

The Role of Echocardiogram in Outpatient Paediatric Chest Pain

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ABSTRACT

Chest pain in children usually makes both patients and family anxious, but - in contrast to among adults - mostly is not caused by serious disease. American College of Cardiology published criteria for initial transthoracic echocardiography in outpatient setting.

Keywords: Appropriate, chest pain, echocardiogram, outpatient paediatric

ABSTRAK

Nyeri dada pada anak pada umumnya menimbulkan kecemasan baik pada anak maupun keluarga; tetapi biasanya tidak disebabkan oleh penyakit serius, lain halnya dengan pada orang dewasa. *American College of Cardiology* mengeluarkan *Appropriate Use Criteria* penggunaan ekokardiografi transtorakal pada pasien rawat jalan kardiologi. **Richo Wijaya, Tina C. L. Tobing. Peran Ekokardiogram pada Nyeri Dada Pediatrik di Poliklinik**.

Kata kunci: Appropriate, ekokardiogram, nyeri dada, pasien rawat jalan

INTRODUCTION

Chest pain in children and adolescents are common and produces anxiety in both patients and family. Although alarming to parents, chest pain in children usually is not caused by a serious disease, in contrast to chest pain in adults, which raises concern for coronary ischemia.¹⁻³ Chest pain may lead to emergency department (ED) visits (0.5% of all paediatric visits), school absences, and unnecessary restriction of activities.²

A recent review found that only 1.2% of patients presenting to a major paediatric cardiology centre for chest pain had identifiable cardiac etiologies.⁴ Pediatric chest pain account for more than 650,000 physician visits per year in patients 10 to 21 years of age.³ Most data on the frequency of causes of chest pain in children come from studies in paediatric ED and cardiology clinics. It accounts for approximately 0.3% to 0.6% of paediatric ED visits.² The two largest studies looked at a total of 8,136 children presenting to ED with chest pain and found that only 0.6 - 1% of these had a cardiac cause for their pain.⁵ Saleeb, et al, reviewed the records of 3,700 patients seen for chest pain and found

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only 1% had a cardiac etiology. The majority of patients were diagnosed with non-cardiac causes (99%), including idiopathic (52%), musculoskeletal (36%), respiratory (7%), and gastrointestinal (3%).⁶ A review reported that children younger than 12 years of age were twice more likely to have an organic cause, adolescents were 2.5 times more likely to have a psychogenic cause.¹

The first paediatric Appropriate Use Criteria (AUC) were recently published by the American College of Cardiology. It consists of common clinical scenarios for which a transthoracic echocardiogram is ordered in outpatient setting. Echocardiography is the most common modality used in the outpatient setting to assess children with suspected heart disease. However, it is known that echocardiography is not cost-effective or high yield for some common indications such as chest pain, syncope, and murmurs.⁷ The use of echocardiography has grown significantly over the past 15 years in both paediatric and adults in parallel with an increase in the use of all diagnostic imaging in the United States.⁴ A study reported that echocardiogram used in patients meeting the "rarely appropriate"

indication criteria is of little diagnostic utility and contributes to additional cost to patient and health care system.⁴

The purpose of the AUC was to guide physicians about "when and how often" to order a given procedure in an individual patient on the basis of current scientific evidence health care resources. The overall goals of the AUC are to evaluate patterns of care by physicians and to serve as a framework for appropriateness of care.⁷

Chest Pain in Children

Chest pain is a common symptom that results in visits to the ED or outpatient paediatric clinics.^{3,8} After cardiac murmurs, this is the second most common symptom that is referred to paediatric cardiolgist.^{3,8} Chest pain accounts for approximately 0.3 to 0.6% of paediatric ED visits.^{2,9} In EDs treating children up to 18 years of age, the median age for presentation with chest pain was 12 to 13 years. The reported male to female ratio is fairly even, ranging from 1:1 to 1.6:1. In adolescents, relatively more girls present with chest pain. Many ED studies report that the most children present with acute pain of less



than 1 day in duration. In contrast, a study in Turkey reported that 59% patients described pain more than 1 month in duration.⁹

The main causes of chest pain are idiopathic and musculoskeletal disease; most patients with chest pain do not require any special treatment. Moreover, patients who show symptoms of chest pain may not need to undergo all available tests, including echocardiography. Careful examination of medical history, performance to relevant medical tests, and physical examination are required to diagnose such diseases.^{1,3,8} The frequency of various causes of paediatric chest pain comes from studies in paediatric ED and cardiology clinics. In general, the most frequent cause reported is musculoskeletal pain, including costochondritis. Respiratory causes including asthma are the second most common organic etiology, representing 13 to 24% of cases. Gastrointestinal and psychogenic causes are identified in less than 10% of cases and a cardiac cause is found infrequently, representing no more than 5% of cases. An idiophatic etiology was frequently assigned in several studies, accounting for 20% to 61% of diganoses.^{1,3,9} The frequency of chest pain causes in children was attached in table 1.

Paediatric chest pain can be classified broadly into cardiac chest pain or non-cardiac chest pain. Non-cardiac chest pain is the most common cause of chest pain in children and adolescents.^{1,3,6} The cardiac cause was explained in table 2 and 3.

Appropriate Use of Echocardiograms

The most common cause of paediatric chest pain was non-cardiac disease or idiophatic.^{3,9} A cross-sectional study in Iran reported that 51% patient who underwent echocardiograms was diagnosed as idiophatic chest pain, other associated disorders were musculoskeletal (30%), gastrointestinal (11.5%), cardiac (4.5%), and pulmonary (3%).¹⁰ However, as the use of echocardiography was increased and parallel to higher cost, the American College of Cardiology (ACC) joined with the American Society of Echocardiography establish and evaluate AUC for the initial use of outpatient paediatric echocardiography.¹¹

The AUC has been established to guide physician decision-making or served as a guide to help clinicians in the care of children. It identified 113 indications based on common clinical scenarios and/or published clinical practical guidelines, and each indication was classified into 1 of 9 categories of common clinical presentations, including palpitations, syncope, chest pain, and murmur.^{7,11} Each indication was rated and classified as either "Appropriate Care", "May Be Appropriate Care", or "Rarely Appropriate Care" based on AUC development process. The rates or scores indication as follows:¹¹

- 1. Median score 7 to 9: appropriate (A) test for specific indication. Test is generally acceptable and reasonable approach for the indication.
- Median score 4 to 6: may be appropriate (M) test for specific indication. Test may be generally acceptable and reasonable

approach for the indication. May be appropriate also implies that more research and/or patient information is needed to classify the indication definitively.

3. Median score 1 to 3: rarely appropriate (R) test for specific indication test. Test is not generally acceptable and not a reasonable approach for the indication.

The scores for paediatric echocardiography are listed by indication in table 3 to 11 and figure 1 to 4.¹¹ The chart is a simple guideline for clinician. For example, R(2) means that the use of echo was Rarely appropriate test. "2" means, they identified 2 report.

The AUC have served as an important tool for quality improvement and education in adult cardiology. Similiar to the adult AUC, the paediatric AUC will need to be revised.⁷ Future studies to evaluate implementation of these AUC in clinical care will be helpful not only in identifying and deficiencies but also in defining ordering patterns for individual practitioners and understanding variations in delivery of care.¹¹

Summary

The etiology of chest pain in paediatrics was mostly musculoskeletal pain and idopathic. American College of Cardiology published Appropriate Use Criteria for echocardiography examination. The AUC could guide physicians on when and how often to use echocardiography in paediatric setting.

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Table 1. Frequency of causes of chest pain in children⁹

Cause	Emergency Department or Paediatric Clinic (%)	Cardiology Clinic (%)
Idiopathic / cause unknown	12-61	37-54
Musculoskeletal/costochondritis	7-69	1-89
Respiratory/asthma	13-24	1-12
Gastrointestinal/gastroesophageal reflux disease	3-7	3-12
Psychogenic	5-9	4-19
Cardiac	2-5	3-7

Table 2. Cardiac causes of paediatric chest pain³

Inflammatory: Pericarditis, Myocarditis
Infective: viruses, bacteria
Noninfective: Systemic Lupus Erythematosus, Crohn disease, postpericardiotomy syndrome
Increased Myocardial Demand or Decreased Supply
Cardiomyopathy: dilated or hypertrophic
Left ventricular outflow tract obstruction: aortic stenosis, subaortic stenosis, supravalvar aortic stenosis
Arrhythmias
Coronary Artery Abnormalities
Congenital: Anomalous origin of the left coronary artery from pulmonary artery, Anomalous left coronary artery from right coronary sinus, coronary fistula
Acquired: Kawasaki disease, postsurgical (after arterial switch operation, after Ross procedure), posttransplant coronary vasculopathy, familial hypercholesterolemia
Miscellaneous
Aortic dissection
Rupture of aortic aneurysm
Pulmonary hypertension
Mitral valve prolapse
Atrial myxomas
Cardiac device/stent complications
Drugs
Cocaine
Sympathomimetic overdose

Table 3. Palpitations and arrhythmias

Indication		Appropriate Use Rating
Palpitations		
1.	Palpitations with no other symptoms or signs of cardiovascular disease, a benign family history, and no recent electrocardiography (ECG)	R (2)
2.	Palpitations with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	R (1)
3.	Palpitations with abnormal ECG	M (6)
4.	Palpitations with family history of a channelopathy	R (3)
5.	Palpitations in a patient with known channelopathy	M (4)
б.	Palpitations with family history at a young age (before the age of 50 years) of sudden cardiac arrest of death and/or pacemaker of implantable defibrillator placement	A (7)
7.	Palpitations with family history of cardiomyopathy	A (9)
8.	Palpitations in a patient with known cardiomyopathy	A (9)
ECG Findings		
9.	Premature atrial contraction (PAC) in the prenatal or neonatal period	R (3)
10.	PAC after the neonatal period	R (3)
11.	Supraventricular tachycardia	A (7)
12.	Premature ventricular contraction (PVC) in the prenatal or neonatal period	M (6)
13.	PVC after the neonatal period	M (6)
14.	Ventricular tachycardia	A (6)
15.	Sinus bradycardia	R (2)
16.	Sinus arrhythmia	R (1)



Table 4. Syncope

Indication		Appropriate Use Rating
17.	Syncope with or without palpitations and with no recent ECG	R (3)
18.	Syncope with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	R (2)
19.	Syncope with abnormal ECG	A (7)
20.	Syncope with family history of channelopathy	M (5)
21.	Syncope with family history at a young age (before the age of 50 years) of sudden cardiac arrest or death and/or pacemaker or implantable defibrillator placement	A (9)
22.	Syncope with family history of caridomyopathy	A (9)
23.	Probable neurocardiogenic (vasovagal) syncope	R (2)
24.	Unexplained pre-syncope	M (4)
25.	Exertional syncope	A (9)
26.	Unexplained post-exertional syncope	A (7)
27.	Syncope or pre-syncope with a known non-cardiovascular cause	R (2)

Table 5. Chest pain

Indication		Appropriate Use Rating
28.	Chest pain with no other symptoms or sign of cardiovascular disease, a benign family history, and a normal ECG	R (2)
29.	Chest pain with other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	M (6)
30.	Exertional chest pain	A (8)
31.	Non-exertional chest pain with no recent ECG	R (3)
32.	Non-exertional chest pain with normal ECG	R (1)
33.	Non-exertional chest pain with abnormal ECG	A (7)
34.	Chest pain with family history of sudden unexplained death or cardiomyopathy	A (8)
35.	Chest pain with family history of premature coronary artery disease	M (4)
36.	Chest pain with recent onset of fever	M (6)
37.	Reproducible chest pain with palpitation or deep insipiration	R (1)
38.	Chest pain with recent illicit drug use	M (6)

Table 6. Murmur

Indication		Appropriate Use Rating
39.	Presumptively innocent murmur with no symptoms, signs of findings of cardiovascular disease and a benign family history	R (1)
40.	Presumptively innocent murmur with signs, symptoms, or findings of cardiovascular disease	A (7)
41.	Pathologic murmur	A (9)

Table 7. Other symptoms and signs

Indication		Appropriate Use Rating
42.	Symptoms and/or signs suggestive of congestive heart failure, including but not limited to respiratory distress, poor peripheral pulses, feeding difficulty, decreased urine output, edema and/or hepatomegaly	A (9)
43.	Chest wall deformities and scollosis pre-operatively	M (6)
44.	Fatigue with no other signs and symptoms of cardiovascular disease, a normal ECG, and a benign family history	R (3)
45.	Signs and symptoms of endocarditis in the absence of blood culture data or a negative blood culture	A (8)
46.	Unexplained fever without other evidence for cardiovascular or systemic involement	M (5)
47.	Central cyanosis	A (8)
48.	Isolated acrocyanosis	R (1)



Table 8. Prior test results

Indication		Appropriate Use Rating
49.	Known channelopathy	M (4)
50.	Genotype positive for cardiomyopathy	A (9)
51.	Abnormal chest X-ray findings suggestive of cardiovascular disease	A (9)
52.	Abnormal ECG without symptoms	A (7)
53.	Desaturation based on pulse oximetry	A (9)
54.	Previously normal echocardiogram with no change in cardiovascular status or family history	R (1)
55.	Previously normal echocardiogram with a change in cardiovascular status and/or a new family history suggestive of heritable heart disease	A (7)
56.	Elevated anti-streptolysin O titer without suspicion for rheumatic fever	R (3)
57.	Chromosomal abnormality known to be associated with cardiovascular disease	A (9)
58.	Chromosomal abnormality with undefined risk for cardiovascular disease	M (5)
59.	Positive blood cultures suggestive of infective endocarditis	A (9)
60.	Abnormal cardiac biomarkers	A (9)
61.	Abnormal barium swallow or bronchoscopy suggesting vascular ring	A (7)

Table 9. Systemic disorders

Indication		Appropriate Use Rating
62.	Cancer with chemotherapy	M (5)
63.	Prior to or during chemotherapy in cancer	A (8)
64.	Sickle cell disease and other hemoglobinopathies	A (8)
65.	Connective tissue disorder such as Marfan, Loeys Dietz, and other aortopathy syndromes	A (9)
66.	Suspected connective tissue disorder	A (7)
67.	Clinically suspected syndrome or extracardiac congenital known to be associated with congenital heart disease	A (9)
68.	Human immunodeficiency virus infection	A (8)
69.	Suspected or confirmed Kawasaki disease	A (9)
70.	Suspected or confirmed Takayasu arteritis	A (9)
71.	Suspected or confirmed acute rheumatic fever	A (9)
72.	Systemic lupus erythematosis and autoimmune disorders	A (7)
73.	Muscular dystrophy	A (9)
74.	Systemic hypertension	A (9)
75.	Renal failure	A(7)
76.	Obesity without other cardiovascular risk factors	R (2)
77.	Obesity with obstructive sleep apnea	M (6)
78.	Obesity with other cardiovascular risk factors	M (6)
79.	Diabetes mellitus	R (3)
80.	Lipid disorders	R (3)
81.	Stroke	A (8)
82.	Seizures, other neurologic disorders, or psychiatric disorders	R (2)
83.	Suspected pulmonary hypertension	A (9)
84.	Gastrointestinal disorders, not otherwise specified	R (2)
85.	Hepatic disorders	M (4)
86.	Failure to thrive	M (5)
87.	Storage disease, mitochondrial and metabolic disorders	A (8)
88.	Abnormalities of visceral or cardiac situs	A (9)



Table 10. Family history of cardiovascular disease in patients without signs or symptoms and without confirmed cardiac diagnosis

Indication		Appropriate Use Rating
89.	Unexplained sudden death before the age of 50 years	M (6)
90.	Premature coronary artery disease before the age of 50 years	R (2)
91.	Channelopathy	R (3)
92.	Hypertrophic cardiomyopathy	A (9)
93.	Non-ischemic dilated cardiomyopathy	A (9)
94.	Other cardiomyopathies	A (8)
95.	Unspecified cardiovascular disease	R (3)
96.	Disease high risk for cardiovascular involvement, including but not limited to diabetes, systemic hypertension, obesity, stroke, and peripheral vascular disease	R (2)
97.	Genetic disorder at high risk for cardiovascular involvement	A (7)
98.	Marfan or Loeys Dietz syndrome	A (7)
99.	Connective tissue disorder other than Marfan or Loeys Dietz syndrome	M (6)
100.	Congenital left-sided heart lesion, including but not limited to mitral stenosis, left ventricular outflow tract obstruction, bicuspid aortic valve, aortic coarctation, and/or hypoplastic left heart syndome	M (6)
101.	Congenital heart disease other than the congenital left-sided heart lesions	M (5)
102.	Idiopathic pulmonary arterial hypertension	M (5)
103.	Heritable pulmonary arterial hypertension	A (8)
104.	Pulmonary arterial hypertension other than idiopathic and heritable	R (3)
105.	Consanguinity	R (3)

Table 11. Outpatient neonate without post-natal cardiology evaluation

Indication		Appropriate Use Rating
106.	Suspected cardiovascular abnormality on fetal echocardiogram	A (9)
107.	Isolated echogenic focus on fetal ultrasound	R (2)
108.	Maternal infection during pregnancy or delivery with potential fetal/neonatal cardiac sequale	A (7)
109.	Maternal diabetes with no prior fetal echocardiogram	M (6)
110.	Maternal diabetes with a normal fetal echocardiogram	M (4)
111.	Maternal phenylketonuria	A (7)
112.	Maternal autoimmune disorder	M (5)
113.	Maternal teratogen exposure	M (6)





Figure 1. Chest pain flow chart



Figure 2. Syncope flow chart







Figure 4. Murmur flow chart