Cigna Medical Coverage Policies – Radiology
Pediatric Cardiac Imaging Guidelines

Effective April 1, 2023

Instructions for use

The following coverage policy applies to health benefit plans administered by Cigna. Coverage policies are intended to provide guidance in interpreting certain standard Cigna benefit plans and are used by medical directors and other health care professionals in making medical necessity and other coverage determinations. Please note the terms of a customer's particular benefit plan document may differ significantly from the standard benefit plans upon which these coverage policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a coverage policy.

In the event of a conflict, a customer's benefit plan document always supersedes the information in the coverage policy. In the absence of federal or state coverage mandates, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of:

1. The terms of the applicable benefit plan document in effect on the date of service
2. Any applicable laws and regulations
3. Any relevant collateral source materials including coverage policies
4. The specific facts of the particular situation

Coverage policies relate exclusively to the administration of health benefit plans. Coverage policies are not recommendations for treatment and should never be used as treatment guidelines.

This evidence-based medical coverage policy has been developed by eviCore, Inc. Some information in this coverage policy may not apply to all benefit plans administered by Cigna.

These guidelines include procedures eviCore does not review for Cigna. Please refer to the Cigna CPT code list for the current list of high-tech imaging procedures that eviCore reviews for Cigna.

Table of Contents

Guideline Page

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

General Information

General Guidelines (PEDCD-1.0)

CDP.GG.0001.0.A
v1.0.2023

Heart disease in the pediatric population involves predominantly congenital lesions. Pediatric individuals can have acquired heart disease unique to children. For those diseases which occur in both pediatric and adult populations, differences exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.

Pediatric Cardiac Imaging Appropriate Clinical Evaluation

- Prior to considering advanced imaging (CT, MR, Nuclear Medicine) or echocardiogram, a pertinent clinical evaluation should be performed, including the following (both):
  - A detailed history, physical examination or meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging)
  - A review of appropriate diagnostic studies (laboratory, EKG, echo, and other diagnostic imaging)
- A recent clinical evaluation is not needed prior to advanced imaging (CT, MR, Nuclear Medicine) or echocardiogram if **any** of the following apply:
  - Individual is undergoing guideline-supported scheduled imaging evaluation
  - Echocardiogram is being performed at the first cardiology visit for an appropriate indication as stated in these guidelines
  - Routine imaging is anticipated at the next visit (e.g., one year follow-up echo for a 10 year old with a VSD)
- Advanced imaging of the heart and echocardiogram are medically necessary in any of the following:
  - Individuals who have documented active clinical signs or symptoms of disease involving the heart
  - As follow-up for findings on echocardiograms.
  - See Initial Transthoracic Echocardiography (TTE) Indications (PEDCD-8.2) for indications for initial echos in asymptomatic individuals
- Repeat imaging studies of the heart are **not** medically necessary unless one of the following applies:
  - Repeat imaging is indicated in a specific guideline section
  - There is evidence for progression of disease

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

  - There is new onset of disease with documentation of how repeat imaging will affect individual management or treatment decisions
  - See Repeat Transthoracic Echocardiography Indications (PEDCD-8.3) for indications for repeat echos in asymptomatic individuals
- Asymptomatic individuals with exposure to cardiotoxic drugs can have serial echocardiograms as per **Cardiotoxicity and Echocardiography (PEDONC-19.2)** in the Pediatric Oncology Imaging Guideline
- Advanced imaging and echocardiogram is **not** indicated, in the absence of other appropriate indications listed in these guidelines, for **any** of the following:
  - Individuals starting ADHD medications
  - To screen asymptomatic individuals for disorders involving the heart

Pediatric Cardiac Imaging Modality General Considerations

- MRI
  - MRI and MRA studies are frequently indicated for evaluation of congenital heart defects not well visualized on echocardiography, thoracic arteries and veins not visualized on echocardiography, cardiomyopathies, and right ventricular disease, as well as in follow-up for these indications.
  - Due to the length of time for image acquisition and the need for the individual to be motionless during the acquisition, anesthesia is required for almost all infants and young children (age <7 years), as well as older children with delays in development or maturity. In this population, MRI imaging sessions should be planned with a goal of avoiding a short-interval repeat anesthesia exposure due to insufficient information using the following considerations:
    - MRI is typically performed without and with contrast.
    - If multiple body areas are supported by eviCore guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.
- CT
  - CT is primarily used to evaluate the coronary and great vessels in congenital heart disease if cardiac MR is contraindicated.
  - Coding considerations are listed in CT Heart and Coronary Computed Tomography Angiography (CCTA) – Other Indications (PEDCD-10)
- Ultrasound

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

- Echocardiography is the primary modality used to evaluate the anatomy and function of the pediatric heart, and is generally indicated before considering other imaging modalities.
- Coding considerations are listed in **Echocardiography - Other Indications (PEDCD-8)**

- Nuclear Medicine
  - SPECT, PET stress may be indicated for individuals with anomalous CA, angina chest pain, and follow-up for Kawasaki and MIS-C. See specific sections for those indications.
  - Multi Gated Acquisition (MUGA) studies (CPT® 78472, CPT® 78473, CPT® 78481, CPT® 78483, CPT® 78494, or CPT® 78496) are rarely performed in pediatrics, but can be approved for the following:
    - Certain pediatric oncology individuals when echocardiography is insufficient: See: **Appropriate Clinical Evaluations (PEDONC-1.2)** for imaging guidelines.
    - Quantitation of left ventricular function when recent echocardiogram shows ejection fraction of <50% and MUGA results will impact acute patient care decisions.
  - SPECT/CT fusion imaging involves SPECT (MPI) imaging and CT for optimizing location, accuracy, and attenuation correction combines functional and anatomic information.
    - There is currently no evidence-based data to formulate appropriateness criteria for SPECT/CT fusion imaging.
    - Combined use of nuclear imaging, including SPECT, along with diagnostic CT (fused SPECT/CT) is considered investigational.
  - Central C-V Hemodynamics (CPT® 78414) is not an imaging study and is an outdated examination
  - Cardiac Shunt Detection (CPT® 78428) is rarely performed in pediatrics but can be approved for individuals in whom Cardiac MR is not diagnostic
    - Calculation of left and right ventricular ejection fractions
    - Assessment of wall motion
    - Quantitation of right to left shunts
  - Myocardial Tc-99m Pyrophosphate Imaging
    - Infarct Avid Myocardial Imaging studies (CPT® 78466, CPT® 78468, and CPT® 78469), historically this method of imaging the myocardium, Myocardial Tc-99m Pyrophosphate Imaging, was used to identify recent infarction, hence, the term "infarct-avid scan." Although still available, the

sensitivity and specificity for identifying infarcted myocardial tissue is variable and the current use for this indication is limited

- CPT® 78466, CPT® 78468, and CPT® 78469, CPT® 78800 or CPT® 78803 may be used, for identification of myocardial ATTR (transthyretin) amyloidosis. See **Myocardial Tc-99m Pyrophosphate Imaging (CD-3.7)** and **Cardiac Amyloidosis (CD-3.8)**

MUGA (Multi Gated Acquisition) – Blood Pool Imaging	CPT®
Myocardial Imaging, infarct avid, planar, qualitative or quantitative	78466
Myocardial Imaging, infarct avid, planar, qualitative or quantitative with ejection fraction by first pass technique	78468
Myocardial Imaging, infarct avid, planar, qualitative or quantitative tomographic SPECT with or without quantification	78469
Radiopharmaceutical Localization Imaging Limited area	78800
Radiopharmaceutical Localization Imaging SPECT Note: When reporting CPT® 78803, planar imaging of a limited area or multiple areas should be included with the SPECT	78803

Supporting Information

Individuals who are <18 years old should be imaged according to the Pediatric Cardiac Imaging Guidelines and the general Cardiac Imaging Guidelines. Individuals who are age ≥18 years should be imaged according to the Cardiac Imaging Guidelines, except where directed otherwise by a specific guideline section. Adult individuals who also have congenital heart disease should be imaged by **Adult Congenital Heart Disease (CD-11)** in the general Cardiac Imaging Guidelines.

The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

Procedure Codes Associated With Cardiac or PVD Imaging

MRI/MRA	CPT ®
Cardiac magnetic resonance imaging for morphology and function without contrast material	75557
Cardiac magnetic resonance imaging for morphology and function without contrast material; with stress imaging	75559
Cardiac magnetic resonance imaging for morphology and function without contrast material(s), followed by contrast material(s) and further sequences	75561
Cardiac magnetic resonance imaging for morphology and function without contrast material(s), followed by contrast material(s) and further sequences; with stress imaging	75563
Cardiac magnetic resonance imaging for velocity flow mapping (List separately in addition to code for primary procedure)	75565

CT	CPT ®
Computed tomography, heart, without contrast material, with quantitative evaluation of coronary calcium	75571
Computed tomography, heart, with contrast material, for evaluation of cardiac structure and morphology (including 3D image postprocessing, assessment of cardiac function, and evaluation of venous structures, if performed)	75572
Computed tomography, heart, with contrast material, for evaluation of cardiac structure and morphology in the setting of congenital heart disease (including 3D image postprocessing, assessment of LV cardiac function, RV structure and function and evaluation of venous structures, if	75573

CT	CPT ®
performed)	
Noninvasive estimated coronary fractional flow reserve (FFR) derived from coronary computed tomography angiography data using computation fluid dynamics physiologic simulation software analysis of functional data to assess the severity of coronary artery disease; data preparation and transmission, analysis of fluid dynamics and simulated maximal coronary hyperemia, generation of estimated FFR model, with anatomical data review in comparison with estimated FFR model to reconcile discordant data, interpretation and report	0501T
Noninvasive estimated coronary fractional flow reserve (FFR) derived from coronary computed tomography angiography data using computation fluid dynamics physiologic simulation software analysis of functional data to assess the severity of coronary artery disease; data preparation and transmission	0502T
Noninvasive estimated coronary fractional flow reserve (FFR) derived from coronary computed tomography angiography data using computation fluid dynamics physiologic simulation software analysis of functional data to assess the severity of coronary artery disease; analysis of fluid dynamics and simulated maximal coronary hyperemia, and generation of estimated FFR model	0503T
Noninvasive estimated coronary fractional flow reserve (FFR) derived from coronary computed tomography angiography data using computation fluid dynamics physiologic simulation software analysis of functional data to	0504T

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

CT	CPT ®
assess the severity of coronary artery disease; anatomical data review in comparison with estimated FFR model to reconcile discordant data, interpretation and report	

CTA	CPT ®
Computed tomographic angiography, heart, coronary arteries and bypass grafts (when present), with contrast material, including 3D image postprocessing (including evaluation of cardiac structure and morphology, assessment of cardiac function, and evaluation of venous structures, if performed)	75574
Computed tomographic angiography, abdominal aorta and bilateral iliofemoral lower extremity runoff, with contrast material(s), including noncontrast images, if performed, and image postprocessing	75635

Nuclear Medicine	CPT ®
Determination of central c-v hemodynamics (non-imaging) (eg, ejection fraction with probe technique) with or without pharmacologic intervention or exercise, single or multiple determinations	78414
Cardiac shunt detection	78428
Myocardial imaging, positron emission tomography (PET), metabolic evaluation study (including ventricular wall motion[s] and/or ejection fraction[s], when performed), single study; with concurrently acquired computed tomography transmission scan	78429
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s]	78430

Nuclear Medicine	CPT ®
and/or ejection fraction[s], when performed); single study, at rest or stress (exercise or pharmacologic), with concurrently acquired computed tomography transmission scan	
Myocardial imaging, positron emission tomography (PET), perfusion study (including ventricular wall motion[s] and/or ejection fraction[s], when performed); multiple studies at rest and stress (exercise or pharmacologic), with concurrently acquired computed tomography transmission scan	78431
Myocardial imaging, positron emission tomography (PET), combined perfusion with metabolic evaluation study (including ventricular wall motion[s] and/or ejection fraction[s], when performed), dual radiotracer (eg, myocardial viability);	78432
Myocardial imaging, positron emission tomography (PET), combined perfusion with metabolic evaluation study (including ventricular wall motion[s] and/or ejection fraction[s], when performed), dual radiotracer (eg, myocardial viability); with concurrently acquired computed tomography transmission scan	78433
Absolute quantitation of myocardial blood flow (AQMBF), positron emission tomography (PET), rest and pharmacologic stress (List separately in addition to code for primary procedure)	78434
Myocardial perfusion imaging, tomographic (SPECT) (including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); single study, at rest	78451

Nuclear Medicine	CPT ®
or stress (exercise or pharmacologic)	
Myocardial perfusion imaging, tomographic (SPECT) (including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); multiple studies, at rest and/or stress (exercise or pharmacologic) and/or redistribution and/or rest reinjection	78452
Myocardial perfusion imaging, planar (including qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); single study, at rest or stress (exercise or pharmacologic)	78453
Myocardial perfusion imaging, planar (including qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); multiple studies, at rest and/or stress (exercise or pharmacologic) and/or redistribution and/or rest reinjection	78454
Myocardial imaging, positron emission tomography (PET), metabolic evaluation study (including ventricular wall motion and/or ejection fraction, when performed), single study	78459
Myocardial imaging, infarct avid, planar; qualitative or quantitative	78466
Myocardial imaging, infarct avid, planar; with ejection fraction by first pass technique	78468
Myocardial imaging, infarct avid, planar; tomographic SPECT with or without quantification	78469
Cardiac blood pool imaging, gated equilibrium; planar, single study at	78472

Nuclear Medicine	CPT ®
rest or stress (exercise and/or pharmacologic), wall motion study plus ejection fraction, with or without additional quantitative processing	
Cardiac blood pool imaging, gated equilibrium; multiple studies, wall motion study plus ejection fraction, at rest and stress (exercise and/or pharmacologic), with or without additional quantification	78473
Cardiac blood pool imaging (planar), first pass technique; single study, at rest or with stress (exercise and/or pharmacologic), wall motion study plus ejection fraction, with or without quantification	78481
Cardiac blood pool imaging (planar), first pass technique; multiple studies, at rest and with stress (exercise and/or pharmacologic), wall motion study plus ejection fraction, with or without quantification	78483
Myocardial imaging, positron emission tomography (PET), perfusion (including ventricular wall motion and/or ejection fraction, when performed); single study at rest or stress (exercise or pharmacologic)	78491
Myocardial imaging, positron emission tomography (PET), perfusion (including ventricular wall motion and/or ejection fraction, when performed); multiple studies at rest and/or stress (exercise or pharmacologic)	78492
Cardiac blood pool imaging, gated equilibrium, SPECT, at rest, wall motion study plus ejection fraction, with or without quantitative processing	78494
Cardiac blood pool imaging, gated equilibrium, single study, at rest, with right ventricular ejection fraction by first pass technique (List separately in	78496

Nuclear Medicine	CPT ®
addition to code for primary procedure)	
Quantitative differential pulmonary perfusion, including imaging when performed	78597
Quantitative differential pulmonary perfusion and ventilation (eg, aerosol or gas), including imaging when performed	78598
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, single area (eg, head, neck, chest, pelvis), single day imaging	78800
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, 2 or more areas (eg, abdomen and pelvis, head and chest), 1 or more days imaging or single area imaging over 2 or more days	78801
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, whole body, single day imaging	78802
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT), single area (eg, head, neck, chest, pelvis), single day imaging	78803
Radiopharmaceutical localization of tumor, inflammatory process or	78804

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Nuclear Medicine	CPT ®
distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, whole body, requiring 2 or more days imaging	
Radiopharmaceutical localization of tumor, inflammatory process or distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); tomographic (SPECT) with concurrently acquired computed tomography (CT) transmission scan for anatomical review, localization and determination/detection of pathology, single area (eg, head, neck, chest, pelvis), single day imaging	78830
Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment	0331T
Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment; with tomographic SPECT	0332T

Ultrasound	CPT ®
Transthoracic echocardiography for congenital cardiac anomalies; complete	93303
Transthoracic echocardiography for congenital cardiac anomalies; follow-up or limited study	93304
Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, complete, with spectral Doppler echocardiography, and with color flow Doppler echocardiography	93306
Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, complete, without spectral	93307

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Ultrasound	CPT ®
or color Doppler echocardiography	
Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, follow-up or limited study	93308
Echocardiography, transesophageal, real-time with image documentation (2D) (with or without M-mode recording); including probe placement, image acquisition, interpretation and report	93312
Echocardiography, transesophageal, real-time with image documentation (2D) (with or without M-mode recording); placement of transesophageal probe only	93313
Echocardiography, transesophageal, real-time with image documentation (2D) (with or without M-mode recording); image acquisition, interpretation and report only	93314
Transesophageal echocardiography for congenital cardiac anomalies; including probe placement, image acquisition, interpretation and report	93315
Transesophageal echocardiography (TEE) for congenital cardiac anomalies; placement of transesophageal probe only	93316
Transesophageal echocardiography for congenital cardiac anomalies; placement of transesophageal probe only	93317
3D echocardiographic imaging and postprocessing during transesophageal echocardiography, or during transthoracic echocardiography for congenital cardiac anomalies, for the assessment of cardiac structure(s) (eg, cardiac chambers and valves, left	+93319

Ultrasound	CPT ®
atrial appendage, interatrial septum, interventricular septum) and function, when performed (List separately in addition to code for echocardiographic imaging) Code with (93303-93304, 93312, 93314, 93315, 93317, 93350-93351)	
Doppler echocardiography, pulsed wave and/or continuous wave with spectral display (List separately in addition to codes for echocardiographic imaging); complete	+93320
Doppler echocardiography, pulsed wave and/or continuous wave with spectral display (List separately in addition to codes for echocardiographic imaging); follow-up or limited study (List separately in addition to codes for echocardiographic imaging)	+93321
Doppler echocardiography color flow velocity mapping (List separately in addition to codes for echocardiography)	+93325
Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation and report;	93350
Echocardiography, transthoracic, real-time with image documentation (2D), includes M-mode recording, when performed, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation and report; including performance of continuous electrocardiographic monitoring, with supervision by a physician or other	93351

Ultrasound	CPT ®
qualified health care professional	
Use of echocardiographic contrast agent during stress echocardiography (List separately in addition to code for primary procedure)	+93352
Myocardial strain imaging using speckle tracking-derived assessment of myocardial mechanics (List separately in addition to codes for echocardiography imaging)	+93356
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, for congenital cardiac anomalies; complete	C8921
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, for congenital cardiac anomalies; follow-up or limited study	C8922
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, real-time with image documentation (2D), includes M-mode recording, when performed, complete, without spectral or color doppler echocardiography	C8923
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, real-time with image documentation (2D), includes M-mode recording when performed, follow-up or limited study	C8924
Transesophageal echocardiography (TEE) with contrast, or without contrast followed by with contrast, real time with image documentation (2D) (with or without M-mode recording); including probe placement, image acquisition, interpretation and report	C8925
Transesophageal echocardiography (TEE) with contrast, or without	C8926

Ultrasound	CPT ®
contrast followed by with contrast, for congenital cardiac anomalies; including probe placement, image acquisition, interpretation and report	
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, real-time with image documentation (2D), includes M-mode recording, when performed, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation and report	C8928
Transthoracic echocardiography with contrast, or without contrast followed by with contrast, real-time with image documentation (2D), includes M-mode recording, when performed, complete, with spectral doppler echocardiography, and with color flow doppler echocardiography	C8929
Transthoracic echocardiography, with contrast, or without contrast followed by with contrast, real-time with image documentation (2D), includes M-mode recording, when performed, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation and report; including performance of continuous electrocardiographic monitoring, with physician supervision	C8930
Myocardial contrast perfusion echocardiography, at rest or with stress, for assessment of myocardial ischemia or viability (List separately in addition to code for primary procedure)	+0439T

Cardiac Cath Procedure	CPT ®
Congenital Heart Disease Code “Set”	93593-93597

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Cardiac Cath Procedure	CPT ®
Right heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone; normal native connections	93593
Right heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone; abnormal native connections	93594
Left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone, normal or abnormal native connections	93595
Right and left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone(s); normal native connections	93596
Right and left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone(s); abnormal native connections	93597
Anomalous coronary arteries, patent foramen ovale, mitral valve prolapse, and bicuspid aortic valve	93451-93464, 93566-93568
RHC without LHC or coronaries	93451
LHC without RHC or coronaries	93452
RHC and retrograde LHC without coronaries	93453
Native coronary artery catheterization;	93454
with bypass grafts	93455
with RHC	93456
with RHC and bypass grafts	93457
with LHC	93458
with LHC and bypass grafts	93459

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Cardiac Cath Procedure	CPT ®
with RHC and LHC	93460
with RHC and LHC and bypass grafts	93461
LHC by trans-septal or apical puncture	+93462
Angiography of non-coronary arteries and veins performed as a distinct service	Select appropriate codes from the Radiology and Vascular Injection Procedures sections.

References

1. Karmazyn BK, John SD, Siegel MJ, et al. ACR–ASER–SCBT-MR–SPR Practice parameter for the performance of pediatric computed tomography (CT). *Am Coll Radiology (ACR)*. Revised 2014 (Resolution 3). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/ct-ped.pdf?la=en.

2. Bridges MD, Berland LL, Kirby AB, et al. ACR Practice Parameter for performing and interpreting magnetic resonance imaging (MRI). *Am Coll Radiology (ACR).* Revised 2017 (Resolution 10). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/mr-perf-interpret.pdf?la=en.

3. Sorantin E and Heinzl B. What every radiologist should know about paediatric echocardiography. *Eur J Radiol.* 2014 Sep;83(9):1519-1528.

4. Ing C, DiMaggio C, Whitehouse A, et al. Long-term differences in language and cognitive function after childhood exposure to anesthesia. *Pediatrics.* 2012 Aug;130(3):e476-e485.

5. Monteleone M, Khandji A, Cappell J, et al. Anesthesia in children: perspectives from nonsurgical pediatric specialists. *J Neurosurg Anesthesiol,* 2014 Oct 01;26(4):396-398.

6. DiMaggio C, Sun LS, and Li G. Early childhood exposure to anesthesia and risk of developmental and behavioral disorders in a sibling birth cohort. *Anesth Analg,* 2011 Nov;113(5):1143-1151.

7. Moss AJ, Adams FH, Allen HD, et al.Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. Philadelphia: Wolters Kluwer; 2016

8. Lai WW, Geva T, Shirali GS, et al. Guidelines and Standards for Performance of a Pediatric Echocardiogram: A Report from the Task Force of the Pediatric Council of the American Society of Echocardiography. *J Am Soc Echocardiogr*. 2006;19(12):1413-1430. doi:10.1016/j.echo.2006.09.001.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

9. Doherty JU, Kort S, Mehran R, Schoenhagen P, et al. ACC/AATS/AHA/AYSE/ASNC/HRS/SCAI/SCCT/SCMR/STS 2017 Appropriate Use Criteria for Multimodality Imaging in Valvular Heart Disease. J Nuc Cardiol. 2017;24(6):2043-2063.

10. Chowdhury D, Gurvitz M, Anderson J, et al. Development of Quality Metrics in Ambulatory Pediatric Cardiology. JACC: *J Am Coll Cardiol*. 2017 Feb, 69 (5) 541-555. doi: 10.1016/j.jacc.2016.11.043.

11. Valente AM, Cook S, Festa P, et al. Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography. *J Am Soc Echocardiogr*. 2014;27(2):111-141. doi:10.1016/j.echo.2013.11.009.

12. Simpson J, Lopez L, Acar P, et al. Three-dimensional Echocardiography in Congenital Heart Disease: An Expert Consensus Document from the European Association of Cardiovascular Imaging and the American Society of Echocardiography. *J Am Soc Echocardiogr*. 2017;30(1):1-27. doi:10.1016/j.echo.2016.08.022.

13. Cohen MS, Eidem BW, Cetta F, et al. Multimodality Imaging Guidelines of Patients with Transposition of the Great Arteries: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography. *J Am Soc Echocardiogr*. 2016;29(7):571-621. doi:10.1016/j.echo.2016.04.002.

14. Silvestry FE, Cohen MS, Armsby LB, et al. Guidelines for the Echocardiographic Assessment of Atrial Septal Defect and Patent Foramen Ovale: From the American Society of Echocardiography and Society for Cardiac Angiography and Interventions.*J Am Soc Echocardiogr*. 2015;28(8):910-958. doi:10.1016/j.echo.2015.05.015.

15. Paridon SM. Clinical Stress Testing in the Pediatric Age Group: A Statement from the American Heart Association Council on Cardiovascular Disease in the Young, Committee on Atherosclerosis, Hypertension, and Obesity in Youth. *Circulation*. 2006;113(15):1905-1920. doi:10.1161/CIRCULATIONAHA.106.17437D.

16. Gidding SS, Champagne MA, Ferranti SDD, et al. The Agenda for Familial Hypercholesterolemia. *Circulation*. 2015;132(22):2167-2192. doi:10.1161/CIR.0000000000000297.

17. National Cancer Institute. Radiation Risks and Pediatric Computed Tomography. National Cancer Institute. https://www.cancer.gov/about-cancer/causes-prevention/risk/radiation/pediatric-ct-scans. Published September 4, 2018.

18. Hundley WG, Bluemke DA, Finn JP, et al. ACCF/ACR/AHA/NASCI/SCMR 2010 Expert Consensus Document on Cardiovascular Magnetic Resonance. *Circulation*. 2010;121(22):2462-2508. doi:10.1161/cir.0b013e3181d44a8f.

Congenital Heart Disease

Congenital Heart Disease General Information (PEDCD-2.1)

CDP.CHD.0002.1.A
v1.0.2023

- Congenital heart disease accounts for the majority of cardiac problems occurring in the pediatric population. Individuals may be diagnosed any time spanning prenatal evaluation to adolescence. For individuals over 18 years of age, see **Adult Congenital Heart Disease (CD-11)** in the Cardiac Imaging Guidelines.
- There are a number of variables that influence the modality and timing of imaging individuals with congenital heart disease, which results in a high degree of individuality in determining the schedule for imaging these individuals, including:
  - Gestational age
  - Individual's age
  - Physiologic effects of the defect
  - Status of interventions (catheterization and surgical)
  - Rate of individual's growth
  - Stability of the defect on serial imaging
  - Comorbid conditions
  - Activity level
- Age definitions for pediatric individuals (for purpose of these guidelines)
  - Infant 0-12 months
    - Subcategory of infant: Neonate or newborn 0-28 days
  - Child 1-18 years
    - Subcategory of child: Adolescent 11-18 years
  - "Children" refers to all pediatric individuals ages 0-18 years
- Newborns (neonates) have special considerations as they have potentially rapidly changing physiology
  - Newborns with any concerns for ductal dependent lesion can have echocardiograms at any frequency
  - Newborns have changes in pulmonary vascular resistance that can affect clinical status rapidly, and may require more frequent imaging.
  - Neonatal physiology can extend to the first couple of months of life.

  - Newborns can have one repeat echo, if prior echocardiogram is abnormal or equivocal (either in the hospital or as newborn outpatient)

- Individuals can have an echocardiogram at that time for **Change in clinical status and/or new concerning signs or symptoms**. This can include:
  - Shortness of breath
  - Fatigue
  - Chest discomfort
  - Percentile weight loss
  - Weight gain
  - Poor feeding
  - Tachypnea
  - Tachycardia
  - CHF signs on exam
  - Change in EKG, Pulse ox, laboratory values.
- An additional study can be approved prior to the next routine interval, to assess for more rapid change, if the change in clinical status involves the echocardiogram itself, such as:
  - Increasing stenosis gradient
  - Increasing regurgitation amount
  - Increasing pulmonary vascular resistance
  - Decreased ventricular function
  - Change in ductal status,
- In individuals that can have both Cardiac MR or Cardiac CT and/or MRA Chest or CTA Chest, this is abbreviated as CMR/CT-CMRA/CTA
- Individuals with medication adjustments may require additional imaging at that time.
  - Pediatric dosing tends to be mg/kg or mcg/kg. Adjusting the dose to the same mg/kg would not be considering a dosing change for imaging.
  - Because does adjustments are done by weight, and infants are growing rapidly, they can have changing physiology, pulmonary vascular resistant, ductal size and weight changes, dose response and may require more than one echo during a medication adjustment.
- Heart surgery
  - One month prior to heart surgery, individuals can have TTE (depending on lesion can also include MR/CT Cardiac and/or Chest)

  - Can have an echocardiogram within one month post-operative
  - Cardiac MRI/CCT if prior echo is equivocal

- MRA/CTA Chest can be performed if prior echo is equivocal and there are issues regarding aortic arch or pulmonary arteries or veins
- In individuals who have a documented equivocal echocardiogram due to a technical factor (i.e., poor acoustic windows due to body habitus) which will likely be present on subsequent echocardiograms, a Cardiac MR/CT, or MRA/CTA Chest, may be done with the frequency of echoes, if done instead of an echo.
- MRA/CTA Chest if thoracic issue not seen on echo
- For routine non-invasive imaging for a specific lesion see **Imaging and Surveillance per Congenital lesion (PEDCD-2.4)** .
- For catheterizations see section **Cardiac Catheterization (PEDCD-11)**
- **Individuals with Pulmonary hypertension with CHD** should be reviewed for both their lesion and for PHT in section **Pediatric Pulmonary Hypertension (PEDCD-7)**

Congenital Heart Disease Coding (PEDCD-2.2)

CDP.CHD.0002.2.A
v1.0.2023

Congenital Heart Disease Echocardiography Coding (PEDCD-2.2.1)

- **Any** of the following echocardiography code combinations are appropriate for re-evaluation of individuals with known congenital heart disease:
  - CPT® 93303, CPT® 93320, and CPT® 93325
  - CPT® 93304, CPT® 93321, and CPT® 93325
  - CPT® 93303
  - CPT® 93304
- CPT® 93306 is not indicated in the evaluation of known congenital heart disease.

Congenital Heart Disease Imaging per Modality (PEDCD-2.2.2)

Echocardiogram

- Transthoracic echocardiogram (TTE)
  - TTE for congenital cardiac anomalies; complete (CPT® 93303)
  - TTE for congenital cardiac anomalies; limited study (CPT® 93304)
  - TTE (2D) m-mode recording, complete, with spectral and color flow doppler echocardiography (CPT® 93306)
  - TTE (2D) with or without m-mode recording; complete (CPT® 93307)
  - TTE (2D) with or without m-mode recording; limited study (CPT® 93308)
- Transesophageal echocardiogram (TEE)
  - TEE (2D) including probe placement, imaging, interpretation, and report (CPT® 93312)
  - TEE for congenital cardiac anomalies; including probe placement, imaging, interpretation, and report (CPT® 93315)

MRI

- Cardiac (CMR)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

- Cardiac MRI for morphology and function without contrast (CPT® 75557)
- Cardiac MRI for morphology and function without and with contrast (CPT® 75561)
- Cardiac magnetic resonance imaging for velocity flow mapping (List separately in addition to code for primary procedure) 75565

- MRI Chest
  - MRI Chest without contrast (CPT® 71550)
  - MRI Chest with contrast (CPT® 71551)
  - MRI Chest with & without contrast (CPT® 71552)
- MRI Angiography (MRA)
  - MRA Chest (excluding myocardium) with or without contrast (CPT® 71555)

CT

- Cardiac (CCT)
  - CT, Heart, with contrast material, for evaluation of cardiac structure and morphology (CPT® 75572)
  - CT, Heart, with contrast material, for evaluation of cardiac structure and morphology in the setting of congenital heart disease (CPT® 75573)
- CT Angiography-Cardiac (CCTA)
  - CTA Heart, coronary arteries and bypass grafts (when present), with contrast, including 3D image post processing (CPT® 75574)
- CT-Chest
  - CT Thorax without contrast (CPT® 71250)
  - CT Thorax with contrast (CPT® 71260)
  - CT Thorax without & with contrast (CPT® 71270)
- CT Angiography-Chest (CTA Chest)
  - CTA Chest without and with contrast (CPT® 71275)

Stress Imaging (echo, MRI, MPI)

- Stress echo
  - Echocardiography (TTE), (2D), with or without m-mode, during rest and cardiovascular stress, with interpretation and report (CPT® 93350)

  - Echocardiography (TTE), (2D), m-mode, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation (CPT® 93351)

- Stress MRI
  - Cardiac MRI for morphology and function without contrast, with stress imaging (CPT® 75559)
  - Cardiac MRI for morphology and function without and with contrast, with stress imaging (CPT® 75563)
- Myocardial perfusion imaging (MPI)
  - MPI, tomographic (SPECT) including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); single study, at rest or stress (exercise or pharmacologic) (CPT® 78451)
  - MPI, tomographic (SPECT) (including attenuation correction, qualitative or quantitative wall motion, ejection fraction by first pass or gated technique, additional quantification, when performed); multiple studies, at rest and/or stress (exercise or pharmacologic) and/or redistribution and/or rest reinjection (CPT® 78452)

Pulmonary perfusion imaging

- Pulmonary perfusion imaging (e.g., particulate) (CPT® 78580)
- Pulmonary ventilation (e.g., aerosol or gas) and perfusion imaging (CPT® 78582)
- Quantitative differential pulmonary perfusion, including imaging when performed (CPT® 78597)
- Quantitative differential pulmonary perfusion and ventilation (e.g., aerosol or gas), including imaging when performed (CPT® 78598)

Congenital Heart Disease Modality Considerations (PEDCD-2.3)

CDP.CHD.0002.3.A
v1.0.2023

- Echocardiography is the primary imaging modality used for diagnosing and monitoring congenital heart disease and is generally required before other imaging modalities are indicated unless otherwise indicated in a specific guideline section.
- Cardiac MRI either without contrast (CPT® 75557) or without and with contrast (CPT® 75561) is indicated, when a recent echocardiogram is inconclusive, needs confirmation of findings, or does not completely define the disease (for subsequent follow-up studies, a recent echocardiogram is not a requirement):
    - CPT® 75565 is also indicated for individuals with valvular disease or a need to evaluate intracardiac blood flow. These individuals will usually have CPT® 93320 and CPT® 93325 performed with their echocardiography studies.
    - MRA Chest (CPT® 71555) may be added if the aorta or pulmonary artery needs to be visualized beyond the root, or if aortopulmonary collaterals, pulmonary veins, or systemic veins need to be visualized.
        - MRA Chest alone (CPT® 71555) should be performed if the individual cannot cooperate with full cardiac MRI exam.
- MRA Chest (CPT® 71555) is indicated for assessment of the great arteries, pulmonary veins, and systemic chest veins with inconclusive recent echocardiography findings, including the following:
    - Coarctation of the aorta
    - Tetralogy of Fallot
    - Anomalous pulmonary veins
    - Transposition of the great arteries
    - Truncus arteriosus
    - Vascular rings and other lesions of the great arteries, with inconclusive recent echocardiography findings
- CT imaging is indicated when recent echocardiogram is inconclusive:
    - Report CPT® 75574 for evaluating coronary artery anomalies
    - Report CPT® 75573 for congenital heart disease
    - CPT® 71275 Determination of vascular extra-cardiac anatomy in individuals with complex congenital heart disease
    - Pulmonary artery (PA) and Pulmonary vein (PV) assessment

- CTA of the chest is indicated with inconclusive recent echocardiography findings to assess:
  - Coarctation of the aorta
  - Tetralogy of Fallot
  - Anomalous pulmonary veins and other lesions of the great arteries
  - Vascular rings
- Pulmonary perfusion imaging
  - Pulmonary perfusion imaging (e.g., particulate) (CPT® 78580)(CPT® 78582) (CPT® 78597)(CPT® 78598)
  - In individuals with congenital heart disease or suspected congenital heart diseases, who have clinical questions regarding relative pulmonary blood flow, can have perfusion imaging

Imaging and Surveillance per Congenital Lesion (PEDCD-2.4)

CDP.CHD.0002.4.A
v1.0.2023

- Echocardiography is often repeated frequently throughout a pediatric individuals life, and can generally be approved regardless of symptoms based on the lesion and age of the individual. These are listed in sections in sections below.
    - Modifiers following guidelines.
        - Some congenital conditions may require more frequent testing, especially with more complex heart disease, congestive heart failure, obstructive heart lesions, ductal dependent lesions, changes in clinical status, repeat interventions, and/or in neonates
        - Any individual being treated for heart failure, with consideration for changing medical regimen can have an echocardiogram
- Echocardiography is performed during the physician office visit, and these studies should not be denied because of lack of contact within 60 days.
- Adults 18 years and older who also have congenital heart disease should be imaged according to **Adult Congenital Heart Disease (CD-11)** and the general Cardiac Imaging Guidelines.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Atrial Defects-Secundum ASD, PFO, and Partial Anomalous Pulmonary Venous Return (PAPVR), Sinus Venosus Defect (PEDCD-2.4.1)

CDP.CHD.0241.A
v1.0.2023

See section on AVSD in AVSD (Atrioventricular canal, Endocardial cushion defect) (PEDCD-2.4.3) for primum ASD

PFO (Patent Foreman Ovale)

- Routine surveillance in an asymptomatic individual with PFO is not indicated
  - PFO is a normal variant
  - In infants, a PFO that is difficult to distinguish from an ASD can be imaged with the same guidelines as used in a small unrepaired ASD (with congenital echo).
  - Individuals with PFOs may have an additional indication for an echo and can be imaged according to the echocardiogram guidelines in **Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)** and **Frequency of echocardiography testing (CD-2.3)** in the general Cardiac Imaging Guidelines.
  - Follow-up imaging with an echocardiogram can be approved when there is documentation of the following:
    - New cardiac symptoms
    - There is a concern that the last echo was equivocal for other cardiac issues
    - There is question of a clot/embolism that has gone across the PFO
    - The last echo did not differentiate the PFO from a secundum ASD
- TTE (CPT® 93306- non congenital echocardiogram) is indicated when an individual with a prior history of PFO requires an echocardiogram for any new reason
- Preoperative for PFO closure
  - TTE or TEE
    - Closure is rare in children, but may be indicated in individuals with transient ischemic attacks or strokes with suspected atrial level shunt
    - CMR/CT-CMRA/CTA if unclear findings from echocardiogram.
- Intra-procedural PFO

- Intra-procedural TEE (CPT® 93355) is not in scope for this program

- Post procedure PFO closure
  - Post-surgical imaging as follows (PFO generally requires less frequent monitoring post device than ASDs):
    - TTE one time within 30 days of closure
    - TTE one time within 6 months of PFO closure
  - TTE or TEE is indicated at any time post procedure when there is concern for **any** of the following:
    - Infection
    - Malposition
    - Embolization
    - Persistent shunt.
  - If persistent shunt, **see ASD device criteria** .

ASD and PAPVR Asymptomatic Isolated Atrial Septal Defect (ASD)

- This section reference secundum ASD, sinus Venosus, ASD and unobstructed partial anomalous pulmonary venous return
- Any surgical status
  - TTE is indicated for any of the following:
    - Initial evaluation of a change in clinical status and/or new concerning signs or symptoms
    - Prior to planned cardiac intervention
    - Repeat any time prior to next allowed study if concern for elevated pulmonary vascular resistance/Pulmonary hypertension
  - CMR/CT-CMRA/CTA
    - If anomalous vein or SV defect cannot be assessed on echo
    - To assess shunt or RV for considering of surgery, or if echocardiogram equivocal.
  - Unrepaired
    - Newborn with isolated ASD can have one repeat TTE within 2 months
    - Small asymptomatic isolated ASD with no pulmonary hypertension can have TTE as follows:

- Infant <6 months every three months
- Infant ≥6 months, repeat at one year.
- Child Every 3 years

  - Routine surveillance for ≥moderate ASD or PAPVR >1 vein

    - Infant every 3 months
    - Echo (TTE) every 1 year

- Prior to planned repair of ASD
  - TTE and/or TEE
  - MRI if any residual issues unanswered by echo
- Prior to planned SV defect or PAPVR
  - TTE and/or TEE
  - CMR/CT-CMRA/CTA
- Post- ASD closure with device
  - TTE post device closure
    - 1 week
    - 1 month
    - Every 3 months
    - 1 year
    - Every 2 years
  - May repeat TTE every 3 months until the finding is stable or there is a need for intervention if there is significant residual shunt, valvular or ventricular dysfunction, arrhythmias, and/or pulmonary hypertension
- Post-surgical closure of ASD
  - TTE
    - Within the first month
    - Within the 1st year
    - Every 2 years after the first year study
  - May repeat TTE every 3 months until the finding is stable or there is a need for intervention if significant residual shunt, valvular or ventricular dysfunction, arrhythmias, and/or pulmonary hypertension.

VSD (PEDCD-2.4.2)

CDP.CHD.0242.A
v1.0.2023

All

TTE is indicated for any of the following:

- with change in clinical status and/or new concerning signs or symptoms
- Prior to planned cardiac intervention

Unrepaired

TTE

- Small muscular VSD, No Symptoms, No pulmonary hypertension
  - Newborn 1 repeat within 2 months
  - Infancy every 6 months
  - Childhood every 3 years
- Small VSD in location other than muscular
  - Newborn 1 repeat TTE within 2 months
  - Infant TTE every 6 months
  - Child TTE every year.
- Moderate or large VSD on medical management
  - Newborn TTE every 2 weeks
  - infant every 1 month
  - Child <2 years old TTE every 3 months
  - Child >2 years old TTE every year.

Post Repair VSD

TTE

- One study within one month of surgery

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- One study within one year of surgery,
- After first year of surgery, every 2 years
- Following surgical or device closure in an individual with significant residual shunt, valvular or ventricular dysfunction, arrhythmias, and/or pulmonary hypertension.
    - Child –TTE every 3 months
    - Adolescent- TTE every 6 months

AVSD (Atrioventricular Canal, Endocardial Cushion Defect) (PEDCD-2.4.3)

CDP.CHD.0243.A
v1.0.2023

Any Surgical Status

TTE is indicated for any of the following:

- Change in clinical status and/or new concerning signs or symptoms
- Prior to planned cardiac intervention

Unrepaired

- Partial/transitional Atrioventricular canal (AVC)
  - Newborn one addition study next 2 months.
  - TTE
    - Infancy every 3 months in infancy
    - Child <2 years every 6 months
    - Child ≥2 years, 1 year
- Complete AVC
  - TTE
    - Newborn, TTE repeat within first month
    - Infant <6 weeks, TTE every 2 weeks.
    - Infant ≥6 weeks, TTE monthly

Repaired (TTE)

- Within one month of surgery
- Within 1 year
- Then annually

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- May repeat TTE every 3 months until the finding is stable or there is a need for intervention if residual shunt, valvular LV dysfunction, LVOT obstruction, arrhythmia, arrhythmia or PHT, symptoms of heart failure

PDA (Patent Ductus Arteriosus) (PEDCD-2.4.4)

CDP.CHD.0244.A
v1.0.2023

Any Surgical Status

TTE is indicated for any of the following:

- Initial evaluation of a change in clinical status and/or new concerning signs or symptoms
- Prior to planned cardiac intervention

Unrepaired

- Newborn, one repeat TTE in newborn period
  - None, if spontaneously closed
- >1-year-old
  - No Routine surveillance in an asymptomatic individual with a trivial, silent PDA
- Infant
  - Small TTE every 3 months
  - ≥moderate/ TTE every month
- Child small PDA every 1 year
- Child Moderate PDA every 6 months
- Adolescent every 3 years

Post PDA Device

- Post procedure surveillance (TTE)
  - One echo in first 30 days
  - Annually for first 2 years
  - Every 5 years after first 2 years
- Post procedure LPA stenosis or aortic obstruction

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- Child
  - TTE annually
  - MRA/CTA Chest, or (lung perfusion for LPA stenosis) if questions remain unanswered after TTE
- Adolescents
  - Every two years TTE and
  - MRA/CTA Chest, or (lung perfusion for LPA stenosis) if questions remain unanswered after TTE

TAPVR Total Anomalous Pulmonary Venous Return (PEDCD-2.4.5)

CDP.CHD.0245.A
v1.0.2023

Any Surgical Status

- TTE, TEE, CMR/CT-CMRA/CTA, Lung perfusion scan are indicated for any of the following:
  - Change in clinical status and/or new concerning signs or symptoms
  - Prior to planned cardiac intervention

Unrepaired

- No restrictions

Repaired

- TTE one Post procedure evaluation first 30 days
- TTE every 3 months in infancy
- Child: every 1 year
- Adolescence: TTE every 2 years

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Ebstein Anomaly and TV Dysplasia (PEDCD-2.4.6)

CDP.CHD.0246.A
v1.0.2023

Any Surgical Status

- TTE, TEE, CMR/CT-CMRA/CTA are indicated for any of the following:
  - Change in clinical status and/or new concerning signs or symptoms
  - Prior to planned repair or intervention

Unrepaired

- Newborn Repeat study within 30 days.
- Infant
  - Trivial TR is a normal finding
  - Mild TR- TTE every year
  - ≥moderate TR- TTE every 3 months
- Child
  - Mild TR every year TTE
  - ≥moderate every 6 months

Repaired (TTE)

- Post op within 30 days
- TTE once a year
- TTE every 6 months if Valvular or ventricular dysfunction, or arrhythmias
- Child every year
- Adolescent every 2 years
- Every 3 months if CHF or atrial arrhythmias

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Pulmonary Stenosis (PS) (PEDCD-2.4.7)

CDP.CHD.0247.A
v1.0.2023

Any Surgical Status

- TTE is indicated for any of the following:
  - Change in clinical status and/or new concerning signs or symptoms
  - Prior to planned cardiac procedure
  - If increasing gradient, 1 additional study prior to next allowed study
  - PS in Williams syndrome: See LVOT lesions (PEDCD-2.4.10)

Unrepaired

- Neonate
  - TTE repeat study within 30 days
- Infant PS asymptotic (any severity)
  - TTE every 3 months
- Child
  - TTE every 1 year
- Adolescent
  - TTE every 2 years
  - MRA/CTA Chest if pulmonary artery dilation every 3 years

Post Procedure (TTE)

- Within 30 days
- Infant
  - TTE every 3 months
- Child

  - TTE 1 year
  - Moderate or severe sequelae TTE every 6 months
- Adolescent
  - TTE every 2 years
- Any individual with heart failure, TTE every 3 months

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Pulmonary Atresia with Intact Septum (PAIVS) (PEDCD-2.4.8)

CDP.CHD.0248.A
v1.0.2023

Any Surgical Status

TTE is indicated for any of the following:

- Prior to planed repair
- Change in clinical status and/or new concerning signs or symptoms

Post Procedural: Palliation

- TTE
  - 1 within 30 days
  - Every 1 month until repaired

Post Procedural: Complete Repair

- TTE within 30 days post op
- Any age
  - TTE every three months for CHF
- Infant
  - TTE at 3 months in asymptomatic infant
- Child
  - TTE annually
  - Every 6 months if moderate sequelae
- Adolescent
  - CMR/CT and/or CMRA/CTA every 3 years

Mitral Valve Disease (PEDCD-2.4.9)

CDP.CHD.0249.A
v1.0.2023

Any Surgical Status

- TTE is indicated for any of the following:
  - Prior to planned surgery
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms

Unrepaired Congenital Mitral Valve Stenosis

- Infant in First three months of life
  - Weekly TTE
- After 3 months (TTE)
  - Every 3 months if mild MS
  - Every month if ≥moderate MS
- Child (TTE)
  - With moderate MS every 3 months until a decision is made to intervene
  - Child with mild symptoms annually

Unrepaired: Congenital Mitral Regurgitation (MR) Including Mitral Valve Prolapse

- Infant
  - TTE every 6 months an asymptomatic infant with mild MR
  - TTE every month in asymptomatic infant with ≥moderate MR
- Child
  - TTE every 2 years with mild MR, normal LV size and systolic function
  - TTE every 6 months with ≥moderate MR
  - TTE every 3 years in an asymptomatic with MVP and mild MR

Post Procedure, Surgical or Catheter Based

- TTE within 30 days
- Infant
  - TTE every 3 months, mild MS or MR, and no LV dysfunction
  - TTE every month in ≥moderate MS or MR, dilated LV, and no LV dysfunction
- Child
  - TTE annually
    - In a child with normal prosthetic mitral valve function and no LV dysfunction
    - In a child with mild MS or MR, and no LV dysfunction
  - TTE every 3 months
    - In a child with ≥moderate MS or MR, dilated LV, and no LV dysfunction
    - In a child with prosthetic mitral valve or ventricular dysfunction, and/or arrhythmias

LVOT Lesions (PEDCD-2.4.10)

CDP.CHD.2410.A
v1.0.2023

Subvalvular Aortic stenosis

Any Surgical Status

- TTE, TEE, Cardiac MR/CT are indicated for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Preoperative
- If aortic dimension z score >2
  - TTE or Chest CTA/MRA every 2 years if stable z score
  - TTE or Chest CTA/MRA every 6 months if increasing z score

Unrepaired

- Newborn- No restrictions
- Infant TTE
  - 1 monthly for any subAS, but ≤mild AR
- Child
  - TTE one per year if mild AS and no AR
  - TTE every 6 months ≥moderate Subvalvular AS and/or Mild AR
  - Routine surveillance (6–12 months) in an asymptomatic child with ≥ moderate AS and/or ≥moderate AR

Repaired

- Infant
  - TTE within 30 days
  - TTE every 3 months ≤mild MS and or AR
  - TTE every 1 month ≥moderate AS or AR

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- Child
  - TTE every 1 year ≤Mild AS or AR
  - TTE every 6 months ≥moderate AS or AR
  - TTE every 3 months if heart failure

Aortic Valve Stenosis and/or regurgitation/ BAV (Bicuspid Aortic Valve)

Any Surgical Status

- TTE, TEE, Cardiac MR/CT are indicated for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Preoperative

Unrepaired

- Infant <3 months
  - TTE 1 per week
- Infant >3 months
  - TTE every 3 months
  - TTE every 1 month, if ≥moderate AS or AR
- Child
  - TTE every 1 year with mild AS/AR and no aortic dilation
  - TTE every 6 months with moderate AS/AR, or Aortic dilation.
  - TTE every 3 years if BAV with trivial or mild valvar dysfunction and no aortic root dilation
  - Every 6 months in any as with increasing z score aortic root Ascending Aorta

Post Procedural

- Within 30 days TTE
- Infant

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

- Every 1 month following neonatal intervention with ≥moderate AS or AR or LV dysfunction
- Every 3 months ≤mild AS/AR and no LV dysfunction

- Child (TTE)
  - 6 months echo if ≥moderate AS or AR
  - 1-year echo if ≤mild AS or AR, and/or normal prosthetic valve
  - Every 3 months if CHF or Ventricular dysfunction

Supravalvular AS

Any Surgical Status

- TTE, TEE, Cardiac MR/CT, Chest MRA/CTA are indicated for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Preoperative
  - Williams syndrome
    - Individuals with Williams syndrome can be screened/evaluated for arch abnormalities and pulmonary artery abnormalities and coronary artery abnormalities with the same intervals as TTE referenced below.
  - Stress imaging can be done at initial evaluation and for cardiac symptoms, change in clinical status and/or new concerning signs or symptoms

Unrepaired

- Infant
  - TTE every 3 months
- Child
  - TTE every 1 year
  - TTE every 6 months if moderate AS

Post-operative (TTE)

- Within 30 days
- Every 2 years in mild to moderate AS
- Every 6 months if ≥moderate AS

Aortic Coarctation and IAA (Interrupted Aortic Arch) (PEDCD-2.4.11)

CDP.CHD.2411.A
v1.0.2023

All Individuals

- TTE, MRA/CTA Chest are indicated for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Prior to planned surgery/intervention
- Cardiac MR/CT is indicated for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Prior to planned surgery/intervention if any issues remain not answered on echo

Unrepaired Aortic Coarctation

- Newborn, TTE weekly if assessing for ductal closure
- Infant with mild Coarctation in absence of PDA
  - Echo every 3 months
- Child with mild Coarctation
  - Echo every 1 year
  - MRA Chest, CTA Chest every 3 years

Post Procedure: Surgical or Catheter Based

- TTE
  - Within 30 days of procedure
  - Every 3 months if mild or no sequel in first year

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- Every 6 months if mild or no sequel in the second year
- Every 1 year after the second year
- Every 3 months at any time if CHF symptoms or ≥moderate sequelae
- MRA/CTA Chest every 3 years (include Cardiac MR/CT if issues not clarified on echo)

Coronary Anomalies (PEDCD-2.4.12)

CDP.CHD.2412.A
v1.0.2023

- CCTA or cardiac MRI is indicated for evaluating coronary artery anomalies and other complex congenital heart disease of cardiac chambers or great vessels
  - CPT® 75574 for evaluating coronary artery anomalies
  - CPT® 75573 for congenital heart disease
  - CTA Chest (CPT® 71275) can be added to evaluate great vessels
- Congenital anomalies of the coronary arteries are an important cause of sudden death in pediatric individuals. Coronary arteries may arise from the wrong coronary artery cusp leading to ischemic changes during exercise. These lesions may be found incidentally during a murmur evaluation. Anomalous coronary arteries may be seen on echocardiogram during an evaluation for chest pain or syncope or palpitations. In addition, individuals with no echocardiographic findings, but symptoms concerning for angina chest pain may require stress testing.
  - Individuals who have positive echocardiographic findings, regardless of symptoms, and individuals who have classical typical angina chest pain regardless of echocardiographic findings, may require treadmill stress testing, stress imaging, of advanced imaging such as Cardiac MRI, Stress echocardiogram, PET, Cardiac CT, and/or cardiac catheterization.
- Congenital coronary anomalies include abnormal origin of a coronary artery from the PA, anomalous aortic origin of a coronary artery from a different aortic sinus of Valsalva (left coronary artery from the right sinus of Valsalva or right coronary artery from the left sinus of Valsalva), coronary arteriovenous fistula, and coronary artery ostial atresia, all in the setting of normal conotruncal anatomy.
  - Any surgical status
    - Prior to planned surgery, or change in clinical status and/or new concerning signs or symptoms
      - TTE
      - CMR or CCT
        - Can initially include MRA/CTA Chest.
        - If the origin of the coronaries arteries is below the sinus of valsavla then a chest study is not needed on subsequent imaging.
        - If the origin of the coronary artery is not at the level of the sinus of Valsalva, a MRA/CTA Chest can be included when MR/CT imaging is required
      - Stress imaging- to assess the need for surgery

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- Unrepaired
  - Routine surveillance every 2 years in an asymptomatic individual with anomalous right coronary artery from the left aortic sinus
    - TTE
    - Stress imaging
  - Although typically repaired, in the event that a repair is not completed, anomalous left coronary artery from the right coronary sinus can have imaging
    - TTE annually
    - Stress imaging annually
  - Routine surveillance in an asymptomatic individual with small coronary fistula
    - TTE- every 2 years
  - Routine surveillance in an asymptomatic individual with moderate or large coronary fistula
    - TTE annually
- Post-procedural: surgical or catheter
  - TTE
    - Within 30 days of procedure
    - Monthly the first year following repair
    - Every 3 months after first year of surgery
    - Annually after the second year of surgery
    - Every 3 months if ventricular dysfunction
  - Stress testing
    - EKG stress testing without imaging may be indicated in the first post year, and every 1-2 years depending on level of activity. EKG stress testing does not require PA by eviCore Healthcare
    - Stress testing with imaging
      - First postoperative year
      - If EKG stress test positive of equivocal
- Change in clinical status and/or new concerning signs or symptoms
- Individuals with congenital heart disease such as TOF, Truncus Arteriosus, and , TGA have increased incidence of coronary artery anomalies

- Individuals with Williams syndrome can have coronary artery stenosis.
- Individuals with confirmed coronary artery anomalies may require repeat imaging based on the clinical scenario.
- CCTA to rule out anomalous coronary artery should be limited to one of the following:
    - Individuals who need to have an anomalous coronary artery mapped prior to an invasive procedure.
    - Individuals who have not had a previous imaging study that clearly demonstrates an anomalous coronary artery
    - Individuals with a history that includes one or more of the indications in Indications for CCTA (CPT® 75574) (PEDCD-10.3) .

Tetralogy of Fallot (TOF) (PEDCD-2.4.13)

CDP.CHD.2413.A
v1.0.2023

Any Surgical Status

- TTE, CMR/CT-CMRA/CTA
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Evaluation prior to planed pulmonary valve replacement, cardiac intervention, or surgery

Unrepaired

- Newborn-TTE no limits
- Infant
  - 1 per month

Post Procedure Palliation

- 1 per month following palliative procedure prior to complete repair, valvuloplasty, PDA and/or RVOT stenting, or shunt placement before complete repair

Post-operative TOF (Initial Repair)

- TTE
  - Within 30 days of repair
  - Child-12 months
  - Adolescence every 24 months
  - Every 6 months in an individual with valvular dysfunction other than pulmonary valve, RVOT obstruction, branch pulmonary artery stenosis, arrhythmias, or presence of an RV-to-PA conduit
  - TTE every 3 months if CHF

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- Cardiac MR/CT, MRA/CTA Chest every
  - Routine surveillance (36 months) in an individual with PR and preserved ventricular function
  - 12 months if moderate (≥150 mL/m2) or progressive (increase of >25 mL/m2) RV dilatation or dysfunction (RVEF ≤48% or ≥6% decrease in EF) or nearing imaging criteria for PVR.

Post-surgical or Catheter Based Pulmonary Valve Replacement

- TTE
  - Within 30 days follow-up
  - 1 and 6 months after replacement
  - One year post procedure
  - Annually after replacement
  - Every 6 months if RV-to-PA conduit dysfunction, valvular or ventricular dysfunction, branch pulmonary artery stenosis, or arrhythmias
  - Every 3 months if CHF symptoms
- CMR/CT-CMRA/CTA every 2 years

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Double Outlet Right Ventricle (DORV) (PEDCD-2.4.14)

CDP.CHD.2414.A
v1.0.2023

Any Surgical Status

- TTE, CMR/CT-CMRA/CTA are indicated for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Evaluation prior to repair

Unrepaired

- TTE
  - Newborn no limit
  - Monthly Infant with balanced systemic and pulmonary circulation
  - Every 3 months Child with balanced circulation

Postoperative

- TTE
  - Within 30 days
  - First year postop every 6 months
  - After one year, TTE every 1 year
  - TTE 3 months in an individual with valvular or ventricular dysfunction, right or left ventricular outflow tract obstruction, branch pulmonary artery stenosis, arrhythmias, or presence of an RV-to-PA conduit, heart failure.
- Cardiac MR/CT, MRA/CTA Chest
  - 3 years for asymptomatic individual

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

D-Loop Transposition of the Great Arteries (D-Loop TGA) (PEDCD-2.4.15)

CDP.CHD.2415.A
v1.0.2023

Any Surgical Status

- TTE, **CMR/CT-CMRA/CTA**, Stress imaging are indicated for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Any time after procedure involving coronary arteries
- **CMR/CT-CMRA/CTA** every 5 years.

Unrepaired (TTE)

- No Limits

Post Arterial Switch

- TTE
  - Within 30 days of repair
  - Infant every one month
  - Child every 3 months
  - Child with moderate or greater sequelae TTE every three 3 months (moderate Valvular or ventricular dysfunction, right or left ventricular outflow tract obstruction, branch pulmonary artery stenosis, or Arrhythmias.
- Routine CMR/CT
  - Every 3 years
  - Every year if neo AI
- MRA/CTA Chest
  - Every 3 years
  - Every year if neo AI, or aortic dilation

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- Stress imaging
    - 1 routine test after arterial switch at any time

Post Rastelli

- TTE
    - Within 30 days
    - Every three months following procedure for one year
    - Child Every 6 months following the first year after repair if no or mild sequelae
    - Adolescent annually
    - Every three months if moderate valvular dysfunction, LVOT obstruction, presence of an RV-to-PA conduit, branch, pulmonary artery stenosis, or arrhythmias, or heart failure
- CMR/CT-CMRA/CTA every 3 years

Post Atrial Switch

- TTE Every 1 year if mild to no Symptoms
    - Every 3 months TTE, and CMR MRA CCT CTA if ≥moderate systemic AV, valve regurgitation, systemic RV dysfunction, LVOT obstruction, or arrhythmias, or CHF.
    - Routine **CMR/CT-CMRA/CTA** every 3 years.

Congenitally Corrected Transposition of the Great Arteries (ccTGA, LTGA) (PEDCD-2.4.16)

CDP.CHD.2416.A
v1.0.2023

Any Surgical Status

- TTE, TEE, **CMR/CT-CMRA/CTA** are indicated for any of the following:
  - Change clinical status and/or new concerning signs or symptoms
  - Preoperative evaluation (typically within one month)
- CMR/CT-CMRA/CTA every 3 years

Unrepaired

- TTE
  - Newborn-Weekly
  - Infant
    - Every 3 months if no cardiac symptoms and only mild findings
    - Every 1 month is cardiac symptoms and moderate findings
  - Child
    - <2 years every 3 months
    - >2 years every 1 year
    - Every 6 months if ≥moderate AV regurg
    - Every 3 months if CHF symptoms
  - CMR/CT-CMRA/CTA
    - Every 3 years

Postoperative: Anatomic Repair

- TTE

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

  - Post–operative evaluation (within 30 days)
  - Every 3 months within a year following repair in an asymptomatic individual with no or mild sequelae
  - Every 1 year after the first year following repair in an asymptomatic individual with no or mild sequelae
  - Every 6 months if valvular or ventricular dysfunction, right or left ventricular outflow tract obstruction, or presence of a RV-to-PA conduit
  - Every 3 months if CHF symptoms
- CMR/CT-CMRA/CTA
  - Every 3 years

Postoperative: Physiological Repair with VSD Closure And/or LV-to-PA Conduit

- TTE
  - Postoperative evaluation (within 30 days)
  - Every 3 months within a year following repair in an asymptomatic individual with no or mild sequelae
  - Annually in an asymptomatic individual with no or mild sequelae
  - Every 3 months if in an individual with ≥moderate systemic AV valve regurgitation, systemic RV dysfunction, and/or LV-to-PA conduit dysfunction, or with CHF symptoms
- CMR/CT-CMRA/CTA every 3 months in an individual with ≥moderate systemic AV valve regurgitation, systemic RV dysfunction, and/or LV-to-PA conduit dysfunction, or with CHF symptoms

Truncus Arteriosus (PEDCD-2.4.17)

CDP.CHD.2417.A
v1.0.2023

Any Surgical Status

- TTE, CMR/CT-CMRA/CTA are indicated for any of the following:
  - Initial evaluation of change in clinical status and/or new concerning signs or symptoms
  - Prior to planned intervention or surgery

Postoperative

- TTE
  - Within 30 days
  - Monthly in first year after surgery
  - After first year every 6 months
  - Every 3 months if
    - ≥moderate truncal stenosis or regurgitation
    - Residual VSD or RV to PA conduit or Branch PA obstruction
    - Symptoms of CHF
- CMR/CT-CMRA/CTA
  - Annually if ≥moderate Truncal stenosis or regurgitation

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Single Ventricle (SV) (PEDCD-2.4.18)

CDP.CHD.2418.A

v1.0.2023

SV references individuals not amenable to biventricular repair, including but not limited to hypoplastic left heart syndrome, tricuspid atresia, Double inlet left ventricle, mitral atresia, unbalanced AVSD, and forms of PA/IVS

Any Surgical Status

- Any/All: TTE, TEE, **CMR/CT-CMRA/CTA** are indicated for any of the following:
  - Change clinical status and/or new concerning signs or symptoms
  - Preoperative evaluation (typically within one month)

Unrepaired SV

- TTE allowed one study per week

Stage 1 Palliation (TTE)

- Often called Norwood or Sano, or hybrid cath procedure
- Routine weekly TTE

Stage 2 Palliation (TTE)

- Often referred to as Glen procedure
- Within 30 days after surgical or cath intervention
- 1 per month in infant or child

Stage III, Also Called Fontan.

- TTE within 30 days
- TTE every three months within first post op year
- Every 6 months after first year
- Every 3 years allow **CMR/CT-CMRA/CTA**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- TTE every 3 months until the finding is stable or there is a need for intervention if there is valvular dysfunction, arrhythmias, heart failure

References (PEDCD-2)

v1.0.2023

1. Kliegman R, Lye PS, Bordini BJ, et al.Nelson Pediatric Symptom-Based Diagnosis. Philadelphia, PA: Elsevier; 2018.

2. Riveros R and Riveros-Perez E. Perioperative considerations for children with right ventricular dysfunction. Seminars in Cardiothoracic and Vascular Anesthesia. 2015 Jul 10;19(3):187–202.

3. Lai WW, Geva T, Shirali GS, et al. Guidelines and Standards for Performance of a Pediatric Echocardiogram: A Report from the Task Force of the Pediatric Council of the American Society of Echocardiography. Journal of the American Society of Echocardiography. 2006;19(12):1413-1430. doi:10.1016/j.echo.2006.09.001.

4. Prakash A, Powell AJ, Geva T. Multimodality Noninvasive Imaging for Assessment of Congenital Heart Disease.*Circulation: Cardiovascular Imaging*. 2010;3(1):112-125. doi:10.1161/CIRCIMAGING.109.875021.

5. Simpson J, Lopez L, Acar P, et al. Three-dimensional Echocardiography in Congenital Heart Disease: An Expert Consensus Document from the European Association of Cardiovascular Imaging and the American Society of Echocardiography. *Journal of the American Society of Echocardiography*. 2017;30(1):1-27. doi:10.1016/j.echo.2016.08.022

6. Truong UT, Kutty S, Broberg CS, Sahn DJ. Multimodality Imaging in Congenital Heart Disease: an Update. Current Cardiovascular Imaging Reports. 2012;5(6):481-490. doi:10.1007/s12410-012-9160-6.

7. Wernovsky G, Rome JJ, Tabbutt S, et al. Guidelines for the Outpatient Management of Complex Congenital Heart Disease.*Congenital Heart Disease*. 2006;1(1-2):10-26. doi: doi:10.1111/j.1747-0803.2006.00002.x.

8. Valente AM, Cook S, Festa P, et al. Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography. *Journal of the American Society of Echocardiography*. 2014;27(2):111-141. doi:10.1016/j.echo.2013.11.009.

9. Margossian R, Schwartz ML, Prakash A, et al. Comparison of Echocardiographic and Cardiac Magnetic Resonance Imaging Measurements of Functional Single Ventricular Volumes, Mass, and Ejection Fraction (from the Pediatric Heart Network Fontan Cross-Sectional Study) A list of participating institutions and investigators appears in the Appendix. *The American Journal of Cardiology*. 2009;104(3):419-428. doi:10.1016/j.amjcard.2009.03.058.

10. Hundley WG, Bluemke DA, Finn JP, et al. ACCF/ACR/AHA/NASCI/SCMR 2010 Expert Consensus Document on Cardiovascular Magnetic Resonance. *Circulation*. 2010;121(22):2462-2508. doi:10.1161/cir.0b013e3181d44a8f.

11. Silvestry FE, Cohen MS, Armsby LB, et al. Guidelines for the Echocardiographic Assessment of Atrial Septal Defect and Patent Foramen Ovale: From the American Society of Echocardiography and Society for Cardiac Angiography and Interventions. *Journal of the American Society of Echocardiography*. 2015;28(8):910-958. doi:10.1016/j.echo.2015.05.015.

12. Franklin RCG, Béland MJ, Colan SD, et al. Nomenclature for congenital and paediatric cardiac disease: the International Paediatric and Congenital Cardiac Code (IPCCC) and the Eleventh Iteration of the International Classification of Diseases (ICD-11). *Cardiology in the Young*. 2017;27(10):1872-1938. doi:10.1017/s1047951117002244.

13. Cohen MS, Eidem BW, Cetta F, et al. Multimodality Imaging Guidelines of Patients with Transposition of the Great Arteries: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography. *Journal of the American Society of Echocardiography*. 2016;29(7):571-621. doi:10.1016/j.echo.2016.04.002.

14. Canobbio MM, Warnes CA, Aboulhosn J, et al. Management of Pregnancy in Patients with Complex Congenital Heart Disease: A Scientific Statement for Healthcare Professionals From the American Heart Association. *Circulation*. 2017;135(8). doi:10.1161/cir.0000000000000458.

15. Hare GFV, Ackerman MJ, Evangelista J-AK, et al. Eligibility and Disqualification Recommendations for Competitive Athletes with Cardiovascular Abnormalities: Task Force 4: Congenital Heart Disease. *Journal of the American College of Cardiology*. 2015;66(21):2372-2384. doi:10.1016/j.jacc.2015.09.036.

16. Giglia T, Stagg A. Infant Single Ventricle Monitoring Program (ISVMP): Outpatient Interstage Pathway: Stage I Discharge to Second Operation. Clinical Pathways Program. https://www.chop.edu/clinical-pathway/single-ventricle-fetus-or-newborn-clinical-pathway. Published July 2011. Accessed July 26, 2019. Revised January 2018.

17. Sachdeva R, Valente AM, Armstrong AK, et al. ACC/AHA/ASE/HRS/ISACHD/SCAI/SCCT/SCMR/SOPE 2020 Appropriate Use Criteria for Multimodality Imaging During the Follow-Up Care of Patients with Congenital Heart Disease. *Journal of the American College of Cardiology*. 2020;75(6):657-703. doi:10.1016/j.jacc.2019.10.002.

18. MacCarrick G, Black JH 3rd, Bowdin S, et al. Loeys-Dietz syndrome: a primer for diagnosis and management. Genet Med. 2014;16(8):576–587. doi:10.1038/gim.2014.11.

19. Loeys BL, Dietz HC. Loeys-Dietz Syndrome. GeneReviews® [Internet]. https://www.ncbi.nlm.nih.gov/books/NBK1133/. Published March 1, 2018.

20. Caglayan AO, Dundar M. Inherited diseases and syndromes leading to aortic aneurysms and dissections. *European Journal of Cardio-Thoracic Surgery*. 2009;35(6):931-940. doi:10.1016/j.ejcts.2009.01.006. Collins RT. Cardiovascular Disease in Williams Syndrome. *Circulation*. 2013;127(21):2125-2134. doi:10.1161/circulationaha.112.000064.

21. D'hondt S, Damme TV, Malfait F. Vascular phenotypes in nonvascular subtypes of the Ehlers-Danlos syndrome: a systematic review. *Genetics in Medicine*. 2017;20(6):562-573. doi:10.1038/gim.2017.138.

22. Hiratzka LF, Creager MA, Isselbacher EM, et al. Surgery for Aortic Dilatation in Patients with Bicuspid Aortic Valves. *Journal of the American College of Cardiology*. 2016;67(6):724-731. doi:10.1016/j.jacc.2015.11.006.

23. Knadler JJ, Lemaire S, Mckenzie ED, et.al. Thoracic Aortic, Aortic Valve, and Mitral Valve Surgery in Pediatric and Young Adult Patients With Marfan Syndrome: Characteristics and Outcomes. *Seminars in Thoracic and Cardiovascular Surgery*. 2019;31(4):818-825. doi:10.1053/j.semtcvs.2019.06.005.

24. Landis BJ, Ware SM, James J, Shikany AR, Martin LJ, Hinton RB. Clinical Stratification of Pediatric Patients with Idiopathic Thoracic Aortic Aneurysm. *J Pediatr*. 2015;167(1):131-137. doi:10.1016/j.jpeds.2015.02.042.

25. Oner T, Akgun G, Ergin SO, Karadag H, Yucel IK, Celebi A. Risk Factors Associated with Ascending Aortic Aneurysms and Aortic Elasticity Parameters in Children with a Bicuspid Aortic Valve. Pediatric Cardiology. 2019;40(5):980-986. doi:10.1007/s00246-019-02102-6.

26. Maron BJ, Zipes DP, Kovacs RJ. Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Preamble, Principles, and General Considerations. *Journal of the American College of Cardiology*. 2015;66(21):2343-2349. doi:10.1016/j.jacc.2015.09.032.

27. Meester JAN, Verstraeten A, Schepers D, et. al. Differences in manifestations of Marfan syndrome, Ehlers-Danlos syndrome, and Loeys-Dietz syndrome. *Annals of Cardiothoracic Surgery*. 2017;6(6):582-594. doi:10.21037/acs.2017.11.03.

28. Otto CM, Nishimura RA, Bonow RO, et al. 2020 ACC/AHA Guideline for the Management of Patients With Valvular Heart Disease: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation*. 2021;143(5):e72-e227. doi:10.1161/cir.0000000000000923.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

29. Bonow RO, O'Gara PT, Adams DH, et al. 2020 Focused Update of the 2017 ACC Expert Consensus Decision Pathway on the Management of Mitral Regurgitation. *J Am Coll Cardiol.* 2020;75(17):2236-2270. doi:10.1016/j.jacc.2020.02.005.

30. Nishimura RA, O'Gara PT, Bavaria JE, et al. 2019 AATS/ACC/ASE/SCAI/STS Expert Consensus Systems of Care Document: A Proposal to Optimize Care for Patients With Valvular Heart Disease: A Joint Report of the American Association for Thoracic Surgery, American College of Cardiology, American Society of Echocardiography, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. *J Am Soc Echocardiogr*. 2019;32(6):683-707. doi:10.1016/j.echo.2019.02.016.

31. Doherty JU, Kort S, Mehran R, et al. ACC/AATS/AHA/ASE/ASNC/HRS/SCAI/SCCT/SCMR/STS 2019 Appropriate Use Criteria for Multimodality Imaging in the Assessment of Cardiac Structure and Function in Nonvalvular Heart Disease. *J Am Coll Cardiol*. 2019;73(4):488-516. doi:10.1016/j.jacc.2018.10.038.

Condition Specific Imaging

Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDCD-12)

CDP.CS.0012.A
v1.0.2023

MIS-C General Information (PEDCD-12.1)

SARS-CoV-2 (COVID 19) is usually mild in children. Some children develop a severe inflammatory disease that can present in a similar way to Kawasaki disease or toxic shock syndrome. This syndrome has been defined by the US Centers for Disease Control and Prevention as multisystem inflammatory syndrome in children (MIS-C).

These guidelines are intended for use in the outpatient management of cardiac findings of MIS-C. Additional information can be found in **PEDHD-12.7** for the outpatient management of head imaging.

MIS-C Indications for Cardiac Imaging (PEDCD-12.2)

MIS-C Initial Cardiac Imaging (PEDCD-12.2.1)

- When there is concern for MIS-C, as in atypical or incomplete Kawasaki (see **Kawasaki Disease - Acute Phase (PEDCD-6.2)**) echo (TTE) can be approved
- A cardiac MRI can be approved at the time of diagnosis when there are issues that can affect treatment management not answered by other testing
- Cardiac CCTA can be done if there is incomplete visualization of the coronary arteries
- Repeat echocardiograms may be required and approved if either:
    - Treatment decisions will be affected by results (e.g., treating with IVIg)
    - There are new signs or symptoms

MIS-C Repeat Cardiac Imaging (PEDCD-12.2.2)

The following imaging guidelines reference outpatient management of individuals who have been discharged from the hospital after stability for MIS-C has been established.

- An echo (TTE) can be approved at the time of presentation and followed by serial echos (TTE) until stabilization has been achieved for any of the following:

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

  - New cardiac signs, symptoms, or findings
  - Evidence of recurrence of MIS-C
  - Changes in medication

- Serial echos can be approved based on the ordering cardiologist's discretion or the treating medical provider in consultation with a cardiologist when there is documented cardiac dysfunction.
- Individuals who are discharged from the hospital after MIS-C and have stable findings can have an echo (TTE):
  - Within 1 week of discharge
  - 4 weeks post discharge
  - At 6 months post discharge
  - One year post discharge
- Cardiac CCTA can be done if there is incomplete visualization of the coronary arteries
- A routine cardiac MRI can be done once after 3 months in an individual with evidence of cardiac involvement (e.g., symptoms, EKG, labs, or echocardiogram)
- Individuals with changes, or unanswered questions, on echo (TTE) may have a Cardiac MRI based on **CD 5.2** in the cardiac imaging guidelines

Individuals with dilated coronary arteries can have imaging based on the AHA Kawasaki guidelines.

AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
All			All risk levels 4-6 weeks after acute illness		
1	Normal	Normal	one echo 2-12 months after acute illness	none	none
2	Dilation	Dilation	6 months One year	None	None

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
			If dilation remains echo every 2-5 yrs until resolves.		
		Normal	After acute illness: 2-12 months One echocardiogram at one year. No echocardiogram after one year		
3.1	Small	Small	6 months 12 months then yearly	2-3 years	3-5 years
3.2	Small	Normal or dilated	6 months 12 months then yearly	3-5 years	none
4.1	Medium	Medium	3 months 6 months 12 months every 6-12 months after that	1-3 years	2-5 years
4.2	Medium	Small	6 months and 12 months, every 1 year.	2-3 years	3-5 years
4.3	Medium	Normal	every 1-	2-4	none

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
		Or Dilated	2 yrs.	years	
5.1	Large	Large	1 month 3 months 6 months 9 months 12 months then every 3-6 months	6-12 months	at 2-6 months, every 1-5 years
5.2	Large	Medium	every 6-12 months	yearly	2-5 years
5.3	Large	Small	6-12 month	1-2 years	2-5 years
5.4	Large	Normal Or Dilation	1-2 years	2-5 years	none

Symptomatic Individuals

- Echocardiogram can be performed at any time with new or progressing cardiac symptoms
- Stress imaging when there are new or progressing symptoms of ischemia or ventricular dysfunction
- Invasive or coronary imaging Coronary angiography (CT, MRI, invasive) when the above studies are positive, inconclusive, or otherwise lead to a conclusion that intervention is needed

References

1. Information for Healthcare Providers about Multisystem Inflammatory Syndrome in Children (MIS-C). Centers for Disease Control and Prevention. https://www.cdc.gov/mis-c/hcp/. Published February 17, 2021.

2. Ahmed M, Advani S, Moreira A, et al. Multisystem inflammatory syndrome in children: A systematic review. *EClinicalMedicine*. 2020;26:100527. doi:10.1016/j.eclinm.2020.100527.

3. Lee PY, Day-Lewis M, Henderson LA, et al. Distinct clinical and immunological features of SARS–CoV-2–induced multisystem inflammatory syndrome in children. *J Clin Invest*. 2020;130(11):5942-5950. doi:10.1172/jci141113.

4. Choi NH, Fremed M, Starc T, et al. MIS-C and Cardiac Conduction Abnormalities. *Pediatrics*. 2020;146(6). doi:10.1542/peds.2020-009738.

5. Dionne A, Mah DY, Son MB, et al. Atrioventricular Block in Children with Multisystem Inflammatory Syndrome. *Pediatrics*. 2020;146(5). doi:10.1542/peds.2020-009704.

6. McMurray JC, May JW, Cunningham MW, Jones OY. Multisystem Inflammatory Syndrome in Children (MIS-C), a Post-viral Myocarditis and Systemic Vasculitis—A Critical Review of Its Pathogenesis and Treatment. *Front Pediatr*. 2020;8. doi:10.3389/fped.2020.626182.

7. Multisystem Inflammatory Syndrome in Children (MIS-C). Centers for Disease Control and Prevention. https://www.cdc.gov/mis-c/. Published February 25, 2021.

8. Kaushik S, Aydin SI, Derespina KR, et al. Multisystem Inflammatory Syndrome in Children Associated with Severe Acute Respiratory Syndrome Coronavirus 2 Infection (MIS-C): A Multi-institutional Study from New York City. *J Pediatr*. 2020;224:24-29. doi:10.1016/j.jpeds.2020.06.045.

9. Feldstein LR, Rose EB, Horwitz SM, et al. Multisystem Inflammatory Syndrome in U.S. Children and Adolescents. *N Engl J Med*. 2020;383(4):334-346. doi:10.1056/nejmoa2021680.

10. Ebina-Shibuya R, Namkoong H, Shibuya Y, Horita N. Multisystem Inflammatory Syndrome in Children (MIS-C) with COVID-19: Insights from simultaneous familial Kawasaki Disease cases. *Int J Infect Dis*. 2020;97:371-373. doi:10.1016/j.ijid.2020.06.014.

11. Valverde I, Singh Y, Sanchez-de-Toledo J, et al. Acute Cardiovascular Manifestations in 286 Children With Multisystem Inflammatory Syndrome Associated With COVID-19 Infection in Europe. *Circulation*. 2021;143(1):21-32. doi:10.1161/circulationaha.120.050065.

12. Alsaied T, Tremoulet AH, Burns JC, et al. Review of Cardiac Involvement in Multisystem Inflammatory Syndrome in Children.*Circulation*. 2021;143(1):78-88. doi:10.1161/CIRCULATIONAHA.120.049836.

13. Sperotto F, Friedman KG, Son MB, VanderPluym CJ, Newburger JW, Dionne A. Cardiac manifestations in SARS-CoV-2-associated multisystem inflammatory syndrome in children: a comprehensive review and proposed clinical approach.

European Journal of Pediatrics. 2020;180(2):307-322. doi:10.1007/s00431-020-03766-6.

14. Friedman KG, Harrild DM, Newburger JW. Cardiac Dysfunction in Multisystem Inflammatory Syndrome in Children. *Journal of the American College of Cardiology*. 2020;76(17):1962-1964. doi:10.1016/j.jacc.2020.09.002.

15. McCrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals From the American Heart Association. *Circulation*. 2017;135(17). doi:10.1161/cir.0000000000000484

16. Belhadjer Z, Méot M, Bajolle F, et al. Acute Heart Failure in Multisystem Inflammatory Syndrome in Children in the Context of Global SARS-CoV-2 Pandemic. *Circulation*. 2020;142(5):429-436. doi:10.1161/circulationaha.120.048360.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Heart Murmur (PEDCD-3.1)

CDP.0003.1.C
v1.0.2023

- The following echocardiography code combinations should be approved for evaluation of any pathologic murmur or any innocent murmur with associated cardiac signs or symptoms:
  - CPT® 93303, CPT® 93306, CPT® 93320, and CPT® 93325
  - CPT® 93303, CPT® 93306
  - CPT® 93306, CPT® 93320 and CPT® 93325 are included with CPT® 93306 and should not be approved separately.
- Repeat echocardiography is not indicated if the initial echocardiogram was normal and the murmur has not changed in quality.

Background and Supporting Information

- Heart murmurs are extremely common in pediatric individuals. The thinner chest wall in children allows clearer auscultation of blood flowing through the chambers of the heart, which may result in a murmur on physical exam.
- The majority of murmurs are innocent and do not require further evaluation. More than 30% of children may have an innocent murmur detected during physical examination. Innocent murmurs are typically systolic ejection murmurs with a vibratory or musical quality, and generally change in quality when the individual changes position.
- Other types of murmurs can be pathologic and require additional evaluation, usually by a pediatric cardiologist. Echocardiography is indicated, and is performed as part of the office visit. When evaluating an individual with a murmur for the first time, it will not be known whether the individual has congenital heart disease or not. The cardiologist only submits charges for the procedure actually performed.

References

1. Nelson Textbook of Pediatrics, 20th Edition, Robert M. Kliegman, MD, Bonita M.D. Stanton, MD, Joseph St. Geme, MD and Nina F Schor, MD, PhD, p2182 to p2292.
2. Campbell RM, Douglas PS, Eidem BW, Lai WW, Lopez L, Sachdeva R. ACC/AAP/AHA/ASE/HRS/SCAI/SCCT/SCMR/SOPE 2014 Appropriate Use Criteria for Initial Transthoracic Echocardiography in Outpatient Pediatric Cardiology. *Journal of the*

American College of Cardiology. 2014;64(19):2039-2060. doi:10.1016/j.jacc.2014.08.003.

3. Lai WW, Geva T, Shirali GS, et al. Guidelines and Standards for Performance of a Pediatric Echocardiogram: A Report from the Task Force of the Pediatric Council of the American Society of Echocardiography. *Journal of the American Society of Echocardiography*. 2006;19(12):1413-1430. doi:10.1016/j.echo.2006.09.001.

4. Allen HD, Shaddy RE, Penny DJ, Cetta F, Feltes TF. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 9th ed. Philadelphia, PA: Wolters Kluwer; 2016.

5. Prakash A, Powell AJ, Geva T. Multimodality Noninvasive Imaging for Assessment of Congenital Heart Disease. *Circulation: Cardiovascular Imaging*. 2010;3(1):112-125. doi:10.1161/circimaging.109.875021.

6. Danford DA, McNamara DG. Innocent murmurs and heart sounds. In: The Science and Practice of Pediatric Cardiology, 2nd ed, Garson AJ, Bricker JT, Fisher DJ, Neish SR (Eds), Williams & Wilkins, Baltimore 1998. p.2204

7. Duff DF, McNamara GG. History and physical examination of the cardiovascular system. In: The Science and Practice of Pediatric Cardiology, 2nd ed, Garson AJ, Bricker JT, Fisher DJ, Neish SR (Eds), Williams and Wilkins, Baltimore 1998. p.693

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Chest Pain General (PEDCD-4.1)

CDP.0004.1.C
v1.0.2023

- Echocardiography is indicated for pediatric individuals with chest pain and one or more of the following:
  - Exertional chest pain
  - Non-exertional chest pain with abnormal EKG
  - Chest pain with signs or symptoms of pericarditis
  - First-degree relative with sudden unexplained death or cardiomyopathy
  - Recent onset of fever
  - Recent illicit drug use
  - Other signs or symptoms of cardiovascular disease
- Echocardiography is performed as part of the office visit. When evaluating an individual for the first time, it will not be known whether the individual has congenital heart disease or not. The cardiologist only submits charges for the procedure actually performed.
- The following echocardiography code combinations for evaluation of chest pain:
  - CPT® 93303, CPT® 93306, CPT® 93320, and CPT® 93325
  - CPT® 93303, CPT® 93306
  - CPT® 93306
  - CPT® 93320 and CPT® 93325 are included with CPT® 93306 and should not be approved separately.
- Repeat echocardiography is not indicated if the initial echocardiogram is normal unless one of the following conditions is present:
  - Increased severity or change in quality of the chest pain
  - New signs or symptoms of cardiovascular disease other than pain
  - New abnormality on EKG
- Individuals with CP may undergo an exercise stress test without imaging. This does not require eviCore prior authorization
- Cardiac MR or cardiac CT is indicated for chest pain if prior evaluation suggests:
  - Any coronary artery abnormalities
  - Cardiomyopathy
  - Myocarditis

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- Chest MRA or CTA if pulmonary embolism or aortic dissection is suspected
- Stress imaging is indicated if other imaging suggests coronary artery abnormality, or ETT suggests ischemia. EKG is uninterpretable. Any indication in section **Stress Testing with Imaging – Indications (CD 1.4)** in the Cardiac Imaging Guidelines. This can include Stress SPECT, echo or MR.

Background and Supporting Information

Chest pain in pediatric individuals is caused by a cardiac etiology in < 5% of cases, yet causes great anxiety for parents resulting in requests for testing.

References

1. Kliegman R, Nelson WE. *Nelson Textbook of Pediatrics*. 20th ed. Philadelphia, PA: Elsevier; 2016.

2. Friedman KG, Alexander ME. Chest Pain and Syncope in Children: A Practical Approach to the Diagnosis of Cardiac Disease. *The Journal of Pediatrics*. 2013;163(3). doi:10.1016/j.jpeds.2013.05.001.

3. Campbell RM, Douglas PS, Eidem BW, Lai WW, Lopez L, Sachdeva R. ACC/AAP/AHA/ASE/HRS/SCAI/SCCT/SCMR/SOPE 2014 Appropriate Use Criteria for Initial Transthoracic Echocardiography in Outpatient Pediatric Cardiology. *Journal of the American College of Cardiology*. 2014;64(19):2039-2060. doi:10.1016/j.jacc.2014.08.003.

4. Allen HD, Shaddy RE, Penny DJ, Cetta F, Feltes TF. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 9th ed. Philadelphia, PA: Wolters Kluwer; 2016

5. Friedman KG, Kane DA, Rathod RH, et al. Management of pediatric chest pain using a standardized assessment and management plan. *Pediatrics* 2011; 128:239.

6. Friedman KG, Alexander ME. Chest Pain and Syncope in Children: A Practical Approach to the Diagnosis of Cardiac Disease. *The Journal of Pediatrics*. 2013;163(3). doi:10.1016/j.jpeds.2013.05.001.

7. Barbut G, Needleman JP. Pediatric Chest Pain. *Pediatr Rev*. 2020;41(9):469-480. doi:10.1542/pir.2019-0058.

Syncope (PEDCD-5.1)

CDP.0005.1.C
v1.0.2023

- Echocardiography is not indicated for most individuals with isolated syncope.
- Echocardiography is indicated for pediatric individuals with syncope and one or more of the following:
  - Exertional syncope
  - Unexplained post-exertional syncope
  - Abnormal EKG
  - Absence of prodromal symptoms
  - Presence of preceding palpitations within seconds of loss of consciousness
  - Lack of a prolonged upright posture
  - Syncope in response to auditory or emotional
  - First-degree relative with any of the following before age 50:
    - Sudden cardiac arrest or death
    - Pacemaker or implantable defibrillator placement
  - First-degree relative with cardiomyopathy
  - Known congenital heart disease
  - History of Kawasaki disease, or other coronary pathology.
  - Pathologic murmur, irregular rhythm, gallop, or click on physical examination
- Echocardiography is performed as part of the office visit. When evaluating an individual for the first time, it will not be known whether the individual has congenital heart disease or not. The cardiologist only submits charges for the procedure actually performed.
- The following echocardiography code combinations for evaluation of syncope:
  - CPT® 93303, CPT® 93306, CPT® 93320, and CPT® 93325
  - CPT® 93303, CPT® 93306
  - CPT® 93306
    - CPT® 93320 and CPT® 93325 are included with CPT® 93306 and should not be approved separately.
- Repeat echocardiography is not indicated if the initial echocardiogram is normal unless one of the following conditions is present:
  - Increased severity or change in quality of the syncope

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

  - New signs or symptoms of cardiovascular disease other than syncope
  - Family of history of sudden death, cardiomyopathy
  - New abnormality on EKG

- Individuals with CP may undergo an exercise stress test without imaging. This does not require eviCore prior authorization
- Cardiac MR or Cardiac CT is indicated for chest pain if prior evaluation suggests any coronary artery abnormalities, cardiomyopathy, myocarditis
- MRA or CTA Chest if pulmonary embolism or aortic dissection is suspected
- Stress imaging (SPECT, echo or MR) is indicated (any);
  - If other imaging suggests coronary artery abnormality
  - ETT suggests ischemia
  - EKG is uninterpretable
  - Any indication in section **Stress Testing with Imaging – Indications (CD 1.4)** in the Cardiac Imaging Guideline.

Background and Supporting Information

Syncope in pediatric individuals is common, with up to 15% of individuals experiencing at least one episode by age 21. Syncope is caused by neurocardiogenic syndrome (vasovagal syncope) in 75 to 80% of cases, which is a benign and self-limiting condition. Despite this, syncope causes great anxiety for parents resulting in requests for testing.

References

1. Kliegman R, Nelson WE. Nelson Textbook of Pediatrics. 20th ed. Philadelphia, PA: Elsevier; 2016.

2. Friedman KG, Alexander ME. Chest Pain and Syncope in Children: A Practical Approach to the Diagnosis of Cardiac Disease. *The Journal of Pediatrics*. 2013;163(3). doi:10.1016/j.jpeds.2013.05.001.

3. Campbell RM, Douglas PS, Eidem BW, Lai WW, Lopez L, Sachdeva R. ACC/AAP/AHA/ASE/HRS/SCAI/SCCT/SCMR/SOPE 2014 Appropriate Use Criteria for Initial Transthoracic Echocardiography in Outpatient Pediatric Cardiology. *Journal of the American College of Cardiology*. 2014;64(19):2039-2060. doi:10.1016/j.jacc.2014.08.003.

4. Cannon B, Wackel P. Syncope. *Pediatrics in Review*. 2016;37(4):159-168. doi:10.1542/pir.2014-0109.

5. Allen HD, Shaddy RE, Penny DJ, Cetta F, Feltes TF. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 9th ed. Philadelphia, PA: Wolters Kluwer; 2016.

6. Shen W-K, Sheldon RS, Benditt DG, et. al. 2017 ACC/AHA/HRS guideline for the evaluation and management of patients with syncope: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *Circulation*. 2017;136:e60–e122. DOI: 10.1161/CIR.0000000000000499.

7. Winder MM, Marietta J, Kerr LM, et al. Reducing Unnecessary Diagnostic Testing in Pediatric Syncope: A Quality Improvement Initiative. *Pediatr Cardiol*. 2021. doi:10.1007/s00246-021-02567-4.

.

Pediatric Pulmonary Hypertension General (PEDCD-7.1)

CDP.0007.1.C
v1.0.2023

- Pulmonary hypertension in children can be caused by cardiac, pulmonary, or systemic diseases, and idiopathic disease occurs as well.
- Chest x-ray, EKG, and echocardiography (CPT® 93306, or CPT® 93303, with CPT® 93320, and CPT® 93325, see: **Transthoracic Echocardiography (TTE) Coding (PEDCD-8.1)**for echocardiography coding considerations) for initial evaluation if pulmonary hypertension is suspected.
- Repeat echocardiography intervals are variable depending on age of individual, etiology, and severity.
    - After a comprehensive initial evaluation, echocardiograms using PH-specific protocols may be performed every 4 to 6 months.
    - Echocardiography is indicated at any time for new or worsening symptoms or to evaluate a recent change in therapy.
    - Right heart and /or left heart catheterization may be utilized for PAH individuals, including before and after initiation of PAH-targeted therapy, and for individuals with concomitant congenital heart disease
- CT Chest (CPT® 71250) may be indicated in addition to CTA Chest (CPT® 71275) or MRA Chest (CPT® 71555) for initial evaluation of all pediatric individuals with pulmonary hypertension to evaluate for pulmonary vascular or interstitial disease, or other intrathoracic causes.
- Cardiac MRI without and with contrast (CPT® 75561) is indicated for evaluation of inconclusive echocardiogram findings, or for monitoring right ventricular function during follow-up.
- Stress echocardiograms may be indicated (as in the general cardiac imaging guidelines) see **Stress Echocardiography – Indications, other than ruling out CAD (CD-2.7)** in the Cardiac Imaging Guidelines.

References

1. Abman SH, Hansmann G, Archer SL, et al. Pediatric Pulmonary Hypertension. *Circulation*. 2015;132(21):2037-2099. doi:10.1161/cir.0000000000000329.

2. Latus H, Kuehne T, Beerbaum P, et al. Cardiac MR and CT imaging in children with suspected or confirmed pulmonary hypertension/pulmonary hypertensive vascular disease. Expert consensus statement on the diagnosis and treatment of paediatric pulmonary hypertension. The European Paediatric Pulmonary Vascular Disease

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Network, endorsed by ISHLT and DGPK. *Heart*. 2016;102(Suppl 2):ii30-ii35. doi:10.1136/heartjnl-2015-308246.

3. Allen HD, Shaddy RE, Penny DJ, Cetta F, Feltes TF. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 9th ed. Philadelphia, PA: Wolters Kluwer; 2016.

4. Galiè N, Humbert M, Vachiery J-L, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *European Respiratory Journal*. 2015;46(4):903-975. doi:10.1183/13993003.01032-2015.

Evaluation of Pericardial Effusion or Diagnosis of Pericardial Tamponade (PEDCD-9.5)

CDP.00.A
v1.0.2023

- Echocardiogram is the initial imaging study of choice to evaluate pericardial effusions or diagnose pericardial tamponade.
- If a specific clinical question is left unanswered by another recent imaging study **and** the answer to the clinical question will affect the management of the individual's clinical condition, contrast-enhanced cardiac MRI is useful for evaluating:
  - Pericarditis
  - Neoplastic effusion
  - Tamponade
  - Myocardial infiltration
- Cancers that can metastasize to the pericardium or myocardium and can cause a malignant effusion include lung, breast, renal cell, lymphoma and melanoma.

References

1. Nelson Textbook of Pediatrics, 20th Edition, Robert M. Kliegman, MD, Bonita M.D. Stanton, MD, Joseph St. Geme, MD and Nina F Schor, MD, PhD, p2182 to p2292.

2. Atweh LA, Orth RC, Guillerman RP, Zhang W, Kan JH. MR imaging of children and young adults with classic findings of osteonecrosis on unenhanced MR images: do contrast-enhanced sequences help? *Pediatric Radiology*. 2013;43(11):1502-1506. doi:10.1007/s00247-013-2714-1.

3. Cohen MS, Eidem BW, Cetta F, et al. Multimodality Imaging Guidelines of Patients with Transposition of the Great Arteries: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography. *Journal of the American Society of Echocardiography*. 2016;29(7):571-621. doi:10.1016/j.echo.2016.04.002.

4. Adler Y, Charron P, Imazio M, et al. 2015 ESC Guidelines for the diagnosis and management of pericardial diseases. *European Heart Journal*. 2015;36(42):2921-2964. doi:10.1093/eurheartj/ehv318.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

5. Allen HD, Shaddy RE, Penny DJ, et. al TF. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 9th ed. Philadelphia, PA: Wolters Kluwer; 2016.

6. Hundley WG, Bluemke DA, Finn JP, et al. ACCF/ACR/AHA/NASCI/SCMR 2010 Expert Consensus Document on Cardiovascular Magnetic Resonance. *Circulation*. 2010;121(22):2462-2508. doi:10.1161/cir.0b013e3181d44a8f.

7. Cohen MS, Eidem BW, Cetta F, et al. Multimodality Imaging Guidelines of Patients with Transposition of the Great Arteries: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography. *Journal of the American Society of Echocardiography*. 2016;29(7):571-621. doi:10.1016/j.echo.2016.04.002.

8. Valente AM, Cook S, Festa P, et al. Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography. *Journal of the American Society of Echocardiography*. 2014;27(2):111-141. doi:10.1016/j.echo.2013.11.009.

9. Feingold B, Mahle WT, Auerbach S, et al. Management of Cardiac Involvement Associated With Neuromuscular Diseases: A Scientific Statement From the American Heart Association. Circulation. 2017;136(13). doi:10.1161/cir.0000000000000526.

10. Mah ML, Cripe L, Slawinski MK, et al. Duchenne and Becker muscular dystrophy carriers: Evidence of cardiomyopathy by exercise and cardiac MRI testing. International Journal of Cardiology. 2020;316:257-265. doi:10.1016/j.ijcard.2020.05.052.

11. Power LC, O'Grady GL, Hornung TS, Jefferies C, Gusso S, Hofman PL. Imaging the heart to detect cardiomyopathy in Duchenne muscular dystrophy: A review. Neuromuscular Disorders. 2018;28(9):717-730. doi:10.1016/j.nmd.2018.05.011.

12. Hor KN, Mah ML, Johnston P, Cripe TP, Cripe LH. Advances in the diagnosis and management of cardiomyopathy in Duchenne muscular dystrophy. Neuromuscular Disorders. 2018;28(9):711-716. doi:10.1016/j.nmd.2018.06.014.

13. Buddhe S, Cripe L, Friedland-Little J, et al. Cardiac Management of the Patient With Duchenne Muscular Dystrophy. Pediatrics. 2018;142(Supplement 2). doi:10.1542/peds.2018-0333i.

14. Birnkrant DJ, Bushby K, Bann CM, et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management. The Lancet Neurology. 2018;17(4):347-361. doi:10.1016/s1474-4422(18)30025-5.

Kawasaki Disease (PEDCD-6)

CDP.CS.0006.C
v1.0.2023

Kawasaki Disease Initial Imaging (PEDCD-6.1)

- Echocardiography (CPT® 93306) is indicated for initial assessment for suspected or known Kawasaki disease
  - Coronary CTA (CPT® 75574), Cardiac MRI without contrast (CPT® 75557), Cardiac MRI without and with contrast (CPT® 75561), or MRA Chest (CPT® 71555) are indicated for evaluation of inconclusive echocardiogram findings, or significant coronary artery abnormalities.
  - Screening of other body areas for aneurysms is not routinely indicated in Kawasaki disease, but MRA or CTA (contrast as requested) of the affected body area can be approved for evaluation of signs or symptoms suggesting aneurysm development.
  - See acute and chronic phase for imaging

Background and Supporting Information

- Kawasaki disease (KD) is the leading cause of acquired pediatric cardiac disease in the developed world. It is an acute febrile illness characterized by a medium vessel vasculitis, which predominantly affects the coronary arteries.
  - Individuals who do not fulfill the diagnostic criteria for classic KD may be considered to have incomplete (atypical) KD.
  - If Kawasaki disease is strongly suspected, treatment will often begin even before cardiac evaluation, since early treatment is associated with a lower risk for coronary aneurysm development.

Kawasaki Disease - Acute Phase (PEDCD-6.2)

- Echocardiography should be performed when the diagnosis of KD is considered
  - Uncomplicated individuals, echocardiography can be repeated after treatment **both:**
    - Within 1 to 2 weeks

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

        - Within 4 to 6 weeks
    - For individuals with important and evolving coronary artery abnormalities (Z score >2.5) detected during the acute illness, more frequent echocardiography (at least twice per week) should be performed until luminal dimensions have stopped progressing to determine the risk for and presence of thrombosis.
    - Expanding large or giant aneurysms:
        - Twice per week while dimensions are expanding rapidly
        - Once weekly after dimension is stabilized for the first 45 days of illness
        - Then monthly until the third month after illness onset
- It is reasonable to obtain advanced imaging studies such as computed tomographic angiography (CTA), cardiac magnetic resonance imaging (CMRI), or invasive angiography on individuals' severe proximal coronary artery abnormalities in the acute phase when results will impact management decisions.
- Transesophageal echocardiography, invasive angiography, CMRI, and CTA can be of value in the assessment of selected individuals but are not routinely indicated for diagnosis and management of the acute illness.
    - Invasive angiography is rarely performed during the acute illness.
    - Transesophageal echocardiography, CTA, and CMRI can be useful for the evaluation of older children and adolescents when both:
        - Visualization of the coronary arteries with Transthoracic echocardiography (TTE) is inadequate and
        - Results will impact immediate management decisions.
- Evaluation of potential aneurysmal involvement in other arterial beds can be assessed with CMRI, CTA, and, rarely, invasive angiography after recovery from the acute illness for individuals with severe coronary artery involvement or symptoms or signs, such as the presence of a pulsatile axillary mass.
- Atypical or incomplete Kawasaki. Echo is indicated when atypical KD is being considered, may require repeat echocardiograms if treatment decisions will be affected by results (e.g., treating with IVIg), if new signs or symptoms (such as typical peeling) develop.

Background and Supporting Information

- The acute phase of Kawasaki disease (KD) can last up to 4-6 weeks from the onset of fever until acute systemic inflammation has resolved and coronary artery dimensions are no longer expanding

- Based on AHA recommendations, the following classifications are used in risk stratification of coronary artery abnormalities
    - Z-Score classification accounts for the effects of body size and age through use of baseline coronary dimensions adjusted for body surface area. The Z score value represents the number of standard deviation above the mean. (e.g., z=0 pt. has coronary artery dimension value equal to mean, z=2 person has value 2 standard deviation above the mean, based on age, gender, BSA).
    - Coronary Artery Abnormalities Risk Classification based on Z-Score:
        - 1 - No involvement at any time point (Z score always <2)
        - 2 - Dilation only (Z score 2 to <2.5)
        - 3 - Small aneurysm (Z score ≥2.5 to <5)
            - 3.1 - Current or persistent
            - 3.2 - Decreased to dilation only or normal luminal dimension
        - 4 - Medium aneurysm (Z score ≥5 to <10, and absolute dimension <8 mm)
            - 4.1 - Current or persistent
            - 4.2 - Decreased to small aneurysm
            - 4.3 - Decreased to dilation only or normal luminal dimension
        - 5 - Large and giant aneurysm (Z score ≥10, or absolute dimension ≥8 mm)
            - 5.1 - Current or persistent
            - 5.2 - Decreased to medium aneurysm
            - 5.3 - Decreased to small aneurysm
            - 5.4 - Decreased to dilation only or normal luminal dimension[4]
    - Additional Clinical Features That May Increase the Long-Term Risk of Myocardial Ischemia
        - Greater length and distal location of aneurysms that increase the risk of flow stasis
        - Greater total number of aneurysms
        - Greater number of branches affected
        - Presence of luminal irregularities
        - Abnormal characterization of the vessel walls (calcification, luminal myofibroblastic proliferation)
        - Presence of functional abnormalities (impaired vasodilation, impaired flow reserve)

- Absence or poor quality of collateral vessels
- Previous revascularization performed
- Previous coronary artery thrombosis
- Previous myocardial infarction
- Presence of ventricular dysfunction

**Adapted from: Mccrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals from the American Heart Association. Circulation. 2017;135(17). doi:10.1161/cir.0000000000000484.

Kawasaki Disease - Chronic Phase (PEDCD-6.3)

- Long-term management begins at the end of the acute illness, usually at 4 to 6 weeks after fever onset. Management is based on two pieces of data:
  - The dimensions of the largest Aneurysm at any point during the disease
  - The dimensions of the largest current aneurysm
- Additional risk factors that may be considered for imaging
  - Greater length and distal location of aneurysms that increase the risk of flow stasis
  - Greater total number of aneurysms
  - Greater number of branches affected
  - Presence of luminal irregularities
  - Abnormal characterization of the vessel wall (calcification, luminal myofibroblastic proliferation)
  - Presence of functional abnormalities (impaired vasodilation, impaired flow reserve)
  - Absence or poor quality of collateral vessels
  - Previous revascularization performed
  - Previous coronary artery thrombosis
  - Previous myocardial infarction
  - Presence of ventricular dysfunction
  - Long term routine surveillance in asymptomatic imaging for Kawasaki disease- see chart
- Long term routine surveillance in asymptomatic imaging for Kawasaki disease

AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
All			All risk levels 4-6 weeks after acute illness		
1	Normal	Normal	one echo 2-12 months after acute illness	none	none
2	Dilation	Dilation	6 months One year If dilation remains echo every 2-5 yrs until resolves.	None	None
		Normal	After acute illness: 2-12 months One echocardiogram at one year. No echocardiogram after one year		
3.1	Small	Small	6 months 12 months then yearly	2-3 years	3-5 years
3.2	Small	Normal or dilated	6 months 12 months	3-5 years	none

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

AHA risk level	Largest Aneurysm At Any Point	Largest Current Aneurysm	Routine Echo	Routine Stress Imaging	Routine Coronary Imaging
			then yearly		
4.1	Medium	Medium	3 months 6 months 12 months every 6-12 months after that	1-3 years	2-5 years
4.2	Medium	Small	6 months and 12 months, every 1 year.	2-3 years	3-5 years
4.3	Medium	Normal Or Dilated	every 1-2 yrs.	2-4 years	none
5.1	Large	Large	1 month 3 months 6 months 9 months 12 months then every 3-6 months	6-12 months	at 2-6 months, every 1-5 years
5.2	Large	Medium	every 6-12 months	yearly	2-5 years
5.3	Large	Small	6-12 month	1-2 years	2-5 years
5.4	Large	Normal Or Dilation	1-2 years	2-5 years	none

- Symptomatic individuals
  - Echocardiogram can be performed at any time with new or progressing cardiac symptoms

- Stress imaging when there are new or progressing symptoms of ischemia or ventricular dysfunction
- Invasive or coronary imaging Coronary angiography (CT, MRI, invasive) when the above studies are positive, inconclusive, or otherwise lead to a conclusion that intervention is needed

***Adapted from: Mccrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals from the American Heart Association. Circulation. 2017;135(17). doi:10.1161/cir.0000000000000484*

References

1. Kliegman R, Nelson WE. *Nelson Textbook of Pediatrics*. 20th ed. Philadelphia, PA: Elsevier; 2016.

2. Son MBF, Newburger JW. Kawasaki Disease. *Pediatrics in Review*. 2013;34(4):151-162. doi:10.1542/pir.34-4-151.

3. Campbell RM, Douglas PS, Eidem BW, Lai WW, Lopez L, Sachdeva R. ACC/AAP/AHA/ASE/HRS/SCAI/SCCT/SCMR/SOPE 2014 Appropriate Use Criteria for Initial Transthoracic Echocardiography in Outpatient Pediatric Cardiology. *Journal of the American College of Cardiology*. 2014; 64(19):2039-2060. doi:10.1016/j.jacc.2014.08.003.

4. Mccrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals from the American Heart Association. *Circulation*. 2017; 135(17). doi:10.1161/cir.0000000000000484.

5. Mccrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals from the American Heart Association. *Circulation*. 2017;135(17). doi:10.1161/cir.0000000000000484.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Echocardiography

Transthoracic Echocardiography (TTE) Coding (PEDCD-8.1)

CDP.000.8.2.A
v1.0.2023

- CPT® codes for echocardiography are listed in General Guidelines (PEDCD-1)

Echocardiogram coding Notes	CPT ®
• The most commonly performed study is a complete transthoracic echocardiogram with spectral and color flow Doppler (CPT® 93306). • CPT® 93306 includes CPT® 93320 and CPT® 93325, so those codes should not be approved along with CPT® 93306.	93306
• Doppler codes (CPT® 93320, CPT® 93321, and CPT® 93325) are add-on codes and are assigned in addition to code for the primary procedure, and should not be approved alone.	+93320 +93321 +93325
• For a 2D transthoracic echocardiogram without Doppler, report CPT® 93307.	93307

• A limited transthoracic echocardiogram is reported with CPT® 93308.
  • Limited transthoracic echocardiogram should be billed if the report does not “evaluate or document the attempt to evaluate” all of the required structures.
  • Unlike CPT® 93306, the Doppler CPT® 93321 and CPT® 93325 are not included with CPT® 93308.
  • CPT® 93321 (not CPT® 93320) should be reported with CPT® 93308 if Doppler is included in the study.
  • CPT® 93325 should also be
	93308

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

reported with CPT® 93308 if color flow Doppler is included in the study.	
• For individuals with known congenital heart disease, a limited transthoracic echocardiogram is reported with CPT® 93304, +/- CPT® 93321 and CPT® 93325.	93304

- Providers performing an **initial** echo on a pediatric individual will not know what procedure codes they will be reporting until the initial study is completed.
  - If congenital heart disease is found on the initial echo, a complete echo is reported with codes CPT® 93303, CPT® 93320, and CPT® 93325 because CPT® 93303 does NOT include Doppler and color flow mapping.
  - If no congenital issue is discovered, then CPT® 93306 is reported alone and includes 2-D, Doppler and color flow mapping.
- Since providers may not know the appropriate code/s that will be reported at the time of the pre-authorization request, they may request multiple codes.
  - The following echocardiography code combinations for any **initial** echocardiogram:
    - CPT® 93303, CPT® 93306, CPT® 93320, and CPT® 93325
    - CPT® 93303, CPT® 93306
    - CPT® 93306
      - CPT® 93320 and CPT® 93325 are included with CPT® 93306 and should not be approved separately.
  - Post-service audits may be completed to ensure proper claims submission.
- Correct coding for subsequent echocardiograms
  - If an individual is being followed for known congenital heart disease, and an echocardiogram is indicated, the appropriate codes are (CPT® 93303 or 93304) in addition to appropriate doppler codes(CPT® 93320 or 93321) and CPT® 93325
  - If an individual has documented normal anatomy, or acquired heart disease, and an echocardiogram is indicated, non-congenital codes are appropriate CPT® 93306 (includes all Doppler codes) or CPT® 93308 with CPT® 93321 and CPT® 93325
  - For individuals with newborn physiology (e.g., ASD versus PFO, or PDA) the final echocardiogram that documents normal anatomy can be coded as

congenital. However, any subsequent echocardiograms after that, which would be completed for a new indication, (e.g., shortness of breath) would be coded as non-congenital

Initial Transthoracic Echocardiography (TTE) Indications (PEDCD-8.2)

CDP.000.8.2.A
v1.0.2023

- In addition to indications listed in previous guideline sections, initial TTE evaluation is indicated for any of the following:
  - Any signs/symptoms that are possibly cardiac in nature, including (but not limited to) central cyanosis, dyspnea, edema, poor peripheral pulses, feeding difficulty, decreased urine output, hepatomegaly, or desaturation on pulse oximetry.
  - Abnormal EKG or cardiac biomarkers
  - Abnormal chest x-ray suggesting cardiovascular disease
  - First-degree relative with any of the following before age 50:
    - Sudden cardiac arrest or death
    - Pacemaker or implantable defibrillator placement
    - First-degree relative with cardiomyopathy
  - Supraventricular Tachycardia (SVT), Ventricular Tachycardia, or Premature Ventricular Contractions (PVCs)
  - Known or suspected valvular dysfunction
  - Persistent systemic hypertension
  - Individuals with new onset hypertension
    - TTE indicated to assess for cardiac target organ damage (LV mass, geometry, and function) at the time of consideration of pharmacologic treatment of systemic hypertension
  - Obesity (BMI >30) with additional cardiovascular risk factors
  - Stroke
  - Renal failure
  - Preoperative evaluation of individuals with chest wall deformities or scoliosis
  - Known or suspected vascular ring
  - Planned administration of cardiotoxic chemotherapy
    - Generally anthracyclines (doxorubicin, daunorubicin, mitoxantrone, idarubicin, epirubicin)
  - Planned radiation therapy involving heart muscle or hematopoietic stem cell transplant

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- Sickle cell disease or other hemoglobinopathy causing chronic anemia
- Known or suspected vasculitis, acute rheumatic fever, or other systemic autoimmune disease
- Aortopathy (such as Marfan, Ehlers-Danlos, Loeys-Dietz)
  - Positive personal diagnosis
  - First degree relative
  - Positive gene
  - Finding suggestive of aortopathy such as xray showing aortic dilation
- Muscular dystrophy
  - Positive personal diagnosis
  - First degree relative
  - Positive gene
  - Any findings suggestive of MD, such as neurological exam
- Cardiomyopathy
  - Diagnosed by other modality (such as cardiac MR)
  - First degree relative
  - Positive genetic testing
  - Findings suggestive of, such as cardiomegaly on x-ray
- Metabolic, mitochondrial, and storage disorders
  - Positive personal diagnosis
  - First degree relative
  - Positive genetic testing
  - Findings suggestive of on exam or lab findings
- Abnormalities of cardiac or other viscera situs
- Signs, symptoms, or blood culture suggestive of endocarditis
- Known or suspected mass lesion involving the heart or great vessels
- Known or suspected clot in atrium or ventricle
- Known or suspected pulmonary hypertension
- Known or suspected pericardial effusion
- Complications during prenatal development:
  - Known or suspected cardiovascular abnormality on fetal echocardiogram

- Maternal phenylketonuria (PKU)
- Maternal diabetes with no fetal echo
- Maternal teratogen exposure
- Maternal infection during pregnancy with potential cardiac sequelae

- Genetic abnormality known to be associated with cardiovascular disease such as
  - Down syndrome
  - Turner syndrome
  - 22q11 deletion syndrome
  - Williams syndrome
  - Noonan syndrome
- First-degree relative family history of:
  - Unexplained sudden death before age 50
  - Hypertrophic cardiomyopathy
  - Non-ischemic dilated cardiomyopathy
  - Genetic abnormality known to be associated with cardiovascular disease
  - Congenital left-sided heart lesion
  - Heritable pulmonary arterial hypertension

Repeat Transthoracic Echocardiography Indications (PEDCD-8.3)

CDP.EC.0008.3.A
v1.0.2023

- Repeat echocardiograms may be required for individuals with no new symptoms.
- In addition to indications listed in previous guideline sections, repeat TTE evaluation is indicated for any of the following:
    - In an individual with known cardiac disease and a previously normal echocardiogram when there is documentation of any of the following:
        - New or worsening cardiac symptoms
        - New EKG abnormality
        - Newly recognized family history suggestive of heritable heart disease
    - In an individual with prior normal evaluation
        - New or worsening symptoms
        - New EKG finding
        - New murmur
        - New finding of inheritable disease in first degree relative
    - Individuals with first-degree family history of cardiomyopathy (such as, hypertrophic, dilated, arrhythmogenic) or aortopathy.
        - Repeat echo every 12 months
        - Repeat echo can be done at the additional intervals when the family history or gene mutation is associated with neonatal or fetal disease:
            - At birth
            - Within the first 6 weeks
            - At 3 months
            - At 6 months
            - At one year
            - Then yearly
        - Repeat imaging is **not** indicated in individuals with first degree relative with known mutation when **both** of the following apply:
            - Individual has been tested and does not have that mutation

- Individual has a normal echocardiogram

- If there are abnormal findings on screening/surveillance imaging, a repeat echo is allowed to assess stability of findings

- Individual with a phenotype positive cardiomyopathy (with or without a positive gene) can be imaged as follows:
  - Infants (under one year)
    - TTE is indicated at frequency requested by pediatric cardiology or provider in consultation with pediatric cardiology
  - Children (over one year) yearly testing is indicated as follows:
    - Repeat TTE every 12 months
    - Repeat TTE to assess stability at discretion of pediatric cardiology, or provider in consultation with pediatric cardiology, after any new or changed clinical finding.
    - TTE at any time with documented new or changing symptoms.
- Individual with a known mutation associated with cardiomyopathy or aortopathy and no previous abnormal imaging
  - Repeat echo every 12 months
    - Individuals whose gene mutation is associated with neonatal or fetal disease or there is a family history of neonatal or fetal disease can have repeat echo at the following intervals:
      - At birth
      - Within the first 6 weeks
      - Then at 3 months
      - At 6 months
      - At one year
      - Then yearly
  - If there are abnormal findings on screening/surveillance imaging, a repeat echo is allowed to assess stability of findings.
- Individuals who are status post heart transplant can have echocardiograms repeated as often as requested by heart transplant team.
- Every 12 months for individuals receiving active therapy for ventricular hypertrophy, valvular dysfunction, cardiomyopathy.
  - One time repeat TTE can be approved at 6 months to assess response to a change in therapy.

- Every 12 months for individuals with chronic pericardial effusions
- Every 12 months routine surveillance in asymptomatic individuals with muscular dystrophy (may be replaced by cardiac MRI CPT® 75557 or 75561 at 6 years of life)
- Every 12 months for sickle cell disease or other hemoglobinopathy causing chronic anemia and one of the following:
  - High risk genotype (Hgb SS or Sß^0, severe thalassemia, etc.)
  - History of acute chest syndrome or intrinsic lung disease
  - History of stroke
  - Receiving chronic transfusion therapy
- As needed for monitoring cardiotoxicity during chemotherapy administration
- After completion of chemotherapy and/or radiation therapy. See **Cardiotoxicity and Echocardiography (PEDONC-19.2)** for imaging guidelines.
- Aortopathies See **Thoracic Aortic Disease (PEDPVD-4.1)** in the Pediatric Peripheral Vascular Disease Imaging Guidelines
- TTE follow-up for systemic hypertension
  - Individuals with evidence of end organ damage (Includes LVH, or decreased EF) can have echo every 6 months until echocardiogram normalizes.
  - Individuals without LV target organ injury (no LVH, normal EF) at initial echocardiographic assessment, repeat echocardiography at yearly intervals may be considered in those with persistent hypertension. (stage 2 HTN, or chronic stage 1 HTN incompletely treated (noncompliance or drug resistance)

Transesophageal Echocardiography (TEE) (PEDCD-8.4)

CDP.EC.0008.4.A
v1.0.2023

- Transesophageal echocardiography imaging indications in pediatric individuals are identical to those for adult individuals. See **Transesophageal Echocardiography (TEE) – Indications (CD-2.5)** in the Cardiac Imaging Guidelines.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Fetal Echo (PEDCD-8.5)

CDP.EC.0008.5.A
v1.0.2023

Fetal Echocardiography - Coding

- Supported fetal echocardiography (echo) codes include:
  - Initial Fetal Echo, CPT® 76825 and Doppler Echo CPT® 76827 are performed only once per fetus/per facility (i.e. Maternal Fetal Medicine versus Pediatric Cardiology request)
  - Follow-up-Fetal echo and/or Follow-up Doppler echo (CPT® 76826/CPT® 76828)
  - CPT® 93325 for Doppler color flow velocity mapping
- An initial fetal echo is usually not performed prior to 16 weeks.
- Doppler echo procedure codes (CPT® 76827 or CPT® 76828) include the evaluation of veins, arteries, and valves. Guidelines do not support the billing of additional codes (CPT® 76820 and/or CPT® 76821)

Background and Supporting Information

- The minimal use of color Doppler (CPT® 93325) alone, when performed for anatomical structure identification during a standard ultrasound procedure, is not separately reimbursable

Fetal Echocardiography - Indications for Fetal Conditions

Initial Fetal echocardiography (CPT® 76825) and/or Doppler echocardiography (CPT® 76827) with or without Doppler color flow velocity mapping (CPT® 93325) can be performed if ≥16 weeks, for the indications listed below (See **Fetal Echocardiography – Coding (OB-12.1)**):

Fetal Echocardiography - Indications for Fetal Conditions

• Known or suspected abnormal fetal cardiac evaluation on fetal anatomic scan.
  • Known or suspected abnormality must be documented as hard copy or acknowledged verbally by provider of known or suspected fetal cardiac evaluation
  • Suboptimal cardiac evaluation alone is not an indication for fetal echogram. If the 4-chamber view is adequate and there is no other suspicion of a cardiac

Fetal Echocardiography - Indications for Fetal Conditions
abnormality, a fetal echocardiogram is not considered medically necessary. A follow up ultrasound (CPT® 76815 or CPT® 76816) is indicated for suboptimal visualization. If the follow-up ultrasound fails to show a 4-chamber view or there is suspicion of a cardiac abnormality, fetal echocardiogram is indicated.
• Fetal cardiac arrhythmia; persistent fetal tachycardia or bradycardia
• Major fetal extra-cardiac anomaly, (excluding soft markers for aneuploidy: for example shortened long bones, pyelectasis, echogenic bowel, hypoplastic nasal bone, cardiac echogenic foci and choroid plexus cyst) See **High Risk Group Two – Ultrasound Findings (OB-9.2)**
• Congenital heart disease (CHD) in a 1st degree relative of the fetus (i.e. CHD in the mother, father, or sibling of the fetus)
• Known fetal chromosomal abnormalities (fetal aneuploidy) or ultrasound findings of a suspected chromosomal abnormality (excluding soft markers as only ultrasound findings)   • Early onset FGR (<32 weeks) may be a sign of fetal aneuploidy[11,12]
• Single umbilical artery • Chorioangioma or Umbilical cord varix (if suspicion of fetal hydrops) • Fetal intra-abdominal venous anomaly (persistent right umbilical vein)
• Fetal effusion (pericardial, pleural effusion, ascites, etc.) • Fetal hydrops, See **Alloimmunization/Rh Isoimmunization/Other Causes of Fetal Anemia/Parvo/Hydrops (OB-16)**
• Monochorionic twins/TTTS
• Abnormal Fetal Nuchal Translucency scan (NT ≥3.0mm or above the 95th percentile for the CRL) during current pregnancy. • Abnormal ductus venosus waveform[5]
• Fetal echocardiography may be indicated with severe or unexplained polyhydramnios, or if there are also other suspicious findings on an anatomy scan

Fetal Echocardiography - Indications for Maternal Conditions

Initial Fetal echocardiography (CPT® 76825) and/or Doppler echocardiography (CPT® 76827) with or without Doppler color flow velocity mapping (CPT® 93325) can be performed if ≥16 weeks, for the indications listed below (See **Fetal Echocardiography – Coding (OB-12.1)**):

Maternal Conditions:

• Maternal pre-gestational DM or early diagnosed GDM (1st or early 2nd trimester)
• Maternal gestational diabetes mellitus on medication, if HbA1C >6% [in the third trimester (≥32 weeks)]
• Maternal connective tissue disease (SLE, RA, Sjogrens) with Anti-Ro/SSA or anti-La/SSB antibodies present
  • Weekly follow-up Fetal echocardiography (CPT® 76826) and/or Doppler fetal echocardiography (CPT® 76828) or CPT® 76815 from the 18th through the 26th week of pregnancy and then every other week until 30 weeks
• Phenylketonuria
• Infections associated with cardiac anomalies (such as parvovirus, Rubella, Coxsackie virus)
• Genetic conditions associated with CHD in a first degree relative of the fetus (e.g. Marfan syndrome, 22q11.2 deletion syndrome (DiGeorge Syndrome) or Noonan syndrome)
• Prior child with CHD born to mother and/or father of the fetus[5]
• Pregnancy conceived by assisted reproductive technology:[1]
  • In Vitro Fertilization (IVF)
  • Intracytoplasmic sperm injection (ICSI)[1]

Background and Supporting Information

If diabetes is diagnosed prior to pregnancy or in the first or early second trimester (typically before 20 weeks gestation) with standard diagnostic criteria of: HbA1C ≥6.5%, fasting plasma glucose ≥126 mg/dL, or 2-hour glucose ≥200 mg/dL on a 75-g oral glucose tolerance test, then image as above

For those with GDM on medication, if HbA1c levels are >6%, fetal echocardiogram in the third trimester to assess for ventricular hypertrophy can be performed.

In cases of extreme obesity (BMI≥40-50) where the 4-chamber view is inadequately documented after 2 separate ultrasound visits with MFM, fetal echo can be performed.

With positive SSA/SSB antibodies, the most vulnerable period for the fetus is during the period from 18 to 24 weeks gestation. Normal sinus rhythm can progress to complete block in seven days during this high-risk period. New onset of heart block is less likely during the 26th through the 30th week, and it rarely develops after 30 weeks of pregnancy.

Fetal Echocardiography - Indications for Medication or Drug Exposure

Initial Fetal echocardiography (CPT® 76825) and/or Doppler echocardiography (CPT® 76827) with or without Doppler color flow velocity mapping (CPT® 93325) can be performed if ≥16 weeks, for the indications listed below (See **Fetal Echocardiography – Coding (OB-12.1)**):

- Ace inhibitors
- Alcohol (excessive quantities)
- Anti-seizure medication, e.g. carbamazepine, hydantoin, valproate
- Folate antagonists (methotrexate)
- Lithium
- NSAIDS (Ibuprofen, Indomethacin) 2nd and 3rd trimester
- Paroxetine (Paxil)
- Retinoids (e.g Isotretinoin, Retinoic acid, Vitamin A -over 10,000 IU per day, etc.)
- Thalidomide
- Venlafaxine (Effexor)
- This may not be an all-inclusive list, however, exposure to other potential teratogens associated with cardiac anomalies in the fetus are typically adequately assessed with a detailed fetal anatomy ultrasound. (CPT® 76811) (See **Potentially Teratogenic Medications/Substances (OB 10.1)**)

References (PEDCD-8)

v1.0.2023

1. Campbell RM, Douglas PS, Eidem BW, Lai WW, Lopez L, Sachdeva R. ACC/AAP/AHA/ASE/HRS/SCAI/SCCT/SCMR/SOPE 2014 Appropriate Use Criteria for Initial Transthoracic Echocardiography in Outpatient Pediatric Cardiology. *J Am Coll Cardiol*. 2014;64(19):2039-2060. doi:10.1016/j.jacc.2014.08.003.

2. Ambrusko SJ, Gunawardena S, Sakara A, et al. Elevation of tricuspid regurgitant jet velocity, a marker for pulmonary hypertension in children with sickle cell disease. *Pediatric Blood & Cancer*. 2006;47(7):907-913. doi:10.1002/pbc.20791.

3. Klings ES, Machado RF, Barst RJ, et al. An Official American Thoracic Society Clinical Practice Guideline: Diagnosis, Risk Stratification, and Management of Pulmonary Hypertension of Sickle Cell Disease. *American Journal of Respiratory and Critical Care Medicine*. 2014;189(6):727-740. doi:10.1164/rccm.201401-0065st.

4. Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014. *Pediatrics*. 2014;134(6). doi:10.1542/peds.2014-2986.

5. Allen HD, Shaddy RE, Penny DJ, Cetta F, Feltes TF. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 9th ed. Philadelphia, PA: Wolters Kluwer; 2016.

6. Flynn JT, Kaelber DC, Baker-Smith CM, et al. Clinical Practice Guideline for Screening and Management of High Blood Pressure in Children and Adolescents. *Pediatrics*. 2017;140(3). doi:10.1542/peds.2017-1904.

7. Sweet M, Taylor MR, Mestroni L. Diagnosis, prevalence, and screening of familial dilated cardiomyopathy. *Expert Opin Orphan Drugs*. 2015;3(8):869-876. doi:10.1517/21678707.2015.1057498.

8. TeRiele, Anneline, James, Cynthia, Approach to family screening in arrhythmogenic right ventricular dysplasia/ Cardiomyopathy. *European Heart Journal* (2016) 37, 755-763 doi:10.1093/eurheartj/ehv387.

9. Ommen SR, Mital S, Burke MA, et al. 2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients With Hypertrophic Cardiomyopathy: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2020;76(25):e159-e240. doi:10.1016/j.jacc.2020.08.045.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

References (Fetal Echo)

1. Donofrio MT, Moon-Grady AJ, Hornberger LK, et al. Diagnosis and Treatment of Fetal Cardiac Disease. Circulation. 2014;129(21):2183-2242. doi:10.1161/01.cir.0000437597.44550.5d

2. Brucato A. Prevention of congenital heart block in children of SSA-positive mothers. *Rheumatology*. 2008;47(Supplement 3):iii35-iii37. doi:10.1093/rheumatology/ken153

3. Mcbride KL, Garg V. Impact of Mendelian inheritance in cardiovascular disease. Annals of the New York Academy of Sciences. 2010;1214(1):122-137. doi:10.1111/j.1749-6632.2010.05791.x

4. Reddy UM, Abuhamad AZ, Levine D, et al. Fetal imaging: executive summary of a joint Eunice Kennedy Shriver National Institute Child Health and Human Development, Society for Maternal-Fetal Medicine, American Institute of Ultrasound in Medicine, American College of Obstetricians and Gynecologists, American College of Radiology, Society of Pediatric Radiology, and Society of Radiologists in Ultrasound Fetal Imaging Workshop. Obstet Gynecol Survey. 2014;69(8):453-455

5. Lee W, Allan L, Carvalho JS, et al. ISUOG consensus statement: what constitutes a fetal echocardiogram? *Ultrasound in Obstetrics and Gynecology*. 2008;32(2):239-242. doi:10.1002/uog.6115

6. Friedman DM, Kim MY, Copel JA, et al. Utility of Cardiac Monitoring in Fetuses at Risk for Congenital Heart Block. The PR Interval and Dexamethasone Evaluation (PRIDE) Prospective Study. Circulation. 2008;117(4):485-493. doi:10.1161/circulationaha.107.707661

7. AIUM Practice Parameter for the Performance of Fetal Echocardiography. *Journal of Ultrasound in Medicine*. 2019;39(1). doi:10.1002/jum.15188

8. Sammaritano LR, Bermas BL, Chakravarty EE, et al. 2020 American College of Rheumatology Guideline for the Management of Reproductive Health in Rheumatic and Musculoskeletal Diseases. *Arthritis Care & Research*. 2020;72(4):461-488. doi:10.1002/acr.24130

9. Jenkins KJ, Correa A, Feinstein JA, et al. Noninherited Risk Factors and Congenital Cardiovascular Defects: Current Knowledge. *Circulation*. 2007;115(23):2995-3014. doi:10.1161/circulationaha.106.183216

10. Anderson KN, Lind JN, Simeone RM, Bobo WV, Mitchell AA, Riehle-Colarusso T, Polen KN, Reefhuis. Maternal Use of Specific Antidepressant Medications During Early Pregnancy and the Risk of Selected Birth Defects. *J.JAMA Psychiatry.* 2020 Aug 5;77(12):1246-55. doi: 10.1001/jamapsychiatry.2020.2453

11. ACOG Practice Bulletin No. 227: Fetal Growth Restriction. *Obstet Gynecol*. 2021;137(2):e16-e28 doi: 10.1097/AOG.0000000000004251

12. Martins JG, Biggio, JR, Abuhamad, A. Society for Maternal-Fetal Medicine (SMFM) Consult Series #52: Diagnosis and Management of Fetal Growth Restriction. 2020 doi:10.1016/j.ajog.2015.01.059

Computed Tomography

CT Heart and Coronary Computed Tomography Angiography (CCTA) General Considerations (PEDCD-10.1)

CDP.00.A
v1.0.2023

- Metal artifact reduces the accuracy of CCTA. Devices that can cause this issue include, but are not limited to:
  - Surgical clips
  - Pacemaker devices
  - Defibrillator devices
  - Tissue expanders
- Cardiac testing that does not involve exposure to ionizing radiation should be strongly considered.

Radiation Dose (PEDCD-10.2)

- ACR–NASCI–SPR Practice Parameter for the Performance and Interpretation of Cardiac Computed Tomography (CT) states “Cardiac CT should be performed only for a valid medical indication and with the minimum radiation exposure that provides diagnostic image quality”
- ACR–NASCI–SPR Practice Parameter for the Performance of Quantification of Cardiovascular Computed Tomography (Ct) And Magnetic Resonance Imaging (MRI) states, “In younger patients, MRI may be the preferred modality, particularly when functional assessment with CT would require retrospective ECG gating and relatively high radiation doses. Further, the use of time-resolved MRA and phase contrast MRI methods offer significant advantages whose relative importance will depend on the specific application”
  - See table: Practice Estimate of Effective Radiation Dose chart for Selected Imaging Studies in **General Guidelines (CD-1)** in the general Cardiac Imaging Guidelines

Indications for CCTA (CPT® 75574) (PEDCD-10.3)

CDP.00.A
v1.0.2023

- In addition to indications listed in previous guideline sections, CCTA is indicated for any of the following, when a recent TTE and/or MRI is inconclusive:
  - Persistent exertional chest pain and normal stress test
  - Full sibling(s) with history of sudden death syndrome before age 30 or with documented anomalous coronary artery
  - Resuscitated sudden death and contraindication to conventional coronary angiography
  - Unexplained new onset of heart failure if CCTA will replace conventional invasive coronary angiography
  - Documented ventricular tachycardia (6 beat runs or greater) if CCTA will replace conventional invasive coronary angiography
  - Equivocal coronary artery anatomy on conventional cardiac catheterization
  - In infants: otherwise unexplained dyspnea, tachypnea, wheezing, episodic pallor, irritability, sweating, poor feeding, and/or failure to thrive
    - The presence of other congenital heart disease is not a separate indication for CCTA to rule out anomalous coronary artery (except when coronary artery surgery is pending, i.e., Transposition of the great arteries, Tetralogy of Fallot, Truncus arteriosis, aortic root surgery)
  - Evaluation of the arterial supply and venous drainage in children with bronchopulmonary sequestration
- See Coronary Anomalies (PEDCD-2.4.12)

Indications for Cardiac CT (CPT® 75572) (PEDCD-10.4)

CDP.00.A
v1.0.2023

- In addition to indications listed in previous guideline sections, CCT is indicated for any of the following, when a recent TTE and/or MRI is inconclusive:
  - Cardiac or pericardial mass
  - Pericarditis
  - Complications of cardiac surgery or evaluation of post-operative anatomy
  - Cardiac thrombus in individuals with technically limited TTE, TEE, or MRI
  - Clinical suspicion of arrhythmogenic right ventricular dysplasia (ARVD) or arrhythmogenic cardiomyopathy (ARVC)
  - Native aortic abnormalities if echocardiogram is indeterminate
  - Intracardiac anatomy unclear after TTE or CMRI
  - A CTA Chest may also be indicated during a Cardiac CT if there are issues regarding the chest vessels that are inconclusive after echocardiogram or Cardiac MRI

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Indications for Chest CTA with Cardiac CT or CTA (PEDCD-10.5)

CDP.00.A
v1.0.2023

- A Chest CTA may be indicated in individuals who require Cardiac CT or Cardiac CTA when:
  - A TTE or MRI is inconclusive for issues regarding chest vasculature
  - Routine imaging is indicated based on Imaging and Surveillance per Congenital lesion (PEDCD-2.4)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

References (PEDCD-10)

v1.0.2023

1. Nelson Textbook of Pediatrics, 20th Edition, Robert M. Kliegman, MD, Bonita M.D. Stanton, MD, Joseph St. Geme, MD and Nina F Schor, MD, PhD p2182 to p2292.

2. American College of Radiology. ACR–ASER–SCBT-MR–SPR Practice Parameter for the Performance of Pediatric Computed Tomography (CT). American College of Radiology | Practice Parameters by Modality: https://www.acr.org/Clinical-Resources/Practice-Parameters-and-Technical-Standards/Practice-Parameters-by-Modality. Published 2014.

3. American College of Radiology. ACR–NASCI–SPR Practice Parameter for the Performance of Quantification of Cardiovascular Computed Tomography (CT) and Magnetic Resonance Imaging (MRI). American College of Radiology | Practice Parameters by Modality. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/Quant-CardiacCT-MR.pdf. Published 2017.

4. Einstein AJ, Henzlova MJ, and Rajagopalan S. Estimating risk of cancer associated with radiation exposure from 64-slice computer tomography coronary angiography. JAMA, 2007;298(3):317-323.

5. Baumgartner H, Bonhoeffer P, Groot NMSD, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010): The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC). *European Heart Journal*. 2010;31(23):2915-2957. doi:10.1093/eurheartj/ehq249.

6. Bhatt AB, Foster E, Kuehl K, et al. Congenital Heart Disease in the Older Adult. *Circulation*. 2015;131(21):1884-1931. doi:10.1161/cir.0000000000000204.

7. Feltes TF, Bacha E, Beekman RH, et al. Indications for Cardiac Catheterization and Intervention in Pediatric Cardiac Disease: A Scientific Statement from the American Heart Association. *Circulation*. 2011;123(22):2607-2652. doi:10.1161/cir.0b013e31821b1f10.

8. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease. *Circulation*. 2008;118(23). doi:10.1161/circulationaha.108.190690.

9. Cifra B, Dragulescu A, Border WL, Mertens L. Stress echocardiography in paediatric cardiology. *European Heart Journal – Cardiovascular Imaging*. 2015;16(10):1051-1059. doi:10.1093/ehjci/jev159.

10. Allen HD, Shaddy RE, Penny DJ, Cetta F, Feltes TF. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 9th ed. Philadelphia, PA: Wolters Kluwer; 2016. American College of

Radiology. ACR–NASCI–SPR Practice Parameter for the Performance and Interpretation of Cardiac Computed Tomography (CT). American College of Radiology | American College of Radiology. https://www.acr.org/Clinical-Resources/Practice-Parameters-and-Technical-Standards/Practice-Parameters-by-Modality. Published 2017.

11. American College of Radiology. Practice Parameters by Modality: ACR–NASCI–SPR Practice Parameter for the Performance and Interpretation of Cardiac Computed Tomography (CT). American College of Radiology | American College of Radiology. https://www.acr.org/Clinical-Resources/Practice-Parameters-and-Technical-Standards/Practice-Parameters-by-Modality. Published 2016.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Magnetic Resonance Imaging

Cardiac MRI Coding (PEDCD-9.2)

CDP.00.A
v1.0.2023

Cardiac Imaging Procedure Codes	
Cardiac MRI	**CPT ®**
Cardiac magnetic resonance imaging for morphology and function without contrast.	75557
Cardiac magnetic resonance imaging for morphology and function without and with contrast and further sequences.	75561
Cardiac magnetic resonance imaging for morphology and function without contrast; with stress imaging (rarely used in pediatrics).	75559
Cardiac magnetic resonance imaging for morphology and function without and with contrast and further sequences; with stress imaging (rarely used in pediatrics).	75563
Cardiac magnetic resonance imaging for velocity flow mapping (List separately in addition to code for primary procedure).	+75565

- Only one procedure code from the set: CPT® 75557, CPT® 75559, CPT® 75561, and CPT® 75563 should be reported per session.
- Only one flow velocity measurement (CPT® +75565) should be reported per session.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Indications for Cardiac MRI (PEDCD-9.3)

CDP.00.A
v1.0.2023

- In addition to indications listed in previous guideline sections, Cardiac MRI evaluation is indicated for any of the following, when a recent TTE is inconclusive:
  - Assessment of global ventricular function and mass if a specific clinical question is left unanswered by recent TTE and the MRI results will affect management of the individual's condition
  - Individuals with complex congenital heart disease (e.g., Tetralogy of Fallot [TOF], single ventricle, truncus arteriosis, Transposition of the Great Arteries [TGA]) may require a baseline MRI, or routine Cardiac MRI, especially as they approach their teenage years, due to poor imaging windows on echocardiogram, and the need for specific clinical information not seen on prior echocardiograms due to these known limitations. Once these individuals reach age 18, they can be imaged by adult congenital heart disease guideline.
  - Clinical suspicion of arrhythmogenic right ventricular dysplasia (ARVD) or arrhythmogenic cardiomyopathy (ARVC).
  - For pericardial disease (including constrictive pericarditis, restrictive pericarditis, and perimyocarditis), MRI should not be utilized to diagnose pericarditis but only to answer the question regarding possible constriction or restriction suggested clinically or by other techniques (TTE, etc.)
    - MRI without and with contrast (CPT® 75561) is considered the optimal test for this disorder.
  - Evaluate cardiac tumor or mass
    - MRI without and with contrast (CPT® 75561) is considered the optimal test for this disorder.
  - Evaluate anomalous coronary artery
    - MRI without and with contrast (CPT® 75561) or CCTA (CPT® 75574) after echocardiogram is considered the optimal test for this disorder.
  - For Fabry's disease, late enhancement MRI may predict the effect of enzyme replacement therapy on myocardial changes that occur with this disease.
    - MRI without and with contrast (CPT® 75561) is considered the preferred test for this disorder.
  - Cardiac MRI can be performed to evaluate individuals with congenital cardiomyopathy (muscular dystrophy, glycogen storage disease, fatty acid oxidation disorders, mitochondrial disorders, etc.) or unexplained cases of cardiomyopathy in order to characterize the myocardium.

- Cardiac stress perfusion study (CPT® 75559 or CPT® 75563) can be considered on a case by case basis for individuals with any of the following:
  - Anomalous coronary artery
  - Kawasaki disease
  - TGA
  - Ross operation
  - Other disorder with the potential for coronary ischemia
  - Individuals in whom an exercise stress test (EST) without imaging is indicated, but the individual is not able to perform an EST.
  - Individuals in whom an exercise stress test (EST) is equivocal, positive, or concern for a false negative
- Assessment of cardiac iron overload such as in hemochromatosis, thalassemia, sickle cell (either CPT® 75557 or CPT® 71550, T2* MRI, contrast not necessary).
  - Screening imaging may be approved every 12 months
  - Imaging may be approved every 3 months for treatment response in individuals receiving active treatment (chelation +/- phlebotomy)
  - Frequently performed along with MRI Abdomen (CPT® 74181) to assess liver iron deposition. See **Transfusion-Associated (Secondary) Hemochromatosis (PEDAB-18.2)** in the Pediatric Abdomen Imaging Guidelines.

- Asymptomatic individuals with Duchenne Muscular Dystrophy (DMD), every year starting at 6 years, if done instead of echocardiogram for surveillance. Female carriers, would not typically be imaged until ≥18 years of age, and should be imaged according to general Cardiac Imaging guidelines. MRI for DMD would be either CPT® 75557 or CPT® 75561. CPT® 75565 or CPT® 71555 would not be indicated unless there was an independent indication for either of those codes.

Indications for Chest MRA for Congenital Heart Disease (PEDCD-9.4)

CDP.00.A
v1.0.2023

- For Familial Aortopathies See Section **Thoracic Aortic Disease (PEDPVD-4.1)** in the Pediatric Peripheral Vascular Disease Imaging Guidelines
- For individuals with known CHD for routine imaging Imaging and Surveillance per Congenital lesion (PEDCD-2.4)
- For individuals who have both cardiac and ascending aorta abnormalities (e.g., truncus arteriosus), the following studies may be indicated following an inconclusive TTE:
  - Cardiac MRI (CPT® 75557 or CPT® 75561)
  - And MRI Chest (CPT® 71552) or MRA Chest (CPT® 71555) if aorta is involved
- For individuals with aortic abnormalities without cardiac abnormalities (i.e. normal intracardiac anatomy, but coarctation or peripheral pulmonary artery stenosis), the following studies may be indicated following an inconclusive TTE:
  - MRI Chest (CPT® 71552)
  - MRA Chest (CPT® 71555)
- MRA Chest is indicated for individuals with cardiomyopathy or isolated abnormal intracardiac anatomy, when there are inconclusive images on echocardiogram related to chest vessels (e.g. aortic arch, pulmonary arteries, pulmonary veins, systemic veins).

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

References (PEDCD-9)

v1.0.2023

1. Nelson Textbook of Pediatrics, 20th Edition, Robert M. Kliegman, MD, Bonita M.D. Stanton, MD, Joseph St. Geme, MD and Nina F Schor, MD, PhD, p2182 to p2292.

2. Atweh LA, Orth RC, Guillerman RP, Zhang W, Kan JH. MR imaging of children and young adults with classic findings of osteonecrosis on unenhanced MR images: do contrast-enhanced sequences help? *Pediatric Radiology*. 2013;43(11):1502-1506. doi:10.1007/s00247-013-2714-1.

3. Cohen MS, Eidem BW, Cetta F, et al. Multimodality Imaging Guidelines of Patients with Transposition of the Great Arteries: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography. *Journal of the American Society of Echocardiography*. 2016;29(7):571-621. doi:10.1016/j.echo.2016.04.002.

4. Adler Y, Charron P, Imazio M, et al. 2015 ESC Guidelines for the diagnosis and management of pericardial diseases. *European Heart Journal*. 2015;36(42):2921-2964. doi:10.1093/eurheartj/ehv318.

5. Allen HD, Shaddy RE, Penny DJ, et. al TF. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 9th ed. Philadelphia, PA: Wolters Kluwer; 2016.

6. Hundley WG, Bluemke DA, Finn JP, et al. ACCF/ACR/AHA/NASCI/SCMR 2010 Expert Consensus Document on Cardiovascular Magnetic Resonance. *Circulation*. 2010;121(22):2462-2508. doi:10.1161/cir.0b013e3181d44a8f.

7. Cohen MS, Eidem BW, Cetta F, et al. Multimodality Imaging Guidelines of Patients with Transposition of the Great Arteries: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography. *Journal of the American Society of Echocardiography*. 2016;29(7):571-621. doi:10.1016/j.echo.2016.04.002.

8. Valente AM, Cook S, Festa P, et al. Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography. *Journal of the American Society of Echocardiography*. 2014;27(2):111-141. doi:10.1016/j.echo.2013.11.009.

9. Feingold B, Mahle WT, Auerbach S, et al. Management of Cardiac Involvement Associated With Neuromuscular Diseases: A Scientific Statement From the American Heart Association. *Circulation*. 2017;136(13). doi:10.1161/cir.0000000000000526.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

10. Mah ML, Cripe L, Slawinski MK, et al. Duchenne and Becker muscular dystrophy carriers: Evidence of cardiomyopathy by exercise and cardiac MRI testing. *International Journal of Cardiology*. 2020;316:257-265. doi:10.1016/j.ijcard.2020.05.052.

11. Power LC, O'Grady GL, Hornung TS, Jefferies C, Gusso S, Hofman PL. Imaging the heart to detect cardiomyopathy in Duchenne muscular dystrophy: A review. *Neuromuscular Disorders*. 2018;28(9):717-730. doi:10.1016/j.nmd.2018.05.011.

12. Hor KN, Mah ML, Johnston P, Cripe TP, Cripe LH. Advances in the diagnosis and management of cardiomyopathy in Duchenne muscular dystrophy. *Neuromuscular Disorders*. 2018;28(9):711-716. doi:10.1016/j.nmd.2018.06.014.

13. Buddhe S, Cripe L, Friedland-Little J, et al. Cardiac Management of the Patient With Duchenne Muscular Dystrophy. *Pediatrics*. 2018;142(Supplement 2). doi:10.1542/peds.2018-0333i.

14. Birnkrant DJ, Bushby K, Bann CM, et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management. *The Lancet Neurology*. 2018;17(4):347-361. doi:10.1016/s1474-4422(18)30025-5.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Diagnostic Heart Catheterization

Cardiac Catheterization General Information (PEDCD-11.1)

CDP.DHC.0011.A
v1.0.2023

Cardiac Catheterization Procedure Codes	
Cardiac Cath Procedures	**CPT ®**
Congenital Heart Disease Code “Set”	93593-93597
Right heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone; normal native connections	93593
Right heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone; abnormal native connections	93594
Left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone, normal or abnormal native connections	93595
Right and left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone(s); normal native connections	93596
Right and left heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone(s); abnormal native connections	93597
Anomalous coronary arteries, patent foramen ovale, mitral valve prolapse, and bicuspid aortic valve	93451-93464, 93566-93568
RHC without LHC or coronaries	93451
LHC without RHC or coronaries	93452
RHC and retrograde LHC without	93453

Cardiac Catheterization Procedure Codes	
Cardiac Cath Procedures	**CPT ®**
coronaries	
Native coronary artery catheterization;	93454
with bypass grafts	93455
with RHC	93456
with RHC and bypass grafts	93457
with LHC	93458
with LHC and bypass grafts	93459
with RHC and LHC	93460
with RHC and LHC and bypass grafts	93461
LHC by trans-septal or apical puncture	+93462
Angiography of non-coronary arteries and veins performed as a distinct service	Select appropriate codes from the Radiology and Vascular Injection Procedures sections.
CPT® 93593 to 93597 are appropriate for invasive evaluation of congenital heart disease	

- These guidelines apply to individuals with stable conditions and who are not in the acute setting. Individuals in acute settings or with unstable angina should be handled as medical emergencies.
- Pediatric catheterizations are done for many purposes, including diagnosis and intervention of congenital and acquired heart disease.
- When device placement is planned (ASD/VSD device, transcatheter valve implantation, pda device), the procedure codes for those devices include all cardiac catheterization(s), intraprocedural contrast injection(s), fluoroscopic radiological supervision and interpretation, and imaging guidance performed to complete the procedure. A diagnostic cath may be considered on a case-by-case basis if there are unanswered issues via noninvasive imaging.
- Stenting or ballooning of coarctation of the aorta is an endovascular procedure. A right and/or left heart cath can be approved during a coarctation stenting when ordered by the cardiologist ordering or performing the procedure.
- A right heart cath can be approved for pulmonary artery interventions (e.g., stents, coils).

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Background and Supporting Information

- As stated in the echo section, a peri-procedural TEE (CPT® 93355) does not require eviCore prior authorization

Cardiac Catheterization Indications (PEDCD-11.2)

CDP.DHC.0011.2.A
v1.0.2023

- Diagnostic catheterization is indicated:
  - When other advanced imaging has failed to resolve a clinical issue and results will impact the individual's management
    - For example, a cath to assess Ventricular pressures and shunt to determine if VSD surgery is required
  - For preoperative assessment in complex heart disease
    - Norwood procedure
    - Bidirectional Glenn shunt
    - Fontan procedure
    - Pulmonary atresia
  - Pulmonary hypertension
  - During some interventions such as:
    - Valvuloplasty
    - Pulmonary artery or vein stents
  - See **Kawasaki Disease Initial Imaging (PEDCD-6.1)** for specific intervals in Kawasaki Disease
  - On an individual who is having a device placed when:
    - A diagnostic catheterization, or stenting is needed in addition to the device
    - The diagnostic catheterization is indicated separate from the device placement
  - Individuals with anomalous coronary arteries, or with syndromes associated with abnormal coronary arteries (i.e., Williams syndrome) or acquired CAD (i.e., KD- see **Kawasaki Disease Initial Imaging (PEDCD-6.1)**
    - When diagnostic images are not adequate or evaluation or treatment decision
    - Preoperative for cardiac surgery
    - New symptoms concerning for ischemia

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

References (PEDCD-11)

v1.0.2023

1. Optum360®EncoderPro.com. EncoderPro.com Online Medical Coding Software | Optum360Coding.com. https://www.encoderpro.com. Published 2019.

2. Feltes TF, Bacha E, Beekman RH, et al. Indications for Cardiac Catheterization and Intervention in Pediatric Cardiac Disease: A Scientific Statement from the American Heart Association. *Circulation*. 2011;123(22):2607-2652. doi:10.1161/cir.0b013e31821b1f10.

3. LaDisa JF, Taylor CA, Feinstein JA. (2006) Endovascular Treatment Strategies for Coarctation of the Aorta. In: Rousseau H, Verhoye JP, Heautot JF. (eds) Thoracic Aortic Diseases. Springer, Berlin, Heidelberg.

4. Bergersen L, Foerster S, Marshall AC, Meadows J. *Congenital Heart Disease The Catheterization Manual*. New York, NY: Springer US; 2009.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924