CHEST PAIN IN PEDIATRICS

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Chest pain is an alarming complaint in children, leading an often frightened and concerned family to a pediatrician or emergency room and commonly to a subsequent referral to a pediatric cardiologist. Because of the well-known association of chest pain with significant cardiovascular disease and sudden death in adult patients, medical personnel commonly share heightened concerns over pediatric patients presenting with chest pain. Although the differential diagnosis of chest pain is exhaustive, chest pain in children is least likely to be cardiac in origin. Organ systems responsible for causing chest pain in children include:

- Idiopathic (12%-85%)
- Musculoskeletal (15%-31%)
- Pulmonary (12%-21%)
- Other (4%-21%)
- Psychiatric (5%-17%)
- Gastrointestinal (4%-7%)
- Cardiac (4%-6%)

Furthermore, chest pain in the pediatric population is rarely associated with life-threatening disease; however, when present, prompt recognition, diagnostic evaluation, and intervention are necessary to prevent an adverse outcome. This article presents a comprehensive list of differential diagnostic possibilities of chest pain in pediatric patients, discusses the common causes in further detail, and outlines a rational diagnostic evaluation and treatment plan.

Chest pain, a common complaint of pediatric patients, is often idiopathic in etiology and commonly chronic in nature. In one study, chest pain accounted for 6 in 1000 visits to an urban pediatric emergency room. In addition, chest pain is the second most common reason for referral to pediatric cardiologists. Chest pain is found equally in male and female patients, with an average

*References 13, 17, 23, 27, 32, 35, 44, 48, 49, 63-67, 74, and 78.

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age of presentation of 13 years. Chest pain in the pediatric population is most commonly idiopathic in etiology, representing 23% to 45% of all cases. Children fewer than 12 years of age are more likely to have a cardiorespiratory cause of their chest pain compared with children more than 12 years of age, who are more likely to have a psychogenic cause. In addition, patients diagnosed with psychogenic chest pain or costochondritis are more likely to be female. Chest pain is a chronic condition in the pediatric population, with persistent symptoms in 45% to 69% of patients and 19% of patients having symptoms lasting for more than 3 years. The corollary, of course, is that in 81% of patients with persistent symptoms, the chest pain resolves with time. Clinically reassuring is that none of the patients followed over 3 years developed any severe disease process. Because of the chronic nature and often unconfirmed cause of chest pain, pediatricians must develop a trusting, working relationship with these children and their families and be prepared to work jointly over time in diagnosing and treating patients with this very common and predominantly benign complaint.

ANATOMY OF MAJOR THORACIC STRUCTURES CAUSING CHEST PAIN

Figure 1 demonstrates the common sites of origin of chest pain in a schematic diagram of the major structures in the chest and abdomen in the anteroposterior and lateral planes.

HISTORY

A thorough and complete history is essential for the accurate diagnosis of chest pain. One must methodically gather historical data and avoid a quick run to judgment or an exhaustive laboratory investigation when evaluating chest pain. A comprehensive time series of events leading up to, through, and after the episode of chest pain should be documented. The pain should be described with respect to time of onset, duration, frequency, nature, intensity, location, points of radiation, precipitants, and relieving factors. The relationship to meals and body position should be noted, as well as the impact the pain has had on the child's lifestyle and activity level. The patient's past medical and surgical history should be taken and specific inquiry made into any underlying cardiac disease or heart surgery. Medications the child is taking should be recorded, with attention to nonprescription medications, oral contraceptives, and those causing gastric irritation. A thorough family history must be elicited, particularly of sudden death or cardiovascular disease. A comprehensive social history, including use of alcohol, tobacco, intoxicants, stimulants, cocaine, or other drugs must be obtained. This is usually done in private with the patient separate from other family members. The type of work or play the child participates in, together with specific sports activity, should be recorded. Any recent family discord, family stressors, change in peer socialization, or change in the child's school performance should be noted. Finally, a complete review of systems should be assessed to elicit symptoms of chronic disease, such as fever, malaise, fatigue, weight loss, and night sweats.
Figure 1. Common sites of origin of chest pain in the major structures in chest and abdomen in anteroposterior (A) and lateral (B) planes.
PHYSICAL EXAMINATION

A thorough and complete physical examination is fundamental to an accurate diagnosis of chest pain. During the physical examination, particular attention must be given to the child’s vital signs (including blood pressure), general appearance and state (e.g., cyanotic, comfortable, distressed, panicked, anxious, or shock). The entire chest wall, musculature, breasts, sternum, xiphoid, and abdomen should be palpated in an attempt to identify the source and reproducibility of the pain. The examination should include cardiac findings (e.g., murmurs or gallop rhythm), pulmonary signs (e.g., depth and rate of breathing, use of accessory respiratory muscles, presence of rales, rhonchi, wheezes, or decreased breath sounds), abdominal examination (i.e., size of liver and points of tenderness), femoral pulses, and distal extremities (i.e., temperature, cyanosis, clubbing, and edema). Finally, the child’s psychological state should be assessed.

Certain historical events, physical signs, and symptoms that should alert the clinician to one of the rare but potentially life-threatening causes of chest pain include:

Cardiac
Underlying congenital or acquired cardiac disease
Arrhythmias
Crushing sternal chest pain with or without radiation to left arm or neck
Exercise-induced chest pain
Persistent tachycardia
Persistent hypertension
Hypotension
Gallop rhythm
Syncope
Pulmonary
Hemoptysis
Dyspnea
Rales
Cyanosis
Gastrointestinal
Hematemesis
Hematochezia
Melena
Other
Febrile
Life-threatening psychiatric illness, such as psychosis or suicidal ideation

When present, prompt evaluation and intervention are necessary.

DIFFERENTIAL DIAGNOSIS

A detailed list of differential diagnoses for chest pain in children includes:

Cardiac \((m = \text{murmur present on auscultation})\)
Anatomic lesions
Aortic stenosis \((m)\)
Aortic aneurysm with dissection \((\pm m)\)
Subaortic stenosis \((m)\)
Supravalvar aortic stenosis \((m)\)
Ruptured sinus of Valsalva \((m)\)
Coarctation of the aorta (m)
Anomalous left, right, or both coronary arteries from pulmonary artery
(± m)
Coronary artery ostia stenosis or atresia
Intramural coronary artery
Left coronary artery arising from anterior cusp
Congenital coronary artery aneurysm
Coronary artery fistula (m)
Mitral valve prolapse (m)
Severe pulmonary stenosis (m)
Arrhythmogenic right ventricular dysplasia

Acquired lesions
Cardiomyopathy—dilated, hypertrophic, restrictive (m)
Endocarditis (± m)
Myocarditis (± m)
Rheumatic fever (± m)
Myocardial infarction (± m)
Coronary vasospasm
Kawasaki disease (± m)
Accelerated atherosclerotic coronary artery disease (diabetes mellitus, familial dyslipoproteinemias)
Dissecting aortic aneurysm (Marfan syndrome) (± m)
Pericarditis—infectious, autoimmune, posttraumatic, constrictive (m)
Postpericardiotomy syndrome (m)
Partial absence of the pericardium
Pulmonary hypertension (primary or secondary) (± m)
Eisenmenger syndrome (± m)
Takayasu arteritis
Cardiac tumors (± m)
Pericardial neoplasm (primary or secondary) (m)
Cardiac transplant—rejection or accelerated coronary artery disease (± m)

Arrhythmias
Premature atrial contractions
Atrial flutter
Atrial fibrillation
Supraventricular tachycardia (reentry or automatic)
Premature ventricular contractions
Ventricular tachycardia

Pulmonary
Reactive airway disease
Pneumothorax or pneumomediastinum
Pneumonia (viral, bacterial, mycobacterium, fungal, or parasitic)
Chronic cough
Pleural effusion
Pleurodynia
Pulmonary embolism
Foreign body aspiration
Cystic adenomatoid malformation
Primary or secondary adenoma or carcinoma

Gastrointestinal
Esophagitis
Esophageal diverticulum
Esophageal spasm
Esophageal rupture (Boerhaave syndrome)
Mallory-Weiss tear
Achalasia
Gastroesophageal reflux
Gastritis
Peptic ulcer disease
Zollinger-Ellison syndrome
Hiatal hernia
Foreign body ingestion
Cholecystitis
Subdiaphragmatic abscess
Fitz-Hugh-Curtis syndrome
Pancreatitis

Musculoskeletal
Muscle pain from overuse
  Pectoralis major and minor muscle
  Intercostal muscle
  Latissimus dorsi muscle
  Serratus muscle
  Trapezius muscle
Costochondritis
Tietze syndrome
Cervical ribs (C7)
Slipping rib (8–10)
Precordial catch syndrome
Trauma
Child abuse
Fractured or contused clavicle or ribs
Xiphoidalgia
Osteomyelitis
Rhabdomyosarcoma
Myositis
Thoracic outlet obstruction
Ankylosing spondylitis
Spondylolisthesis, spondyloyisis
Discitis
Herniated disc
Transverse myelitis

Psychiatric
Somatoform disorder
Stress
Depression
Hyperventilation syndrome
Panic attacks
Bulimia nervosa
Munchhausen syndrome

Ingestion
Cocaine
Tobacco
Methamphetamine
Sympathomimetic decongestants

Breast disease
Gynecomastia
Thelarche
Mastitis
Fibrocystic disease
Adenocarcinoma

**Mediastinal tumors**
- Hodgkin's disease
- T-cell lymphoma
- Thymoma
- Thymolipoma
- Teratoma
- Germ cell tumor
- Liposarcoma

**Miscellaneous**
- Diabetes mellitus
- Hyperthyroidism
- Cystic fibrosis
- Neurofibromatosis
- Marfan syndrome
- Ehlers-Danlos syndrome
- Homocysteinuria
- SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, osteitis)
- Sickle cell disease with vaso-occlusive crises
- Sickle cell disease
- Shingles (herpes zoster)
- Echinococcosis
- Mediterranean fever
- Hypercoagulation syndromes (protein S/C deficiency, antithrombin 3 deficiency, heparin cofactor 2 deficiency, plasminogen deficiency, factor 5 Lieden, anticardiolipin antibodies, lupus anticoagulant)
- Spinal cord meningioma
- Spinal cord nerve root compression

Numerous review articles have been written on the topic, but very little new research on the topic has been published in recent years. Following is a brief discussion of the clinical features of several pertinent diagnoses.

**Cardiac**

Although a rare cause of chest pain in pediatric patients (4–6%), this group remains the most concerning because an error in diagnosis may lead to significant morbidity or mortality. The three major etiologic categories are: (1) anatomic lesions, (2) acquired lesions, and (3) arrhythmias. Following are descriptions of the diagnostic features of many of the cardiac anomalies within each of the major categories.*

**Anatomic Lesions**

Anatomic lesions frequently have a pathologic heart murmur present. However, one must be careful not to interpret commonly occurring innocent murmurs

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*References 1, 3, 8, 11, 14, 18, 20, 26, 30, 38, 39, 41, 49, 59–61, 68, 73, 76, and 77.
as pathologic. A resting 12-lead ECG should be obtained for all patients with anatomic lesions to evaluate for the expected lesion specific ECG changes and for evidence of ischemia. A chest radiograph may be helpful for comparison of the cardiac size to previous examinations. In most cases, the diagnosis is confirmed and specific features detailed by echocardiography. Cardiac catheterization is now rarely required for proper diagnosis. Consultation with a pediatric cardiologist is necessary because the chest pain may represent life-threatening myocardial ischemia. In the majority of cases, a discussion and review of the case with the pediatric cardiologist is all that is necessary. Occasionally, the cardiologist may want to see the patient immediately for further evaluation and testing. Chest pain is common in children with mitral valve prolapse (31\%), although it is rarely life threatening.\(^1,3,36,56\) These patients have an increased incidence of arrhythmias (18\%), some of which are life threatening and should be excluded.\(^35,34\)

**Acquired Lesions**

Acquired cardiac lesions are diverse in their presentation. A dilated cardiomyopathy should be suspected when a patient presents with fatigue, decreased exercise tolerance, and palpitations. A gallop rhythm is heard on auscultation and may be accompanied by the murmur of mitral regurgitation. In contrast, hypertrophic cardiomyopathy presents with a systolic ejection murmur that becomes louder with an increase in heart rate (exercise) or decrease in preload (Valsalva maneuver). Patients with idiopathic hypertrophic subaortic stenosis often relate a family history of sudden death.

In patients with acquired cardiac lesions, ECG changes are found, cardiomegaly is seen by chest radiography, and an echocardiogram confirms the diagnosis. Patients with endocarditis are febrile and appear acutely ill. Serial blood cultures are positive in 92\% of children with endocarditis. An echocardiogram, particularly via the transesophageal route, may assist in the diagnosis of endocarditis. Children with myocarditis present with fatigue, dyspnea, and frequently cardiovascular collapse. These patients commonly report a recent viral upper respiratory tract infection. Although an echocardiogram demonstrates the decreased cardiac function, cardiac catheterization with endomyocardial biopsy is often required. Rheumatic fever is diagnosed using the modified Jones' criteria. Although myocardial infarction is extremely rare in the pediatric population, it is diagnosed by the presence of classic symptoms (i.e., persistent crushing sternal chest pain with or without radiation to neck or arm, dyspnea, and diaphoresis). A chest radiograph, serial ECG, and cardiac enzymes (i.e., creatinine kinase with MB fraction or troponin T) are necessary for diagnosis. Coronary vasospasm or accelerated atherosclerotic coronary artery disease results in myocardial ischemia with symptoms of angina pectoralis. Patients with Kawasaki disease present acutely with the classic features of mucocutaneous lymph node syndrome and pancarditis. Later, they develop coronary aneurysms that may thrombose, resulting in myocardial infarction. Patients with Marfan syndrome who present with acute tearing chest pain must have dissection of the aorta excluded because it is a life-threatening emergency. MR imaging techniques or transesophageal echocardiography are the methods for rapid diagnosis of aortic dissection. Pericarditis is multifactorial in origin. On auscultation, distant heart sounds or a pericardial friction rub is typically heard. A pulsus paradoxicus is present. A chest radiograph reveals cardiomegaly, and an echocardiogram confirms the diagnosis. Postpericardiotomy syndrome is the development of a pericardial effusion days to months after cardiac surgery. Fatigue, decreased
exercise tolerance, palpitations, and syncope are common complaints of patients with pulmonary hypertension. Chronic pulmonary hypertension results in right ventricular dilatation and hypertrophy, with the resulting physical findings of a narrowed second heart sound, hepatomegaly, and cyanosis if an atrial or ventricular septal defect is present. A chest radiograph reveals cardiomegaly, and an echocardiogram confirms the diagnosis, but cardiac catheterization is frequently performed with a therapeutic trial of pulmonary vasodilating agents. Eisenmenger syndrome is severe pulmonary hypertension resulting from uncorrected congenital heart disease with left-to-right shunting and subsequent reversal of the shunt causing severe cyanosis. Takayasu arteritis is the “pulseless disease” affecting the aorta and great vessels in young women. Cardiac and pericardial tumors are extremely rare in children.

Arrhythmias

Arrhythmias are not uncommon in children. Most are benign (e.g., premature atrial contractions and unifocal premature ventricular contractions), but a few (e.g., supraventricular tachycardia or ventricular tachycardia) are life threatening. Children with arrhythmias complain of palpitations and chest pain. If altered vital signs, shock, congestive heart failure, or syncope occur, immediate life-saving treatment should begin, together with consultation with a pediatric cardiologist. For stable patients, an electrocardiogram and 24-hour Holter monitor are diagnostically useful. For significant arrhythmias, referral should be made to a pediatric cardiologist. Patients with hyperthyroidism frequently have cardiac arrhythmias. Palpation of the thyroid gland should be included in the physical examination, and thyroid function tests may be ordered as part of the laboratory investigation.

Pulmonary

Pulmonary causes of chest pain are common, occurring in 12% to 21% of cases, particularly in children fewer than 12 years of age. Reactive airway disease and pneumonia are principally diagnosed by auscultation and confirmed when necessary by chest radiography. Palpation of subcutaneous crepitans and crunching heart sounds (Hamman’s sign) are suggestive of a pneumothorax or pneumomediastinum and confirmed by chest radiography. Decreased breath sounds coupled with dullness to percussion are indicative of a pleural effusion and posterior-anterior and decubitus chest radiographs or sonography are diagnostic. Pleurodynia, caused by Coxsackie virus, results in excruciating spasms of sharp chest pain, pleural friction rub, and fever. Pulmonary embolism in children is rare but should be suspected in female adolescents on oral contraceptives and children with hypercoagulation syndromes who present with dyspnea and cyanosis. The ECG reveals the typical pattern of right heart strain with ST changes, whereas chest radiography is usually nonspecific. A ventilation perfusion scan is usually diagnostic, although cardiac catheterization with angiography is sometimes necessary. Treatment with thrombolytic agents is life saving, although surgical thromboembolectomy is occasionally necessary. A foreign body should be suspected by history and physical examination. Inspiratory and expiratory chest radiographs can be helpful, although bronchoscopy is both diagnostic and therapeutic.
Gastrointestinal

Gastrointestinal causes of chest pain are relatively rare, accounting for only 4% to 7% of cases. Historical references to diet, relationship to meals, and body position are important diagnostically. Trials of antacids or hydrogen ion blockers are helpful diagnostically and therapeutically for patients with gastritis, gastrointestinal reflux, and peptic ulcer disease. If symptoms persist, referral may be made to a pediatric gastroenterologist for more intensive diagnostic procedures, such as endoscopy, pH probe, or esophageal manometry. Altered vital signs, hematemesis, hematochezia, melena, or esophageal rupture are life-threatening events, and immediate referral and intervention are indicated. A hiatal hernia is easily diagnosed when suspected clinically by a barium upper gastrointestinal series. A foreign body is suspected by history, possibly confirmed by plain radiograph, and is usually allowed to pass spontaneously. If the foreign body is lodged in the esophagus, endoscopy is necessary for removal. Cholecystitis presents with postprandial pain referred to the right upper quadrant, and an abdominal sonogram is diagnostic.6 22, 23, 25, 29, 47, 52, 53

Musculoskeletal

Musculoskeletal causes of chest pain are fairly common, representing 15% to 31% of pediatric patients with chest pain. A typical history is that of a deconditioned adolescent participating in sports activity and afterward complaining of chest pain. Palpation and contraction of the specific muscle group reproduces the pain, and treatment is symptomatic. Costochondritis is fairly common, particularly in girls. The chest pain is reproduced by palpation, and treatment is symptomatic. Tietze syndrome, characterized by swelling of the costochondral joints, is rare in children. Trauma, either accidental or intentional, can result in chest wall injury and chest pain. Examination localizes the source of pain, and radiographs are helpful in excluding fractures and pulmonary contusions. Slipping rib syndrome results from excess mobility of the 8th to 10th ribs, which do not directly insert into the sternum. Precordial catch syndrome is a positional change in the chest (“slouching”), resulting in acute sharp chest pain.40, 43, 45, 58, 75

Psychiatric

Psychiatric reasons for chest pain are common and account for 5% to 17% of cases of chest pain. Patients frequently identify a life stressor (e.g., death in family, family discord, divorce of parents, poor school performance, or nonacceptance from peers) preceding the onset of chest pain. Symptoms of depression are common. Hyperventilation may be one of the presenting signs. A positive rapport and trusting working relationship with the family is essential for the diagnosis and treatment of these patients, and this will contribute to the eventual relief of the chest pain. Patients with bulimia nervosa can develop esophagitis or an esophageal tear from frequent bouts of emesis, resulting in chest pain. Rarely, disturbed parents inflict trauma to a child’s chest area, resulting in chest pain.50, 51, 69
Other

A whole host of other disease processes can result in chest pain. Cocaine use can result in palpitations, coronary vasospasm, or even myocardial infarction. Adolescents who smoke tobacco products have an increased incidence of chronic cough and chest pain. Male adolescents who develop gynecomastia can develop chest pain, as can female adolescents with fibrocystic breast disease. Mediastinal tumors are exceptionally rare. Young adults with advanced stages of diabetes mellitus can develop premature coronary artery disease. Children with sickle cell disease can develop vaso-occlusive crises, resulting in chest pain. The hypercoagulation syndromes result in thrombus formation, which can produce a pulmonary embolism or myocardial infarction.*

Laboratory Investigation

The potential laboratory tests available in evaluating and diagnosing even the most unusual causes of chest pain include:

**Cardiac:** ECG, chest radiograph, echocardiogram, Holter monitor, exercise stress test, dobutamine stress test, thallium scan, serum creatinine kinase with MB fraction, serum troponin T, fractionated serum lipid profile, pericardiocentesis, endomyocardial biopsy with polymerase chain reaction analysis, cardiac catheterization with or without selective coronary angiography

**Pulmonary:** Chest radiograph, CT scan of chest, MR imaging of chest, pulmonary function testing with and without methacholine challenge, bronchoscopy, ventilation perfusion scan, sweat test

**Gastrointestinal:** Gastric lavage; pH probe; upper gastrointestinal series; upper endoscopy; esophageal manometry; abdominal sonography, including liver, pancreas, and gallbladder; serum liver function tests; serum lipase and amylase; serum gastrin level; stool guaiac testing

**Musculoskeletal:** Skeletal radiographs, CT scan of spine, MR imaging of spine, nuclear bone scan, creatinine kinase with MM fraction

**Miscellaneous:** Urine and serum toxicology screen, serum complement levels, antinuclear antibodies, glycosylated hemoglobin A1c, hemoglobin electrophoresis, coagulation studies, mammography, breast sonography, breast biopsy, thyroid function tests, complete blood cell count, Westergren erythrocyte sedimentation rate, cultures (i.e., blood, pericardial fluid, and sputum), viral or bacterial antibody titers (i.e., IgM, and IgG), Tine testing, psychological testing

This list is given for reference only and should not be used as a checklist for evaluating routine chest pain in children. As mentioned previously, laboratory testing is most commonly unnecessary for appropriately diagnosing and treating chest pain in children.

**SUMMARY**

Chest pain in the pediatric population is a common and mostly benign occurrence. A thorough history and physical examination are usually all that are necessary in excluding the rare, life-threatening causes of chest pain. These rare,

*References 12, 16, 28, 31, 42, 46, 53, 54, 62, 79, 81, and 82.
life-threatening events require immediate evaluation, treatment, and subspecialty consultation. Idiopathic chest pain is the most common diagnosis, and the symptoms are typically chronic. Laboratory testing is usually nondiagnostic, costly, and burdensome to patients and therefore unnecessary. A long-term, trusting relationship with the patients and their families is needed to reassure them and allow symptoms to resolve.

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