cardiac cause for their chest pain in this study had suggestive symptoms, especially exertional chest pain, a concerning personal or family history, and abnormal physical examination or ECG findings. (50)

Finally, if a cardiac etiology of pain is suspected by the patient or family, the care provider should keep these perceptions in mind because this is a common fear, especially in adolescents. (3)(4) Awareness on the part of the care provider and specifically addressing these concerns with families are important in preventing continued pain and anxiety. Small studies have been performed showing the effectiveness of psychoeducation and various coping mechanisms in decreasing chest pain if a psychogenic cause has been identified. (53)

COMPLICATIONS

Thoracic trauma leading to pulmonary contusion or penetrating chest wall trauma may lead to the development

of pneumonia, which can separately lead to respiratory failure in affected patients. (17) Rib fractures may lead to atelectasis as well as pneumonia, and their prevention should be a high priority in affected children using incentive spirometry and antibiotics when indicated. (17)

In considering cardiac etiologies for chest pain in pediatric patients, providers should also be aware of conditions that predispose children to sudden cardiac death. There are 4 commonly considered conditions that may predispose a child to sudden cardiac arrest: hypertrophic cardiomyopathy, long QT syndrome, catecholaminergic polymorphic ventricular tachycardia (a ventricular tachycardia brought on by exercise or emotional stress in affected individuals), and anomalous origin of the left coronary artery from the right sinus of Valsalva. (54) A large-scale multicenter study in 2016 assessed clinical presentation of these conditions, finding that long QT syndrome, hypertrophic cardiomyopathy, and catecholaminergic polymorphic ventricular tachycardia commonly presented with a known or concerning family history, (54) again enforcing the importance of thorough history taking as discussed. Notably, one-fifth of patients with long QT syndrome and nearly one-third with hypertrophic cardiomyopathy had a significant family history that was missed during their initial evaluation by primary providers but was elucidated

at subsequent evaluation by pediatric cardiologists. (54) The American Academy of Pediatrics released a policy statement regarding pediatric sudden cardiac arrest in 2012 that specifically outlines 4 questions that the medical provider should ask during assessment for risk of sudden cardiac death (55):

- I. Have you ever fainted, passed out, or had a seizure suddenly and without warning, especially during exercise or in response to auditory triggers such as doorbells, alarm clocks, and ringing telephones?
- 2. Have you ever had exercise-induced chest pain or shortness of breath?
- 3. Are you related to anyone with sudden, unexplained, and unexpected death before age 50 years?
- 4. Are you related to anyone who has been diagnosed as having a sudden death-predisposing heart condition such as hypertrophic cardiomyopathy, long QT syndrome, or catecholaminergic polymorphic ventricular tachycardia?

The policy statement includes 10 recommendations in both assessment of children for sudden cardiac warnings and policy and ideologies to support further education and prevention of sudden cardiac death in children. (54)

SUMMARY

- The most common identifiable etiologies of chest pain in children include musculoskeletal causes, followed by pulmonary, gastroenterologic, and psychogenic causes.
- Chest pain in children due to cardiac etiologies represents an exceedingly low proportion of chest pain despite being a common fear in families and older children.
- Based on strong research evidence, (2)(6)(7) initial assessment of pediatric patients presenting with chest pain should be composed of a thorough history and physical examination as well as ECG.
 Further diagnostic testing, including echocardiography and referral to pediatric cardiology, should be reserved for patients with positive findings on initial assessment.
- Based on strong research evidence, (34) appropriate use criteria have been created for echocardiography and based on some research evidence as well as consensus (5)(6)(51)(52) SCAMPs have also shown promise in determining the best use of echocardiography.
- Based on some research evidence and consensus, (55) specific questions regarding personal and family history may help elucidate children at risk for sudden cardiac death.

References for this article are at http://pedsinreview.aappublications.org/content/41/9/469.

PIR Quiz

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- 1. A 14-year-old previously healthy girl presents to the emergency department with constant REQUIREMENTS: Learners midsternal chest pain for the past 8 hours. Her parents are concerned about heart disease. In narrowing down the wide variety of causes of chest pain in pediatric patients, which of the following is the most appropriate next step in the initial evaluation of this patient?
 - A. Chest radiography and troponins.
 - B. Electrocardiography (ECG) and chest radiography.
 - C. ECG and echocardiography.
 - D. History and physical examination.
 - E. Stress testing and ECG.
- 2. Further assessment of the patient in guestion 1 shows that her chest pain has been severe and unremitting and has caused her to take rapid, shallow breaths. There has been no history of trauma, fever, wheezing, or symptoms suggestive of pneumonia or upper respiratory tract infection. Her medical history is normal; she has no history of chest pain or syncope with exertion and no history of cardiac disease or murmurs. There is no family history of unexpected sudden death before age 50 years, and no family history of cardiac disease. Her physical examination findings are normal, including a heart examination with a regular rate and rhythm and no murmurs, and lung examination findings are normal. There is marked tenderness along the right costochondral junctions from rib 3 to rib 6 that reproduces her pain. Which of the following is the most likely diagnosis for the chest pain in this patient?
 - A. Costochondritis.
 - B. Mitral regurgitation.
 - C. Pericarditis.
 - D. Pneumonia.
 - E. Pneumothorax.
- 3. A 10-year-old boy is evaluated for chest pain occurring on and off for more than a month. The pain is in the center of his chest and is described as a burning sensation that lasts several hours. There is no associated shortness of breath. He has no history of exertional chest pain or dyspnea. He was administered ranitidine for 14 days with no relief of the pain. His cardiac examination reveals a regular rate and rhythm, no murmurs or gallops, normal capillary refill, and normal pulses. The remainder of his examination findings are normal. This clinical presentation is most consistent with which of the following diagnoses?
 - A. Costochondritis.
 - B. Eosinophilic esophagitis.
 - C. Gastroesophageal reflux.
 - D. Pneumonia.
 - E. Precordial catch syndrome.
- 4. A 12-year-old girl presents with a history of intermittent chest pain over the past 3 months. She has been evaluated by an urgent care physician who recommended antacids, which have not helped resolve her symptoms. The patient describes some exertional dyspnea, as well as palpitations. She is quite anxious about her symptoms. Her physical examination findings are largely normal except for the heart examination, which showed a regular rhythm and a high-pitched late systolic murmur heard along the upper left sternal border, II/VI in intensity, preceded by a mid-to-late systolic click. The click occurs earlier when she is sitting or standing. She has normal, physiologic splitting of her second heart sounds. Her capillary refill time is normal. An ECG is completely normal. Which of the following is the most likely diagnosis in this patient?

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- A. Aortic stenosis.
- B. Kawasaki disease.
- C. Large atrial septal defect.
- D. Mitral valve prolapse.
- E. Tricuspid stenosis.
- 5. A 13-year-old boy is brought to the clinic for evaluation of chest pain. His medical history is significant for being a former 27-week premature infant. The patient is overweight with a history of chronic renovascular hypertension and secondary cardiomegaly. In assessing the causes of his chest pain, which of the following historical findings is most suggestive of ischemic causes of chest pain?
 - A. Pain associated with deep inspiration.
 - B. Pain that improves with use of nonsteroidal anti-inflammatory medications.
 - C. Pain that occurs during sleep.
 - D. Pain that occurs in conjunction with a systolic murmur.
 - E. Pain that occurs most frequently during exercise.