

# EVALUATING CHEST PAIN IN CHILDREN

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## INTRODUCTION

Chest pain in children represents approximately 5% of all referrals to paediatric cardiology services in a tertiary referral hospital.<sup>1</sup> It is a common symptom in children and adolescents. Although the aetiology is benign and non-cardiac in the majority of cases, chest pain as a complaint often leads to anxiety both in the child and parent, school absenteeism and restrictions on physical activity. Extensive public awareness on the association of chest pain with ischaemic heart disease and myocardial infarction in the adult has led to an erroneous assumption that the same holds true for the paediatric patient. In Driscoll's prospective study of chest pain in children, 52% interviewed by a child psychiatrist believed their chest pain to be cardiac in origin.<sup>2</sup>

## EPIDEMIOLOGY AND PRESENTATION

Paediatric chest pain as a primary complaint to the accident and emergency department has an occurrence rate of 0.249% and 0.288% per patient visit.<sup>2,3</sup> There is no gender predilection in patients presenting and in the largest prospective study to date the average age at presentation with this complaint is at a mean age of 11.9 years and a median age of 12.5 years.<sup>1,4</sup> In the same study by Selbst et al, chest pain was described as acute (less than 48 hours duration) in 43% of cases and chronic (greater than six months' duration) in 7%. In contrast to these figures, a study by Pantell et al in adolescents (mean age 16.2 years, 67% female) presenting to an out-patient's department setting described an incidence of chronic chest pain of 36%.<sup>5</sup>

## TYPES OF CHEST CAGE AND CHEST WALL PAIN

### Costochondritis

Costochondritis involves two to four contiguous costochondral or costosternal junctions. Usually it is unilateral. More commonly it involves the more cephalad joints. The pain is described as sharp, lasting several seconds to several minutes, and it is exacerbated by deep breathing. The joints are not inflamed and there is no swelling of the joints. Pushing

on the joint can reproduce the pain.

### Tietze's syndrome

This syndrome is quite uncommon in children. It involves inflammation of one costochondral junction. The area involved is warm, swollen and tender.

### Non-specific chest wall pain (idiopathic chest pain)

Non-specific chest wall pain may be the most common type of chest pain in children and adolescents. The pain is described as sharp. When asked to point to the pain, the patient will usually point to the centre of the chest or the infranipple area. The pain lasts several seconds to several minutes and is exacerbated by deep breathing. Sometimes squeezing the thoracic cage or gently pressing the sternum can reproduce the pain. Frequently the pain cannot be reproduced by palpation or pushing on chest structures. The costochondral and costosternal joints are non-tender.

### Precordial catch syndrome

Precordial catch syndrome consists of a brief (several seconds), sharp pain inferior to the left nipple or at the lower sternal border. It is frequently pleuritic and can be accentuated by bending forward. This syndrome frequently forces the patient to breathe shallowly. Its cause is unknown.

### Slipping rib syndrome

Slipping rib syndrome is quite rare, but it does produce rather intense pain, usually involving the eighth, ninth and tenth ribs. These ribs do not attach directly to the sternum but, rather, attach to each other. It has been postulated that trauma to the chest results in disruption of the connection of these ribs to each other and subsequent movement produces pain. A positive 'hooking manoeuvre' is said to be characteristic of this problem. The hooking manoeuvre is performed by the examiner putting his or her fingers under the inferior rib margin and pulling inferiorly. This action will reproduce the pain and may produce a clicking sound.

### Hypersensitive xiphoid syndrome

Hypersensitive xiphoid syndrome is uncommon in children. It can be diagnosed easily by digital pressure on the xiphoid reproducing the pain.

### Trauma and muscle strain

Obviously, injury to the chest wall produces chest wall pain. The history of prior trauma is suggestive and usually the pain can be reproduced by palpation of the chest wall. The same trauma, of course, could also produce myocardial contusion and, possibly, a haemopericardium, both of which can cause chest pain.

### Sickle cell disease

Sickle cell crisis can produce chest wall bone pain. In addition, chest pain in children and adolescents with sickle cell disease can be of cardiac and pulmonary origin.

## OTHER CAUSES OF CHEST PAIN

### Asthma

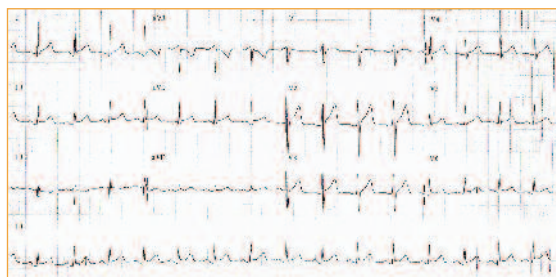
Wiens et al described findings of reversible airways disease in 72.7% of children who had been referred to a cardiology out-patients' clinic with no prior history of asthma or congenital heart disease.<sup>6</sup> The mean age of children who underwent the treadmill test was 12.4 years old. Inhaled salbutamol resulted in a subjective improvement in chest pain in 97% and an objective improvement in pulmonary function tests in 70%. Reactive airways disease should be considered in patients with chest pain, particularly if there is a history of asthma, eczema, shortness of breath with exercise, exercise-associated chest pain, exertional cough, wheezing or a family history of atopy.

### Infection

A number of infective processes can be associated with chest pain. Lower respiratory infections of all types can produce chest pain. Herpes zoster can produce chest pain frequently before the appearance of the typical vesicopustular rash.

### Pericarditis

Classically, pericarditis is associated with chest pain, whether due to an infectious aetiology or a non-infectious inflammatory cause. In general, the pain associated with pericarditis is described as fairly sudden in onset, located over the anterior chest wall and more severe than other forms of chest pain. It is frequently pleuritic in nature, being sharp and exacerbated with deep breathing. The pain may decrease in intensity when the patient sits up and leans forward and may radiate, especially to one or both trapezius ridges. Pericarditis is associated with typical electrocardiographic findings of generalised ST segment elevation.



ECG showing ST segment elevation in chest leads V2-V6 characteristic of acute pericarditis.

### Gastrointestinal

Gastro-oesophageal reflux disease (GORD) and oesophagitis can and frequently do cause chest pain. Additional gastrointestinal (GI) symptoms (e.g. reflux, heartburn) at the time of clinical presentation can greatly aid and are quite specific in the diagnosis of GORD as the aetiology of the chest pain. In an adult study by Mousavi et al, GORD was found in 44.8% of patients presenting with non-cardiac chest pain.<sup>7</sup>

Berezin et al, investigating idiopathic chest pain in children, described a GI cause in 78%.<sup>8</sup> Oesophageal endoscopy and manometry may detect abnormalities in children with chest pain, even without other GI symptoms. This was demonstrated in a study of 83 children with chest pain who underwent these procedures.<sup>9</sup> A total of 57% had normal oesophageal histology and normal motility. Among the others, 18% had oesophagitis on histology but normal motility, 15.6% had normal histology but gut dysmotility and 9.6% had both oesophagitis and dysmotility. It is likely that if GI causes of chest pain were sought more vigorously, this diagnosis would be made more frequently.

### Pneumothorax

Among patients with chest pain, pneumothorax is uncommon; however, a pneumothorax is very frequently associated with chest pain, and the abrupt onset of severe chest pain with or without dyspnoea should alert the clinician to this differential, especially as the clinical signs of a small pneumothorax can be difficult to appreciate.

### Pulmonary embolism

The most frequent discharge diagnosis in patients who initially presented with a clinically suspected (and subsequently out-ruled) pulmonary embolus is non-specific chest pain.<sup>10</sup> By the time of presentation to hospital, it can frequently be difficult to separate clinically pulmonary embolus from lower respiratory tract infection; however, careful history may give the clues to the initial symptoms of a pulmonary embolus being dyspnoea and/or pleuritic chest pain.<sup>11</sup>

In a retrospective study by Bernstein et al over a 15-year period, the incidence of pulmonary embolism in adolescents was reported as 78 per 100,000 hospitalised adolescents.<sup>12</sup> Females presented with twice the frequency of males. Common complaints were chest pain, dyspnoea, cough and haemoptysis. Major risk factors were oral contraceptive use and elective abortion in 75% of female patients and trauma in 67% of male patients. In children, the presence of a central venous catheter is the most frequent underlying risk factor.<sup>13</sup> Pulmonary angiography is still the gold standard in diagnosing pulmonary embolism with anticoagulation the mainstay of therapy.

## CARDIAC CAUSES OF CHEST PAIN

Cardiac conditions are a rare but potentially serious cause of chest pain in children. Cardiac disease is more likely if chest pain occurs during exertion and is recurrent. Most conditions will be associated with an abnormal cardiac examination or co-existing symptoms. In patients with known heart disease, chest pain may indicate progression of the underlying condition.

- Severe left ventricular outflow tract obstruction caused by aortic stenosis (subvalvar, valvar or supra-valvar), obstructive cardiomyopathy or coarctation of the aorta.
- Aortic root dissection associated with Marfan syndrome, Turner syndrome, Ehlers-Danlos syndrome, chronic systemic hypertension, homo-cysteinuria, rare familial aortopathies or cystic medial necrosis.
- Pericarditis and myocarditis, in which chest pain typically occurs with concomitant pericarditis.
- Coronary artery abnormalities, including congenital disorders or acquired conditions (e.g., coronary aneurysm or stenosis following Kawasaki disease; coronary stenosis after coronary re-implantation following arterial switch operation). Anomalous origin of the left coronary artery from the main pulmonary artery (ALCAPA) usually presents in infancy but can become symptomatic later in childhood.
- Ruptured sinus of valsalva aneurysm, a rare condition caused by congenital absence of media in the aortic wall behind the sinus of valsalva. The aneurysm typically ruptures into the right ventricle or right atrium, leading to intra-cardiac shunting and myocardial ischaemia.
- Tachyarrhythmias (e.g. supraventricular tachycardia with or without underlying Wolff-Parkinson-White syndrome, ventricular tachycardia).
- Coronary thrombosis and acute myocardial infarct can occur in premature arteriosclerosis, paradoxical embolus or hypercoagulable state.

## MITRAL VALVE PROLAPSE

Whether mitral valve prolapse (MVP) is associated with chest pain is controversial. Greenwood et al studied

331 children with isolated MVP for one month to eight years (mean 2.7 years).<sup>14</sup> Chest pain developed in 12 children. Ohara et al studied the incidence of symptoms in 108 children with MVP.<sup>15</sup> Chest pain was the most common symptom, occurring in 11 children (10.2%). The chest pain was non-exertional, located in the left chest and intermittent. More recently, Van der Ham et al describes a cohort of 45 children with echocardiographically-proven mitral valve prolapse.<sup>16</sup> There was no statistical difference in incidence among the sexes. The most commonly reported symptoms were shortness of breath and fatigue and not chest pain. Most of the children were asymptomatic.

## TOXIC EXPOSURE

Exposure to vasoconstrictive agents, such as cocaine, can cause chest pain that is likely ischaemic in origin. Hollander et al described a prospective cohort of 246 adult patients (median age 33 years) who had chest pain following cocaine use.<sup>17</sup> A total of 5.7% had suffered myocardial infarcts. Chest pain began a median of 60 minutes after cocaine use and persisted for a median of 120 minutes. Chest pain was most frequently described as substernal (71.3%) and pressure-like (46.7%)



## MEDICAL EVALUATION

The evaluation of chest pain requires a thorough history and careful physical examination.

The details of the history should be obtained, paying specific attention to the onset, frequency, and precipitating and relieving factors, as well as the characteristic duration and location of the chest pain. Associated features that would heighten suspicion for a

true cardiac aetiology would be exercise intolerance, palpitations or shortness of breath with activity, presyncope or syncope, or a family history of congenital heart disease or sudden cardiac death. In addition, it may be helpful to know whether other family members have chest pain, such as a parent or grandparent who experiences angina. This information might heighten the concerns of chest pain in the child.

A complete cardiovascular, respiratory and abdominal examination should be performed. The physical examination of the patient presenting with chest pain should initially focus on the vital signs. After documenting a stable regular heart rate and rhythm, respirations and blood pressure, a thorough physical examination should focus on finding non-cardiac causes. The initial evaluation should include inspecting for trauma and bruises or abrasions on the chest wall. Palpation should focus on bony abnormalities and localised chest swellings and on the site of the pain indicated by the patient. There should be an attempt to reproduce the pain by palpation of the location indicated by the patient. Reproducible pain, particularly at the costochondral junction or over a rib, points to costochondritis as the aetiology of the pain. A cardiac cause of chest pain may be suggested by auscultation of abnormal heart sounds or a cardiac murmur or abnormal pulse or blood pressure. Signs of left ventricular outflow obstruction include a systolic ejection murmur at the right upper sternal border and occasionally along the left sternal border. Co-actation of the aorta is associated with elevated blood pressure in the arms and a lower blood pressure in the legs. If the co-actation is long standing (present for more than five to seven years), collateral vessels may form that connect the upper and lower portions of the aorta; these vessels create a continuous murmur over the lateral aspect of the ribs.

In patients with pericarditis, pain increases when manual pressure is applied to the sternal region. The pain typically improves with sitting up and leaning forward. Signs of pericarditis depend upon the size of the pericardial effusion. Patients with a small effusion typically have an audible pericardial friction rub, caused by rubbing together of the inflamed parietal and visceral pericardial surfaces. The rub is often continuous in systole and diastole. It is easier to hear with the diaphragm of the stethoscope when the patient is sitting and leaning forward. A rub will not be heard if the effusion is large because the two pericardial surfaces of the pericardium are not in contact with each other. A large effusion may result in cardiac tamponade, manifested by a narrow pulse pressure, elevated pulsus paradoxus (>10mmHg), elevated jugular venous pressure, distant heart sounds, hepatomegaly, ascites, and peripheral oedema.

Signs of MVP are a constant, mid-systolic apical click and, occasionally, an apical systolic murmur of mitral regurgitation. Both auscultatory findings are more prominent when the patient is in the standing rather than supine position.

In most cases, the aetiology of the pain will be apparent after the history and physical examination.

## DIAGNOSTIC TESTS

Most patients with chest pain have a normal physical examination or findings consistent with a musculo-skeletal aetiology. Further investigations are not needed in those cases.

Diagnostic studies may help establish a diagnosis in patients with abnormal physical findings or with associated symptoms that suggest organic disease. Although cardiac causes of chest pain are uncommon in children, patients with dyspnoea, palpitations, anginal pain and pain with exertion that cannot be attributed to respiratory disease or syncope should be referred to a paediatric cardiologist for further evaluation.

- A chest x-ray may show cardiomegaly, pulmonary vasculature, infective infiltrates, hyperinflation and pneumothoraces.
- An electrocardiogram can aid in the diagnosis of arrhythmias, left ventricular outflow tract obstruction, pericarditis, ALCAPA, pulmonary hypertension and pulmonary embolus. If an arrhythmia is intermittent, a 24-hour Holter monitor or King of Hearts event recorder may be needed.
- In diagnoses of suspected cardiac aetiology, an echocardiogram can confirm cardiac structure, pericardial effusions and tamponade, cardiac function, anomalous coronary anatomy, sinus of valsalva aneurismal rupture and aortic root pathology.
- GI evaluation in children with chest pain should be performed under the remit of a paediatric gastroenterologist. Investigation of the upper GI tract may reveal histological oesophagitis or gut dysmotility.
- Other tests should be based upon associated signs and symptoms and clinical suspicion. Further evaluation may include pulmonary function testing, a ventilation-perfusion scan, cardiac catheterisation, exercise stress testing, a full blood count, serum reactive markers, appropriate cultures and toxicology screening.

## OUTCOME OF CHILDREN WITH CHEST PAIN

Two studies of the outcome of chest pain in children have been reported. Selbst et al reported on the outcome of an initial 407 children with chest pain seen in the accident and emergency department.<sup>18</sup> Thirty-four per cent of the original diagnoses were altered, usually in favour of a non-organic aetiology. A new organic aetiology was uncovered in only 12 of 149

cases, with only one having a heart abnormality (MVP). Chest pain resolved in 57% of those followed. Driscoll et al had a similar rate of resolution of chest pain in his cohort of 43 patients, with 60% pain-free four to eight weeks later on telephone follow-up.<sup>2</sup> Of note, in the group of Driscoll's patients with a diagnosis of idiopathic chest pain, only 30% of them had resolution of chest pain at follow-up.

## IN SUMMARY

The primary role of the evaluating cardiologist is to rule out the unlikely prospect of serious cardiac pathology. An equally important role is to provide reassurance and support to the patient and family. Most patients will not have a serious underlying medical problem. A thorough and thoughtful history and physical examination help allay fears and are important in reassuring the patient and family.

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