

# Review Article

## Approach to Chest Pain in Children and Adolescents

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### Abstract

Chest pain in children seems to be is one of the most routinely seen presenting complaint that brings lot of anxiety and stress to patients, parents and treating physician, though most of them have a benign cause .

A simple history taking and detailed physical examination is sufficient in most of the times to identify the origin as well as the source which causes chest pain and diagnostic testing and cardiologist referral can be reserved for selected cases as routine referral still increases the family concern .

This article aims to acknowledge patients, parents fear and to give appropriate reassurance to the patients by the physician after differentiating benign from a serious cardiac condition as the possible etiology for the chest pain.

**Key Words:** Chest, Cardiac ,Benign

### Introduction

Chest pain in children appears to be one of the most frequent cause for referral to the physicians and emergency department accounting for >6,50,000 visits every year ,especially noted in patients age group of 10-21 years.<sup>1</sup> It is also found to be a most common etiology after murmur, for referral to a pediatric cardiologist.<sup>2</sup>

Fortunately chest pain experienced by children and adolescents is found to be rarely associated with cardiac cause, with the prevalence being less than 6%.<sup>3</sup>

Although chest pain does not indicate serious cardiac cause in most pediatric cases, the anxiety surrounding chest pain most often is due to its association with cardiac ischemia in adults, and a society with high prevalence of atherosclerotic cardiovascular disease. and media showing sudden cardiac deaths.

There is a long list of etiologies for chest pain in children, most causes seem to be benign and also self limiting and need only symptomatic treatment and reassurance. The physicians should be aware of the differential diagnosis for chest pain in children and should make prompt efforts to determine the cause by simple history taking and physical examination before making a referral for diagnostic testing, cardiology referral and also for providing reassurance to the patients and parents .

This article helps us in understanding the various cardiac and non cardiac causes for chest pain, its approach, evaluation and need for referral to the cardiologist.

***Despite the low possibility of association of cardiac disease, chest pain remains a most significant complaint in most out-patients of pediatric cardiology which needs prompt evaluation. The possible causes would be as follows:***

### Infectious or inflammatory causes:

Children with pericarditis, myocarditis, or endocarditis usually present with sharp retrosternal chest pain that worsens with deep breaths or when lying flat. Pericarditis, may be due to an infections like Acute rheumatic fever, coxsackie virus infection, ECHO virus, or associated with conditions such as collagen vascular disorder as in systemic lupus erythematosus, in uremia or due to neoplasm, or trauma to the chest. It can also occur as a part of the postpericardiotomy syndrome seen after heart surgery. Myocarditis again due to viral or rheumatic etiology usually occurs with pericarditis. Embolization and dissemination of infected vegetation will lead to various organ injuries and their associated symptoms.<sup>3,5</sup>

### Coronary artery abnormalities:

Chest pain as a result of myocardial ischemia occurs in patients with abnormal coronary artery anatomy, that includes congenital anomalies of the coronary artery, coronary artery fistulas, and sometimes stenosis or atresia of the coronary artery ostium.

Coronary anomalies may precipitate myocardial ischemia or sudden cardiac death. Children presenting with chest pain along with a history of cardiac surgery or transplant are found to be at risk of myocardial ischemia or transplantation rejection leading to accelerated coronary artery vasculopathy or tachyarrhythmias. In addition, heart surgeries that potentially affect the coronary arteries (Example: corrective surgery of transposition of the great arteries) increase the risk of developing coronary artery stenosis.

Patients with a history of Kawasaki disease and associated coronary artery aneurysms are known to be at higher risk for developing coronary artery stenosis, rupture, or sometimes thrombosis. The risk of coronary aneurysm is usually seen highest during the first 5 weeks after the diagnosis of Kawasaki disease.<sup>6</sup> Kato and associates, showed in his 10- to 21-year follow-up study, that 46% of patients with Kawasaki disease and who had history of giant coronary aneurysms were found to have stenosis or full complete obstruction and 67% had myocardial infarction, with a mortality rate of 50%. Risk factors as studied for developing coronary artery aneurysms, thrombosis, and stenosis were the extremes of age as early as < 6months , or older age (>5 years) at the time of diagnosis and males were more at risk.<sup>7</sup>

Coronary artery disease may also be encountered in patients who have a significant family history of hypercholesterolemia, but it is seen that children and adolescents rarely have enough obstruction so as to cause chest pain due to ischemia. Coronary artery disease presents in patients born with homozygous familial hypercholesterolemia within the first 2 decades, in contrast to those who have heterozygous familial hypercholesterolemia, in whom it presents commonly after the fifth decade of life. According to the American Academy of Pediatrics (AAP) guidelines for preventive health - care , every child needs to undergo a risk assessment for dyslipidemia usually beginning at 2 years of age, and assessment then has to be repeated every 2 years until the child is 10 years old and then annually.<sup>8</sup>

### Structural abnormalities :

Intrinsic or structural abnormalities of the heart such as septal defects, left - to - right shunts, or cardiomyopathies [dilated /hypertrophic]may present as chest pain, but are usually associated with other symptoms like palpitations, fatigue, exercise intolerance, dizziness, or exertional syncope.<sup>3</sup>

### Arrhythmogenic causes:

Young children who are not able to describe palpitations as a result of arrhythmias usually complain

of chest pain and point to the chest. In children under 12 years, the most common etiology for Supraventricular tachycardia is usually an accessory atrioventricular pathway, whereas in teenagers, it tends to be more of atrioventricular node re-entry tachycardia. Patients with congenital QT syndrome may present in the pre-teens or teenage years with seizure or syncope along with chestpain.<sup>3</sup>

### Syndromic causes:

Children who have Marfan syndrome, Turner syndrome, type IV Ehlers-Danlos syndrome, and homocystinuria are at a higher risk for a dissecting aortic aneurysm that presents with the sudden onset of severe chest pain.

**Table 1 - Differential Diagnosis for Chest Pain in Children:**<sup>3,9-12,21-23</sup>

| NON CARDIAC CAUSES FOR CHEST PAIN <sup>3,9-12,21-23</sup> |  |
|---|--|
| MUSCULOSKELETAL: [24-56%]                                 |  |
| Costochondritis   | ▪ Sharp stabbing , anterior chest wall pain ,involving costosternal and multiple costochondral junctions, usually unilateral, exaggerated by breathing movements, reproducible tenderness over 2 to 5 ribs is commonly seen. Self limiting ,with frequent exacerbation during adolescence. |
| Teitze syndrome   | ▪ Sharp pain localized to one costochondral junction usually affecting the second and third rib, seen in adolescents and young adults, presents as tender, swollen [1-4 cm mass] over the ribs.  |
| Chest wall deformity                                      | ▪ Seen in Marfans syndrome ,Ehler Danlos ,chest pain associated with pectus excavatum /Carinatum.  |
| Non specific /idiopathic chest wall pain                  | ▪ Sharp pain lasting for few seconds to minutes, localized to mid line of sternum sometimes infra mammary area is also involved, exacerbated on deep breathing and on manual pressure on the ribs without any signs of inflammation.   |
| Slipping rib syndrome                                     | ▪ Intense pain localized to lower chest area,due to trauma or dislocation of 8th and/or 9th and 10th ribs usually seen in athletes . It increases on sudden upward movement or on flexion of the trunk and by hooking manoeuvre Pain can be reproduced along with a clicking sound.        |
| Trauma  | ▪ Severe chest pain with shortness of breath and other features of contusion, hemothorax, hemopericardium.   |

| PULMONARY/RESPIRATORY CAUSES:[7-20%]                              |   |
|---|---|
| Bronchial asthma  | ▪ Chest pain is bilateral described as chest tightness, associated with wheeze and dyspnea. In exercise induced asthma, exercise itself causes chest pain even in absence of wheeze.  |
| Infections  | ▪ Pneumonia, pleurisy, bronchitis   |
| Pulmonary embolism  | ▪ Pleural effusion, empyema all cause acute chest pain associated with fever, dyspnea, grunting and clinically on chest examination they have pertaining signs .  |
| Pneumothorax :  | <p>▪ Acute severe chest pain , with dyspnea ,seen at risk individuals like hypercoagulability, immobilization, medications.</p> <p>▪ Seen in tall, thin built, asthma patients, substance abuse ,sudden sharp pain ,with dyspnea and radiating to ipsilateral shoulder ,hyper resonant note on percussion and decreased breath sounds on auscultation</p> |
| GASTROINTESTINAL CAUSES:[3-6%]                                    |   |
| Gastro esophageal reflux disease, oesophageal spasm, peptic ulcer | ▪ Retro sternal burning pain, water brash, ascending pain , pain associated with eating , sometimes presents with dyspepsia, epigastric tenderness present on examination.  |
| Cholecystitis   | ▪ Sharp, pain with fever, vomiting, tenderness over right hypochondrium   |
| PSYCHOGENIC CAUSES[1-9%]  |   |
| Anxiety / hyper ventilation                                       | ▪ Pain is often fleeting, vague,with history of recent stressful events, recurrent somatic complaints, sleep disturbances   |
| MISCELLANEOUS[ 4-11%]   |   |
| Herpes zoster Pleurodynia [devils-grip]: .                        | ▪ Seen in Coxsackie B infection, sudden episodes of sharp pain in the chest and abdomen   |
| Breast related conditions .                                       | ▪ Teenage boys present usually with gynaecomastia, post pubertal females present with mastitis, fibroadenosis   |

| HEMATOLOGIC & ONCOLOGIC CAUSES              |  |
|---|--|
| Acute chest syndrome [sickle cell disease]: | ▪ Severe, sharp pain with icterus associated with fever, dehydration, and dactylitis, hematuria                |
| Mediastinal/thoracic wall tumor: .          | ▪ Dull aching chest pain, with features of mediastinal syndrome like headache, facial edema, venous congestion |
| NEUROLOGIC CAUSES                           |  |
| Migraine                                    | ▪ Frequent episodes of headache with vomiting with aura.   |
| Spinal cord compression                     | ▪ Associated with motor or sensory deficits.   |
| IDIOPATHIC CAUSES [12-52%]                  |  |

**Table 2 - Cardiac Causes For Chest Pain** <sup>3,9-12,21-23</sup>

| Condition                                       | Presentation  |
|---|---|
| INFLAMMATORY                                    |   |
| Pericarditis                                    | <p>▪ Positional chest pain</p> <p>▪ Predisposing factors: Rheumatologic conditions, Malignancy, Mediastinal radiation, Infection (HIV, tuberculosis, viral), Renal failure, Recent cardiac surgery.</p> <p>▪ Cardiac rub Tachycardia / tachypnea, Distant heart sounds, JVP raised.</p> |
| Myocarditis                                     | <p>▪ Fever Viral prodrome, Short duration of symptoms, New onset heart failure symptoms.</p> <p>▪ Tachycardia, Tachypnea With or without gallop rhythm, Cardiovascular collapse</p>   |
| INCREASED MYOCARDIAL DEMAND OR DECREASED SUPPLY |   |
| Arrhythmia                                      | <p>▪ Palpitations, Syncope, Positive family history</p> <p>▪ Irregular rhythm</p>   |
| HCM   | <p>▪ Positive family history, Exercise intolerance Exertional chest pain, Syncope and / or arrhythmia</p> <p>▪ Dynamic systolic murmur on auscultation.</p>   |
| Dilated cardiomyopathy                          | <p>▪ Family history Decreased exercise tolerance, syncope , Heart failure symptoms</p> <p>▪ Gallop rhythm , Mitral regurgitation murmur</p>   |

|   |   |
|---|---|
| Severe left ventricular outflow tract obstruction   | <ul style="list-style-type: none"> <li>Exertional symptoms</li> <li>Exertional syncope</li> <li>Loud systolic murmur</li> </ul>   |
| <b>CORONARY ARTERY ABNORMALITIES</b>                |   |
| Anomalous coronary artery origin, coronary fistula. | <ul style="list-style-type: none"> <li>Exertional chest pain,</li> <li>Exertional syncope</li> </ul>  |
| Coronary ischemia                                   | <ul style="list-style-type: none"> <li>Predisposing conditions</li> <li>History of Kawasaki disease</li> <li>Cardiac surgery or heart transplant</li> <li>Systemic arteriopathy (Williams syndrome)</li> <li>Severe familial hypercholesterolemia</li> <li>Drug use: cocaine, sympathomimetics</li> <li>Anginal chest pain.</li> <li>Tachycardia, Tachypnea, New murmur or gallop on examination</li> </ul> |
| <b>MISCELLANEOUS</b>                                |   |
| Aortic dissection, Rupture of aortic aneurysm       | <ul style="list-style-type: none"> <li>Personal or family history of bicuspid aortic valve or connective tissue disorders (Marfan, Loey - Dietz, Ehlers-Danlos type IV, others)</li> <li>Acute onset sharp or tearing type of pain.</li> <li>Marfanoid body habitus</li> </ul>  |
| MVP, Atrial myxomas                                 | <ul style="list-style-type: none"> <li>Positive family history</li> <li>Thin built, thoracic wall deformity, mid systolic click with or without late systolic murmur</li> </ul>   |

**Table 3 - Findings from Various Studies Who Evaluated Only Cardiac Cause for Chest Pain .**

| Study [year]                        | Total no. of patients | Cardiac |
|-------------------------------------|-----------------------|---------|
| Danduran et al (2008) <sup>13</sup> | 263                   | 0%      |
| Saleeb et al (2011) <sup>14</sup>   | 3700                  | 1%      |
| Friedman et al (2011) <sup>15</sup> | 406                   | 5%      |

**Table 4 - Findings from Various Studies Which Looked Into Causes of Chest Pain.**

| Study [year]                           | Total no. of patients | Non Cardiac cause for chest pain | Cardiac cause for chest pain |
|--|-----------------------|----------------------------------|------------------------------|
| Lin CH [2011] <sup>16</sup>            | 103                   | 98%                              | 2%                           |
| Drossner DM [2011] <sup>17</sup>       | 4436                  | 99.4%                            | 0.6%                         |
| Carleen L. Hanson [2011] <sup>18</sup> | 135                   | 99.3%                            | 0.7%                         |
| Ahmet Sert et al [2013] <sup>19</sup>  | 380                   | 99.7%                            | 0.3%                         |
| P.Babu et al [2015] <sup>20</sup>      | 1126                  | 99%                              | 1%                           |

### Basic Approach <sup>3,21-23</sup> History :

- 1. Description of chest pain:** Typical anginal pain is retrosternal, compressive or burning type which can radiate to left arm, shoulder or jaw; associated with perspiration and worsens on exertion; sharp localising pain is usually due to musculoskeletal problems; Respiratory conditions like pneumonia can produce localised pleuritic chest pain worsening on deep breathing or coughing; acute severe pain can be due to either pulmonary embolism or a pneumothorax. Gastroesophageal diseases can produce a burning pain, but usually show a variation in intensity with food intake or may be associated with vomiting or abdominal pain
- 2. Past medical history:** Asthma, Sickle cell disease, Kawasaki disease, Congenital or acquired cardiac disease, hypercholesterolemia are to be ruled out to exclude specific conditions
- 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. Others :** History of trauma, intense physical activity, drug abuse (eg, cocaine), psychological stressors

### General Physical Examination:

- Check Vital signs: Temperature to be recorded, Pulse rate, rhythm, character, volume, radio femoral / radio radial delay to be noted and all peripheral pulses felt, blood pressure has to be measured in all 4 limbs and oxygen saturation to be checked.
- Asses the general appearance of the child like colour [any cyanosis/pallor], level of consciousness, any breathlessness and is there any evidence of anxiety / distress in the child is the child hyperventilating, are there any dysmorphic features.

- Range of movement tests of the arms may be done in order to elucidate any relationship to pain as seen in muscular strain.

### Systemic examination :

- On inspection of chest look for any signs of trauma to the chest, bruising, asymmetry of the chest and localised swelling.
- Palpate the chest to see for tenderness (particularly at the location where they described the pain), any crepitus, parasternal eaves or thrills felt . Hooking maneuver where we hook fingers under lower costal margin and pull anteriorly as this produces pain in slipping rib syndrome
- Auscultate the lung fields and note air entry bilaterally, any added sounds like wheeze, crackles and pleural rub.
- Auscultation of precordium usually done for abnormal loud second heart sound, systolic clicks or murmurs, gallops, and pericardial rub.

▪ On abdominal examination look for signs of tenderness (particularly in the epigastric region), any trauma and palpate to see organomegaly.

❖ STEP 3: Here the clinician looks for other psychogenic cause, other systems, idiopathic causes.

| DIAGNOSTIC TEST  | INDICATIONS  |
|--|--|
| CHEST XRAY   | Acute onset of severe pain, Pain that causes awakening from sleep, History of drooling, foreign body ingestion, Cough, Fever, Dyspnoea, History/signs of significant trauma, Abnormal pulmonary / cardiac auscultation.  |
| TRIAL ANTI - REFLUX MEDICATION / pH / IMPEDANCE MONITORING | Gastrointestinal - type pain, Epigastric tenderness  |
| ELECTROCARDIOGRAM  | Cardiac-type chest pain, Cardiac red flags [text] Pericarditic pain. Any abnormal chest sensation/pain in a preschool child, Palpitations, Abnormal cardiac auscultation or diminished pulses, Abnormal heart rate or rhythm, Family history of sudden death, inherited arrhythmias, cardiomyopathy or ICD/pacemaker insertion |
| ECHOCARDIOGRAPHY   | Abnormal physical exam, or ECG, family history, or exertional chest pain, Anomalous coronary artery origins, cardiomyopathy, myocarditis, pericarditis, pulmonary HTN, left ventricular outflow obstruction.   |
| TROPONIN TESTING   | Suspected ischemia; myocarditis or pericarditis  |
| AMBULATORY ECG   | Chest pain and palpitations.   |
| EXERCISE STRESS TEST                                       | Exertional chest pain and exertional syncope or palpitations   |

### Simplified Three Step Approach <sup>23</sup>

- ❖ Considering the vast etiology, simple three step approach which will help to arrive at a diagnosis is considered:
- ❖ STEP 1: Here, clinician looks for the three most common and frequently encountered cause for chest pain in children [45-65%]: Costochondritis, other musculoskeletal disorders and respiratory causes for chest pain after complete history and physical examination.
- ❖ STEP 2: Here, the clinician looks for a cardiac cause [0-4%], if no clue from Step 1, he asks for an X-ray, ECG, and cardiologist opinion following a comprehensive history taking and physical examination.

### Red flags that points to cardiac cause for chest pain that needs referral to cardiologist. <sup>3,21</sup>

- ♦ Any abnormal cardiac findings noted on Examination.
- ♦ Chest pain on exertion/strain
- ♦ Syncope on exertion /strain
- ♦ Any Chest pain associated with palpitations
- ♦ ECG showing any abnormalities
- ♦ Family history being significant like history of arrhythmias, sudden death, or genetic disorders.
- ♦ History of congenital heart disease or acquired heart disease in the past /present.
- ♦ History of cardiac surgery or interventions
- ♦ Any Orthotopic heart transplant, Any Implantable cardioverter defibrillators in situ
- ♦ History of Kawasaki disease, Connective tissue disorders

### Points to remember

Chest pain in children is most commonly due to non cardiac cause.

- ❖ Every child who presents with chest pain justifies a detailed evaluation with, the elaborate history and thorough physical examination which is usually sufficient to diagnose the cause of the chest pain.
- ❖ Family and the child are to be addressed and reassurance to be given about the benign nature of chest pain.
- ❖ Any symptoms that suggests myocardial ischemia or any abnormal cardiac finding on examination should pave the way for immediate referring the child to a pediatric cardiologist.

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