

Chest Pain and Syncope in Children: A Practical Approach to the Diagnosis of Cardiac Disease

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Chest pain and syncope are common in children, accounting for numerous provider visits and leading to a considerable amount of testing.¹⁻⁷ The Centers for Medicare and Medicaid Services projects that health care spending will exceed 20% of the US Gross Domestic Product by 2018.⁸ Considering the looming crisis in health care finance and impending changes in the US health care delivery system, provider-driven methods to optimize resource utilization are critical. Careful evaluation of current practices, with particular attention to use of expensive and overutilized resources, is needed.

Pediatric chest pain and syncope share a paradox in that both are common and benign in the vast majority of cases, but can be cardinal symptoms of rare and serious cardiac conditions. The prevalence of serious cardiac pathology in children presenting with these symptoms is low, but extensive and costly cardiac testing is common and may be unnecessary in many cases.^{4,5} Outpatient visits for chest pain and syncope at Boston Children's Hospital have more than doubled over the last 10 years and now account for ~15% of all outpatient visits (Figure 1; available at www.jpeds.com). In this article, we discuss causes of pediatric syncope and chest pain and propose an approach focused on careful history and physical exam that aims to optimize testing and referral patterns while maintaining a high detection rate for serious heart disease.

Chest Pain

Case

An 18-year-old male presented to the pediatrician with a 2-week history of exertional chest pain. The onset of chest pain coincided with the beginning of track season. The chest pain occurred only during high levels of exertion, and forced the patient to stop competing in track. Physical examination findings were normal.

What are the concerning symptoms for a cardiac etiology of chest pain in this patient? What further evaluation is warranted?

Chest pain is a common presenting complaint to pediatricians, pediatric cardiologists, and pediatric emergency depart-

ments (EDs),^{1-5,9-11} accounting for at least 650 000 physician visits annually in patients aged 10-21 years.^{9,10} The overwhelming majority of cases of chest pain in otherwise healthy children have a noncardiac etiology.^{1-4,9-11} Numerous previous studies in multiple practice settings have demonstrated a cardiac etiology in only 0-5% of children and adolescents presenting with chest pain (Table I; available at www.jpeds.com). In contrast to adults, in whom chest pain often signals a significant cardiac problem, the most common etiologies of chest pain in children are benign and include musculoskeletal, gastrointestinal, pulmonary, idiopathic, and psychogenic causes (Table II).^{2-5,10-12} In the largest review of patients seen in a cardiology clinic for chest pain, Saleeb et al¹¹ reported on a cohort of ~3700 patients seen over a 10-year period. The most common causes of chest pain were idiopathic (52%), musculoskeletal (36%), respiratory (7%), and gastrointestinal (3%). Only 37 of the 3700 patients seen (1%) had a cardiac etiology for chest pain, with supraventricular arrhythmias (n = 16) and pericarditis (n = 10) the most frequent causes. Only 9 patients (0.24%) had a cardiac conditions that put them at increased risk for sudden death, including myocarditis (n = 4), anomalous coronary origin from incorrect sinus of Valsalva (n = 3), hypertrophic cardiomyopathy (HCM; n = 1), and dilated cardiomyopathy (n = 1). The potentially life-threatening and more benign causes of cardiac chest pain were all identifiable by history, physical examination, and selected use of electrocardiography (ECG). Importantly, in more than 18 000 patient-years of follow-up of this cohort, there have been no missed diagnoses leading to cardiac death.

Despite the low prevalence of serious cardiac pathology in this population, chest pain generates considerable anxiety among patients, parents, and providers and often leads to evaluations that are low-yield, extensive, and costly. Owing largely to anxiety and misconceptions about the causes of chest pain in children, patients and their families frequently seek medical attention, and providers may have difficulty providing reassurance to families. Patients and their families vastly overestimate the prevalence of cardiac causes of chest pain while underestimating the more benign causes of chest pain, with families reporting that they suspect a cardiac etiology in more than 50% of cases, compared with the actual

ECG	Electrocardiography
ED	Emergency department
EST	Exercise stress testing
HCM	Hypertrophic cardiomyopathy
HTN	Hypertension
NCS	Neurocardiogenic syncope
SCAMP	Standardized Clinical Assessment and Management Plan

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Table II. Noncardiac etiologies of chest pain

Cause	Estimated incidence	Examples
Musculoskeletal	50%-68%	Costochondritis Precordial catch syndrome Muscle strain Trauma
Respiratory	3%-12%	Slipping rib syndrome Asthma Pneumonia Bronchitis Pleuritis Pulmonary embolus Pneumothorax
Gastrointestinal	2%-8%	Gastroesophageal reflux disorder Esophagitis Gastritis Pancreatitis Gastric ulcer Biliary disease
Psychogenic	10%-30%	Conversion disorder Panic/anxiety attack
Other	<10%	Skin infection Breast disease Acute chest syndrome in sickle cell disease

incidence of 0-5%.^{1,2,13} In a study of 100 consecutive patients seen in a pediatric practice for noncardiac chest pain, 69% reported restricting their activity, 40% had school absences related to chest pain, 44% thought they were having a heart attack, and 12% feared they had cancer.¹⁴ High levels of anxiety disorder have been documented in children with noncardiac chest pain and their parents, with 1 study finding psychiatric disorders listed in the *Diagnostic and Statistical Manual of Mental Disorders, 4th Edition* (predominantly anx-

ety disorder) in 60% of pediatric patients with noncardiac chest pain.^{14,15} Compared with patients seen for innocent murmurs, those with noncardiac chest pain have been shown to have greater levels of functional disability, somatization, and psychosocial distress, and to use significantly more health care services.^{15,16}

Understanding the cardiac and noncardiac etiologies of pediatric chest pain helps guide evaluation and determine the need for referrals and additional testing. Causes of noncardiac chest pain are listed in a **Table II**. Musculoskeletal chest pain is the most common cause, accounting for up to 70% of cases; it is characteristically sharp, fleeting, and exacerbated by deep inspiration, and may be reproducible on palpation. The major cardiac causes of pediatric chest pain, and key red flag physical examination and history findings associated with each diagnosis, are listed in **Table III**.

Owing to the acuity and severity of symptoms, most patients with chest pain related to serious cardiac pathology present to the ED, with a smaller number, typically with less acute chest pain, presenting in outpatient clinics. In a 10-year review, Kane et al¹⁷ identified 171 patients with a serious cardiac condition who presented with chest pain. Only 41 of these patients (24%) presented through an outpatient clinic, 32 with a potentially serious cardiac diagnosis, included coronary anomalies (n = 21), HCM (n = 3), myocarditis (n = 1), pericarditis (n = 4), and pulmonary artery hypertension (HTN) (n = 1). The most common diagnoses in the ED diagnoses were pericarditis (n = 58), myocarditis (n = 45), and pulmonary embolus (n = 13). All patients who presented through outpatient clinics were identifiable by the presence of at least 1 of the following: exertional chest pain (n = 20), abnormal physical exam findings (n = 12),

Table III. Utility and indications for cardiac testing in pediatric chest pain and syncope

Symptom/test	Indication for test	Usefulness	Conditions diagnosed
Chest pain			
ECG	Abnormal physical exam, ECG, exertional chest pain, or palpitations	Selected patients	Cardiomyopathy, myocarditis, pericarditis, with or without pulmonary HTN
Echocardiography	Abnormal physical exam, ECG, family history, or exertional chest pain	Selected patients	Anomalous coronary artery origins, cardiomyopathy, myocarditis, pericarditis, pulmonary HTN, left ventricular outflow obstruction
Troponin testing	Suspected myocarditis or pericarditis	Selected patients	Myocarditis, pericarditis, coronary ischemia
Ambulatory ECG	Chest pain and palpitations	Rarely useful for chest pain	Atrial or ventricular arrhythmia, intermittent heart block
EST	Exertional chest pain and exertional syncope or palpitations	Rarely useful	Coronary ischemia, exercise-induced asthma (if spirometry included)
Syncope			
ECG	All patients with syncope	First-line diagnostic test	Long QT interval, short QT interval, Wolff-Parkinson-White syndrome, cardiomyopathy, pulmonary HTN, Brugada, heart block, with or without ARVD
Echocardiography	Abnormal physical exam, ECG, family history, or alarming event history	Selected patients	Cardiomyopathy, pulmonary HTN, with or without ARVD, coronary anomalies
Exercise ECG/EST	Exertional syncope or exertional symptoms	Selected patients	Catecholaminergic polymorphic VT, long QT interval
Ambulatory ECG	Syncope plus palpitations, frequent symptoms, exertional syncope	Selected patients	Atrial or ventricular arrhythmia, intermittent heart block, reflex bradycardia
Tilt-table test	Very frequent, recurrent syncope or atypical syncope	Rarely useful	Confirmation of NCS (low specificity), postural orthostatic tachycardia syndrome

ARVD, arrhythmogenic right ventricular dysplasia; VT, ventricular tachycardia.

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