CLINICAL GUIDELINES

Pediatric Cardiac Imaging Guidelines

Version 1.0

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eviCore healthcare Clinical Decision Support Tool Diagnostic Strategies:This tool addresses common symptoms and symptom complexes. Imaging requests for individuals with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician, specialist and/or individual's Primary Care Physician (PCP) may provide additional insight.

Pediatric Cardiac Imaging Guidelines

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PEDCD-1: General Guidelines

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PEDCD-1.0: General Guidelines

- A recent (within 60 days) face-to-face evaluation should be performed prior to considering advanced imaging unless the patient is undergoing guideline-supported scheduled follow-up imaging evaluation. This evaluation should include:
  - A detailed history
  - Physical examination
  - Appropriate laboratory studies
- Advanced imaging of the heart should only be approved in patients who have documented active clinical signs or symptoms of disease involving the heart or as follow-up for findings on echocardiograms.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the heart are not necessary unless:
  - There is evidence for progression of disease
  - New onset of disease and/or documentation of how repeat imaging will affect patient management or treatment decisions.

PEDCD-1.1: Pediatric Cardiac Imaging Age Considerations

- Heart disease in the pediatric population involves predominantly congenital lesions. Pediatric patients 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 patient age, comorbidities, and differences in disease natural history between children and adults.
- Individuals who are < 18 years old should be imaged according to the Pediatric Cardiac Imaging Guidelines, and individuals who are age ≥ 18 years should be imaged according to the Cardiac Imaging Guidelines, except where directed otherwise by a specific guideline section.

PEDCD-1.2: Pediatric Cardiac Imaging Appropriate Clinical Evaluation

- Patients for whom routine imaging is anticipated at the next visit (for example on year follow-up echo for a 10 year old with a VSD) may have these imaging studies approved without face to face evaluation if study was already indicated
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic patients for disorders involving the heart is not supported.
- Patients starting ADHD medications, in the absence of other appropriate indications listed in these guidelines, imaging is not indicated.
- Asymptomatic Patients with known or suspected syndromes, which may be associated with congenital heart disease, can have an initial echocardiogram.
- Asymptomatic patients with family history of aortopathy, cardiomyopathy, congenital heart disease with known inheritance pattern:
  - Can have an echocardiogram as an initial study

- Patients who are genotype positive and phenotype negative, can have annual screening.
- Patients whose first degree relative has a known identified genotype, and the patient has a negative result for that prior identified genotype, do not require cardiac screening.
- Additional studies are determined based on findings.

- Asymptomatic patients with exposure to cardiotoxic drugs can have serial echocardiograms as per **PEDONC-19.2: Cardiotoxicity and Echocardiography** in the Pediatric Oncology imaging guideline

PEDCD-1.3: 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 patient 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 patient 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 **PEDCD-10: CT Heart and Coronary Computed Tomography Angiography (CCTA) – Other Indications)**
- Ultrasound
  - 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 **PEDCD-8: Echocardiography Other Indications**.
- Nuclear Medicine
  - SPECT, PET stress may be indicated for patients with anomalous CA, angina chest pain, and follow-up for Kawasaki. 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 patients when echocardiography is insufficient: See: **PEDONC-1.2: Appropriate Clinical Evaluations** 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 patients 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. Refer to **CD-3.7: Myocardial Tc-99m Pyrophosphate Imaging** and **CD-3.8: Cardiac Amyloidosis**

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

- 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.

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PEDCD-2: Congenital Heart Disease

PEDCD-2.1: Congenital Heart Disease General Information

- Congenital heart disease accounts for the majority of cardiac problems occurring in the pediatric population. Patients may be diagnosed any time spanning prenatal evaluation to adolescence. For patients over 18 year of age, see **CD-11: Adult Congenital Heart Disease** in the Cardiac Imaging Guidelines.
- There are a number of variables that influence the modality and timing of imaging patients with congenital heart disease, which results in a high degree of individuality in determining the schedule for imaging these patients, including:
  - Gestational age
  - Patient age
  - Physiologic effects of the defect
  - Status of interventions (catheterization and surgical)
  - Rate of patient growth
  - Stability of the defect on serial imaging
  - Comorbid conditions
  - Activity level
- Age definitions (for purpose of these guidelines)
  - Neonate (newborn) 0-28 days,
  - Infant 0-12 months
  - Child 1-18 years
  - Adolescents 11-18
- 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)
- Patients 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 patients that can have both cardiac MR or cardiac CT and/or chest MRA or chest CTA, this is abbreviated as CMR/CT-CMRA/CTA
- Patients 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, patients can have TTE (depending on lesion can also include cardiac and or chest MR/CT)
  - Can have an echocardiogram within one month post-operative
  - Cardiac MRI/CCT if prior echo is equivocal
- Chest MRA/CTA can be performed if prior echo is equivocal and there are issues regarding aortic arch or pulmonary arteries or veins
- In patients 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 Chest MRA/CTA, may be done with the frequency of echoes, if done instead of an echo.
- Chest MRA/CTA if thoracic issue not seen on echo
- For routine non-invasive imaging for a specific lesion see **PEDCD-2.4: Imaging and Surveillance per Congenital lesion**.
- For catheterizations see section **PEDCD-11: Cardiac Catheterization**
- Patients with Pulmonary hypertension with CHD should be reviewed for both their lesion and for PHT in section **PEDCD-7: Pediatric Pulmonary Hypertension**

PEDCD-2.2: Congenital Heart Disease Coding

PEDCD-2.2.1: Congenital Heart Disease Echocardiography Coding

- Any of the following echocardiography code combinations are appropriate for re-evaluation of patients 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.
- All requested CPT® combinations other than those listed in this section should be forwarded for Medical Director Review.

PEDCD-2.2.2: Congenital Heart Disease imaging per modality

- 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)
    - 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
  - Chest MRI
    - 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 (chest CTA)
        - 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)

PEDCD-2.3: Congenital Heart Disease Modality Considerations

- 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 patients with valvular disease or a need to evaluate intracardiac blood flow. These patients 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 patient cannot cooperate with full cardiac MRI exam.
- MRA Chest (CPT® 71555) is 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 patients with complex congenital heart disease
  - Pulmonary artery (PA) and Pulmonary vein (PV) assessment
  - CTA of the chest is indicated to assess:
    - Coarctation of the aorta
    - Tetralogy of Fallot
    - Anomalous pulmonary veins and other lesions of the great arteries
    - Vascular rings with inconclusive recent echocardiography findings

PEDCD-2.4: Imaging and Surveillance per Congenital lesion

- Echocardiography is repeated frequently throughout a child's life, and can generally be approved regardless of symptoms based on the lesion and age of the patient. 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 patient 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.

PEDCD-2.4.1: Atrial Defects-Secundum ASD, PFO, and Partial anomalous pulmonary venous return (PAPVR), Sinus Venosus defect

- See section on AVSD in **PEDCD 2.4.3: AVSD (Atrioventricular canal, Endocardial cushion defect)** for primum ASD
- PFO (Patent Foreman Ovale)
  - Routine surveillance in an asymptomatic patient with PFO is not indicated
  - PFO is a normal variant
  - In infants, a PFO that is difficult to distinguish from an ASD should be managed like a small ASD (with congenital echo).
- TTE (CPT® 93306- non congenital echocardiogram) is indicated when a patient 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 patients 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.

PEDCD-2.4.2: VSD

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

PEDCD-2.4.3: AVSD (Atrioventricular canal, Endocardial cushion defect)

- 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
  - 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

PEDCD-2.4.4: PDA (Patent ductus arteriosus)

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

PEDCD-2.4.5: TAPVR Total anomalous pulmonary venous return

- 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
  - Every 1 year in childhood
  - Adolescence
    - TTE every 2 years

PEDCD-2.4.6: Ebstein anomaly and TV dysplasia

- Any surgical status
  - TTE, TTE, 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

PEDCD-2.4.7: Pulmonary Stenosis (PS)

- 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 Section **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
    - Chest MRA/CTA 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 patient with heart failure, TTE every 3 months

PEDCD-2.4.8: Pulmonary Atresia with intact septum (PAIVS)

- 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

PEDCD-2.4.9: Mitral valve disease

- 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

PEDCD-2.4.10: LVOT lesions

Subvalvar 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

- 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

- 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 Ao
- Post procedural
  - Within 30 days TTE
  - Infant
    - 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 f, 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
            - Patients 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

PEDCD-2.4.11: Aortic Coarctation and IAA (interrupted aortic arch)

- All patients
    - TTE, Chest MRA/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 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
        - Chest MRA, Chest CTA 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
        - 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

- Chest MRA/CTA every 3 years (include cardiac MR/CT if issues not clarified on echo)

PEDCD-2.4.12: Coronary Anomalies

- Evaluating coronary artery anomalies and other complex congenital heart disease of cardiac chambers or great vessels is an appropriate indication for CCTA, or cardiac MRI
  - CPT® 75574 for evaluating coronary artery anomalies
  - CPT® 75573 for congenital heart disease
  - Can add CPT® 71275 (chest CTA) to evaluate great vessels
- Congenital anomalies of the coronary arteries are an important cause of sudden death in pediatric patients. 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, patients with no echocardiographic findings, but symptoms concerning for angina chest pain may require stress testing.
  - Patients who have positive echocardiographic findings, regardless of symptoms, and patients 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 chest MRA/CTA.
        - 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 chest MRA/CTA can be included when MR/CT imaging is required
      - Stress imaging- to assess the need for surgery
  - Unrepaired
    - Routine surveillance every 2 years in an asymptomatic patient 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 patient with small coronary fistula
        - TTE- every 2 years
    - Routine surveillance in an asymptomatic patient 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
- Patients with congenital heart disease such as TOF, Truncus Arteriosus, and , TGA have increased incidence of coronary artery anomalies Patients with Williams syndrome can have coronary artery stenosis.
- Patients with confirmed coronary artery anomalies may require repeat imaging based on the clinical scenario.
- The use of CCTA to rule out anomalous coronary artery should be limited to one of the following:
    - Patients who need to have an anomalous coronary artery mapped prior to an invasive procedure.
    - Patients who have not had a previous imaging study that clearly demonstrates an anomalous coronary artery
    - Patients with a history that includes one or more of the indications in **PEDCD-10.3: Indications for CCTA (CPT® 75574)**.

PEDCD-2.4.13: Tetralogy of Fallot (TOF)

- 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
- Unpaired
  - 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 patient 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
  - Cardiac MR/CT, Chest MRA/CTA every
    - Routine surveillance (36 months) in a patient 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

PEDCD-2.4.14: Double Outlet Right Ventricle (DORV)

- 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 a patient 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, Chest MRA/CTA
    - 3 years for asymptomatic patient

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

- 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
    - Chest MRA/CTA
        - Every 3 years
        - Every year if neo AI, or aortic dilation
    - 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.

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

- 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
        - post–operative evaluation (within 30 days)
        - every 3 months within a year following repair in an asymptomatic patient with no or mild sequelae
        - every 1 year after the first year following repair in an asymptomatic patient 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 patient with no or mild sequelae
        - Annually in an asymptomatic patient with no or mild sequelae
        - Every 3 months if in a patient 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 a patient with ≥moderate systemic AV valve regurgitation, systemic RV dysfunction, and/or LV-to-PA conduit dysfunction, or with CHF symptoms

PEDCD-2.4.17: Truncus Arteriosus

- 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

PEDCD-2.4.18: Single Ventricle (SV)

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

PEDCD-2.4.19: Eisenmenger and PHT (with CHD)-

- PHT without CHD is covered in section **PEDCD-7: Pediatric Pulmonary Hypertension**
- These are in addition to studies supported by lesion
- Any surgical status
  - TTE, CMR/CT-CMRA/CTA are indicated for any of the following:
- Initial evaluation (TTE)
  - Change in clinical status and/or new concerning signs or symptoms
  - Before and after PHT therapy
- Eisenmenger Syndrome (ES) patient
  - TTE every 6 months

- PHT associate with CHD
  - Unrepaired Patients with evidence of elevated pulmonary vascular resistance can have echocardiograms based on the frequency requested by the provider
  - TTE and Cardiac CMR/CCT for changes in change in pulmonary arterial hypertension-targeted therapy in a patient with postoperative PH
  - TTE every 3 months in postoperative stable child with PHT

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PEDCD-3: Heart Murmur

PEDCD-3.1: Heart Murmur General

- Heart murmurs are extremely common in pediatric patients. 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 patient 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 a patient with a murmur for the first time, it will not be known whether the patient has congenital heart disease or not. The cardiologist only submits charges for the procedure actually performed.
- 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.

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PEDCD-4: Chest Pain

PEDCD-4.1: Chest Pain General

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

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination, EKG, and appropriate laboratory studies should be performed prior to considering advanced imaging.
- Echocardiography is indicated for pediatric patients 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 a patient for the first time, it will not be known whether the patient has congenital heart disease or not. The cardiologist only submits charges for the procedure actually performed.
- The following echocardiography code combinations should be approved 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
- Patients 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
- Chest MRA or CTA if pulmonary embolism or aortic dissection is suspected

- Stress imaging is indicated if other imaging suggests coronary artery abnormity, or ETT suggests ischemia. EKG is uninterpretable. Any indication in section CD 1.4 This can include Stress SPECT, echo or MR

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PEDCD-5: Syncope

PEDCD-5.1: Syncope

Syncope in pediatric patients is common, with up to 15% of patients 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.

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination, EKG, and appropriate laboratory studies should be performed prior to considering advanced imaging.
- Echocardiography is not indicated for most patients with isolated syncope.
- Echocardiography is indicated for pediatric patients 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 a patient for the first time, it will not be known whether the patient has congenital heart disease or not. The cardiologist only submits charges for the procedure actually performed.
- The following echocardiography code combinations should be approved 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
  - New signs or symptoms of cardiovascular disease other than syncope

- Family of history of sudden death, cardiomyopathy
- New abnormality on EKG

- Patients 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
- Chest MRA or CTA 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 CD 1.4

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PEDCD-6: Kawasaki Disease

PEDCD-6.1: Kawasaki Disease Initial Imaging

- 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.
  - A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination, and appropriate laboratory studies should be performed prior to considering advanced imaging.
  - Scheduled indicated follow-up imaging does not require 60-day contact, if indicated based on the below follow-up schedule.
  - Patients 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.
- 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

PEDCD-6.2: Acute Phase

- 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.*

- Echocardiography should be performed when the diagnosis of KD is considered,
  - Uncomplicated patients, echocardiography can be repeated after treatment **both:**
    - Within 1 to 2 weeks
    - Within 4 to 6 weeks
  - For patients 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 patients' 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 patients 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 in whom visualization of the coronary arteries with transthoracic echocardiography is inadequate and results will impact immediate management decisions.
  - These requests will be forwarded to Medical Director for evaluation
- 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 patients with severe coronary artery involvement or symptoms or signs, such as the presence of a pulsatile axillary mass. All other requests during the acute phase will be forwarded for review
- 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.

PEDCD-6.3: Chronic Phase

- 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 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 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 patients
  - 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*

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PEDCD-7: Pediatric Pulmonary Hypertension

PEDCD-7.1: Pediatric Pulmonary Hypertension General

- Pulmonary hypertension in children can be caused by cardiac, pulmonary, or systemic diseases, and idiopathic disease occurs as well.
- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination, and appropriate laboratory studies should be performed prior to considering advanced imaging.
- If pulmonary hypertension is suspected, initial evaluation should consist of chest x-ray, EKG, and echocardiography (CPT® 93306, or CPT® 93303, with CPT® 93320, and CPT® 93325, see: **PEDCD-8.1: Transthoracic Echocardiography (TTE) Coding** for echocardiography coding considerations).
- Repeat echocardiography intervals are variable depending on age of patient, 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 patients, including before and after initiation of PAH-targeted therapy, and for patients with concomitant congenital heart disease
- Chest CT (CPT® 71250) may be indicated in addition to Chest CTA (CPT® 71275) or Chest MRA (CPT® 71555) for initial evaluation of all pediatric patients 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 adult guidelines) see **CD-2.7: Stress Echocardiography – Indications, other than ruling out CAD**.

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PEDCD-8: Echocardiography – Other Indications

PEDCD-8.1: Transthoracic Echocardiography (TTE) Coding

- CPT® codes for echocardiography are listed in **PEDCD-1: General Guidelines**

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 reported with CPT® 93308 if color flow Doppler is included in the study.	93308
➢ For patients 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 patient 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 should be approved 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.
  - Depending upon individual health plan payer contracts, post-service audits may be completed to ensure proper claims submission.

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PEDCD-8.2: Initial Transthoracic Echocardiography (TTE) Indications

- 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
  - Obesity (BMI > 30) with additional cardiovascular risk factors
  - Stroke
  - Renal failure
  - Preoperative evaluation of patients 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
  - 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 , 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, and 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

PEDCD-8.3: Repeat Transthoracic Echocardiography Indications

- Repeat echocardiograms may be required for patients with no new symptoms.
  - In addition to indications listed in previous guideline sections, repeat TTE evaluation is indicated for any of the following:
    - New or worsening symptoms in a patient with known cardiac disease, previously normal echocardiogram with one of the following:
      - New or worsening cardiac symptoms
      - New EKG abnormality
      - Newly recognized family history suggestive of heritable heart disease
  - Every 12 months for patients age 12 to 18 years with first-degree family history of hypertrophic cardiomyopathy.
  - Patients who are status post heart transplant can have echocardiograms repeated as often as requested by heart transplant team.
  - Every 12 months for patients 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 patients with chronic pericardial effusions
- 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 **PEDONC-19.2: Cardiotoxicity and Echocardiography** for imaging guidelines.
- Aortopathies See **PEDPVD 4.1**

PEDCD-8.4: Transesophageal Echocardiography (TEE)

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

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PEDCD-9: Cardiac MRI – Other Indications

PEDCD-9.1: Cardiac MRI General Guidelines

- Requests for cardiac MRI that contain only one CPT® code can be completed by the Nurse Reviewer. If the request contains more than one cardiac/chest MRI CPT® code, it should be forwarded for Medical Director Review.

PEDCD-9.2: Cardiac MRI Coding

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.

PEDCD-9.3: Indications for Cardiac MRI

- 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 the management of the patient's condition
  - Patients 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, and these studies should be forwards to the medical director. Once these patients reach age 18, they can be imaging 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
  - After echocardiogram, MRI without and with contrast (CPT® 75561) or CCTA (CPT® 75574) 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 patients 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 patients with any of the following:
  - Anomalous coronary artery
  - Kawasaki disease
  - TGA
  - Ross operation
  - or other disorder with the potential for coronary ischemia
  - Patients in whom an exercise stress test (EST)without imaging is indicated, but the patient is not able to perform an EST.
  - Patients 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 patients receiving active treatment (chelation +/- phlebotomy)
  - Frequently performed along with MRI Abdomen (CPT® 74181) to assess liver iron deposition. See **PEDAB-18.2: Transfusion-Associated (Secondary) Hemochromatosis** for additional imaging guidelines.

PEDCD-9.4: Aortic Root and Aorta

- For screening due to family history of aortic aneurysm or dissection, see: **PVD-2.2: Screening for Vascular related genetic connective tissue Disorders (Familial Aneurysm Syndromes/Spontaneous Coronary Artery Dissection (SCAD)/ Ehlers-Danlos/Marfan/Loeys-Dietz)**.
- For patients who have both cardiac and ascending aorta abnormalities (e.g., truncus arteriosus), the following studies may be indicated following TTE:
  - Cardiac MRI (CPT® 75557 or CPT® 75561) when TTE is inconclusive.
  - If aorta is involved, MRI Chest (CPT® 71552) or MRA Chest (CPT® 71555) is also indicated.
- For patients with aortic abnormalities without cardiac abnormalities, any of the following studies is indicated:
  - MRI Chest (CPT® 71552)
  - MRA Chest (CPT® 71555)

PEDCD-9.5: Evaluation of Pericardial Effusion or Diagnosis of Pericardial Tamponade

- 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 patient'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.

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PEDCD-10: CT Heart and Coronary Computed Tomography Angiography (CCTA) – Other Indications

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

- 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, and tissue expanders.
- Cardiac testing that does not involve exposure to ionizing radiation should be strongly considered.

PEDCD-10.2: Radiation Dose

- 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 **CD-1: General Guidelines** in the cardiac imaging guidelines

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

- 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 also section **PEDCD-2.4.12: Coronary Anomalies**

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

➢ 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:
- Cardiac or pericardial mass
- Pericarditis
- Complications of cardiac surgery or evaluation of post-operative anatomy
- Cardiac thrombus in patients 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

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PEDCD-11: Cardiac Catheterization

PEDCD-11.1: Cardiac Catheterization General Information

Cardiac Catheterization Procedure Codes	
Cardiac Cath Procedures	**CPT®**
Congenital Heart Disease Code "Set"	93530-93533
Right Heart Catheterization (CHD)	93530
Right/Left Heart Catheterization (CHD)	93531
Right/Left Heart Catheterization (CHD-TS)	93532
Right/Left Heart Catheterization (CAD-ASD)	93533
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
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® 93530 to 93533 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 required, and should be reviewed by the medical director.
- For coarctation or aortic arch stenting, or other endovascular procedures with no intracardiac issues that require clarification by left heart cath, a left heart cath is not required along with these endovascular procedures. A left heart and right heart cath may be appropriate for some patients with Coarctation, and should be reviewed by the medical director

- A right heart cath can be approved for pulmonary artery interventions (e.g., stents, coils)

Practice Note

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

PEDCD-11.2: Cardiac Catheterization Indications

- Diagnostic catheterization is indicated:
  - When other advanced imaging has failed to resolve a clinical issue and results will impact patient 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 **PEDCD-6.1: Kawasaki Disease Initial Imaging** for specific intervals in Kawasaki Disease
  - On a patient 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
  - Patients with anomalous coronary arteries, or with syndromes associated with abnormal coronary arteries (i.e., Williams syndrome) or acquired CAD (i.e., KD- see **PEDCD-6.1: Kawasaki Disease Initial Imaging**)
    - When diagnostic imagines are not adequate or evaluation or treatment decision
    - Preoperative for cardiac surgery
    - New symptoms concerning for ischemia

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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 performed)	75573
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 assess the severity of coronary artery disease; anatomical data review in comparison with estimated FFR model to reconcile discordant data, interpretation and report	0504T

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] and/or ejection fraction[s], when performed); single study, at rest or stress (exercise or pharmacologic), with concurrently acquired computed tomography transmission scan	78430
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 or stress (exercise or pharmacologic)	78451
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 rest or stress (exercise and/or pharmacologic), wall motion study plus ejection fraction, with or without additional quantitative processing	78472
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 addition to code for primary procedure)	78496
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 distribution of radiopharmaceutical agent(s) (includes vascular flow and blood pool imaging, when performed); planar, whole body, requiring 2 or more days imaging	78804
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 or color Doppler echocardiography	93307
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
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 qualified health care professional	93351
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 contrast followed by with contrast, for congenital cardiac anomalies; including probe placement, image acquisition, interpretation and report	C8926
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 Catheterization Procedure Codes	
Right Heart Catheterization (CHD)	93530
Right/Left Heart Catheterization (CHD)	93531
Right/Left Heart Catheterization (CHD-TS)	93532
Right/Left Heart Catheterization (CAD-ASD)	93533
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
with RHC and LHC	93460
with RHC and LHC and bypass grafts	93461
LHC by transseptal or apical puncture	+93462