This tool addresses common symptoms and symptom complexes. Imaging requests for patients with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician, specialist and/or patient's Primary Care Physician (PCP) may provide additional insight.

**PEDIATRIC AND CONGENITAL IMAGING GUIDELINES**

**CARDIAC and PERIPHERAL VASCULAR**

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MedSolutions, Inc. Clinical Decision Support Tool
for Advanced Diagnostic Imaging

Common symptoms and symptom complexes are addressed by this tool. Imaging requests for patients with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician may provide additional insight.

This version incorporates MSI accepted revisions prior to 7/22/11
### 2011 PEDIATRIC CARDIAC IMAGING GUIDELINE NUMBER & TITLE

<table>
<thead>
<tr>
<th>Guideline Number</th>
<th>Title</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>PACCD-1</td>
<td>General Guidelines</td>
<td>3</td>
</tr>
<tr>
<td>PACCD-2</td>
<td>Echocardiography (ECHO)</td>
<td>4</td>
</tr>
<tr>
<td>PACCD-3</td>
<td>Cardiac MRI</td>
<td>10</td>
</tr>
<tr>
<td>PACCD-4</td>
<td>CT Heart &amp; Computed Tomography Coronary Angiography (CTCA)</td>
<td>14</td>
</tr>
<tr>
<td>PACCD-5</td>
<td>Syncope</td>
<td>18</td>
</tr>
</tbody>
</table>

### EVIDENCE BASED CLINICAL SUPPORT

<table>
<thead>
<tr>
<th>Guideline Number</th>
<th>Title</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>PACCD-3</td>
<td>Cardiac MRI</td>
<td>19</td>
</tr>
</tbody>
</table>

### PEDIATRIC CARDIAC GUIDELINE REFERENCES

<table>
<thead>
<tr>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>20</td>
</tr>
</tbody>
</table>

### 2011 PEDIATRIC & CONGENITAL PERIPHERAL VASCULAR DISEASE (PVD) IMAGING GUIDELINES

<table>
<thead>
<tr>
<th>Guideline Number</th>
<th>Title</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>PACPVD-1</td>
<td>General Guidelines</td>
<td>21</td>
</tr>
<tr>
<td>PACPVD-2</td>
<td>Aortic Disorders, Renal Vascular Disorders, and Visceral Artery Aneurysms</td>
<td>21</td>
</tr>
</tbody>
</table>

### PEDIATRIC PVD GUIDELINE REFERENCES

<table>
<thead>
<tr>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>22</td>
</tr>
</tbody>
</table>
PACCD-1~GENERAL GUIDELINES

- The Cardiac Imaging Guidelines are the same for both the pediatric population and the adult population unless there are specific guidelines listed here in the Pediatric and Congenital Cardiac Imaging Guidelines.
  - These guidelines are based upon using cardiac imaging to answer a specific clinical question that will affect patient management.
    - If the clinical question has already been answered based upon previous clinical evaluation or imaging, then additional cardiac imaging is not indicated.
    - Cardiac imaging is not indicated if the results will not affect patient management decisions.
- SPECT/CT which 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 these hybrid scans.
### PACCD-2~ECHOCARDIOGRAPHY (ECHO)

<table>
<thead>
<tr>
<th>PACCD-2</th>
<th>ECHOCARDIOGRAPHY (ECHO)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2.1</td>
<td>Transthoracic Echocardiography (TTE)</td>
</tr>
<tr>
<td>2.2</td>
<td>Frequency of Echocardiography Testing</td>
</tr>
<tr>
<td>2.3</td>
<td>Transesophageal Echocardiography (TEE)</td>
</tr>
</tbody>
</table>

#### PACCD-2.1 Transthoracic Echocardiography (TTE)

- **CODING NOTES**: Transthoracic Echocardiography (TTE)
  - **2011 Transthoracic Echo CPT® Codes**:
    - CPT®93303 TTE for congenital cardiac anomalies, complete
    - CPT®93304 TTE for congenital cardiac anomalies, follow-up or limited
    - CPT®93306 TTE with 2-D, M-mode, Doppler and color flow, complete
    - CPT®93307 TTE with 2-D, M-mode, without Doppler or color flow
    - CPT®93308 TTE with 2-D, M-mode, follow-up or limited
  - **2011 Doppler Echocardiography CPT® Codes**:
    - CPT®93320 Doppler echo, pulsed wave and/or spectral display
    - CPT®93321 Doppler echo, pulsed wave and/or spectral display, follow-up or limited study
    - CPT®93325 Doppler echo, color flow velocity mapping
      - 93320 and 93321 should not be requested or billed together
  - **The most commonly performed congenital echo study is CPT®93303 plus CPT®93320 and CPT®93325**
  - **The most commonly performed non-congenital echo study is a complete transthoracic echocardiogram with spectral and color flow Doppler (CPT® 93306)**.
    - CPT®93306 includes the Doppler exams, so CPT® codes 93320-93325 should **not** be assigned together with CPT®93306.
  - For a complete transthoracic echocardiogram without Doppler, report CPT®93307.
  - By CPT® criteria, both of the complete transthoracic echo codes (93306 and 93307) require: “2-dimensional and, when performed, selected M-mode examination of the:
    - left and right atria,
    - left and right ventricles,
    - the aortic, mitral, and tricuspid valves,
    - the pericardium, and
    - adjacent portions of the aorta.”
    - If it is impossible to image all the listed structures, the report must indicate the reason.
A limited transthoracic echocardiogram should be billed if the report does not “evaluate or document the attempt to evaluate” all of the required structures listed above.

- A limited transthoracic echocardiogram is reported with code 93308.
- CPT®93321 (not CPT®93320) should be reported with 93308 if Doppler is included in the study. CPT®93325 can be reported with CPT®93308 if color flow Doppler is included in the study.

- A recent documented history consistent with a cardiac disorder, recent physical examination and/or relevant laboratory tests should be considered prior to considering the need for imaging.
- The need for repeat transthoracic echocardiography (TTE) is based upon findings in the original study and documentation of the way in which repeat studies will affect patient management.
- The following are indications for which transthoracic echocardiography (TTE) can be performed at least once:
  
  - **Valve function and structure including:**
    - Mitral valve prolapse
    - Mitral regurgitation
    - Mitral stenosis
    - Aortic regurgitation
    - Aortic stenosis
    - Bicuspid aortic valve
    - Tricuspid valve regurgitation
    - Pulmonary valve regurgitation

  - If valve surgery is being considered, TTE to assess aortic, pulmonary or mitral stenosis or regurgitation can be performed once or twice a year.
    - TTE can accurately assess the severity of valve stenosis but is sometimes less accurate in assessing valve regurgitation.

  - **Ventricular function** including global and segmental wall motion for evaluating ejection fraction (EF) and coronary artery disease.
    - Echo can be performed to evaluate cardiomyopathy due to etiologies such as ischemia, alcohol, viral myocarditis, or idiopathic.
    - Echo can be performed before and after chemotherapy known to affect heart function.

  - **Ventricular structure including:**
    - Infiltrative diseases (e.g. sarcoid, amyloid)
    - Aneurysm with/without thrombus
    - Ventricular septal defect (VSD)
    - Papillary muscle rupture/dysfunction
    - Hypertrophy (including asymmetric septal hypertrophy, spade heart, hypertensive concentric hypertrophy, infiltrative hypertrophy)

  - **Evaluate atrial or ventricular chamber size** (e.g. patients with atrial fibrillation, tachyarrhythmias, or left ventricular dilatation).
    - Yearly TTE may be indicated depending on the clinical circumstance.

  - **Detection of embolic source** in patients with recent Transient Ischemic Attack (TIA), stroke, or peripheral vascular emboli.
- Although transesophageal Echo (TEE) is more accurate in visualizing thrombus in the cardiac chambers and in visualizing the cardiac valves for vegetations (or classic mitral valve fibrinous excrescences), TTE is non-invasive and is indicated as the initial study.
- Intravenous injected sterile saline contrast can be performed for shunt detection in cases of known or suspected atrial and/or ventricular septal defect and/or patent foramen ovale.
  - This is best assessed using TEE, especially in patients with decompression illness, although TTE is still useful in this setting.
- **Evaluation of ASD repair or VSD repair:** within the first year of surgery, if stable clinically, routine imaging is not supported.
  - If patients become newly symptomatic more than one year after successful repair of congenital heart disease, TTE is appropriate
- **Tumor evaluation including myxomas**
- **Clot detection**
- **Evaluation of right ventricular systolic pressure and pulmonary hypertension**
- **Evaluation of pericardial effusion/pericardial disease including pericardial cysts** (cysts are usually benign, most frequently at the right cardiophrenic border, treated by observation [chest CT, cardiac MRI, or TTE] or drainage percutaneously or by open surgery), particularly suspected cardiac tamponade
- **Evaluation of congenital heart disease**
- **Evaluation of endocarditis**
  - TTE or TEE is appropriate when there is fever, positive blood cultures indicating bacteremia, or a new murmur.
  - **Note:** lack of visible vegetations does not eliminate the diagnosis.
  - **TEE** remains a more sensitive technique for identification of small vegetations.
- **Complications of pacemaker insertion** should be monitored by TTE
- **Coarctation of the Aorta**
  - Follow-up (surveillance) imaging after repair of coarctation.
  - **Infants and children:** echo every month for several months, then echo every 6 months to one year thereafter until age 18, then follow surveillance imaging frequency for adults—chest MRA (CPT®71555) every 2 to 3 years and before and after any intervention for re-coarctation.
- **Screening for first-degree relatives of patients with hypertrophic cardiomyopathy (HCM)**
  - First-degree relatives who are 12 to 18 years old should be screened yearly for HCM by 2D- echocardiography and ECG.
  - First-degree relatives who are older than age 18 should have 2D-echo and ECG every five years to screen for delayed adult-onset LVH.
  - Systematic screening is usually not indicated for first-degree relatives who are younger than age 12 unless there is a high-risk family history or the child is involved in particularly intense competitive sports.
Affected individuals identified through family screening or otherwise should be evaluated every 12 to 18 months with 2D-echo, Holter monitor, and blood pressure response during maximal upright exercise.

**Reference:**

**PACCD-2.2 Frequency of Echocardiography Testing**

- Repeat echocardiograms are no longer supported (annually or otherwise) for evaluation of clinically stable syndromes, including valvular heart disease, evaluation of prosthetic valve, cardiomyopathy, and hypertension.

- **Annual testing can be performed for the following when there is evidence for change in clinical status or to assess interventions (medical or surgical):**
  - Assessment of left ventricular hypertrophy progression or regression
  - Assessment of valve dysfunction
  - Assessment of cardiac chamber size in cardiomyopathy and atrial dysrhythmias
  - Assessment of chronic pericardial effusions
  - Assessment of left ventricular contractility/diastolic function prior to planned medical therapy for heart failure or to evaluate the effectiveness of on-going therapy
    - BNP levels are useful and may alone be sufficient for monitoring in many cases
  - Assessment of aortic dissection
  - Assessment of aortic root dilatation

- **Testing twice a year should not be routine but can be performed for the following:**
  - New or changing (not chronic stable) pericardial effusions
  - Assessment of new/changed medical therapy for congestive heart failure
    - BNP levels are useful and may alone be sufficient for monitoring in many cases
  - Assessment of new/changed medical therapy for hypertension if left ventricular hypertrophy was present
  - Assessment of hypertrophic cardiomyopathy when the results of the echo will potentially change patient management
  - Assessment of critical valvular heart disease when the results of the echo will potentially change patient management

- **New Echo can be performed for the following regardless of number of previous Echo studies:**
  - New cardiac murmurs
  - New myocardial infarction or acute coronary syndrome
  - New congestive heart failure (or new symptoms of dyspnea, orthopnea, paroxysmal nocturnal dyspnea, edema, elevated BNP)
  - New pericardial disease
  - New stroke/transient ischemic attack
  - New aortic dissection (TEE is preferred)
  - New decompression illness
- Re-evaluation of prosthetic valve with suspected dysfunction or thrombosis or a change in clinical status
  - Reference:  

**PACCD-2.3 Transesophageal Echocardiography (TEE)**

- **CODING NOTES:** Transesophageal Echocardiography (TEE)
  - **2011 Transesophageal Echo CPT® Codes:**
    - 93312 TEE with 2-D, M-mode, probe placement, image acquisition, interpretation and report
    - 93313 TEE probe placement only
    - 93314 TEE image acquisition, interpretation, and report only
    - 93315 TEE for congenital anomalies with 2-D, M-mode, probe placement, image acquisition, interpretation and report
    - 93316 TEE for congenital anomalies, probe placement only
    - 93317 TEE for congenital anomalies, image acquisition, interpretation and report only
    - 93318 TEE for monitoring purposes, ongoing assessment of cardiac pumping function on an immediate time basis
  - **2011 Doppler Echocardiography CPT® Codes:**
    - 93320 Doppler echo, pulsed wave and/or spectral display
    - 93321 Doppler echo, pulsed wave and/or spectral display, follow-up or limited study
    - 93325 Doppler echo, color flow velocity mapping
      - The Doppler echo codes, if performed, may be reported with TEE codes: 93312, 93314, 93315, and 93317.
  - For coding purposes, there are two parts to the TEE service:
    1. Placement of the transesophageal probe (transducer)
    2. Image acquisition, interpretation, and report
  - The complete transesophageal echocardiogram service, including both probe placement and image acquisition/interpretation, is reported with code 93312.
  - Probe placement only is reported with code 93313.
  - The image acquisition/interpretation only is reported with code 93314.
  - Physicians use codes 93312, 93313, and/or 93314 to report professional services if the test is performed in a hospital or other facility where the physician cannot bill globally.
    - Modifier -26 (professional component) is appended to the appropriate code
    - Codes 93313 and 93314 should never be billed together.
      - If both services are provided, code 93312 is reported.
  - Hospitals should report TEE procedures using code 93312 (the complete service).
    - Codes 93313 and 93314 are not used for hospital billing.
  - Transesophageal echo is frequently used to monitor patients undergoing cardiac surgery and is reported with code 93318.
The following are indications for which transesophageal echocardiography (TEE) can be performed at least once:

- Limited transthoracic echo window
- Detection of embolic source or intracardiac shunting when TTE is inconclusive
  - **Examples:** atrial septal defect, ventricular septal defect, patent foramen ovale, aortic cholesterol plaques, thrombus in cardiac chambers, valve vegetations, tumor
- Evaluation of embolic events when there is an abnormal TTE, abnormal ECG, and a history of atrial fibrillation
- Need to clarify pathology of the atria/atrial appendage, aorta, mitral/aortic valve beyond the information that other imaging studies have provided
- Evaluation of cardiac valve dysfunction
  - Differentiation of tricuspid from bicuspid aortic valve
  - Congenital abnormalities

The need for repeat TEE studies is based upon findings in the original study and documentation of the way in which repeat studies will affect patient management:

TEE is not particularly sensitive for left ventricular assessment since this chamber lies farther from the TEE probe than in transthoracic echo

- **Exceptions:** the base of the heart in evaluating asymmetric septal hypertrophy or membranous ventricular septal defect

**Reference:**

### PACCD-3~CARDIAC MRI

#### PACCD-3.1 General
- All requests for cardiac MRI should be sent for Medical Director review.
- MRI may be contraindicated due to metal implants, claustrophobia, BMI >40 kg/m²
  
  *Am J Cardiol 2009;104:1540-1546*

- MRA of the coronary arteries is not yet adequately sophisticated to replace coronary angiography in evaluating coronary disease and should not be authorized.
  - **EXCEPTIONS:** coronary artery anomalies (refer to CD-8.6 Other Indications for CCTA in the adult Cardiac Imaging Guidelines) and Kawasaki disease are conditions where MRA is considered useful.
    - MRA of the coronary arteries is reported with CPT®76498, unlisted magnetic resonance procedure (e.g., diagnostic, interventional).
    - All requests for unlisted studies should be sent for Medical Director review
    - Requests must be accompanied by detailed notes describing the procedure and indications.

- **NOTE:** Many patients with congenital heart disease are adequately evaluated using echocardiography. Cardiac MRI should be considered if a specific clinical question is left unanswered by another recent cardiac imaging study (usually echo) and the answer to the clinical question will affect management of the patient’s clinical condition.

#### PACCD-3.2 CODING NOTES: CARDIAC MRI
- 2011 Cardiac MRI CPT® codes:
  - **CPT®75557** Cardiac MRI for morphology and function without contrast
  - **CPT®75559** Cardiac MRI for morphology and function without contrast materials; with stress imaging
  - **CPT®75561** Cardiac MRI for morphology and function without contrast materials, followed by contrast material(s) and further sequences
  - **CPT®75563** Cardiac MRI for morphology and function without contrast materials, followed by contrast material(s) and further sequences; with
stress imaging

- **CPT® 75565** Cardiac MRI for velocity flow mapping
  - The add-on code, CPT® 75565, describes the cardiac MRI blood flow measurement procedure and, when performed, is to be reported in conjunction with CPT® 75557, CPT® 75559, CPT® 75561, or CPT® 75563.
  - *Per the AMA:* Only one procedure in the series 75557-75563 is appropriately reported per session and only one flow velocity measurement (CPT® 75565) may be reported per session.*

- **PACCD-3.3 Indications for Cardiac MRI**
  - Many patients with congenital heart disease are adequately evaluated using echocardiography. Cardiac MRI should be considered if a specific clinical question is left unanswered by another recent cardiac imaging study (usually echo) and the answer to the clinical question will affect management of the patient’s clinical condition.
  - **Indications for Cardiac MRI in the Congenital Heart Disease and/or Pediatric Population Include:**
    - **Congenital heart disease assessment** before and after invasive intervention (e.g. Tetralogy of Fallot, patent ductus arteriosus, platypnea, coarctation of the aorta, atrial septal defects, ventricular septal defects [VSD], pulmonary atresia with VSD, transposition of the great arteries, double outlet right ventricle, heterotaxy syndromes, anomalous pulmonary arteries or veins or anomalous coronary arteries, etc.
      - [See PACCD-4.2 Coronary Artery(ies) for criteria regarding anomalous coronary arteries]).
      - **Use one** of the following: CPT® 75557 or CPT® 75561.
      - CPT® 71555 (chest MRA) 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.
      - Chest MRA alone (CPT® 71555) should be performed if the patient cannot cooperate with full cardiac MRI exam.
      - Cardiac MRI (CPT® 75565 in conjunction with CPT® 75557 or CPT® 75561) can be used to evaluate for shunting through a VSD if a recent echo has been done, including a bubble study, and there is documented need to perform cardiac MRI in order to resolve an unanswered question.
    - **Coarctation of the aorta**
      - Follow-up (surveillance) imaging after repair of coarctation:
        - Adults (age 18 and older): chest MRA (CPT® 71555) every 2 to 3 years and before and after any intervention for re-coarctation
        - Infants and children (below 18 years old): echo every month for several months then echo every 6 months to one year thereafter.
      - Typical frequency of follow-up imaging for **Tetralogy of Fallot** is once a year. More frequent imaging may be necessary if clinical symptoms warrant or if imaging is needed following a new interventional
procedure.

- Clinical suspicion of arrhythmogenic right ventricular dysplasia or arrhythmogenic cardiomyopathy (ARVD/ARVC), especially if patient has presyncope or syncope if the clinical suspicion is supported by established criteria for ARVD (see PACCD-3 Evidence Based Clinical Support) or if the patient has documented Brugada’s syndrome. MRI (CPT®75557) is considered the optimal test for this disorder.*
  - If right ventricular abnormalities are already identified by echo or other techniques, MRI may not be necessary.
  - Young patients with right bundle branch block, unexplained syncope, and “normal” echo can undergo cardiac MRI (CPT®75557 or CPT®75561) to rule out ARVD/ARVC.
    * Circulation 2006;113:316-327
    * Eur Heart J 1989;10:127-132
    * Circulation 2005;112(25):3823-3832

- Pericardial disease: (constrictive versus restrictive pericarditis; perimyocarditis): Report CPT®75561. 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.

- Evaluate cardiac tumor or mass: (e.g. in sarcoidosis or tuberous sclerosis). Report CPT®75561

- Anomalous coronary arteries: Cardiac MRI (CPT®75561) or CCTA (CPT®75574) (which is still favored) is much better at detecting this than conventional angiography (see PACCD-4.2 Anomalous Coronary Artery(ies) for criteria regarding anomalous coronary arteries)

- Fabry's disease: late enhancement MRI may predict the effect of enzyme replacement therapy on myocardial changes that occur with this disease. Report CPT®75561

- Cardiomyopathy
  - Cardiac MRI can be performed to evaluate patients with congenital cardiomyopathy (muscular dystrophy, glycogen storage disease, fatty acid oxidation disorders, mitochondrial disorders, etc.)
  - Cardiac MRI can be performed in unexplained cases of cardiomyopathy in order to characterize the myocardium.

- Assessment of global ventricular function and mass if a specific clinical question is left unanswered by another recent cardiac imaging study (e.g. echo, etc.) and the answer to the clinical question will affect management of the patient’s clinical condition.

- Cardiac stress perfusion study: (see PACCD-3.2 CODING NOTES: Cardiac MRI) can be considered on a case by case basis for patients with anomalous coronary artery, Kawasaki disease, or other disorder with the potential for coronary ischemia.
• PACCD-3.4 Aortic Root and Proximal Ascending Aorta
  o The aortic root and proximal ascending aorta can be adequately evaluated during a cardiac MRI.
  o For screening due to family history of aortic aneurysm or dissection see CH-32 Thoracic Aorta in the adult Chest Imaging Guidelines.
  o If a patient (e.g. Marfan’s or Loeys-Dietz syndrome) with known ascending aortic aneurysm needs a cardiac MRI to evaluate another problem and the physician wishes to evaluate the ascending aorta, this evaluation should be included with the cardiac MRI interpretation. If the ascending aortic aneurysm is quite distal, near the arch, it is appropriate to include the chest MRI code (CPT®71551) or chest MRA code (CPT®71555).

• PACCD-3.5 Evaluation of Pericardial Effusion or Diagnosis of Pericardial Tamponade
  o Echocardiogram is the initial imaging study of choice to evaluate pericardial effusions or diagnose pericardial tamponade.
  o However, contrast enhanced cardiac MRI is useful for evaluating pericarditis, neoplastic effusion, tamponade or myocardial infiltration if a specific clinical question is left unanswered by another recent imaging study and the answer to the clinical question will affect management of the patient’s clinical condition.
  o Cancers that can metastasize to the pericardium or myocardium and can cause a malignant effusion include lung, breast, renal cell, lymphoma and melanoma.
PACCD-4 CT HEART & (CTCA)

4.1 GENERAL  14
4.2 Anomalous Coronary Arteries 14
4.3 Other Indications for Cardiac CT/ CTCA 15
4.4 Radiation Dose 16

PACCD-4~CT HEART and CORONARY COMPUTED TOMOGRAPHY ANGIOGRAPHY (CCTA)

- **PACCD-4.1 General**
  - Certain payers consider coronary calcium scoring and/or cardiac CT and Coronary Computed Tomography Angiography (CCTA) investigational, and their coverage policies will take precedence over MedSolutions’ guidelines.
  - Most payers require cardiac CT studies to be performed on a 64-slice CT scanner.
  - 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.
    - Multislice CT is associated with a non-negligible risk for cancer, especially in women and younger patients.*
      - *JAMA 2007;298:317-323
  - Contraindications to CCTA include:
    - Irregular heart rhythms (e.g. atrial fibrillation/flutter, frequent irregular premature ventricular contractions or premature atrial contractions, and high grade heart block)
    - Very obese patients (body mass index >40 kg/m²)
    - Elevated calcium score
      - CCTA should not be performed if there is extensive coronary calcification (calcium score >1000).
    - Renal insufficiency
    - Inability to follow breath holding instructions
    - Heart rate over 75 beats per minute
    - Allergy to iodine contrast material

- **PACCD-4.2 Anomalous Coronary Artery(ies)**
  - Evaluating coronary artery anomalies and other complex congenital heart disease of cardiac chambers or great vessels is an appropriate indication for CCTA.
    - Report CPT®75574 for evaluating coronary artery anomalies
    - Report CPT®75573 for congenital heart disease
      - Can add CPT®71275 (chest CTA) to evaluate great vessels
- In cases of anomalous pulmonary venous return, can add CT abdomen and pelvis.
  - The use of CCTA to rule out anomalous coronary artery(ies) should be limited to patients who need to have an anomalous coronary artery mapped prior to an invasive procedure, or who have not had a previous imaging study that clearly demonstrates an anomalous coronary artery and/or shows the anomalous artery to be patent and who are less than age 40 with a history that includes one or more of the following (aortic root echocardiography or cardiac MRI can also be considered to avoid radiation exposure):
    - 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 contraindications for conventional coronary angiography
    - Unexplained syncope (not presyncope)
      - Patients should have had a thorough negative evaluation for syncope as outlined in HD-31 Syncope in the adult Head Imaging Guidelines and PACCD-5 Syncope (e.g. echocardiogram, cardiac evaluation for postural blood pressure changes, resting low blood pressure, or low heart rate, myocardial perfusion imaging study, exercise treadmill test, stress echocardiogram, or stress MRI, consideration for situational syncope) prior to considering CCTA.
    - Unexplained new onset of heart failure (e.g. without atherosclerotic coronary artery disease or other known causes for cardiomyopathy) 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(ies).

**PACCD-4.3 Other Indications for Cardiac CT/ CCTA:**
- Congenital heart disease assessment (with procedures CPT®75573 or CPT®71275) is indicated in both children and adults for the following:
  - Determination of extra-cardiac anatomy in patients with complex congenital heart disease
    - For example: great vessel relationships, bronchial collateral vessels, abdominal situs, etc.
  - Pulmonary artery (PA) and Pulmonary vein assessment:
    - Pulmonary artery evaluation in children who need preoperative or postoperative evaluation for PA stenosis or PA atresia
    - PA caliber evaluation in children with pulmonary hypoplasia
    - PA evaluation to look for another anatomic structure impinging on the PA, or to look for airway/bronchial compromise by an enlarged PA or
other mediastinal vessel.

- Assessment of the course of drainage of pulmonary veins when chest x-ray suggests anomalous pulmonary venous drainage.
- Coarctation of the aorta or interruption of the aortic arch suspected on echocardiography.
- Evaluation of the arterial supply and venous drainage in children with bronchopulmonary sequestration.

- Vasculitis/Takayasu's/Kawasaki's disease can be imaged with CCTA (CPT®75574).
- Cardiac CT (CPT®75572) can be used to assess cardiac tumor or mass, pericardial mass, pericarditis/ constrictive pericarditis, complications of cardiac surgery, evaluation of post-operative anatomy and surgically corrected systemic-to-pulmonary artery shunts and intra-cardiac baffles, etc.
- Cardiac CT (CPT®75572) can be used to evaluate cardiac thrombus in patients with technically limited echocardiogram, MRI, or transesophageal echocardiogram.
- Cardiac CT (CPT®75572) can be used to evaluate clinical suspicion of arrhythmogenic right ventricular dysplasia or arrhythmogenic cardiomyopathy (ARVD/ARVC), especially if patient has presyncope or syncope if the clinical suspicion is supported by established criteria for ARVD (see PACCD-3 Evidence Based Clinical Support) or if the patient has documented Brugada’s syndrome.
- If right ventricular abnormalities are already identified by echo or other techniques, cardiac CT may not be necessary.
- Young patients with right bundle branch block, unexplained syncope, and “normal” echo can undergo cardiac MRI (CPT®75557 or CPT®75561) or cardiac CT (CPT®75572) to rule out ARVD/ARVC.
- Native aortic abnormalities can be investigated with cardiac CT (CPT®75572) if echocardiogram is indeterminate.

**PACCD-4.4 Radiation Dose**

- Radiation dosage for CCTA varies by facility and the particular protocol used. The American College of Radiology Clinical Statement on Noninvasive Cardiac Imaging states that “as a general rule a multi-detector CT scan encompassing the heart should not result in an effective dose of greater than 12 mSv.”

- 64-slice CT scanners can deliver a radiation does from 15-25 mSv (especially in women due to breast tissue density).
- Multislice CT is associated with a non-negligible risk for cancer, especially in women and younger patients.*

  *JAMA 2007; 298:317-323*

- Sophisticated gating and other techniques can reduce the radiation dose of cardiac CT studies to less than 5 mSv. Application of these techniques is increasing nationwide.*

  *J Am Coll Radiol 2005;2:471-477*

- Dual source scanners decrease radiation exposure by approximately one third.
Conventional coronary angiography typically delivers a radiation dose of 3 to 6 mSv.*

*J Am Coll Cardiol 2007;50(15):1469-1475

Newer imaging technologies will allow for reduced radiation exposure
2011 PEDIATRIC AND CONGENITAL CARDIAC IMAGING GUIDELINES

PACCD-5~SYNCOPE

- Also see HD-31 Syncope in the adult Head Imaging Guidelines
- **Evaluation of syncope:**
  - Echocardiogram should be performed initially to look for valvular or cardiomyopathic dysfunction.
  - Cardiac evaluation for postural blood pressure changes (which have the highest yield and affect management the most*), resting low blood pressure, low heart rate, or serious dysrhythmias should be performed prior to considering advanced imaging or stress testing.
    *Arch Intern Med 2009 July;169(14):1299-1305
- Stress testing should proceed based on the following guidelines in the adult Cardiac Imaging Guidelines:
  - **CD-1.3 Stress Testing**
  - **CD-2.4 Stress Echocardiography (Stress Echo)**
  - **CD-3.2 INDICATIONS FOR MPI**
  - **CD-6.3 Stress Perfusion MRI**
- Cardiac MRI (CPT®75561) or CCTA (see CD-8.10 CODING NOTES: CARDIAC CT and CCTA in the adult Cardiac Imaging Guidelines for procedure codes) can be considered if there is concern for anomalous coronary arteries, infiltrative heart disease or certain types of cardiomyopathy.
- Cardiac MRI (CPT®7557) can be performed to evaluate pre-syncope or syncope in patients with suspected ARVD/ARVC if the clinical suspicion is supported by established criteria for ARVD (see PACCD-3 Evidence Based Clinical Support) or if the patient has documented Brugada’s syndrome.
  - If right ventricular abnormalities are already identified by echo or other techniques, MRI may not be necessary.
  - Young patients with right bundle branch block, unexplained syncope, and “normal” echo can undergo cardiac MRI (CPT®75557 or CPT®75561) to rule out ARVD/ARVC.
- **Duchenne Muscular Dystrophy:** usually imaged by echocardiogram but evaluation for ischemic or cardiomyopathic changes using MPI or (typically) cardiac MRI (CPT®75557 or CPT®75561) can be performed

- **Family history**
  - Familial disease confirmed at necropsy or surgery (Major criterion)
  - Family history of premature sudden death (<35 years old) caused by suspected ARVD (Minor criterion)
  - Family history (clinical diagnosis based on present criteria) (Minor criterion)

- **ECG depolarization/conduction abnormalities**
  - Epsilon waves or localized prolongation (≥110 ms) of the QRS complex in the right precordial leads (V1-V3) (Major criterion)
  - Late potentials seen on signal averaged ECG (Minor criterion)

- **ECG repolarization abnormalities**
  - Inverted T waves in right precordial leads (V2 and V3) in patients aged >12 y and in the absence of right bundle branch block (Minor criterion)

- **Arrhythmias**
  - Sustained or nonsustained left bundle branch block type ventricular tachycardia documented on ECG or Holter monitoring, or during exercise testing (Minor criterion)
  - Frequent ventricular extrasystoles (>1000/24 h on Holter monitoring) (Minor criterion)

- **Global or regional dysfunction and structural alterations**
  - Severe dilatation and reduction of RV ejection fraction with no (or only mild) LV involvement (Major criterion)
  - Localized RV aneurysms (akinet ic or dyskinetic areas with diastolic bulging) (Major criterion)
  - Severe segmental dilatation of the right ventricle (Major criterion)
  - Mild global RV dilatation or ejection fraction reduction with normal left ventricle (Minor criterion)
  - Mild segmental dilatation of the right ventricle (Minor criterion)
  - Regional RV hypokinesia (Minor criterion)

- **Tissue characteristics of walls**
  - Fibrofatty replacement of myocardium on endomyocardial biopsy (Major criterion)

The diagnosis of ARVD/ARVC requires the presence of 2 major criteria or 1 major plus 2 minor or 4 minor criteria.

*The American Journal of Medicine 2008;121:674-681*
PACCD-2~Echocardiography (ECHO)
PACCD-2.1~Transthoracic Echocardiography (TTE)

PACCD-2.2~Frequency of Echocardiography Testing

PACCD-2.3~Transesophageal Echocardiography (TEE)

PACCD-3~Cardiac MRI

PACCD-4~CT of the Heart and CTCA

PACCD-5~Syncope

EVIDENCE BASED CLINICAL SUPPORT
PACCD-3~Cardiac MRI, Evidence Based Clinical Support
PEDIATRIC AND CONGENITAL PVD GUIDELINES

PACPVD-1~GENERAL GUIDELINES

- The Peripheral Vascular Disease Imaging Guidelines are the same for both the pediatric population and the adult population, unless there are specific guidelines listed here in the Pediatric and Congenital Peripheral Vascular Disease Imaging Guidelines.

PACPVD-2~AORTIC DISORDERS, RENAL VASCULAR DISORDERS, and VISCERAL ARTERY ANEURYSMS

- **Thoracic Aortic Disease**
  - Chest CT (CPT®71260 or CPT®71270), chest CTA (CPT®71275), or chest MRA (CPT®71555) can be used for surveillance or follow-up of thoracic aortic abnormalities in patients with Loeys-Deitz syndrome, Marfan’s syndrome, Takayasu’s arteritis, or Kawasaki syndrome.*
    

  - Less lethal disorders such as Turner syndrome and tuberous sclerosis have also been associated with aortic dissection.*
    
    *Clin Cardiol 2006;29:383-386

- **Aortic Congenital Vascular Malformations**
  - Aortic congenital vascular malformations can be seen with chromosomal abnormalities such as Turner syndrome.
  - Malformations can include aortic coarctation and aortic valve abnormalities.
  - Cardiac MRI (CPT®75557 or CPT®75561), chest MRA (CPT®71555), chest CT (CPT®71260), or chest CTA (CPT®71275) may be needed for evaluation.
  - Specialist input is helpful in determining the appropriate imaging pathway.
  - Coarctation is usually detected at younger ages with blood pressure substantially elevated in one or both upper extremities relative to lower extremity blood pressures. Plain chest x-ray in this syndrome may also demonstrate characteristic “rib notching.”

- **Visceral Artery Aneurysms**
  - These include arteries to the spleen, kidney, liver and intestines.
  - Aneurysm of these arteries is defined by an increase of more than 50% of the original arterial diameter.
  - Risk for rupture is high when the aneurysm is greater than 2 cm or is increasing rapidly* 
    
    *Circulation 2006;113:e463-e654

  - Vascular specialist consultation is beneficial in order to determine the time-frame to intervention.
  - Monitoring by ultrasound or CT with contrast is appropriate, although ultrasound should be attempted first.
  - Celiac artery aneurysm can be evaluated by CT abdomen with contrast (CPT®74160), CTA abdomen (CPT®74175), or ultrasound (CPT®76700 or CPT®76705).*
No definitive time period for serial studies has been established.
- Initial evaluation with six month follow-up is reasonable.
- Yearly follow-up in conjunction with vascular specialist consultation should be performed if no significant enlargement is seen.

**Follow-up imaging after stent placement**
- No definitive guidelines have been established for follow-up imaging, but it would be reasonable to follow the same time table as for endovascular aortic repair: CTA of abdomen (CPT®74175), MRA of abdomen (CPT®74185), or CT abdomen (CPT®74160) at 1 month, 6 months, and 12 months following stent placement, then every year. An additional study can be done at 3 months if there was evidence of endoleak on the 1 month study.

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**PEDIATRIC AND CONGENITAL PERIPHERAL VASCULAR DISEASE (PVD) IMAGING GUIDELINES REFERENCES**

PACPVD-2~Aortic Disorders, Renal Vascular Disorders, and Visceral Artery Aneurysms