

Appropriate Use Criteria for Initial Transthoracic Echocardiography in Outpatient Pediatric Cardiology (scores listed by Appropriate Use rating)

Table 1: Appropriate indications (median score 7-9)

Indication		Appropriate Use Score (1-9)
Palpitations		
6	Palpitations 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 (7)
7	Palpitations with family history of cardiomyopathy	A (9)
8	Palpitations in a patient with known cardiomyopathy	A (9)
Arrhythmias/ ECG Findings		
11	Supraventricular tachycardia	A (7)
14	Ventricular tachycardia	A (9)
Syncope		
19	Syncope with abnormal ECG	A (7)
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 cardiomyopathy	A (9)
25	Exertional syncope	A (9)
26	Unexplained post-exertional syncope	A (7)
Chest Pain		
30	Exertional chest pain	A (8)
33	Non-exertional chest pain with abnormal ECG	A (7)
34	Chest pain with family history of sudden unexplained death or cardiomyopathy	A (8)
Murmur		
40	Presumptively innocent murmur with signs, symptoms, or findings of cardiovascular disease	A (7)

41	Pathologic murmur	A (9)
Other Symptoms and Signs		
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)
45	Signs and symptoms of endocarditis in the absence of blood culture data or a negative blood culture	A (8)
47	Central cyanosis	A (8)
Prior Test Results		
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)
55	Previously normal echocardiogram with a change in cardiovascular status and/or a new family history suggestive of heritable heart disease	A (7)
57	Chromosomal abnormality known to be associated with cardiovascular disease	A (9)
59	Positive blood cultures suggestive of infective endocarditis	A (9)
60	Abnormal cardiac enzymes	A (9)
61	Abnormal barium swallow or bronchoscopy suggesting vascular ring	A (7)
Systemic Disorders		
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 anomaly 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 erythematosus and autoimmune disorders	A (7)
73	Muscular dystrophy	A (9)
74	Systemic hypertension	A (9)
75	Renal failure	A (7)
81	Stroke	A (8)
83	Suspected pulmonary hypertension	A (9)
87	Storage diseases, mitochondrial and metabolic disorders	A (8)
88	Abnormalities of visceral or cardiac situs	A (9)
Family History of Cardiovascular Disease in Patients without Signs or Symptoms and without Confirmed Cardiac Diagnosis		
92	Hypertrophic cardiomyopathy	A (9)
93	Non-ischemic dilated cardiomyopathy	A (9)
94	Other cardiomyopathies	A (8)
97	Genetic disorder at high risk for cardiovascular involvement	A (7)
98	Marfan or Loeys Dietz syndrome	A (7)
103	Heritable pulmonary arterial hypertension	A (8)
Outpatient Neonates without Post-Natal Cardiology Evaluation		
106	Suspected cardiovascular abnormality on fetal echocardiogram	A (9)
108	Maternal infection during pregnancy or delivery with potential fetal/ neonatal cardiac sequelae	A (7)
111	Maternal phenylketonuria	A (7)

Table 2: May Be Appropriate indications (median score 4-6)

Indication		Appropriate Use Score (1-9)
Palpitations		
3	Palpitations with abnormal ECG	M (6)
5	Palpitations in a patient with known channelopathy	M (4)
Arrhythmias/ ECG Findings		
12	PVCs in the prenatal or neonatal period	M (6)
13	PVCs after the neonatal period	M (6)
Syncope		
20	Syncope with family history of channelopathy	M (5)
24	Unexplained pre-syncope	M (4)
Chest Pain		
29	Chest pain with other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	M (6)
35	Chest pain with family history of premature coronary artery disease	M (4)
36	Chest pain with recent onset of fever	M (6)
38	Chest pain with recent illicit drug use	M (6)
Other Symptoms and Signs		
43	Chest wall deformities and scoliosis pre-operatively	M (6)
46	Unexplained fever without other evidence for cardiovascular or systemic involvement	M (5)
Prior Test Results		
49	Known channelopathy	M (4)
58	Chromosomal abnormality with undefined risk for cardiovascular disease	M (5)
Systemic Disorders		
62	Cancer without chemotherapy	M (5)
77	Obesity with obstructive sleep apnea	M (6)
78	Obesity with other cardiovascular risk factors	M (6)

85	Hepatic disorders	M (4)
86	Failure to thrive	M (5)
Family History of Cardiovascular Disease in Patients without Signs or Symptoms and without Confirmed Cardiac Diagnosis		
89	Unexplained sudden death before the age of 50 years	M (6)
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 syndrome	M (6)
101	Congenital heart disease other than the congenital left-sided heart lesions	M (5)
102	Idiopathic pulmonary arterial hypertension	M (5)
Outpatient Neonates without Post-Natal Cardiology Evaluation		
109	Maternal diabetes with no prior fetal echocardiogram	M (6)
110	Maternal diabetes with a normal fetal echocardiogram	M (4)
112	Maternal autoimmune disorder	M (5)
113	Maternal teratogen exposure	M (6)

Table 3: Rarely Appropriate indications (median score 1-3)

Indication		Appropriate Use Score (1-9)
Palpitations		
1	Palpitations with no other symptoms or signs of cardiovascular disease, a benign family history, and no recent ECG	R (2)
2	Palpitations with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	R (1)
4	Palpitations with family history of a channelopathy	R (3)
Arrhythmias/ ECG Findings		
9	PACs in the prenatal or neonatal period	R (3)
10	PACs after the neonatal period	R (3)
15	Sinus bradycardia	R (2)
16	Sinus arrhythmia	R (1)

Syncope		
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)
23	Probable neurocardiogenic (vasovagal) syncope	R (2)
27	Syncope or pre-syncope with a known non-cardiovascular cause	R (2)
Chest Pain		
28	Chest pain with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	R (2)
31	Non-exertional chest pain with no recent ECG	R (3)
32	Non-exertional chest pain with normal ECG	R (1)
37	Reproducible chest pain with palpation or deep inspiration	R (1)
Murmur		
39	Presumptively innocent murmur with no symptoms, signs, or findings of cardiovascular disease and a benign family history	R (1)
Other Symptoms and Signs		
44	Fatigue with no other signs and symptoms of cardiovascular disease, a normal ECG, and a benign family history	R (3)
48	Isolated Acrocyanosis	R (1)
Prior Test Results		
54	Previously normal echocardiogram with no change in cardiovascular status or family history ⁵⁴	R (1)
56	Elevated anti-streptolysin O titers without suspicion for rheumatic fever ⁵⁶	R (3)
Systemic Disorders		
76	Obesity without other cardiovascular risk factors	R (2)
79	Diabetes mellitus	R (3)
80	Lipid disorders	R (3)
82	Seizures, other neurologic disorders, or psychiatric disorders	R (2)
84	Gastrointestinal disorders, not otherwise specified	R (2)
Family History of Cardiovascular Disease in Patients without Signs or Symptoms and without Confirmed Cardiac Diagnosis		

90	Premature coronary artery disease before the age of 50 years	R (2)
91	Channelopathy	R (3)
95	Unspecified cardiovascular disease	R (3)
96	Disease at high risk for cardiovascular involvement, including but not limited to diabetes, systemic hypertension, obesity, stroke, and peripheral vascular disease	R (2)
104	Pulmonary arterial hypertension other than idiopathic and heritable	R (3)
105	Consanguinity	R (3)
Outpatient Neonates without Post-Natal Cardiology Evaluation		
107	Isolated echogenic focus on fetal ultrasound	R (2)