

Recurrent chest pain in the well child

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INTRODUCTION

Chest pain is a common symptom in paediatric practice and a frequent cause of attendance to general practitioners and hospital outpatients' departments. Public education campaigns have emphasised its potentially serious cause in adults, namely 'a heart attack', leading to heightened concern when it occurs in a child.^{1,2} However, in children it is typically benign and self-limiting, not usually a manifestation of organic disease, and rarely cardiac in origin.¹⁻¹¹ The patients are often referred by their general practitioner or local accident & emergency department to general paediatricians, respiratory paediatricians or paediatric cardiologists. Chest pain is in fact the second most common cause of referral to paediatric cardiologists in the USA,⁶ murmurs being the largest.¹² The exact cause of chest pain often remains undiagnosed¹³ and is labelled as 'idiopathic'.^{1,3-6,9,11,14}

There are a number of chronic conditions known to be associated with recurrent chest pain, for example cystic fibrosis and sickle cell disease. This article will not discuss these conditions as the underlying diagnosis is usually known. There are a number of acute causes of chest pain, for example pneumothorax or empyema, that are not usually recurrent and where the child is often unwell, so they will also not be discussed. Of course all recurrent cases will, at one point, have to present for the first time, so acute causes will sometimes need to be considered at the initial presentation. This article will highlight the most likely causes of recurrent chest pain in a child who is otherwise well, although the full differential diagnosis is long (table 1).

CAUSES OF RECURRENT CHEST PAIN

There are a number of causes of recurrent chest pain broadly divided into the following categories: cardiac, respiratory, gastrointestinal, musculoskeletal, psychological, miscellaneous, and idiopathic (unknown). The frequency of these causes varies widely between studies.⁶ Generally, idiopathic pain is the most common cause^{1,3-6,9,11,14} followed by musculoskeletal pain, while cardiac pain is the least common.

The gender of the child is not usually relevant to the cause of the pain,⁵ although a psychological cause is more common in girls.^{4,5} Age does play an important role — chest pain in children <12 years old is more likely to be due to organic disease, whereas psychogenic disease is more common in teenagers.^{4,5}

Cardiac

Chest pain, particularly originating in the left precordium, is often assumed by the patient to be cardiac in origin and may even be described

as 'heart pain'. A cardiac cause, although potentially serious, is among the least common causes of chest pain in an otherwise healthy child.¹⁻¹¹ Most children with congenital heart disease do not have chest pain, and conversely, most children with chest pain do not have congenital or acquired heart disease. Cardiogenic pain is usually crushing, left-sided, radiating to the left arm, sometimes with paraesthesia, brought on by exercise and settling at rest, and associated with sweating and nausea. Pericarditic pain differs and is described below. Cardiac chest pain may be precipitated by arrhythmia, acquired heart disease and underlying congenital defects.^{9,10}

Arrhythmia

Children with arrhythmias will occasionally present with chest pain, but normally have tachycardia or an irregular heart beat by history and/or examination.

Acquired heart disease

Acquired lesions can produce chest pain. Pericarditis frequently presents with non-specific chest pain and can follow a viral illness, or be part of rheumatic fever. It is almost always associated with fever and other signs of inflammation and the children are unwell. The pain is often sharp, typically sternal or substernal and worse on deep breathing, coughing or leaning forward.¹⁰ Endocarditis, cardiomyopathies, myocarditis and rheumatic fever are more likely to present with other classic symptoms such as fevers or exercise intolerance, but also can be associated with chest pain. Acquired coronary artery lesions due to Kawasaki disease, accelerated atherosclerotic coronary artery disease and cocaine use,¹⁵ can produce true ischaemic chest pain. Pulmonary hypertension is a rare cause of chest pain; these patients normally have fatigue, exercise intolerance, palpitations and/or syncope.

Congenital heart disease

The vast majority of structural cardiac abnormalities are not associated with chest pain. Patients who have recently undergone cardiac surgery may have postpericardiotomy syndrome. Lesions such as aortic stenosis, that can lead to decreased myocardial perfusion and ischaemia, may cause exercise-induced chest pain. Other less common causes include aortic aneurysm with dissection (as seen in aortopathies such as Marfan syndrome, bicuspid aortic valve or coarctation of the aorta), mitral valve prolapse or congenital coronary artery abnormalities (such as anomalous origin or course).

Respiratory

Respiratory pain can originate from the main airways, parietal pleura, chest wall or diaphragm. It

Table 1 Causes of chest pain by site of origin

Cardiac	Structural lesions Acquired/inflammatory conditions (eg, vasculitis, cardiomyopathy) Arrhythmias
Respiratory	Infection Pleural disease Asthma Foreign body Pneumothorax, pneumomediastinum Inhalant irritation Dysfunctional breathing Thoracic malignancy
Gastrointestinal	Gastro-oesophageal reflux Oesophagitis (reflux, eosinophilic) Gastritis Oesophageal spasm Achalasia Oesophageal foreign body Pancreatitis Subdiaphragmatic abscess
Musculoskeletal	Slipping rib syndrome Tietze's syndrome Costochondritis Vertebral deformities or collapse Myofascial trauma or strain
Miscellaneous	Precordial catch syndrome Stitch Breast development Herpes zoster

should be remembered that respiratory pain can occasionally present as abdominal pain (eg, lower lobe pneumonia), and due to phrenic nerve innervation diaphragmatic irritation can present as ipsilateral shoulder pain. Major airway pain is usually retrosternal, while pleuritic pain is typically sharp and well localised laterally. Respiratory pain is classically characterised by pain on coughing and respiratory disease is often suggested by other symptoms such as cough, wheeze and sputum production, and clinical signs such as asymmetrical air entry, wheeze or crepitations.

Asthma and exercise-induced bronchospasm are common causes of respiratory pain, although it is more often described as chest tightness or discomfort.^{13 16 17} Asthma-associated chest pain is usually benign and may be due to dyspnoea, hyperinflation, cough or muscle strain; rarely it is secondary to more serious acute causes such as a pneumothorax or pneumomediastinum.¹⁶ Children with exercise-induced bronchospasm may have no wheeze, even if they have significant shortness of breath or chest pain,¹³ and a prevalence of 10–20% in athletes has been suggested.¹⁸

Infectious causes such as tracheitis, pneumonia, pleuritis or parapneumonic effusion/empyema are also common causes of respiratory chest pain. More rarely pleurodynia (pleuritic pain without an effusion) due to coxsackie virus (Bornholm's disease) is described.^{16 19} However, in these cases the children are clearly unwell.

Gastrointestinal

Gastrointestinal causes of chest pain are relatively infrequently diagnosed,^{1 5 9 14} however are often confused with pain originating in other sites. Chest pain and abdominal pain are easily confused in smaller children, and even in adults cardiac pain and oesophageal pain can be difficult to distinguish as they have very similar autonomic nerve pathways.^{14 20} Over half of adults with angina-like pain and normal coronary arteriograms are found to have oesophageal disorders.^{14 20}

Although chest pain can be caused by any site along the gastrointestinal tract,²⁰ oesophagitis (reflux or eosinophilic) and gastritis are probably the most common causes.^{8 14} Pain from these sites often presents with the classic symptom of heartburn although may be non-specific.¹⁴ Oesophageal spasm is particularly associated with chest pain,¹⁴ and may be due to an oesophageal motor disorder and/or a primary motor disorder.²⁰ Pain is often associated with posture and eating, and is usually felt in the epigastrium, retrosternally, at the chest bases or at the shoulder tip. Pain associated with dysphagia (difficulty swallowing) is a strong pointer to an oesophageal cause. Tenderness in the epigastrium is a particularly good sign that the pain may be of gastrointestinal origin.⁸ Abdominal/subphrenic abscesses may cause diaphragmatic irritation with chest pain that radiates to the shoulder or lower chest,¹⁶ but the child is unwell. One of the difficulties in diagnosing gastrointestinal chest pain is that the tests are typically invasive and disliked by the children.¹¹

Musculoskeletal

Musculoskeletal chest pain tends to be sharp, well localised and brief, lasting seconds to minutes. It is usually exacerbated by movement of the affected part and so is often worse on breathing or coughing. Localised tenderness is the dominant feature,²¹ and needs to be distinguished from pleuritis, where there is often significant muscular irritation and resultant pain. There may be a history of trauma such as a strain or recent participation in active sports such as swimming, tennis or trampolining.

Fam and Smythe²¹ have categorised localised musculoskeletal chest wall pain into four groups — (1) arising from ribs and articulations, (2) arising from sternum and articulations, (3) arising from myofascial structures, and (4) arising from the thoracic spine, spinal cord and spinal nerves.

Ribs and articulations

Tietze's syndrome. This is due to an isolated painful, swollen costochondral junction that the child localises precisely.²² The second and third junctions are more commonly affected, but lower ribs can be also affected in children.²² The affected area is tender and swollen but not hot.²¹ The cause is usually unknown but may be related to trauma, a history of violent coughing or an upper respiratory tract infection.^{21 22} Investigations (including blood inflammatory markers) tend to be normal, but it is important to exclude septic arthritis, osteomyelitis or malignancy (although in these cases the child will be unwell).²² Chest radiographs and bone scans tend to be normal,²² although a chest CT scan may show osteochondritis of the rib. It can usually be diagnosed on clinical grounds alone however. It tends to run a self-limiting course, typically resolving within a few weeks or months.^{21 22} Treatment is anti-inflammatory medication or occasionally an intercostal block if the pain is severe.^{21 22}

Costochondritis. This differs from Tietze's syndrome in that it is often at multiple sites and there is no swelling.²¹ The pain usually involves the costochondral or costosternal areas of the second to fifth costal cartilages.^{3 21} Its aetiology is unknown but it may be viral or trauma related.^{3 21} The condition is self-limiting but can last for many months.³ Treatment is rest and non-steroidal anti-inflammatory drugs.^{3 21}

Rib trauma. The history is obvious and it is more common in boys.

Slipping rib syndrome. The 8th to 10th ribs do not attach to the sternum directly but to each other via fibrous tissue,

allowing mobility but at the cost of a susceptibility to trauma.²³ Inadequacy or rupture of the fibrous attachments due to direct trauma, or indirectly due to lifting can allow the costal cartilage tips to curl upwards and press on the intercostal nerves.^{21 23 24} The pain can last several months,²¹ and is often worse in situations that cause forceful upward movement of the costal cartilages, such as horse riding; it is also worsened by flexing of the trunk, heavy lifting and even stretching or coughing.²³ The pain can mimic biliary or renal colic.²⁴ The main diagnostic test is the hooking manoeuvre — the drawing forward of the lowest costal cartilages, which reproduces the symptoms and gives a clicking sensation.^{21 24} A chest radiograph is of no value except to rule out other diagnoses.²³ Treatment is analgesia and sometimes local anaesthetic infiltration, or in extreme situations surgical resection of the affected cartilage.^{21 23 24}

Sternum

Sternoclavicular joint pain is worse on shoulder shrugging. Pain and tenderness can also originate from the manubriosternal joint or the xiphisternal joint and xiphoid cartilage.

Myofascial

Pain from the intercostal and thoracic muscles is usually traumatic in origin.²¹ The muscles may be strained during active sports. It presents as localised pain and tenderness over the affected muscles, and is worsened by muscle movement. Myositis due to bacterial and viral agents, such as Bornholm's disease,¹⁹ causes severe, sharp pains in the upper abdomen (more common in children)²¹ or lateral chest wall, with muscular tenderness.¹⁹ It tends to be short lived, lasting 3–7 days.²¹

Spine

Causes include trauma, tumours, infection, thoracic disc disease and arthritis.

Psychogenic

Psychogenic chest pain is a diagnosis of exclusion, and is not necessarily the same as idiopathic pain. It is seen in all ages, but has a higher incidence in teenagers,⁴⁵ particularly girls.^{45 25} Chronic pain of more than 6 months' duration is more likely to be due to psychological problems.⁵ Although there is usually no organic cause for the pain, patients often have a significant degree of functional impairment,²⁶ and for them the pain can be very real.^{16 27} In fact some psychogenic causes can cause genuine physical pain, and vice versa — the child with recent organic disease is at increased risk for stress-related psychological pain.¹⁶ Psychological and organic aetiologies may not always be mutually exclusive.¹⁶ Some patients may have minor physiological symptoms but these are made worse by the belief that they are evidence of severe physical illness. When a group of adolescents was asked what they were afraid of with regard to the cause of their pain, 56% said that they feared heart disease and 12% were worried about cancer; 70% of this same group believed that people of their ages could have heart attacks.¹

Psychogenic pain is much more likely where there is a family history of chest pain or heart disease⁵⁷ — a 'model' or example in the family.²⁷ Studies have shown that there is often a history of other psychological disorders such as panic disorder, anxiety or depression.^{9 16 26} Emotional stress is very common; adverse life events such as changes in the family structure (births, deaths, divorce, etc), school problems and family physical or psychological illness can often be identified as the stressor.^{7 27} A not infrequent situation is the keen sportsperson who has realised they are simply not good enough for top-level

competition, which can lead to psychogenic pain, perhaps as an excuse for being able to stop the sport. Our experience is the child is often accompanied by one or both parents who have strongly encouraged the child in the sport and who insist how much the child loves taking part.

This kind of pain is often fleeting or vague, or localised over the heart and/or left arm. It does not generally get worse on exercise and may occur at rest. Often the parents may be extremely anxious while the child shows a classic 'belle indifférence' seen in many psychosomatic disorders. The physical examination is usually normal.

Dysfunctional breathing. A very common form of psychogenic pain, again particularly common in adolescents,²⁸ is dysfunctional breathing or hyperventilation. It has been suggested that hyperventilation can cause up to 20% of all chest pain in adolescent patients.²⁸ While the origin of the breathing dysfunction is psychological, it can cause very real physical pain. A resultant hypocapnoeic alkalosis can cause coronary artery vasoconstriction, albeit only after up to 30 min of deep breathing.^{1 29} Deep breathing can also cause stomach distension due to aerophagia, spasm of the left hemidiaphragm and transient arrhythmias.¹ Experienced physiotherapists can often make the diagnosis and after retraining the child to breathe 'properly', the symptoms usually disappear. The children often have poor posture accompanying the problem which also needs to be corrected.

Miscellaneous

Breast development

Breast growth during puberty can lead to breast pain in girls. Trauma and mastitis may also lead to localised pain.¹⁶ Boys are not immune to pain from this — gynaecomastia may cause discomfort, even when something as minor as clothing rubbing over the skin occurs.¹⁶

Herpes zoster

Shingles can cause an intercostal neuralgia with a severe sharp, stabbing pain along the area of the nerve, as well as localised tenderness. The pain is often worse on movement, deep breathing and exposure to cold.¹⁶ The pain tends to resolve when the vesicles begin to heal, but occasionally post-herpetic neuralgia may require regional nerve blocks or even surgical intervention.¹⁶ The child is unwell and the rash obvious.

Precordial catch

This is a self-limiting, well-localised, very brief, sharp pain in young healthy individuals — sometimes called Texidor's twinge.^{21 30} It is usually felt in the precordial area but can also occur under the left breast or by the left sternal border, or other sites.^{21 30} It is most common in those aged 6–12 years old.³¹ The origin of the pain is uncertain, but it may come from the parietal pleura (the visceral pleura has no pain receptors) or be due to a muscular spasm.²¹ It is certainly not of cardiac or pericardial origin.³¹ The pain can occur at rest or on exercise, and generally lasts from 30 s to 3 min — it rarely lasts for longer than 1 min.^{21 30 31} The pain does not radiate and is usually made substantially worse on deep inspiration.³¹ The description of this pain is usually so classic that it can be diagnosed on history alone. There is no local tenderness and the physical examination is completely normal. No investigations are necessary. There is normally a sudden and complete resolution of the pain but the pain can sometimes be helped by a change of posture or even forced deep inspiration.^{21 30 31} Usually, however, no treatment is required.³¹

Stitch

A stitch is classically a pain in the lower chest or side of the abdomen occurring during exercise, that is worse after eating or drinking. Its cause is uncertain but may be due to an engorged gut pulling on visceral ligaments where they attach to the diaphragm, or ischaemic pain in the diaphragm due to blood shunting to the gut and limb muscles during exercise.³² Treatment involves delaying exercise for 2–3 h after eating, the wearing of a light, wide belt around the waist and the taking of small amounts of fluid regularly while exercising, bending forward to tighten the abdominal muscles, and breathing through pursed lips.³²

MAKING THE DIAGNOSIS

In keeping with the classical mantra taught at medical school, the most useful diagnostic tools are a careful history and examination.^{7 11 21} In many cases this is all that is required to identify the cause of the pain, removing the need for investigations.

HISTORY

This is the most important part of making the diagnosis as there are often few physical findings and investigations are usually normal.^{2–6 9 11} The pain can occur as an isolated symptom or in association with other symptoms, depending on the cause (table 2). Questions should be directed at the location (including any radiation), severity and character (sharp, dull throbbing, crushing, etc) of the pain. Important also are how long it lasts and any exacerbating or alleviating factors. Benign pain tends to have a sudden onset at rest, lasting seconds to minutes with a sharp character and is often well localised to the chest wall. The absence of a crushing, poorly localised pain with radiation to the left arm, nausea, sweating and association with exercise is reassuring and effectively excludes an ischaemic cause.

The frequency and overall duration of the pain is important because pain of >6 months' duration is more likely to be psychogenic.⁵ In one study that included questions about duration of pain, 36% was for less than 1 day and 33% had lasted from

2 days to 1 month.⁴ A typical presentation is of a child who complains of pain that has lasted (intermittently) for weeks or months but who attends the clinic looking completely well. Chest pain that wakes children from sleep should always be taken seriously as it suggests organic pain (but not necessarily a serious cause).⁵ A history of fever, weight loss or fatigue is also a key pointer to potentially serious pathology.

Patients often have a history of asthma or a previously diagnosed heart murmur.⁴ Similarly a family history of premature ischaemic heart disease, sudden death, arrhythmia and cardiomyopathy should be sought, particularly as patients and families often worry most about cardiac pain, and it is worth finding out what the child and parents are most concerned about. The social history may reveal psychological and emotional factors although these may not be revealed during the initial consultation; they can of course still occur in the presence of organic disease. A drug history is important as some medications can cause gastric irritation⁶ or be associated with embolic disease (eg, the oral contraceptive pill).

EXAMINATION

There may be few physical findings¹⁶ and the physical examination is normal in 37–60% of cases in different series.^{1 5 9 11} Despite this, non-organic pain remains a diagnosis of exclusion and so other reasons for the pain should be sought first. A thorough examination of the cardiac, respiratory and gastrointestinal systems should be performed as well as palpation of the chest wall; if normal this is usually very reassuring to all concerned.

INVESTIGATIONS

Investigations are not often helpful in making the diagnosis^{2–6 9 11} and should be tailored to any abnormalities or suspicions identified after the history and examination. Table 3 gives a comprehensive list of possible investigations, most, if not all of which, will not be required following a careful history and physical examination. Very frequently any abnormal results from investigations are either mild or have been previously noted.^{4 5}

Chest radiographs are also usually normal if there are no other suspicious features from the history or examination and are rarely diagnostic.^{2 4 9–11} They may still be useful though as a normal radiograph often helps to reassure patients and their family. ECGs and echocardiograms have almost always been normal in most studies of paediatric chest pain.^{2 3 5 7 9–11} An ECG can, however, show evidence of ischaemic changes, structural abnormalities or rhythm disturbances (including changes such as the δ wave in Wolff–Parkinson–White syndrome) and should be performed if these are suspected. Simple spirometric lung function or exercise testing with bronchodilator responsiveness may be useful to exclude exercise-induced bronchospasm.

MANAGEMENT

Where an organic cause is found for the pain then specific treatment is commenced. If there is a strong suspicion of a cardiac, respiratory or gastroenterological cause then referral to the appropriate specialist is usually necessary (table 4). Certain symptoms and signs suggest referral to cardiologist is essential (box 1). Depending on the circumstances, a therapeutic trial may be warranted, for example use of an inhaled bronchodilator 15–20 min before exercise, use of adequate antireflux medication for 4 weeks, or regular non-steroidal anti-inflammatory medication for 1–2 weeks.

Table 2 Clues in history to causes of chest pain

Cardiac	Association with exercise Syncope, dizziness, vertigo Colour change (pallor, cyanosis) Palpitations Sweating and nausea Shortness of breath Family history of premature ischaemic heart disease, sudden death, arrhythmia and cardiomyopathy Illicit drug use, particularly cocaine
Respiratory	Worse on inspiration, exercise History of asthma Sputum, haemoptysis
Gastrointestinal	Worse with eating or posture Belching Tenderness or pain — chest base, shoulder tip, retrosternal, epigastric Heartburn, water-brash Dysphagia Haematemesis, melaena
Musculoskeletal	Pain elicited by palpation, squeezing of chest wall Pain worse on movement, breathing, coughing History of trauma, sprain, strain Well-localised pain
Psychological	Chronic pain Pain as isolated symptom Family history of heart disease Stressful life events

Table 3 Investigations to be considered for chest pain

Cardiac	ECG Chest radiograph Echocardiogram Exercise tolerance test 24 h ECG monitoring Serum fasting lipids Cardiac enzymes, troponins
Respiratory	Chest radiograph Spirometric lung function testing Exercise testing and bronchodilator responsiveness Very occasionally — CT chest scan
Gastrointestinal	24 h pH probe Isotope milk scan Impedance studies Barium swallow Endoscopy Oesophageal manometry
Musculoskeletal	None
Psychological	None

It must be stressed that as most causes of chest pain can be determined from a careful history and examination, few if any of the investigations in this comprehensive list need to be undertaken in practice.

Table 4 Recommendations of when to refer to a paediatric subspecialist

Cardiology	Presence of other significant cardiac symptoms and/or signs, particularly colour change (cyanosis or pallor), syncope, dizziness, palpitations, sweating, nausea, dyspnoea Presence of a crushing, poorly localised pain with radiation to the left arm Exercise-induced pain (assuming not due to asthma) Strong family history of heart disease (especially hypertrophic obstructive cardiomyopathy, arrhythmias, sudden death or premature ischaemic heart disease) Suspicion of Marfan syndrome or other connective tissue disorders
Respiratory	Presence of other significant respiratory symptoms and/or signs, particularly breathlessness, cough, noisy breathing, sputum production, haemoptysis When asthma is likely but has not responded to standard asthma therapy, especially when pain is exercise induced
Gastroenterology	Presence of other significant gastrointestinal symptoms and/or signs, particularly dysphagia When gastro-oesophageal reflux is likely but has not responded to adequate reflux therapy

This assumes no underlying diagnosis has been made, as referral may be required depending on the presence of an identified cause. Referral may sometimes be necessary to offer further reassurance, even if a significant diagnosis is unlikely.

Where an organic cause is not considered to be present, initial management involves understanding of the child's and family's worries, with patient explanation and reassurance that the problem is common and benign with a good prognosis.³³ Often reassurance that the pain is unlikely to be cardiac can reduce anxiety and decrease the severity of the symptoms.⁷ Physiotherapists may be most helpful in retraining breathing patterns when dysfunctional breathing is the issue. They can also help children 'withdraw with honour' so they can point to a treatment that helped them recover. Occasionally a clinical psychologist may need to be involved. Specialist treatment such as cognitive behavioural therapy may be required, especially for those with continuing symptoms and disability, particularly if they are associated with other psychological problems.³³

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Box 1 The following are symptoms and signs that suggest referral to a cardiologist is mandatory:

- ▶ Crushing left-sided precordial pain
- ▶ Pain radiating to left arm or jaw ± paraesthesia
- ▶ Onset with exercise, subsiding at rest (with asthma excluded)
- ▶ Dizziness or syncope
- ▶ Nausea or sweatiness
- ▶ Palpitations
- ▶ Cyanosis
- ▶ Abnormal cardiovascular examination
- ▶ Known structural heart disease or pulmonary hypertension

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