2014 Appropriate Use Criteria for Initial Transthoracic Echocardiography in Outpatient Pediatric Cardiology: Guideline Mapping and References

Table 1: Palpitations and Arrhythmias

Indication #		Guideline Recommendation
Palpitations		
1.	Palpitations with no other symptoms or signs of cardiovascular disease, a benign family history, and no recent ECG	None
2.	Palpitations with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	None
3.	Palpitations with abnormal ECG	None
4.	Palpitations with family history of a channelopathy	None
5.	Palpitations in a patient with known channelopathy	None
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	None
7.	Palpitations with family history of cardiomyopathy	None
8.	Palpitations in a patient with known cardiomyopathy	None
ECG Findings		
9.	PACs in the prenatal or neonatal period	None
10.	PACs after the neonatal period	None
11.	Supraventricular tachycardia	None
12.	PVCs in the prenatal or neonatal period	None
13.	PVCs after the neonatal period	None
14.	Ventricular tachycardia	None

15.	Sinus bradycardia	None
16.	Sinus arrhythmia	None

References

Cardiovascular Monitoring of Children and Adolescents With Heart Disease Receiving Medications for Attention Deficit/Hyperactivity Disorder. A Scientific Statement From the American Heart Association Council on Cardiovascular Disease in the Young Congenital Cardiac Defects Committee and the Council on Cardiovascular Nursing. *Circulation*. 2008;117:2407-2423

Iwamoto M, Niimura I, Shibata T, Yasui K, Takigiku K, Nishizawa T, Akaike T, Yokota S Long-term course and clinical characteristics of ventricular tachycardia detected in children by school-based heart disease screening Circ J. 2005 Mar;69(3):273-6.

Sun Y, Blom NA, Yu Y, Ma P, Wang Y, Han X, Swenne CA, van der Wall EE. The influence of premature ventricular contractions on left ventricular function in asymptomatic children without structural heart disease: an echocardiographic evaluation. Int J Cardiovasc Imaging. 2003 Aug;19(4):295-9.

Table 2: Syncope

Indication #		Guideline Recommendation
17.	Syncope with or without palpitations and with no recent ECG	None
18.	Syncope with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	None
19.	Syncope with abnormal ECG	None
20.	Syncope with family history of channelopathy	None
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	None
22.	Syncope with family history of cardiomyopathy	None
23.	Probable neurocardiogenic (vasovagal) syncope	None

24.	Unexplained pre-syncope	None
25.	Exertional syncope	**Cheitlin et al :ACC/AHA Guidelines for the Clinical Application of Echocardiography 1997; 95: 1686-1744** Table 51 Exercise-induced precordial chest pain or syncope (Class I)
26.	Unexplained post-exertional syncope	None
27.	Syncope or pre-syncope with a known non-cardiovascular cause	None

References

Anderson JB, Czosek RJ, Cnota J, Meganathan K, Knilans TK, Heaton PC. Pediatric Syncope: National Hospital Ambulatory Medical Care Survey Results. The Journal of Emergency Medicine. 2012. 1-9

Benditt DG, Ferguson DW, Grubb BP, Kapoor WN, Kugler J, Lerman BB, et al. ACC expert consensus document: tilt table testing for assessing syncope. J Am Coll Cardiol 1996;28: 263-75.

Bo I, Carano N, Agnetti A, Tchana B, Allegri V, Sommi M, Squarcia U. Syncope in children and adolescents: a two-year experience at the Department of Pediatrics in Parma. Acta Biomed. 2009. 80:36-41

Briugnole M, Alboni P, Benditt DG, Bergfeldt L, Blanc J, Thomsen PEB, van Dijk JG, Fitzpatrick A, Hohnloser S, Janousek J, Kapoor W, Kenny RA, Kulakowski P, Masotti G, Moya A, Raviele A, Sutton R, Theodorakis G, Ungar A, Wieling W. Guidelines on management (diagnosis and treatment) of syncope--update 2004.European Heart Journal. 2004. 25:2054-2072.

Calkins H, Shyr Y, Frumin H, Schork A, Morady F. The Value of the Clinical History in the Differentiation of Syncope Due to Ventricular Tachycardia, Atrioventricular Block, and Neurocardiogenic Syncope. The American Journal of Medicine. 1995. 98:365-373.

Cheitlin MD, Alpert JS, Armstrong WF, Aurigemma GP, Beller GA, Bierman FZ, Davidson TW, Davis JL, Douglas PS, Gillam LD. ACC/AHA Guidelines for the Clinical Application of Echocardiography. A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Committee on Clinical Application of Echocardiography). Developed in collaboration with the American Society of Echocardiography. Circulation. 1997 Mar 18;95(6):1686-744.

Johnsrude CL. Current Approach to Pediatric Syncope. Pediatr Cardiol. 2000. 21:522-531.

Kessler C, Tristano JM, De Lorenzo R. The Emergency Department Approach to Syncope: Evidence-based Guidelines and Prediction Rules. Emerg Med Clin N Am. 2010. 28:487-500.

Kuriachan V, Sheldon RS, Platonov M. Evidence –based Treatment for Vasovagal Syncope. Heart Rhythm. 2008. 5:1609-1614

MacCormick JM, Crawford JR, Chung SK, Shelling AN, Evans CA, Rees MI, Smith WM, Crozier IG, McAlister H, Skinner JR. Symtpoms and Signs Assocaited with Syncope in Young People with Primary Cardiac Arrhythmias. Heart, Lung, and Circulation. 2011. 20:593-598.

Massin MM, Malekzadeh-milani S, Benatar A. Cardiac Syncope in Pediatric Patients. Clin. Cardiol. 2007. 30:81–85.

Medow MS, Stewart JM, Sanyal Sanjukta, Mumtaz A, Sica D, Frishman WH. Pathophysiology, Diagnosis, and Treatment of Orthostatic Hypotension and Vasovagal Syncope. Cardiology in Review. January/February 2008. 16(1):4-20.

Ouyang H, Quinn J. Diagnosis and Evaluation of Syncope in the Emergency Department. Emerg Med Clin N Am. 2010. 28:471-485.
Ritter S, Tani LY, Etheridge SP, Williams RV, Craig JE, Minich L. What is the Yield of Screening Echocardiography in Pediatric Syncope? Pediatrics. 2000. 105;e58

Saarel EV, Stefanelli CB, Fischbach PS, Serwer GA, Rosenthal A, Dick M. Transtelephonic Electrocardiographic Monitors for Evaluation of Children and Adolescents with Suspected Arrhythmias. Pediatrics. 2004. 113;248-251.

Sheldon R, Rose S, Ritchie D, Connolly SJ, Koshman ML, Lee MA, Frenneaux M, Fisher M, Murphy W. Historical Criteria That Distinguish Syncope from Seizures. Journal of the American College of Cardiology. 2002. 40:142-148

Steinberg LA, Knilans TK. Syncope in Children: Diagnostic Tests have a High Cost and Low Yield. J Pediatr. 2005. 146:355-358.

Strickberger SA, Benson WD, Biaggioni I, Callans DJ, Cohen MI, Ellenbogen KA, Epstein AE, Friedman P, Goldberger J, Heidenreich PA, Klein GJ, Knight BP, Morillo CA, Myerburg RJ, Sila CA. AHA/ACCF Scientific Statement on the Evaluation of Syncope. JACC. 2006 Vol. 47, No. 2:473–84

Zhang Q, Zhu L, Wang C, Du Z, Hu X, Tian H, Todd O, Zhang F, Du J, Jin H. Value of history taking in children and adolescents with cardiac syncope. Cardiology in the Young. 2012. 1-7.

Table 3: Chest Pain

Indication #		Guideline Recommendation
28.	Chest pain with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	None
29.	Chest pain with other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	None
30.	Exertional chest pain	**Cheitlin et al :ACC/AHA Guidelines for the Clinical Application of Echocardiography 1997; 95: 1686-1744** Table 51 Exercise-induced precordial chest pain or syncope (Class I)
31.	Non-exertional chest pain with no recent ECG	None
32.	Non-exertional chest pain with normal ECG	None
33.	Non-exertional chest pain with abnormal ECG	None
34.	Chest pain with family history of sudden unexplained death or cardiomyopathy	None
35.	Chest pain with family history of premature coronary artery disease	None
36.	Chest pain with recent onset of fever	None
37.	Reproducible chest pain with palpation or deep inspiration	None
38.	Chest pain with recent illicit drug use	None

References

Drossner DM, Hirsh DA, Sturm JJ, Mahle WT, Goo DJ, Massey R. Simon HK. Cardiac disease in pediatric patients presenting to a pediatric ED with chest pain. American Journal of Emergency Medicine 2011 Jul;29(6):632-8

Friedman KG, Kane DA, Rathod RH, Renaud A, Farias M, Geggel R et al. Management of pediatric chest pain using a standardized assessment and management plan. Pediatrics. 2011 Aug;128(2):239-45

Kane DA, Fulton DR, Saleeb S, Zhou J, Lock JE, Geggel R. Needles in hay: chest pain as the presenting symptom in children with serious underlying cardiac pathology. Congenital Heart Disease 2010; 5: 366-373.

Saleeb S.F.Li WY, WarrenSZ, Lock JE. Effectiveness of screening for life threatening chest pain in children Pediatrics 2011;128 e1062-8.

Zavaras-Angelidou KA, Weinhouse E, Nelson DB. Review of 180 episodes of chest pain in 134 children. Pediatr Emerg Care. 1992; Aug;8(4):189-93.

Table 4: Murmur

Indication #		Guideline Recommendation
39.	Presumptively innocent murmur with no symptoms, signs, or findings of cardiovascular disease and a benign family history	**Cheitlin et al :ACC/AHA Guidelines for the Clinical Application of Echocardiography 1997; 95: 1686-1744 Table 51** In a child or adolescent, an asymptomatic heart murmur identified by an experienced observer as functional or an insignificant cardiovascular abnormality (Class III).
40.	Presumptively innocent murmur with signs, symptoms, or findings of cardiovascular disease	None
41.	Pathologic murmur	**Cheitlin et al :ACC/AHA Guidelines for the Clinical Application of Echocardiography 1997; 95: 1686-1744 Table 51** Atypical or pathological murmur or other abnormal cardiac finding in an infant or older child. (Class I)

References

Biancaniello T. Innocent murmurs. Circulation. 2005;111:e20-e22

Danford DA, Martin AB, Fletcher SE, Gumbiner CH: Echocardiographic yield in children when innocent murmur seems likely but doubts linger. Pediatric Cardiology 2002;23(4):410-4
Kwiatkowski D, Wang Y, Cnota J. The utility of outpatient echocardiography for evaluation of asymptomatic murmurs in children. Congenit Heart Dis. 2012 May-Jun;7(3):283-8

McCrindle BW, Shaffer KM, Kan JS, Zahka KG, Rowe SA, Kidd L. Cardinal clinical signs in the differentiation of heart murmurs in children. Arch Pediatr Adolesc Med. 1996 Feb;150(2):169-74.

Rosenthal A. How to distinguish between innocent and pathologic heart murmurs in children. The pediatric Clinics of North America, Dec 1984, page 129
Saunders NR. Innocent heart murmurs in children. Taking a diagnostic approach. Can Fam Physician. 1995 Sep;41:1507-12

Yi MS, Kimball TR, Tsevat J, Mrus JM, Kotagal UR. Evaluation of heart murmurs in children: cost-effectiveness and practical implications. J Pediatr. 2002 Oct;141(4):504-11.

Table 5: Other Symptoms and Signs

Indication #		Guideline Recommendation
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	None
43.	Chest wall deformities and scoliosis pre-operatively	None
44.	Fatigue with no other signs and symptoms of cardiovascular disease, a normal ECG, and a benign family history	None
45.	Signs and symptoms of endocarditis in the absence of blood culture data or a negative blood culture	None
46.	Unexplained fever without other evidence for cardiovascular or systemic involvement	None
47.	Central cyanosis	None

48.	Isolated Acrocyanosis	**Cheitlin et al ACC/AHA/ASE 2003 Guideline Update for the clinical Application of Echocardiography: Summary Article. Circulation 2003: 1145-1162** Class III Acrocyanosis with normal upper- and lower extremity pulsed oximetry oxygen saturations

References

Cheitlin MD, Armstrong WF, Aurigemma GP, Beller GA, Bierman FZ, Davis JL, Douglas PS, Faxon DP, Gillam LD, Kimball TR, Kussmaul WG, Pearlman AS, Philbrick JT, Rakowski H, Thys DM, Antman EM, Smith SC Jr, Alpert JS, Gregoratos G, Anderson JL, Hiratzka LF, Hunt SA, Fuster V, Jacobs AK, Gibbons RJ, Russell RO; American College of Cardiology; American Heart Association; American Society of Echocardiography. ACC/AHA/ASE 2003 guideline update for the clinical application of echocardiography: summary article: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (ACC/AHA/ASE Committee to Update the 1997 Guidelines for the Clinical Application of Echocardiography). Circulation. 2003 Sep 2;108(9):1146-62.

Table 6: Prior Test Results

Indication #		Guideline Recommendation
49.	Known channelopathy	None
50.	Genotype positive for cardiomyopathy	None
51.	Abnormal chest X-ray findings suggestive of cardiovascular disease	**Cheitlin et al :ACC/AHA Guidelines for the Clinical Application of Echocardiography 1997; 95: 1686-1744** Table 51 Cardiomegaly on chest radiograph (Class I)
52.	Abnormal ECG without symptoms	None

53.	Desaturation based on pulse oximetry	**Mahle WT, et al. Role of pulse oximetry in examining newborns for congenital heart disease: a scientific statement from the AHA and AAP. Pediatrics. 2009 Aug;124(2):823-36.** When neonates are identified as having hypoxemia (SpO2 ≤ 95%), it is necessary to evaluate them for CCHD. Although physical examination, chest radiography, and electrocardiography can assist in this process, echocardiography is now considered the definitive diagnostic modality. Whenever possible, the echocardiograms should be interpreted by pediatric cardiologists; major errors in the interpretation of a newborn echocardiogram by trained pediatric cardiologists are rare. **Kemper AR, et al. Strategies for implementing screening for critical congenital heart disease. Pediatrics. 2011; 128(5):e1259-67** A screen result would be considered positive if (1) any oxygen saturation measure is ≤90%, (2) oxygen saturation is ≤95% in both extremities on 3 measures, each separated by 1 hour, or (3) there is a ≥3% absolute difference in oxygen saturation between the right hand and foot on 3 measures, each separated by 1 hour.
54.	Previously normal echocardiogram with no change in cardiovascular status or family history	None
55.	Previously normal echocardiogram with a change in cardiovascular status and/or a new family history suggestive of heritable heart disease	None

56.	Elevated anti-streptolysin O titers without suspicion for rheumatic fever	None
57.	Chromosomal abnormality known to be associated with cardiovascular disease	**Cheitlin et al ACC/AHA/ASE 2003 Guideline Update for the clinical Application of Echocardiography: Summary Article. Circulation 2003: 1145-1162** Page 1155 Presence of a syndrome associated with cardiovascular disease and dominant inheritance or multiple affected family members (eg, Marfan syndrome or Ehlers-Danlos syndrome). (Class I)
58.	Chromosomal abnormality with undefined risk for cardiovascular disease	None
59.	Positive blood cultures suggestive of infective endocarditis	**Baddour LM et al. Infective endocarditis: diagnosis, antimicrobial therapy, and management of complications: a statement for healthcare professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, and the Councils on Clinical Cardiology, Stroke, and Cardiovascular Surgery and Anesthesia, American Heart Association: endorsed by the Infectious Diseases Society of America. Circulation. 2005; 111: e394-e434.** Echocardiography should be performed in all cases of suspected IE (Class I, Level of Evidence: A).
60.	Abnormal cardiac enzymes	None
61.	Abnormal barium swallow or bronchoscopy suggesting vascular ring	None

References

Baddour LM, Wilson WR, Bayer AS, Fowler VG Jr, Bolger AF, Levison ME, Ferrieri P, Gerber MA, Tani LY, Gewitz MH, Tong DC, Steckelberg JM, Baltimore RS, Shulman ST, Burns JC, Falace DA, Newburger JW, Pallasch TJ, Takahashi M, Taubert KA. Infective endocarditis: diagnosis, antimicrobial therapy, and

management of complications: a statement for healthcare professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, and the Councils on Clinical Cardiology, Stroke, and Cardiovascular Surgery and Anesthesia, American Heart Association: endorsed by the Infectious Diseases Society of America. Circulation. 2005; 111: e394-e434.

Kemper AR, Mahle WT, Martin GR, Cooley WC, Kumar P, Morrow WR, Kelm K, Pearson GD, Glidewell J, Grosse SD, Howell. Strategies for implementing screening for critical congenital heart disease. Pediatrics. 2011; 128(5):e1259-67

Mahle WT, Newburger JW, Matherne GP, Smith FC, Hoke TR, Koppel R, Gidding SS, Beekman RH 3rd, Grosse SD. Role of pulse oximetry in examining newborns for congenital heart disease: a scientific statement from the AHA and AAP. Pediatrics. 2009 Aug;124(2):823-36

Thangaratinam S, Brown K, Zamora J, Khan KS, Ewer AK. Pulse oximetry screening for critical congenital heart defects in asymptomatic newborn babies: a systematic review and meta-analysis. Lancet. 2012 Jun 30;379(9835):2459-64. doi: 10.1016/S0140-6736(12)60107-X. Epub 2012 May 2.

Table 7: Systemic Disorders

Indication #		Guideline Recommendation
62.	Cancer without chemotherapy	None
63.	Prior to or during chemotherapy in cancer	**Steinherz LJ, Graham T, Hurwitz R, et al. Guidelines for cardiac monitoring of children during and after anthracycline therapy: report of the Cardiology Committee of the Children's Cancer Study Group. Pediatrics 1992; 89 (5 Pt 1): 942-9.** Pg 946 All patients expected to receive doxorubicin and/or daunorubicin should have baseline cardiac evaluation with ECG, echocardiography, and when available, radionuclide angiocardiography. **Cheitlin et al ACC/AHA/ASE 2003 Guideline Update for the clinical Application of Echocardiography: Summary Article. Circulation 2003: 1145-1162. Section XV-H** Baseline and re-evaluation examinations of patients receiving cardiotoxic chemotherapeutic agents. (Class I)

64.	Sickle cell disease and other hemoglobinopathies	**Galie N, et al. Guidelines for the diagnosis and treatment of pulmonary hypertension. Eur Respir J. 2009;34:6,1219-1263.** Pg 1223: Chronic haemolytic anaemia such as sickle cell disease [18], thalassaemia, hereditary spherocytosis, stomatocytosis and microangiopathic haemolytic anaemia may result in PAH and are included in the APAH forms. **Humbert M, et al. Early detection and management of pulmonary arterial hypertension. Eur Respir Rev. 2012;21:126,306-312.** Pg 306: Screening programmes play an important role in PAH detection and expert opinion favours echocardiographic screening of asymptomatic patients who may be predisposed to the development of PAH (i.e. those with systemic sclerosis or sickle cell disease), although current guidelines only recommend annual echocardiographic screening in symptomatic patients.
65.	Connective tissue disorder such as Marfan, Loeys Dietz, and other aortopathy syndromes	**Cheitlin et al ACC/AHA/ASE 2003 Guideline Update for the clinical Application of Echocardiography: Summary Article. Circulation 2003: 1145-1162** Page 1155 Presence of a syndrome associated with cardiovascular disease and dominant inheritance or multiple affected family members (eg, Marfan syndrome or Ehlers-Danlos syndrome). (Class I)
66.	Suspected connective tissue disorder	None

67.	Clinically suspected syndrome or extracardiac congenital anomaly known to be associated with congenital heart disease	**Cheitlin et al :ACC/AHA Guidelines for the Clinical Application of Echocardiography 1997; 95: 1686-1744** Table 51 Presence of a syndrome associated with cardiovascular disease and dominant inheritance or multiple affected family members (Class I) **Bondy CA, Turner Syndrome Study Group. Care of girls and women with Turner syndrome: a guideline of the Turner Syndrome Study Group. J Clin Endocrinol Metab 2007; 92 (1): 10-25.** Pg 13. A comprehensive postnatal echocardiogram should be evaluated by a pediatric cardiologist in all infants diagnosed with TS, even in those who had an apparently normal fetal echocardiogram. **Bull MJ and the Committee on Genetics:American Academy of Pediatrics: Health supervision for children with Down syndrome. Pediatrics 2011; 128:393-406. Pg 396.** Perform an echocardiogram, to be read by a pediatric cardiologist, regardless of whether a fetal echocardiogram was performed.
68.	Human immunodeficiency virus infection	None

69.	Suspected or confirmed Kawasaki disease	**Newburger JW, Takahashi M, Gerber MA, et al. Diagnosis, treatment, and long-term management of Kawasaki Disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Circulation 2004; 110(17): 2747-71**. Pg 2754. Because it is noninvasive and has a high sensitivity and specificity for the detection of abnormalities of the proximal LMCA and RCA,echocardiography is the ideal imaging modality for cardiac assessment (evidence level C).
70.	Suspected or confirmed Takayasu arteritis	None
71.	Suspected or confirmed acute rheumatic fever	**Carapetis, J., Parr, J. & Cherian, T. Standardization of epidemiologic protocols for surveillance of post-streptococcal sequelae: acute rheumatic fever, rheumatic heart disease and acute post-streptococcal glomerulonephritis. Department of Health and Human Services, National Institutes of Health [online] http://www.niaid.nih.gov/topics/strepThroat/Documents/groupasequelae.pdf (2010**). Page 12. Echocardiography in the diagnosis of rheumatic valvular disease (Needed)
72.	Systemic lupus erythematosis and autoimmune disorders	None

73.	Muscular dystrophy	**Cheitlin et al :ACC/AHA Guidelines for the Clinical Application of Echocardiography 1997; 95: 1686-1744** Table 51 Baseline and follow-up examinations of patients with neuromuscular disorders having known myocardial involvement (Class I)
74.	Systemic hypertension	**Cheitlin et al ACC/AHA/ASE 2003 Guideline Update for the clinical Application of Echocardiography: Summary Article. Circulation 2003: 1145-1162** Pg 1154 Patients with severe renal disease and/or systemic Hypertension (Class I) **National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents. The fourth report on the diagnosis, evaluation, and treatment of high blood pressure in children and adolescents. Pediatrics 2004;114(2) (suppl 4th report):555-576.** Pediatric patients with established hypertension should have echocardiographic assessment of left ventricular mass at diagnosis and periodically thereafter.
75.	Renal failure	**Cheitlin et al ACC/AHA/ASE 2003 Guideline Update for the clinical Application of Echocardiography: Summary Article. Circulation 2003: 1145-1162** Pg 1154. Patients with severe renal disease and/or systemic Hypertension (Class I)

76.	Obesity without other cardiovascular risk factors	None
77.	Obesity with obstructive sleep apnea	None
78.	Obesity with other cardiovascular risk factors	None
79.	Diabetes mellitus	None
80.	Lipid disorders	None
81.	Stroke	None
82.	Seizures, other neurologic disorders, or psychiatric disorders	None
83.	Suspected pulmonary hypertension	**McLaughlin VV et al. ACCF/AHA 2009 Expert Consensus Document on Pulmonary Hypertension. J Am Coll Cardiol. 2009 Apr 28;53(17):1573-619.** Pg 1585 If PH is suspected based on the history, risk factor assessment, and physical examination, an echocardiogram is the next appropriate study.
84.	Gastrointestinal disorders, not otherwise specified	None
85.	Hepatic disorders	None
86.	Failure to thrive	**Cheitlin et al :ACC/AHA Guidelines for the Clinical Application of Echocardiography 1997; 95: 1686-1744** Table 51 Failure to thrive in the absence of definite abnormal clinical findings (Class IIb)
87.	Storage diseases, mitochondrial and metabolic disorders	None

88.	Abnormalities of visceral or cardiac situs	**Cheitlin et al :ACC/AHA Guidelines for the Clinical Application of Echocardiography 1997; 95: 1686-1744** Table 51 Dextrocardia, abnormal pulmonary or visceral situs on clinical, electrocardiographic, or radiographic examination. (Class I)

References

Bondy CA, Turner Syndrome Study Group. Care of girls and women with Turner syndrome: a guideline of the Turner Syndrome Study Group. JClin Endocrinol Metab 2007; 92 (1): 10-25.

Bull MJ and the Committee on Genetics:American Academy of Pediatrics: Health supervision for children with Down syndrome. Pediatrics 2011; 128:393-406. Guidelines specify during first month: Perform an echocardiogram, to be read by a pediatric cardiologist, regardless of whether a fetal echocardiogram was performed.Cox GF. Diagnostic Approaches to Pediatric Cardiomyopathy of Metabolic Genetic Etiologies and Their Relation to Therapy. Prog Pediatr Cardiol. 2007;24(1):15-25.

Carapetis, J., Parr, J. & Cherian, T. Standardization of epidemiologic protocols for surveillance of post-streptococcal sequelae: acute rheumatic fever, rheumatic heart disease and acute post-streptococcal glomerulonephritis. Department of Health and Human Services, National Institutes of Health [online] http://www.niaid.nih.gov/topics/strepThroat/Documents/groupasequelae.pdf (2010

DC Knockaert. Cardiac involvement in systemic inflammatory diseases. Eur Heart J 2007; 28:1797–1804
Expert Panel on Integrated Guidelines for Cardiovascular Health and Risk Reduction in Children and Adolescents: Summary Report. Pediatrics 2011;128:S213-S256.

Galie N, Hoeper MM, Humbert M, et al. Guidelines for the diagnosis and treatment of pulmonary hypertension. Eur Respir J. 2009;34:6,1219-1263.

Humbert M, Coghlan JG, Khanna D. Early detection and management of pulmonary arterial hypertension. Eur Respir Rev. 2012;21:126,306-312.

Moder KG, Miller TD, Tazellar HD. Cardiac involvement in systemic lupus erythematosis. Mayo Clin Proc 1999;74:275-284.

McLaughlin VV, Archer SL, Badesch DB, Barst RJ, Farber HW, Lindner JR, Mathier MA, McGoon MD, Park MH, Rosenson RS, Rubin LJ, Tapson VF, Varga J; ACCF/AHA 2009 expert consensus document on pulmonary hypertension a report of the American College of Cardiology Foundation Task Force on Expert

Consensus Documents and the American Heart Association developed in collaboration with the American College of Chest Physicians; American Thoracic Society, Inc.; and the Pulmonary Hypertension Association. J Am Coll Cardiol. 2009 Apr 28;53(17):1573-619.

McNeice KL, Gupta-Malhotra M, Samuals J, et al. Left ventricular hypertrophy in hypertensive adolescents: analysis of risk by 2004 National High Blood Pressure Education Education Program Working Group staging criteria. Hypertension 2007; 50 (2): 392-5.

ME Pierpont, CT Basson, DW Benson, BD Gelb, TM Giglia, E Goldmuntz, G McGee, CA Sable, D Srivastava, CL Webb. Genetic basis for congenital heart defects: Current knowledge : A scientific statement from the American Heart Association Congenital Cardiac Defects Committee, Council on Cardiovascular Disease in the Young (Endorsed by the American Academy of Pediatrics). Circulation 2007;115:3015-3038

Moak JP, Kaski JP. Hypertrophic cardiomyopathy in children. Heart. 2012 Jul;98(14):1044-54. doi: 10.1136/heartjnl-2011-300531. Epub 2012 May 16.
National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents. The fourth report on the diagnosis, evaluation, and treatment of high blood pressure in children and adolescents. Pediatrics 2004;114(2) (suppl 4th report):555-576.

National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents. The fourth report on the diagnosis, evaluation, and treatment of high blood pressure in children and adolescents. Pediatrics 2004;114(2) (suppl 4th report):555-576.

Newburger JW, Takahashi M, Gerber MA, Gewitz MH, Tani LY, Burns JC, Shulman ST, Bolger AF, Ferrieri P, Baltimore RS, Wilson WR, Baddour LM, Levison ME, Pallasch TJ, Falace DA, Taubert KA. Diagnosis, treatment, and long-term management of Kawasaki disease: A statement for health professionals from the Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Circulation 2004;110:2747-2771

Kavey RW, Allada V, Daniels SR, Hayman LL, McCrindle BW, Newburger JW, Parekh RS, Steinberger J. Cardiovascular risk reduction in high-risk pediatric patients : A scientific statement From the American Heart Association Expert Panel on Population and Prevention Science; the Councils on Cardiovascular Disease in the Young, Epidemiology and Prevention, Nutrition, Physical Activity and Metabolism, High Blood Pressure Research, Cardiovascular Nursing, and the Kidney in Heart Disease; and the Interdisciplinary Working Group on Quality of Care and Outcomes Research: endorsed by the American Academy of Pediatrics. Circulation 2006;114:2710-2738

Naoman SG, Nouraie M, Castro OL, Nwokolo C, Fadojutimi-Akinsiku M, Diaz S, GWillie-Carnegie, Andrews N, Gordeuk VR. Echocardiographic findings in patients with sickle cell disease. Ann Hematol 2010;89:61-66.

Starc TJ, Lipshultz SE, Easley KA, et al. Incidence of cardiac abnormalities in children with human immunodeficiency virus infection: The prospective P2C2 HIV Study. J Pediatrics 2002; 141 (3): 327-34.

Steinherz LJ, Graham T, Hurwitz R, et al. Guidelines for cardiac monitoring of children during and after anthracycline therapy: report of the Cardiology Committee of the Children's Cancer Study Group. Pediatrics 1992; 89 (5 Pt 1): 942-9.

Tulloh RM, Tansey SP, Parashar K, et al. Echocardiographic screening in neonates undergoing surgery for selected gastrointestinal malformations. Arch Dis Child Fetal Neonat Ed 1994; 70 (3): F206-8.

Wilkinson JD, Sleeper LA, Alvarez JA, Bublik N, Lipshultz SE; the Pediatric Cardiomyopathy Study Group The Pediatric Cardiomyopathy Registry: 1995-2007. Prog Pediatr Cardiol. 2008 Apr;25(1):31-36.

Table 8: Family History of Cardiovascular Disease in Patients without Signs or Symptoms and without Confirmed Cardiac Diagnosis

Indication #		Guideline Recommendation
89.	Unexplained sudden death before the age of 50 years	None
90.	Premature coronary artery disease before the age of 50 years	None
91.	Channelopathy	None
92.	Hypertrophic cardiomyopathy	**Gersh et al. 2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy** **5.3. Imaging** **5.3.1. Echocardiography—Recommendations** CLASS I 2. A TTE is recommended as a component of the screening algorithm for family members of patients with HCM unless the family member is genotype negative in a family with known definitive mutations. (Level of Evidence: B)
93.	Non-ischemic dilated cardiomyopathy	**Rosenthal D, Chrisant MR, Edens E, et al. International Society for Heart and Lung Transplantation: Practice guidelines for management of heart failure in children. J Heart Lung Transplant. 2004;23:1313-33.** Pg 1317 Screening of first-degree relatives should be considered in patients with new-onset ventricular dysfunction due to DCM (HF Stages B, C or D). (Level of Evidence C; Strength of Recommendation I)
94.	Other cardiomyopathies	None
95.	Unspecified cardiovascular disease	None
96.	Disease at high risk for cardiovascular involvement, including but not limited to diabetes, systemic hypertension, obesity, stroke, and peripheral vascular disease	None

97.	Genetic disorder at high risk for cardiovascular involvement	**Cheitlin et al ACC/AHA/ASE 2003 Guideline Update for the clinical Application of Echocardiography: Summary Article. Circulation 2003: 1145-1162**. The presence of a syndrome associated with a high incidence of congenital heart disease for which there are no abnormal cardiac findings and no urgency of management decisions was classified as a Class IIa indication for echocardiography in neonates in this publication.
98.	Marfan or Loeys Dietz syndrome	**Hiratzka LF et al Guidelines for the diagnosis and management of patients with thoracic aortic disease J Am Coll Cardiol. 2010 Apr 6;55(14):e27-e129** Aortic imaging is recommended for first-degree relatives of patients with thoracic aortic aneurysm and/or dissection to identify those with asymptomatic disease. (Class I; Level of Evidence B) If one or more first-degree relatives of a patient with known thoracic aortic aneurysm and/or dissection are found to have thoracic aortic dilatation, aneurysm, or dissection, then imaging of second-degree relatives is reasonable. (Class IIa; Level of Evidence B)
99.	Connective tissue disorder other than Marfan or Loeys Dietz syndrome	None
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	None
101.	Congenital heart disease other than the congenital left-sided heart lesions	None
102.	Idiopathic pulmonary arterial hypertension	None

103.	Heritable pulmonary arterial hypertension	**McGoon M et al Screening, Early Detection, and Diagnosis of Pulmonary Arterial Hypertension ACCP Evidence-Based Clinical Practice Guidelines. CHEST 2004; 126:14S–34S** (Pg 21S) In asymptomatic patients at high risk, Doppler echocardiography should be performed to detect elevated pulmonary arterial pressure. Quality of evidence: expert opinion; benefit: intermediate; strength of recommendation: E/B. Risk groups warranting screening for PH include the following: patients with known genetic mutations predisposing to PH, first-degree relatives in a FPAH family, patients with scleroderma spectrum of disease, patients with portal hypertension prior to liver transplantation, and patients with congenital heart disease with systemic-to-pulmonary shunts.
104.	Pulmonary arterial hypertension other than idiopathic and heritable	None
105.	Consanguinity	None

References

Cripe L, Andelfinger G, Martin LJ, Shooner K, Benson DW. Bicuspid aortic valve is heritable. J Am Coll Cardiol. 2004 Jul 7;44(1):138-43

Gersh BJ, Maron BJ, Bonow RO, Dearani JA, Fifer MA, Link MS, Naidu SS, Nishimura RA, Ommen SR, Rakowski H, Seidman CE, Towbin JA, Udelson JE, Yancy CW. 2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Thorac Cardiovasc Surg. 2011 Dec;142(6):e153-203.

Hinton RB Jr, Martin LJ, Tabangin ME, Mazwi ML, Cripe LH, Benson DW. Hypoplastic left heart syndrome is heritable. J Am Coll Cardiol. 2007 Oct 16;50(16):1590-5

Hiratzka LF, Bakris GL, Beckman JA, Bersin RM, Carr VF, Casey DE Jr et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the diagnosis and management of patients with thoracic aortic disease. A Report of the American College of Cardiology Foundation/American Heart Association Task

Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology,American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons,and Society for Vascular Medicine. J Am Coll Cardiol. 2010 Apr 6;55(14):e27-e129

Hershberger RE, Siegfried JD. Update 2011: Clinical and Genetic Issues in Familial Dilated Cardiomyopathy. J Am Coll Cardiol 2011;57:1641–9.

Lewin MB, McBride KL, Pignatelli R, Fernbach S, Combes A, Menesses A, Lam W, Bezold LI, Kaplan N, Towbin JA, Belmont JW. Echocardiographic evaluation of asymptomatic parental and sibling cardiovascular anomalies associated with congenital left ventricular outflow tract lesions. Pediatrics. 2004 Sep;114(3):691-6.

McGoon M, Gutterman D, Steen V, Barst R, McCrory DC, Fortin TA, Loyd JE. Screening, early detection, and diagnosis of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. Chest. 2004 Jul;126(1 Suppl):14S-34S.

Pereira L, Levran O, Ramirez F, Lynch JR, Sykes B, Pyeritz RE, Dietz HC. A molecular approach to the stratification of cardiovascular risk in families with Marfan's syndrome. N Engl J Med. 1994 Jul 21;331(3):148-53

Rosenthal D, Chrisant MR, Edens E, et al. International Society for Heart and Lung Transplantation: Practice guidelines for management of heart failure in children. J Heart Lung Transplant. 2004;23:1313-33.

Table 9: Outpatient Neonates without post-natal cardiology evaluation

Indication #		Guideline Recommendation
106.	Suspected cardiovascular abnormality on fetal echocardiogram	None
107.	Isolated echogenic focus on fetal ultrasound	None
108.	Maternal infection during pregnancy or delivery with potential fetal/ neonatal cardiac sequelae	None
109.	Maternal diabetes with no prior fetal echocardiogram	None
110.	Maternal diabetes with a normal fetal echocardiogram	None
111.	Maternal phenylketonuria	None
112.	Maternal autoimmune disorder	None

113.	Maternal teratogen exposure	None

References

Hornberger L, Rajaa A. Spectrum of Cardiac Involvement in Neonatal Lupus. Scandinavian Journal of Immunology. 2010;72,189-197.

Jenkins KJ, Correa A, Feinstein JA, et al. Noninherited risk factors and congenital cardiovascular defects: current knowledge: a scientific statement from the American Heart Association Council on Cardiovascular Disease in the Young: endorsed by the American Academy of Pediatrics. Circulation. 2007;115:2995-3014.

Levy HL, Guldberg P, Guttler F, et al. Congenital heart disease in maternal phenylketonuria: report from the Maternal PKU Collaborative Study. Pediatr Res. 2001;49:636-42.

Lisowski LA, Verheijen PM, Copel JA, et al. Congenital heart disease in pregnancies complicated by maternal diabetes mellitus. An international clinical collaboration, literature review, and meta-analysis. Herz. 2010;35:19-26.

Pruetz JD, Sklansky M, Detterich J, et al. Twin-twin transfusion syndrome treated with laser surgery: postnatal prevalence of congenital heart disease in surviving recipients and donors. Prenat Diagn. 2011;31:973-7.