

MedSolutions, Inc.
Clinical Decision Support Tool
Diagnostic Strategies

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 11/30/08

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PEDIATRIC AND CONGENITAL CARDIAC IMAGING GUIDELINES

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.
- Hybrid imaging (e.g SPECT/CT) which involve 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~CARDIAC MRI

- All requests for cardiac MRI should be sent for Medical Director review.
- 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 and Kawasaki disease are conditions where MRA (CPT 75561) is considered useful.
- **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-2.1 (See NOTE above) Indications for cardiac MRI in the 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-3.2 Coronary Artery(ies) for criteria regarding anomalous coronary arteries]).
 - Use one of the following: CPT 75557, 75561, 75558, or 75562.
 - 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 75560) 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.
 - 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-2 Evidence Based Clinical Support) or if the patient has documented Brugada's syndrome. MRI (CPT 75557) is considered the optimal test for this disorder.*

* *Circulation* 2006;113:316-327

* *Am J Med* 1994;97:78-88

* *Eur Heart J* 1989;10:127-132

* *Circulation* 2005;112(25):3823-3832

- Pericardial disease (constrictive versus restrictive pericarditis; perimyocarditis). Use CPT 75561.
 - Evaluate cardiac tumor or mass (e.g. in sarcoidosis or tuberous sclerosis). Use CPT 75561.
 - Anomalous coronary arteries: Cardiac MRI (CPT 75561) or CTCA (CPT 0146T) (which is still favored) is much better at detecting this than conventional angiography (see PACCD-3.2 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. Use 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 CD-6.1 Cardiac MRI Coding in the adult Cardiac guidelines) 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-2.2 The aortic root and proximal ascending aorta** can usually be adequately evaluated during a cardiac MRI, but in cases of bicuspid aortic valve and congenital heart disease, both cardiac MRI and chest MRA (CPT 71555) may be needed.
 - For screening due to family history of aortic aneurysm or dissection see CH-30 Thoracic Aortic Dissection or Aneurysm in the adult Chest guidelines.
 - 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-2.3 Echocardiogram is the initial imaging study of choice to evaluate pericardial effusions or diagnose pericardial tamponade.**
 - 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.
 - Cancers that can metastasize to the pericardium or myocardium and can cause a malignant effusion include lung, breast, renal cell, lymphoma and melanoma.

PACCD-3~CT OF THE HEART and COMPUTED TOMOGRAPHY CORONARY ANGIOGRAPHY (CTCA)

PACCD-3.1 General

- Certain payers consider coronary calcium scoring and/or cardiac CT and Computed Tomography Coronary Angiography (CTCA) investigational, and their coverage policies will take precedence over MedSolutions' guidelines. Prior authorization does not guarantee payment of the study.
- Most payers require cardiac CT studies to be performed on a 64-slice CT scanner.
- Cardiac testing that does not involve exposure to ionizing radiation should be strongly considered in individuals under age 40.

PACCD-3.2 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 CTCA.
 - Use CPT 0146T for evaluating coronary artery anomalies
 - Use CPT 0150T for congenital heart disease
 - Can add CPT 71275 (chest CTA) to evaluate great vessels
 - In cases of anomalous pulmonary venous return, can add CTV abdomen (CPT 74175).
 - The use of CTCA 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 (cardiac MRI can also be considered to avoid radiation exposure):
 - angina or myocardial infarction without high atherosclerosis risk
 - full sibling(s) with history of sudden death syndrome before age 30 or with documented anomalous coronary artery
 - resuscitated sudden death
 - unexplained syncope (not presyncope)
 - Patients should have had a thorough negative evaluation for syncope

as outlined in HD-32 Syncope in the adult Head Guidelines and PACCD-4 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, or stress echocardiogram, consideration for situational syncope) prior to considering CTCA.

- unexplained new onset of heart failure (e.g. without atherosclerotic coronary artery disease or other causes for cardiomyopathy)
- documented ventricular tachycardia (6 beat runs or greater)
- 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 CTCA to rule out anomalous coronary artery(ies).

PACCD-3.3 Other Indications for Cardiac CT/ CTCA:

- Congenital heart disease assessment using CPT 0150T or 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 CTCA (CPT 0148T).
- Cardiac CT (CPT 0145T) 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 0145T) can be used to evaluate cardiac thrombus in patients with technically limited echocardiogram, MRI, or transesophageal echocardiogram.
- Cardiac CT (CPT 0145T) 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-2 Evidence

- Based Clinical Support) or if the patient has documented Brugada's syndrome.
- Proximal native aortic abnormalities can be investigated with cardiac CT (CPT 0145T) if echocardiogram is indeterminate.

PACCD-3.4 Radiation Dose

- Radiation dosage for CTCA 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."^{*}
 - Current 16-slice CT scanners usually keep the radiation dose <13 mSv.
 - 64-slice CT scanners can deliver a radiation dose from 15-25 mSv (especially in women due to needing to penetrate breast tissue).
 - Dual source scanners decrease radiation exposure by approximately one third.
 - 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.
 - Conventional coronary angiography typically delivers a radiation dose of 3 to 6 mSv^{*}

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

PACCD-4~SYNCOPE

- Also see HD-32 Syncope in the adult Head guidelines
- Evaluation of syncope:**
 - Echocardiogram should be performed initially to look for valvular or cardiomyopathic dysfunction.
 - Cardiac evaluation for postural blood pressure changes, resting low blood pressure, low heart rate, or serious dysrhythmias should be performed prior to considering advanced imaging or stress testing.
- Stress testing should proceed based on CD-1.3 Stress Testing, CD-2.4 Stress Echocardiography (Stress Echo), and CD-3.2 Indications for MPI in the adult Cardiac guidelines.
- Cardiac MRI (CPT 75561) or CTCA (see CD-8.10 CPT Coding in the adult Cardiac guidelines for CPT codes) can be considered if there is concern for anomalous coronary arteries, infiltrative heart disease or certain types of cardiomyopathy.
- Cardiac MRI (CPT 75557) 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-2 Evidence Based Clinical Support) or if the patient has documented Brugada's syndrome.
- Duchenne muscular dystrophy:** usually imaged by echocardiogram but evaluation for ischemic or cardiomyopathic changes using MPI or (typically) cardiac MRI (CPT 75557 or 75561) can be performed

Evidence Based Clinical Support PACCD-2~CARDIAC MRI

- Proposed diagnostic criteria for Arrhythmogenic Right Ventricular Cardiomyopathy from the Task Force of the Working Group on Myocardial and Pericardial Disease of the European Society of Cardiology and of the Scientific Council on Cardiomyopathies of the International Society and Federation of Cardiology.*
 - 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 (akinetic 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

PEDIATRIC AND CONGENITAL CARDIAC IMAGING GUIDELINE REFERENCES

PACCD-2~Cardiac MRI

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PACCD-3~CT of the Heart and CTCA

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PACCD-2~Cardiac MRI, Evidence Based Clinical Support

- Ramaraj R, Sorrell VL, Marcus F, et al. Recently defined cardiomyopathies: A clinician's update. *The American Journal of Medicine* 2008;121:674-681.

PEDIATRIC AND CONGENITAL PERIPHERAL VASCULAR DISEASE (PVD) IMAGING 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), 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 syndrome, Takayasu disease, or Kawasaki syndrome.*
**N Engl J Med 2006 August;355:788-798*
 - 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** can be seen with chromosomal abnormalities such as Turner syndrome.
 - Malformations can include aortic coarctation and aortic valve abnormalities.
 - Cardiac MRI (CPT 75557 or 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*
** Visceral artery aneurysms. UC Davis Vascular Center.
<http://www.ucdmc.ucdavis.edu>. Accessed September 27, 2007*
 - 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.*
** Arch Surg 2002;137:670-674*

- 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.

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

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