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Surendranath R. Veeram Reddy and Harinder R. Singh

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Chest Pain in Children and Adolescents

Surendranath R. Veeram Reddy, MD,* Harinder R. Singh, MD*

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

1. Enumerate the most common causes of chest pain in pediatric patients.
2. Differentiate cardiac chest pain from that of noncardiac cause.
3. Describe the detailed evaluation of a pediatric patient who has chest pain.
4. Screen and identify patients who require a referral to a pediatric cardiologist or other specialist.
5. Explain the management of the common causes of pediatric chest pain.

Case Studies

Case 1

During an annual physical examination, a 12-year-old girl complains of intermittent chest pain for the past 5 days that localizes to the left upper sternal border. The pain is sharp and stabbing, is 5/10 in intensity, increases with deep breathing, and lasts for less than 1 minute. The patient has no history of fever, cough, exercise intolerance, palpitations, dizziness, or syncope. On physical examination, the young girl is in no pain or distress and has normal vital signs for her age. Examination of her chest reveals no signs of inflammation over the sternum or rib cage. Palpation elicits mild-to-moderate tenderness over the left second and third costochondral junctions. The patient reports that the pain during the physical examination is similar to the chest pain she has experienced for the past 5 days. A detailed cardiovascular and other organ system examination yields normal results. What is the most likely cause of this patient's chest pain? What will you recommend for this patient? Does she need to see a pediatric cardiologist?

Case 2

A 17-year-old boy is playing soccer on a Saturday afternoon and has a syncopal event on the field. He regains consciousness within a few seconds, does not require resuscitation, and is taken to the emergency department. The patient informs the physician that he developed sudden midsternal chest pain and lightheadedness prior to passing out. His vital signs are normal for age, and he is alert, oriented, and in no apparent pain or distress. Physical examination reveals an ejection click and a harsh, grade 3/6 systolic ejection murmur at the base of his heart and right upper sternal border with radiation to both carotid arteries. The rest of the physical examination findings are normal, except for minor abrasions on his elbows and knees caused by the fall. Twelve-lead electrocardiography (ECG) reveals left ventricular hypertrophy with ST segment depression in leads V5 and V6. What is the diagnosis? What would you recommend for this patient? Does he need a referral to a pediatric cardiologist?

Abbreviations

AAP: American Academy of Pediatrics
d-TGA: d-transposition of great arteries
ECG: electrocardiography
GERD: gastroesophageal reflux disease
NSAID: nonsteroidal anti-inflammatory drug

Introduction

Chest pain in children is one of the most common reasons for an unscheduled visit to the primary care physician's office and the emergency department, accounting for more than 650,000 physician visits per year in patients 10 to 21 years of age. (1) Although alarming to parents, chest pain in children usually is not caused by a serious disease, in contrast to chest pain in adults, which raises concern for coronary ischemia. Chest pain is second only to heart murmur for referral to a

*Division of Cardiology, The Carman and Ann Adams Department of Pediatrics, Children's Hospital of Michigan, Wayne State University School of Medicine, Detroit, Mich.

pediatric cardiologist. (2) Pediatric chest pain can be classified broadly into cardiac chest pain or noncardiac chest pain. Noncardiac chest pain is, by far, the most common cause of chest pain in children and adolescents. This article reviews most causes of chest pain in children and adolescents, emphasizing cardiac causes, and provides a guideline for evaluating a child or adolescent who has chest pain.

Noncardiac Chest Pain

Chest pain is noncardiac in origin in more than 98% of children and adolescents who complain of it. (3) Noncardiac causes of chest pain can be classified into musculoskeletal, pulmonary, gastrointestinal, and miscellaneous (Table 1).

Musculoskeletal or Chest-wall Pain

The most common cause of chest pain in children and adolescents is musculoskeletal or chest-wall pain. The prevalence of musculoskeletal chest pain is between 15% and 31%. (6)(7) There are various types of musculoskeletal pain.

COSTOCHONDRITIS. Costochondritis, or costosternal syndrome, is characterized by unilateral sharp, stabbing pain along the upper two or more contiguous costochondral joints. The pain usually is exacerbated by deep breathing and lasts from a few seconds to a few minutes. There is no sign of inflammation, although chest-wall tenderness can be reproduced by manual palpation over the affected area. In most patients, pain due to costochondritis is self-limited, with intermittent exacerbations occurring during adolescence.

TIETZE SYNDROME. Tietze syndrome is a localized nonsuppurative inflammation of the costochondral, costosternal, or sternoclavicular joint seen in adolescents and young adults. The cause is unknown, but recent upper respiratory tract infection with excessive coughing has been implicated. (8) Tietze syndrome usually involves a single joint, commonly at the second or third rib. Unlike the more diffuse costochondritis, Tietze syndrome is distinguished by localized involvement of a single joint along with signs of inflammation in the form of warmth, swelling, and tenderness.

IDIOPATHIC CHEST-WALL PAIN. Also known as nonspecific chest-wall pain, this disorder is one of the common causes of chest pain in children. The pain is sharp, lasts for a few seconds to minutes, localizes to the middle of the sternum or the inframammary area, and is exacerbated by deep breathing or by manual pressure on the

Table 1. Noncardiac Causes of Chest Pain in Children (4–6)

Musculoskeletal

- Costochondritis/costosternal syndrome
- Tietze syndrome
- Nonspecific or idiopathic chest-wall pain
- Slipping rib syndrome
- Trauma and muscle strain–overuse injury
- Xiphoid pain (xiphoidalgia)
- Sickle cell vaso-occlusive crisis

Pulmonary or Airway-related

- Bronchial asthma
- Exercise-induced or cough variant asthma
- Bronchitis
- Pleurisy
- Pneumonia
- Pneumothorax
- Pulmonary embolism
- Acute chest syndrome

Gastrointestinal

- Gastroesophageal reflux disease
- Esophageal spasm
- Peptic ulcer disease
- Drug-induced esophagitis/gastritis
- Cholecystitis

Miscellaneous

- Panic disorder
- Hyperventilation
- Breast-related conditions
- Herpes zoster
- Spinal cord or nerve root compression

sternum or rib cage. Signs of inflammation are absent.

SLIPPING-RIB SYNDROME. Also known as the lower rib pain syndrome, slipping-rib syndrome occurs infrequently in children, and the exact prevalence is unknown. Slipping-rib syndrome is characterized by intense pain in the lower chest or upper abdominal area caused by trauma or dislocation of the 8th, 9th, and 10th ribs. Because these ribs do not attach to the sternum directly but rather are attached to each other via a cartilaginous cap or fibrous band, they can be hypermobile and prone to trauma. Slipping-rib syndrome pain can be reproduced by the “hooking maneuver,” in which the examiner places his or her fingers under the inferior rib margin and pulls the rib edge outward and upward. (9) Occasionally, a clicking sound accompanies the pain during this maneuver.

TRAUMA AND MUSCLE STRAIN. Teenagers active in gymnastics and most other sports are prone to chest-wall trauma. In one series, skeletal trauma was the cause of chest pain in 2% of children. (6) Chest-wall injury can produce localized pain and tenderness and usually is associated with swelling or erythema at the site of injury. Teenagers who experience chest muscle strain usually give a history of weightlifting. For patients who have a history of significant trauma to the chest, the findings of severe chest pain, arrhythmia, and shortness of breath may indicate myocardial contusion and hemopericardium.

PRECORDIAL CATCH. The pain of precordial catch, also known as “Texidor twinge,” usually is sudden and sharp, lasts for a few seconds, and localizes to one intercostal space along the left lower sternal border or to the cardiac apex. (10) The origin of the pain is unknown, but precordial catch has been associated with poor posture and may be caused by a pinched nerve. The pain occurs either at rest or during mild activity and is exacerbated with inspiration, often leading to shallow breathing in an effort to alleviate pain.

XIPHOID PAIN OR XIPHODYNIA. Also known as hypersensitive xiphoid syndrome, xiphodynia is localized pain or discomfort over the xiphoid process of the sternum that can be exacerbated by eating a heavy meal, coughing, and bending or rotating movements. (11) The cause of pain is unknown. Digital compression of the xiphoid elicits dull pain.

TREATMENT. Reassurance, rest, and analgesia are the primary treatments for musculoskeletal chest pain. In most circumstances, allaying the fears of the patient and parents by counseling them about the benign nature of the condition helps to relieve concern for and reduce the degree of chest pain. For patients who have severe pain, applying warm compresses and administering nonsteroidal anti-inflammatory drugs (NSAIDs) for 1 week may be helpful.

Pulmonary and Airway-related Causes

The prevalence of chest pain due to respiratory causes is approximately 2% to 11%. (12) Bronchial asthma is the most common pulmonary cause of chest pain. Exercise-induced asthma frequently causes chest pain, even in patients who have no audible wheezing. Weins and associates (13) analyzed pulmonary function during treadmill testing in a group of 88 otherwise healthy children and adolescents who had chest pain and found that

approximately 73% had laboratory evidence of asthma, suggesting that the incidence of exercise-induced asthma is greater than previously reported. Patients who have chest pain due to reactive airway disease should be treated with inhaled bronchodilators.

Infections of the bronchial tree or lung, including bronchitis, pleurisy, pleural effusion, pneumonia, empyema, bronchiectasis, and lung abscess, can cause acute chest pain. Intense chest pain with hypoxia is the manifesting feature of pulmonary embolism. Patients who have sickle cell disease may present with chest pain due to acute chest syndrome or pulmonary infarctions.

Gastrointestinal Causes

Evangelista and colleagues (14) found the prevalence of chest pain due to gastrointestinal causes to be about 8%. Common gastrointestinal causes of chest pain in children are gastroesophageal reflux disease (GERD), peptic ulcer disease, esophageal spasm or inflammation, and cholecystitis. Rare gastrointestinal causes include esophageal strictures, foreign body, and ingestion of caustic substances. Chest pain caused by GERD typically is described as a burning pain in the epigastric area that frequently has a temporal relationship to food intake. Histamine-2 blockers or proton pump inhibitors are the mainstay of treatment for GERD. If symptoms suggest cholecystitis, prompt referral to a specialist and treatment with antibiotics are indicated.

Miscellaneous

Psychogenic chest pain in older children occasionally can result from anxiety or a conversion disorder triggered by recent stressors in personal or family life. Pantell and Goodman (6) reported that approximately one third of adolescents who presented to the outpatient clinic complaining of chest pain had a history of stressful events either in the family or at school. Psychogenic chest pain often is associated with other somatic complaints as well as with sleep disturbances.

Hyperventilation, either due to anxiety or a panic disorder, can cause chest pain that is accompanied by complaints of difficulty in breathing, dizziness, or paresthesias. Spasm of the diaphragm, gastric distension caused by aerophagia, and coronary vasoconstriction due to hypocapnic alkalosis are postulated explanations for chest pain during hyperventilation.

Chest pain due to breast-related causes has a prevalence of 1% to 5%. (6) Postmenarchal girls may complain of throbbing or burning chest pain caused by mastitis, fibrocystic disease, or pregnancy. Teenage males who

have gynecomastia occasionally complain of unilateral or bilateral chest pain.

Herpes zoster infection of the chest wall might present with burning pain or paresthesia in a dermatomal pattern, sometimes preceding the rash by a few days.

Children who have scoliosis or other deformities that lead to spinal cord or nerve root compression also may complain of chest pain as their initial presentation and should be referred to an orthopedist for additional evaluation and treatment.

Cardiac Chest Pain

Chest pain due to a cardiac condition is rare in children and adolescents, with a prevalence of less than 6%. (2)(5) Table 2 lists the common causes of cardiac chest pain.

Inflammatory Causes

Pericarditis with or without pericardial effusion usually is infectious in origin. Pericarditis presents as a sharp retrosternal chest pain that often radiates to the left shoulder, is aggravated when the patient lies supine or takes a deep breath, and is relieved by bending forward. Myocarditis or endocarditis also can present with chest pain.

Increased Myocardial Demand or Decreased Oxygen Supply

Chest pain may be the first complaint that points to an unsuspected anatomic heart defect. Pain associated with palpitations, dizziness, and panic attacks may be the presenting symptoms in some patients who have mitral valve prolapse (Barlow syndrome). Mitral valve prolapse usually is associated with a midsystolic click and, occasionally, an apical mid-to-late systolic honking murmur. Bisset and colleagues (15) reported that only 18% of patients in a group of 119 children who had mitral valve prolapse complained of atypical chest pain.

Patients who have left ventricular outflow tract obstruction in the form of stenosis of the aortic valve, subaortic valve area, or supra-aortic valve area or who have coarctation of the aorta may present with chest pain associated with dizziness and fatigue. In aortic valve stenosis, a harsh ejection systolic murmur with radiation to the neck is heard on auscultation. An ejection click from a stenosed bicuspid aortic valve may be heard.

Chest pain in association with exercise intolerance and fatigue may be the initial presenting complaint of patients who have hypertrophic or dilated cardiomyopathy.

Coronary Artery Abnormalities

Chest pain due to myocardial ischemia can occur in patients who have abnormal coronary artery anatomy,

Table 2. Cardiac Causes of Pediatric Chest Pain

Inflammatory: Pericarditis, Myocarditis

- Infective: viruses, bacteria
- Noninfective: SLE, Crohn disease, postpericardiotomy syndrome

Increased Myocardial Demand or Decreased Supply

- Cardiomyopathy: dilated or hypertrophic
- LVOT obstruction: aortic stenosis, subaortic stenosis, supravalvar aortic stenosis
- Arrhythmias

Coronary Artery Abnormalities

- Congenital: ALCAPA, ALCA from right coronary sinus, coronary fistula
- Acquired: Kawasaki disease, postsurgical (after arterial switch operation, after Ross procedure), posttransplant coronary vasculopathy, familial hypercholesterolemia

Miscellaneous

- Aortic dissection
- Rupture of aortic aneurysm
- Pulmonary hypertension
- Mitral valve prolapse
- Atrial myxomas
- Cardiac device/stent complications

Drugs

- Cocaine
- Sympathomimetic overdose

ALCA=anomalous left coronary artery, ALCAPA=anomalous origin of the left coronary artery from pulmonary artery, LVOT=left ventricular outflow tract, SLE=systemic lupus erythematosus

including congenital anomalies of the coronary artery, coronary artery fistulas, and stenosis or atresia of the coronary artery ostium. Coronary artery abnormalities are second only to hypertrophic cardiomyopathy in causing sudden cardiac deaths in adolescents. Unfortunately, sudden death may be the first and only presentation of coronary artery abnormalities. However, a significant number of patients who have abnormal coronary artery connections present initially with anginal chest pain, usually associated with exertion.

Patients describe ischemic chest pain as a squeezing sensation, tightness, pressure, constriction, burning, or fullness in the chest. One example of chest pain due to myocardial ischemia is a condition in which the left main coronary artery or left anterior descending coronary artery arises from the right sinus of Valsalva or right coro-

nary artery and courses between the aorta and pulmonary artery. (16)(17) During exertion, the aorta and pulmonary artery squeeze the left main coronary artery or left anterior descending artery, leading to myocardial ischemia and, occasionally, sudden death in adolescents. Infants who have coronary insufficiency due to anomalous origin of the left coronary artery from the pulmonary artery usually present with irritability, drawing of their knees up to their abdomens after feeding, pallor, diaphoresis, and circulatory shock. These babies often are misdiagnosed as having colic.

Children who have a history of cardiac surgery and heart transplantation also are susceptible to myocardial ischemia and may present with chest pain as the initial complaint. Beyond the first year after transplantation, secondary malignancy and transplant coronary vasculopathy are the most common causes of mortality. (18) Among patients who have had orthotopic heart transplantation, chest pain may be a sign of rejection or accelerated coronary artery vasculopathy with myocardial ischemia. Patients who have had an arterial switch operation for d-transposition of the great arteries (d-TGA) are at risk of developing coronary artery ostial stenoses.

A long-term complication of Kawasaki disease is coronary artery stenosis. Giant coronary artery aneurysms in Kawasaki disease have a high risk for rupture, occlusion due to thrombosis, or stenosis causing myocardial ischemia or infarction. (19) Kato and associates, (20) in a 10- to 21-year follow-up study, reported that 46% of patients who had Kawasaki disease and a history of giant coronary aneurysms developed stenosis or complete obstruction and 67% experienced myocardial infarction, with a 50% mortality rate. Risk factors for developing coronary artery aneurysms, thrombosis, and stenosis are male sex, early age (<6 months), or older age (>5 years) at the time of diagnosis. (19)

Coronary artery disease also may be seen in patients who have a family history of hypercholesterolemia, but children and adolescents seldom have enough obstruction to cause chest pain from ischemia. Coronary artery disease may present within the first 2 decades in patients born with homozygous familial hypercholesterolemia, in contrast to those who have heterozygous familial hypercholesterolemia, in whom coronary artery disease commonly occurs after the fifth decade of life. According to the preventive health-care guidelines from the American Academy of Pediatrics (AAP), every child should undergo a dyslipidemia risk assessment beginning at 2 years of age, and assessment should be repeated every 2 years until 10 years of age and then annually. (21) For children and adolescents who have a family history of hypercho-

lesterolemia, the AAP recommends measurement of a nonfasting serum total cholesterol concentration as the initial screening test, followed by a fasting lipid profile if the total cholesterol value is abnormal. (21)

Miscellaneous Causes

When a patient complains of an extremely severe and tearing midsternal chest pain radiating to the back, aortic dissection should be considered. Pediatric patients who have Marfan syndrome are at risk for a dissecting aortic aneurysm that may present with the sudden onset of severe chest pain. Aortic root dissection also can be seen in patients who have Turner syndrome, type IV Ehlers-Danlos syndrome, and homocystinuria.

Young children who are unable to describe palpitations caused by arrhythmias may complain of chest pain and point to the sternum. Chest pain in children or adolescents who have congenital heart disease and undergo Fontan palliation or Mustard or Senning palliation for d-TGA may have intra-atrial re-entry arrhythmias (slow atrial flutter).

Primary or secondary pulmonary hypertension may present with chest pain in association with fatigue and exertional dyspnea or syncope.

Toxic Drug-induced Chest Pain

Toxic exposure to cocaine, marijuana, methamphetamines, and sympathomimetic decongestants may be the cause of chest pain due to myocardial ischemia or arrhythmias.

Management of Chest Pain in Children and Adolescents

Although cardiac disease rarely presents as chest pain, primary care physicians should evaluate every patient complaining of chest pain to rule out any significant underlying disease. The history and physical examination are the first steps in diagnosing the cause of such pain in most pediatric patients (Table 3).

History

The history should include a description of the pain; associated complaints; and precipitating, aggravating, and relieving factors. Acute chest pain can be caused by trauma, pulmonary embolism, asthma, and cardiac causes, including aortic dissection or ischemic pain due to coronary artery anomalies. Chronic chest pain usually is noncardiac, and causes can be musculoskeletal, gastrointestinal, psychogenic, or idiopathic.

Location of pain sometimes may aid in differentiating the cause. Localized or pinpoint pain usually is chest-wall

Table 3. History and Physical Examination of a Pediatric Patient Who Has Chest Pain

History

1. Description of chest pain: duration, onset, location, quality, severity, radiation, precipitating and relieving factors
2. Past medical history: asthma, sickle cell disease, Kawasaki disease, cardiac disease, hypercholesterolemia
3. Surgical history: any previous surgeries of the chest or abdomen
4. Family history: early/sudden cardiac deaths of unknown cause, arrhythmias, cardiomyopathy, hypercholesterolemia
5. Genetic disorders: Marfan syndrome, Turner syndrome, type IV Ehlers-Danlos syndrome
6. History of trauma, drug abuse (eg, cocaine), psychological stressors

Physical Examination

Vital signs, dysmorphic features, peripheral pulses, chest inspection, reproducible chest pain, hyperdynamic precordium, irregular heart beats, distant heart sounds, abnormal loud second heart sound, systolic clicks or murmurs, gallops, absent femoral pulses

or pleural; diffuse chest pain usually is due to underlying visceral disease of the lung or heart. Radiation of chest pain to a certain area may suggest a specific cause. Chest pain due to myocardial ischemia may radiate to the neck, throat, lower jaw, teeth, upper extremity, or shoulder. In acute cholecystitis, chest pain radiates to the right shoulder. Patients who have pericarditis complain of chest pain radiating to the left shoulder. Chest pain from aortic dissection frequently radiates to the interscapular space in the back.

Certain precipitating and aggravating factors might help identify the cause of chest pain. Musculoskeletal chest pain may worsen with certain body positions, movement, and deep breathing. Chest pain aggravated by eating or associated with vomiting, regurgitation, or swallowing suggests a gastrointestinal cause. Chest pain precipitated by exertion and associated with dyspnea could be due to cardiac or respiratory disease. A recent history of upper respiratory tract infection with fever and current symptoms of chest pain should raise suspicion of pericarditis. Chest pain associated with recurrent multiple somatic complaints such as headaches, abdominal

pain, or extremity pain could be psychogenic. Chest pain with lightheadedness and paresthesias is associated with hyperventilation. Arrhythmias may present as chest pain with palpitations.

A history of asthma should alert the physician to bronchospasm as the cause of chest pain. Exercise-induced asthma should be considered if the patient complains of cough, chest pain, and difficulty in breathing after physical activity.

Adolescents who have chest pain may give a recent history of having trauma, lifting heavy weights, or pulling a muscle during sports or exercise. History of any recent use of tobacco or of recreational drugs should be sought. Bronchitis is a common cause of chest pain in tobacco smokers. Cocaine and other sympathomimetic drugs are potent vasoconstrictors, and their use may lead to myocardial ischemia.

A past history of Kawasaki disease or arterial switch operation for d-TGA should alert the physician to the possibility of coronary ostial stenosis causing myocardial ischemia. Patients who have undergone transcatheter stent placements or device closure of atrial or ventricular septal defects may complain of chest pain related to device embolization or impingement on adjacent structures. Anginal chest pain may be the presenting symptom in patients born with anomalies of the coronary artery, coronary vasculopathy in heart transplant patients, and coronary artery narrowing due to familial hypercholesterolemia. Acute chest syndrome in patients who have sickle cell disease may be the cause of chest pain. A past history of Crohn disease, systemic lupus erythematosus, or other autoimmune disease should alert the physician to pericarditis as the cause of chest pain.

A family history of certain genetic or connective tissue disorders, such as Marfan syndrome, should prompt evaluation for specific disease-related features.

Physical Examination

The physical examination of a child complaining of chest pain always should begin with vital signs and anthropometric measurements. Tall stature might point toward certain genetic causes (Marfan syndrome). Fever and tachypnea might help to confirm an infectious disorder, usually of the lung and rarely the heart, especially if the pain is associated with tachycardia and worsens with lying down (pericarditis with or without effusion). Elevated jugular venous pressure or hypotension should point toward systolic or diastolic dysfunction of the heart (myocarditis or cardiomyopathy). Any facial dysmorphism or abnormality of the lens (Marfan syndrome) should be noted. Visual inspection of the chest wall is

important to look for any bony abnormalities such as pectus excavatum or carinatum, scoliosis, or surgical scars. Thelarche in girls or gynecomastia in adolescent boys can cause chest pain. Warmth, redness, and tenderness around the nipple and of the breast occur with mastitis. Particular attention should be paid to eliciting any reproducible tenderness, similar to the experienced chest pain, on palpation of the chest wall. Cough, crackles, or wheezes might suggest a hyperreactive bronchial airway (asthma or exercise-induced asthma) or a pneumonic process of the lung.

A detailed cardiovascular examination should include palpation of the precordium for heaves (right ventricular heave in pulmonary hypertension) or thrills (obstructive valvular lesions). Particular attention should be paid to heart sounds. When the heart sounds are distant or not strongly audible, a diagnosis of pericardial effusion should be entertained. A loud second heart sound along the left upper sternal border suggests pulmonary hypertension. Pericardial rub may be heard in mild-to-moderate pericardial effusions.

Patients who have myocarditis may present with tachycardia, muffled heart sounds, a gallop rhythm, and a mitral regurgitation murmur. An ejection systolic click is heard in aortic stenosis and a midsystolic click in mitral valve prolapse. A harsh, medium-pitched midsystolic murmur is heard in patients who have a fixed aortic obstruction due to valvular, subvalvular, or supravalvular stenosis or hypertrophic cardiomyopathy. In patients who have mitral valve prolapse, a midsystolic click may be accompanied by an apical mid- to late-systolic honking murmur. A continuous murmur is heard in patients who have coronary artery fistulas or ruptured sinus of Valsalva. Weak or absent femoral pulses with upper limb hypertension and a continuous flow murmur at the left lower scapular margin should prompt evaluation for coarctation of the aorta. Hepatomegaly, ascites, and peripheral edema should alert the physician to look for causes of congestive heart failure.

Investigations

Patients who have the clinical features of musculoskeletal chest pain and no other noteworthy findings do not require additional evaluation or referral. Those who have a significant history or abnormal findings on physical examination should have additional diagnostic evaluation and a referral to a pediatric cardiologist if cardiac disease is suspected. Evaluation is individualized, based on associated symptoms and findings. A chest radiograph should be performed to look for bony lesions, cardiomegaly, airways, and lung parenchymal or pleural lesions.

ECG is useful for evaluation of rate and rhythm and signs of ischemia, pericarditis, or chamber hypertrophy. Additional testing should be left to the discretion of the pediatric cardiologist. When baseline ECG readings are normal, an exercise stress test may be necessary to assess the development of arrhythmia or ischemia during exertion. Exercise stress testing along with pulmonary function testing may be warranted in patients who experience exertional chest pain and other cardiovascular symptoms.

Echocardiography, performed in a pediatric center and supervised by a pediatric cardiologist, is a very valuable tool for delineating cardiac anatomy. (22) The diagnosis of pericardial effusion, aortic root dissection, left-sided obstructive lesions, cardiomyopathy, pulmonary hypertension, and ventricular dysfunction may be established by two-dimensional echocardiography. Echocardiography also can help to identify coronary abnormalities, including abnormal origin and course of the coronary arteries, coronary aneurysms, and coronary artery fistulas. Historically, coronary angiography has been the gold standard for evaluation of the origin and course of the coronary arteries. However, newer non-invasive techniques such as 64-slice computed tomography scan or magnetic resonance angiography are being used increasingly to define abnormal coronary artery anatomy. Occasionally, electrophysiologic studies may help diagnose underlying arrhythmias.

Treatment of Chest Pain

Reassurance, analgesia, rest, or any combination of these three measures is the best treatment for patients experiencing noncardiac chest pain. NSAIDs administered for 1 week often decrease inflammation and pain. Patients who have pericarditis and pericardial effusion should be treated with ibuprofen. Administration of steroids may be considered in refractory cases of pericarditis and pericardial effusions. (23) The appropriate specialist should treat specific cardiac, pulmonary, gastrointestinal, and psychogenic causes of chest pain. Most, but not all, patients who experience chest pain and are awaiting a cardiology consultation should have their physical activity restricted pending their final cardiology evaluation.

Referring a Patient to a Pediatric Cardiologist

Consultation with a pediatric cardiologist is highly recommended for any child or adolescent patient who has chest pain associated with exertion, palpitations, sudden syncope (especially during exercise) or abnormal findings on cardiac examination or ECG; a history of past cardiac

Table 4. Reasons to Refer Children Who Have Chest Pain to a Cardiologist

1. Abnormal cardiac findings
2. Exertional chest pain
3. Exertional syncope
4. Chest pain with palpitations
5. Electrocardiographic abnormalities
6. Significant family history of arrhythmias, sudden death, or genetic disorders
7. History of cardiac surgery or interventions
8. Orthotopic heart transplant
9. History of Kawasaki disease
10. First-degree relatives have familial hypercholesterolemia

surgery or intervention; or a family history of genetic syndrome, arrhythmias, sudden cardiac death, or high risk for coronary artery disease (Table 4). Patients who have a past history of Kawasaki disease, congenital heart disease, cardiac surgery, or heart transplantation are at risk for coronary artery disease. Heart transplant patients who experience myocardial ischemia may not complain of chest pain because of sympathetic denervation of the transplanted heart but may present with nonspecific symptoms such as nausea, vomiting, and an elevated resting heart rate. Patients who have had a Mustard or Senning procedure for d-TGA or who have had Fontan palliation for a single ventricle are at high risk for intra-atrial re-entry tachycardias (also called slow atrial flutters) and should be referred promptly to a pediatric cardiologist.

Children or adolescents who have had a recent transcatheter cardiac intervention, including device closure or stent placements, should be evaluated by a cardiologist for device embolization or compression of adjacent structures. Complaints of chest pain in patients who recently had congenital heart disease correction should be taken seriously. The chest should be inspected to rule out any infection of the sternotomy incision and chest tube sites. History of fever, chest pain worsening when supine, and distant heart sounds on auscultation should raise the suspicion of postpericardiotomy syndrome. Patients who have had arterial switch operation for d-TGA and those having a history of Kawasaki disease are at risk for coronary artery stenosis or atresia and subsequent myocardial ischemia or infarction. Obtaining ECG is a good starting point to look for signs of ischemia.

Case Discussion

Case 1

The 12-year-old child described in this case has a classic history for costochondritis, a benign condition that is a very common cause of chest pain in the pediatric population. On physical examination, most affected patients have reproducible tenderness on palpation of the chest. The absence of signs of inflammation differentiates costochondritis from Tietze syndrome. This patient does not need any additional investigations or referral to a pediatric cardiologist. Reassuring her and her family about the benign nature of the condition is sufficient. For patients experiencing severe pain, NSAID therapy and warm compresses applied for a few days may be beneficial.

Case 2

The 17-year-old boy described in this case has exertional chest pain followed by a syncopal event. Any patient experiencing exertional chest pain and syncope should be restricted from all strenuous physical activities and referred to a pediatric cardiologist immediately for additional evaluation and treatment. In this patient, 12-lead ECG revealed left ventricular hypertrophy and ST segment depressions in the left-sided precordial leads, suggesting a strain pattern caused by transient left ventricular subendocardial ischemia. Echocardiography performed the next day demonstrated a bicuspid aortic valve with moderate-to-severe aortic stenosis and left ventricular hypertrophy. The patient was taken to the cardiac catheterization laboratory for balloon aortic valvuloplasty.

Summary

- Every patient who has chest pain warrants a thorough evaluation. Usually, the history and physical examination are sufficient to diagnose the cause of the pain.
- Based on strong research evidence, musculoskeletal chest pain is the most common cause of chest pain in the pediatric population. (3) Educating and reassuring both the patient and the family about the benign nature of chest pain is of utmost importance.
- Based on some research evidence as well as on consensus, a cardiac cause for chest pain is more likely if the pain occurs during exertion. (3) Any coexisting symptoms suggestive of myocardial ischemia or an abnormal cardiac finding should prompt immediate referral to a pediatric cardiologist.

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