

# Cigna Medical Coverage Policies – Radiology Cardiac Imaging Guidelines

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## Instructions for use

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

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

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

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

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

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

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# General Information

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# General Information

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## Abbreviations for the Cardiac Imaging Guidelines

<b>ACC</b>	American College of Cardiology
<b>ACS</b>	acute coronary syndrome
<b>AHA</b>	American Heart Association
<b>ASCOT</b>	Anglo-Scandinavian Cardiac Outcomes Trial
<b>ASD</b>	atrial septal defect
<b>BMI</b>	body mass index
<b>CABG</b>	coronary artery bypass grafting
<b>CAD</b>	coronary artery disease
<b>CHF</b>	congestive heart failure
<b>COPD</b>	chronic obstructive pulmonary disease
<b>CT</b>	computed tomography
<b>CCTA</b>	coronary computed tomography angiography
<b>CTA</b>	computed tomography angiography
<b>EBCT</b>	electron beam computed tomography
<b>ECP</b>	external counterpulsation (also known as EECP)
<b>ECG</b>	electrocardiogram
<b>ECP</b>	external counterpulsation
<b>ETT</b>	exercise treadmill stress test
<b>FDG</b>	Fluorodeoxyglucose, a radiopharmaceutical used to measure myocardial metabolism
<b>HCM</b>	hypertrophic cardiomyopathy
<b>IV</b>	intravenous
<b>LAD</b>	left anterior descending coronary artery
<b>LDL-C</b>	low density lipoprotein cholesterol

<b>LHC</b>	left heart catheterization
<b>LV</b>	left ventricle
<b>LVEF</b>	left ventricular ejection fraction
<b>MI</b>	myocardial infarction
<b>MPI</b>	myocardial perfusion imaging (SPECT study, nuclear cardiac study)
<b>MRA</b>	magnetic resonance angiography
<b>MRI</b>	magnetic resonance imaging
<b>mSv</b>	millisievert (a unit of radiation exposure) equal to an effective dose of a joule of energy per kilogram of recipient mass
<b>MUGA</b>	multi gated acquisition scan of the cardiac blood pool
<b>PCI</b>	percutaneous coronary intervention (includes percutaneous coronary angioplasty (PTCA) and coronary artery stenting)
<b>PET</b>	positron emission tomography
<b>PTCA</b>	percutaneous coronary angioplasty
<b>RHC</b>	right heart catheterization
<b>SPECT</b>	single photon emission computed tomography
<b>TEE</b>	transesophageal echocardiogram
<b>TIA</b>	Transient Ischemic Attack
<b>VSD</b>	ventricular septal defect

## Glossary

**Agatston Score:** a nationally recognized calcium score for the coronary arteries based on Hounsfield units and size (area) of the coronary calcium

**Angina:** principally chest discomfort, exertional (or with emotional stress) and relieved by rest or nitroglycerin

**Anginal variants or equivalents:** a manifestation of myocardial ischemia which is perceived by individuals to be (otherwise unexplained) dyspnea, unusual fatigue, more often seen in females and may be unassociated with chest pain

**ARVD/ARVC – Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy:** a potentially lethal inherited disease with syncope and rhythm disturbances, including sudden death, as presenting manifestations

**BNP:** B-type natriuretic peptide, blood test used to diagnose and track heart failure (n-T-pro-BNP is a variant of this test)

**Brugada Syndrome:** an electrocardiographic pattern that is unique and might be a marker for significant life-threatening dysrhythmias

**Double Product (Rate Pressure Product):** an index of cardiac oxygen consumption, is the systolic blood pressure times heart rate, generally calculated at peak exercise; over 25000 means an adequate stress load was performed

**Fabry's Disease:** an infiltrative cardiomyopathy, can cause heart failure and arrhythmias

**Hibernating myocardium:** viable but poorly functioning or non-functioning myocardium which likely could benefit from intervention to improve myocardial blood supply

**Optimized Medical Therapy** should include (where tolerated): antiplatelet agents, calcium channel antagonists, partial fatty acid oxidase inhibitors (e.g. ranolazine), statins, short-acting nitrates as needed, long-acting nitrates up to 6 months after an acute coronary syndrome episode, beta blocker drugs (optional), angiotensin-converting enzyme (ACE) inhibitors/angiotensin receptor blocking (ARB) agents (optional)

**Platypnea:** shortness of breath when upright or seated (the opposite of orthopnea) and can indicate cardiac malformations, shunt or tumor

**Silent ischemia:** cardiac ischemia discovered by testing only and not presenting as a syndrome or symptoms

**Syncope:** loss of consciousness; near-syncope is not syncope

**Takotsubo cardiomyopathy:** apical dyskinesia oftentimes associated with extreme stress and usually thought to be reversible

**Troponin:** a marker for ischemic injury, primarily cardiac



**Practice Estimate of Effective Radiation Dose chart for Selected Imaging Studies**

Imaging Study	Estimate of Effective Radiation Dose
Sestamibi myocardial perfusion study (MPI)	9-12 mSv
PET myocardial perfusion study:	3 mSv
Rubidium-82	2 mSv
NH3	
Thallium myocardial perfusion study (MPI)	22-31 mSv
Diagnostic conventional coronary angiogram (cath)	5-10 mSv
Computed tomography coronary angiography (CTCA)	5-15 mSv
(with prospective gating)	Less than 5 mSv
CT Abdomen and Pelvis	8-14 mSv
Chest x-ray	<0.1 mSv

# General Guidelines (CD-1.0)

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- A current clinical evaluation (within 60 days) is required prior to considering advanced imaging, which includes:
  - Relevant history and physical examination and appropriate laboratory studies and non-advanced imaging modalities, such as recent ECG (within 60 days), chest x-ray or ECHO/ultrasound, after symptoms started or worsened.
    - Effort should be made to obtain copies of reported “abnormal” ECG studies in order to determine whether the ECG is uninterpretable for ischemia on ETT
    - Most recent previous stress testing and its findings should be obtained
    - Other meaningful contact (telephone call, electronic mail or messaging) by an established individual can substitute for a face-to-face clinical evaluation.
  - Vital signs, height and weight, or BMI, or description of general habitus is needed.
  - Clinical question to be answered by advanced imaging that will affect management of the individual’s clinical condition.
- Cardiac imaging is not indicated if the results will not affect clinical management decisions. If a decision to perform cardiac catheterization or other angiography has already been made, there is often no need for imaging stress testing
- Assessment of ischemic symptoms can be determined by **Table-1**

## Clinical pre-test probability of CAD in individuals with stable chest pain symptoms

Clinical pre-test probability of CAD is a statistical tool used in the initial assessment of stable chest pain syndromes to estimate the likelihood that the symptoms are caused by obstructive coronary artery disease using the individual's description of the symptoms, their age, and sex assigned at birth. The pre-test probability for obstructive coronary artery disease as the cause of the symptoms is categorized as the following:

- **High** ≥85% pre-test probability
- **Intermediate/high** between 66-85% pre-test probability
- **Intermediate** between 15-65% pre-test probability
- **Low** <15% pre-test probability

Table - 1

Clinical pretest probability of CAD in individuals with stable chest pain symptoms				
Age (years)	Sex at birth	Type of symptoms		
		Cardiac (chest pain/pressure/tightness)	Possibly cardiac (including dyspnea/fatigue)	Non-cardiac/non-ischemic symptoms
30-39	Male	Intermediate	Intermediate	Intermediate
	Female	Intermediate	Low	Low
40-49	Male	Intermediate/High	Intermediate	Intermediate
	Female	Intermediate	Low	Low
50-59	Male	Intermediate/High	Intermediate/High	Intermediate
	Female	Intermediate	Intermediate	Low
60-69	Male	Intermediate/High	Intermediate/High	Intermediate
	Female	Intermediate/High	Intermediate/High	Intermediate
70-79	Male	High	Intermediate/High	Intermediate
	Female	Intermediate/High	Intermediate/High	Intermediate
>80	Male	High	Intermediate/High	Intermediate
	Female	Intermediate/High	Intermediate/High	Intermediate

- For purposes in this guideline ischemic symptoms can be defined as the following:
  - Cardiac chest pain/pressure/tightness** (typical angina):
    - Angina pectoris is classified as typical when all of the following are present
      - Retrosternal chest discomfort (generally described as pressure, heaviness, burning, constriction, squeezing, or tightness)
      - Brought on by exertion or emotional stress
      - Relieved by rest or nitroglycerin

- May radiate to the left arm or jaw
- When clinical information is received indicating that an individual is experiencing chest pain that is "exertional" or "due to emotional stress" and relieved with rest, this meets the cardiac chest pain (typical angina) definition under the Pre-Test Probability Grid. No further description of the chest pain is required (location within the chest is not required).
- The Clinical pretest probability of CAD ( *Table-1* ) is based on age, sex assigned at birth, and symptoms. All factors must be considered in order to approve for stress testing with imaging using the Pre-Test Probability Grid.
- **Possibly cardiac chest pain** (atypical angina):
  - Chest pain or discomfort (arm or jaw pain) that lacks one of the characteristics of cardiac chest pain.
  - DOE
- **Non-cardiac/non-ischemic chest pain:**
  - Chest pain or discomfort that meets one or none of the possibly cardiac characteristics.
- **Anginal equivalents**
  - Symptoms consistent with individual's known angina pattern in an individual with a history of CABG or PCI.
- Other signs and symptoms suggestive of potential cardiac etiology:
  - Dyspnea
  - Orthopnea
  - Paroxysmal nocturnal dyspnea
  - Heartburn unrelated to meals/nausea and vomiting
  - Palpitations
  - Syncope
  - Heart failure
- Chest pain remains the predominant symptom reported by females among those diagnosed with an acute coronary syndrome.
- For the purpose of this guideline, evidence documenting the presence of CAD includes any of the following:
  - Prior heart catheterization or CCTA revealing any of the following:
    - $\geq 40\%$  stenosis of the left main coronary artery
    - $\geq 50\%$  stenosis for other coronary arteries
    - Significant stenosis defined by an FFR of  $<0.80$
    - History of a prior PCI or CABG
- For the purpose of this guideline, evidence documenting the presence of non-obstructive CAD includes prior heart catheterization or CCTA revealing any of the following:
  - $<40\%$  stenosis of the left main coronary artery

- <50% stenosis for other coronary arteries
- FFR >0.8
- For the purposes of this guideline, evidence documenting a prior MI includes any of the following:
  - Presence of diagnostic Q waves on an ECG
  - A fixed perfusion defect on MPI
  - Akinetic or dyskinetic wall motion on echocardiogram
  - Area of Late Gadolinium Enhancement (LGE) on cardiac MRI suggesting scar
- Findings that may alter the ECG changes during exercise or are uninterpretable for ischemia on a stress test:
  - Complete Left Bundle Branch Block (bifascicular block involving right bundle branch and left anterior hemiblock does not render ECG uninterpretable for ischemia)
  - Ventricular paced rhythm
  - Pre-excitation pattern such as Wolff-Parkinson-White
  - Greater or equal to 1.0 mm ST segment depression (NOT nonspecific ST/T wave changes)
  - LVH with repolarization abnormalities, also called LVH with strain (NOT without repolarization abnormalities or by voltage criteria)
  - T wave inversion in the inferior and/or lateral leads. This includes leads II, AVF, V5 or V6. (T wave inversion isolated in lead III or T wave inversion in lead V1 and V2 are not included).
  - Individual on digitalis preparation

## References (CD-1)

1. Fihn SD, Gardin JM, Abrams J, et al. 2012 ACCF/AHA/ACP/AATS/PCNA/SCAI/STS Guideline for the Diagnosis and Management of Patients with Stable Ischemic Heart Disease: Executive Summary. *Circulation*. 2012;126(25):3097-3137. doi:10.1161/cir.0b013e3182776f83.
2. Qaseem A, Fihn SD, Williams S, et al. A. Diagnosis of Stable Ischemic Heart Disease: Summary of a Clinical Practice Guideline from the American College of Physicians/American College of Cardiology Foundation/American Heart Association/American Association for Thoracic Surgery/Preventive Cardiovascular Nurses Association/Society of Thoracic Surgeons. *Annals of Internal Medicine*. 2012;157(10):729. doi:10.7326/0003-4819-157-10-201211200-00010.
3. Rybicki FJ, Udelsom JE, Peacock WF, et al. 2015 ACR/ACC/AHA/AATS/ACEP/ASNC/NASCI/SAEM/SCCT/SCMR/SCPC/SNMMI/STR/STS Appropriate Utilization of Cardiovascular Imaging in Emergency Department Patients with Chest Pain. *J Am Coll Cardiol*. 2016;67(7):853-879. doi:10.1016/j.jacc.2015.09.011.
4. Fleisher LA, Fleischmann KE, Auerbach AD, et al. 2014 ACC/AHA Guideline on Perioperative Cardiovascular Evaluation and Management of Patients Undergoing Noncardiac Surgery. *J Am Coll Cardiol*. 2014;64(22):e77-e137. doi:10.1016/j.jacc.2014.07.944.
5. Ho PM, Rumsfeld JS, Peterson PN. Chest pain on exercise treadmill test predicts future cardiac hospitalizations. *Clin Cardiol* 2007; 30:505-510. doi:10.1002/clc.20139.
6. Sechtem U. Do heart transplant recipients need annual coronary angiography? *European Heart Journal* 2001; 22:895-897. doi:10.1053/euhj.2001.2660.
7. Tavel ME. Stress testing in cardiac evaluation: Current concepts with emphasis on the ECG. *Chest* 2001; 119:907-925. doi:10.1378/chest.119.3.907.
8. Wolk MJ, Bailey SR, Doherty JU, et al. ACCF/AHA/ASE/ASNC/HFSA/HRS/SCAI/SCCT/SCMR/STS. 2013 Multi-modality appropriate use criteria for the detection and risk assessment of stable ischemic heart disease: a report of the American College of Cardiology Foundation, Appropriate Use Criteria Task Force,

- American Heart Association, American Society of Echocardiography, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, Society for Cardiovascular Magnetic Resonance, and Society of Thoracic Surgeons. *J Am Coll Cardiol* 2014; 63: forthcoming. doi:10.1016/j.jacc.2013.11.009.
9. Blank P, Scheopf UJ, Leipsic JA. CT in transcatheter aortic valve replacement. *Radiology*, 2013; 269(3). doi:10.1148/radiol.13120696.
  10. Leipsic JA, Blanke P, Hanley M, et al. ACR Appropriateness Criteria® Imaging for Transcatheter Aortic Valve Replacement. *Journal of the American College of Radiology*. 2017;14(11). doi:10.1016/j.jacr.2017.08.046.
  11. Mieres JH, Gulati M, Bairey Merz N, et al. American Heart Association Cardiac Imaging Committee of the Council on Clinical Cardiology, Cardiovascular Imaging and Intervention Committee of the Council on Cardiovascular Radiology. Role of Noninvasive Testing in the Clinical Evaluation of Women with Suspected Ischemic Heart Disease A Consensus Statement from the American Heart Association. *Circulation*. 2014; 130(4):350. doi:10.1161/CIR.0000000000000061.
  12. American College of Cardiology Foundation Appropriate Use Criteria Task Force, American Society of Echocardiography, American Heart Association, et al. ACCF/AHA/ASA/ASNC/HFSA/HRS/SCAI/SCCM/SCCT/SCMR 2011 Appropriate Use Criteria for Echocardiography. A Report of the American College of Cardiology Foundation Appropriate Use Criteria Task Force, American Society of Echocardiography, American Heart Association, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, Society of Critical Care Medicine, Society of Cardiovascular Computed Tomography, and Society for Cardiovascular Magnetic Resonance Endorsed by the American College of Chest Physicians. *J Am Coll Cardiol* 2011; 57:1126. doi:10.1016/j.echo.2010.12.008.
  13. Melon CC, Eshtiahi P, Luksun WJ, et al. Validated questionnaire vs physicians' judgment to estimate preoperative exercise capacity. *JAMA Intern Med* 2014; 174:1507. doi:10.1001/jamainternmed.2014.2914.
  14. Taqueti V, Dorbala S, Wolinsky D. Myocardial perfusion imaging in women for the evaluation of stable ischemic heart disease— state-of-the-evidence and clinical recommendations. *Journal of Nuclear Cardiology*. June 2017. doi.org/10.1007/s12350-017-0926-8.
  15. Chamberlain JJ, Johnson EL, Leal S, et al. Cardiovascular Disease and Risk Management: Review of the American Diabetes Association Standards of Medical Care in Diabetes 2018. *Annals of Internal Medicine*. 2018;168(9):640. doi:10.7326/m18-0222.
  16. Bateman TM, Dilsizian V, Beanlands RS, Depuey EG, Heller GV, Wolinsky DA. American Society of Nuclear Cardiology and Society of Nuclear Medicine and Molecular Imaging Joint Position Statement on the Clinical Indications for Myocardial Perfusion PET. *Journal of Nuclear Medicine*. 2016;57(10):1654-1656. doi:10.2967/jnumed.116.180448.
  17. U.S. Food and Drug Administration. PROLEUKIN® (aldesleukin) for injection, for intravenous infusion. U.S. Food and Drug Administration Website. <https://www.accessdata.fda.gov>.
  18. Gulati M, Levy PD, Mukherjee D, et al. 2021 AHA/ACC/ASE/CHEST/SAEM/SCCT/SCMR Guideline for the Evaluation and Diagnosis of Chest Pain: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines [published correction appears in *Circulation*. 2021 Nov 30;144(22):e455]. *Circulation*. 2021;144(22):e368-e454. doi:10.1161/CIR.0000000000001029.
  19. Kidney Disease: Improving Global Outcomes (KDIGO) Kidney Transplant Candidate Work Group. KDIGO Clinical Practice Guideline on the Evaluation and Management of Candidates for Kidney Transplantation. Transplantation. 2020;104: S1 – S103.
  20. Wenger NK, Lloyd-Jones DM, Elkind MSV, et al. Call to Action for Cardiovascular Disease in Women: Epidemiology, Awareness, Access, and Delivery of Equitable Health Care: A Presidential Advisory From the American Heart Association. *Circulation*. 2022;145(23):e1059-e1071. doi:10.1161/CIR.0000000000001071

# Stress Testing

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# Stress Testing without Imaging – Procedures (CD-1.2)

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**The Exercise Treadmill Test (ETT) is without imaging.**

- Necessary components of an ETT include:
  - ECG that can be interpreted for ischemia.
  - Individual capable of exercise to achieve target heart rate on a treadmill or similar device (5 METs or greater; see functional capacity below). Target heart rate is calculated as 85% of the maximum age predicted heart rate (MPHR). MPHR is estimated as 220 minus the individual's age.
- An abnormal ETT (exercise treadmill test) includes any one of the following:
  - ST segment depression (usually described as horizontal or downsloping,  $\geq 1.0$  mm below baseline)
  - Development of chest pain
  - Significant arrhythmia (especially ventricular arrhythmia)
  - Hypotension during exercise
- Functional capacity  $\geq 5$  METs equates to the following:
  - Can walk four blocks without stopping
  - Can walk up a hill
  - Can climb one flight of stairs without stopping
  - Can perform heavy work around the house
  - Can walk 4mph at a brisk pace

***Background and supporting information***

An observational study found that, compared with the Duke Activity Status Index, subjective assessment by clinicians generally underestimated exercise capacity



# Stress Testing with Imaging – Procedures (CD-1.3)

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- Imaging Stress Tests include any one of the following:
  - Stress Echocardiography see **Stress Echocardiography (Stress Echo) – Coding (CD-2.7)**
  - SPECT MPI see **Myocardial Perfusion Imaging (MPI) – Coding (CD-3.1)**
  - Stress perfusion MRI see **Cardiac MRI – Indications for Stress MRI (CD-5.3)**
  - PET Perfusion see **Cardiac PET-Perfusion-Indications (CD-6.2)**
- Stress testing with imaging can be performed with maximal exercise or chemical stress (adenosine, dipyridamole, dobutamine, or regadenoson) and does not alter the CPT® codes used to report these studies.

# Stress Testing with Imaging – Indications (CD-1.4)

CD.ST.0001.4.A

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- Stress echo, SPECT MPI or stress MRI, can be considered if there are **new, recurrent, or worsening** symptoms consistent with ischemia and **any** of the following:
  - Intermediate-high or High pretest probability (>66% probability of CAD) per **Table-1**
  - A history of obstructive CAD as defined in **General Guidelines (CD-1.0)**
  - Evidence of ventricular tachycardia
  - High suspicion of ventricular tachycardia such as unheralded syncope (not near syncope)
  - Age 40 years or greater and known diabetes mellitus
  - Coronary calcium score  $\geq 100$
  - Poorly controlled hypertension defined as systolic BP  $\geq 180$ mmHg, if provider feels strongly that CAD needs evaluation prior to BP being controlled.
  - ECG is uninterpretable for ischemia due to any one of the following:
    - Complete Left Bundle Branch Block (bifascicular block involving right bundle branch and left anterior hemiblock does not render ECG uninterpretable for ischemia)
    - Ventricular paced rhythm
    - Pre-excitation pattern such as Wolff-Parkinson-White
    - Greater or equal to 1.0 mm ST segment depression (NOT nonspecific ST/T wave changes)
    - LVH with repolarization abnormalities, also called LVH with strain (NOT without repolarization abnormalities or by voltage criteria)
    - T wave inversion in the inferior and/or lateral leads. This includes leads II, AVF, V5 or V6. (T wave inversion isolated in lead III or T wave inversion in lead V1 and V2 are not included).
    - Individual on digitalis preparation
  - Continuing symptoms in an individual who had a normal or submaximal exercise treadmill test and there is suspicion of a false negative result.
  - Individuals with recent equivocal, borderline, or abnormal stress testing where ischemia remains a concern. See **Stress Testing without Imaging – Procedures (CD-1.2)**.
  - Heart rate <50 bpm in individuals, including those on beta blocker, calcium channel blocker, or amiodarone, where it is felt that the individual may not achieve an adequate workload for a diagnostic exercise study.

- Inability to safely use a treadmill or exercise bicycle, for example, the need for ambulatory assistance (wheelchair, cane, walker, etc.) or significant neurologic or orthopedic issue
- ETT inadequate due to one of the following:
  - Physical (musculoskeletal or neurological) inability to achieve target heart rate- 85% MPHR (220-age). See **Stress Testing without Imaging – Procedures (CD-1.2)** for necessary components for ETT.
  - History of false positive exercise treadmill test: a false positive ETT is one that is abnormal however the abnormality does not appear to be due to macrovascular CAD.
- Stress echo, SPECT MPI or stress MRI, can be considered regardless of symptoms for **any** of the following:
  - One imaging stress test can be performed within 3 months of an acute coronary syndrome (e.g. ST segment elevation MI [STEMI], unstable angina, non-ST segment elevation MI [NSTEMI]), to evaluate for inducible ischemia if ALL of the following related to the most recent acute coronary event apply:
    - Individual is hemodynamically stable
    - No recurrent chest pain symptoms and no signs of heart failure
    - No prior coronary angiography or imaging stress test since the current episode of symptoms
  - Assessing myocardial viability in individuals with significant ischemic ventricular dysfunction (suspected hibernating myocardium) and persistent symptoms or heart failure such that revascularization would be considered.
    - MRI, cardiac PET, SPECT MPI, or Dobutamine stress echo can be used to assess myocardial viability depending on physician preference.
    - See **Cardiac PET – Metabolic – Indications (CD-6.4)**.
  - Asymptomatic individual with an uninterpretable ECG as described in **General Guidelines (CD 1.0)** that either:
    - Has never been evaluated
    - Is a new uninterpretable change
  - Individual with an elevated cardiac troponin.
  - One routine study 2 years or more after a stent
    - Except with a left main stent where it can be done at 1 year.
  - One routine study at 5 years or more after CABG, without cardiac symptoms.
  - Every 2 years if there was documentation of previous “silent ischemia” on the imaging portion of a stress test but not on the ECG portion.
  - To assess for CAD prior to starting a Class IC antiarrhythmic agent (flecainide or propafenone) and annually while taking the medication.
  - Prior to starting Interleukin-2
  - Prior anatomic imaging study (coronary angiogram or CCTA) demonstrating coronary stenosis in the proximal or mid-portion of a major coronary branch,

which is of uncertain functional significance, can have one stress test with imaging.

- Asymptomatic individual who has an ischemic EKG response on ETT (horizontal or downsloping ST depression  $\geq 1.0$  mm below baseline).
- Evaluating new, recurrent, or worsening left ventricular systolic dysfunction
- Cardiac perfusion PET (CPT® 78430, 78431, 78491, 78492) can be considered in place of stress echo, SPECT MPI, or stress MRI when any of the above indications for stress testing with imaging have been met and there is documentation of one of the following:
  - Individual is severely obese (for example BMI  $\geq 40$  kg/m<sup>2</sup>) or
  - Individual has large breasts or implants
  - Individual incapable of exercise due to physical (musculoskeletal or neurological) inability to achieve target heart rate. Target heart rate is calculated as 85% of the maximum age predicted heart rate (MPHR). MPHR is estimated as 220 minus the individual's age
- See **Cardiac PET – Perfusion – Indications (CD-6.2)** for additional indications for cardiac PET perfusion

# Stress Testing with Imaging – Preoperative (CD-1.5)

CD.ST.0001.5.A

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- There are **2** steps that determine the need for imaging stress testing in (stable) pre-operative individuals:
  - Step1: Would the individual qualify for imaging stress testing independent of planned surgery?
    - If yes, proceed to stress testing guidelines **Stress Testing with Imaging – Indications (CD-1.4)**
    - If no, go to step 2
  - Step 2: Is the surgery considered high, moderate or low risk? (see **Table-2**) If high or moderate-risk, proceed below. If low-risk, there is no evidence to determine a need for preoperative cardiac testing.
    - **High Risk Surgery:** All individuals in this category should receive an imaging stress test if there has not been an imaging stress test within 1 year\* unless the individual has developed new cardiac symptoms or a new change in the EKG since the last stress test.
    - **Intermediate Surgery:** One or more risk factors and unable to perform an ETT per guidelines if there has not been an imaging stress test within 1 year\* unless the individual has developed new cardiac symptoms or a new change in the EKG since the last stress test.
    - **Low Risk:**Preoperative imaging stress testing is not supported.
  - Clinical Risk Factors (for cardiac death & non-fatal MI at time of non-cardiac surgery)
    - Planned high-risk surgery (open surgery on the aorta or open peripheral vascular surgery)
    - History of ischemic heart disease (previous MI, previous positive stress test, use of nitroglycerin, typical angina, ECG Q waves, previous PCI or CABG)
    - History of compensated previous congestive heart failure (history of heart failure, previous pulmonary edema, third heart sound, bilateral rales, chest x-ray showing heart failure)
    - History of previous TIA or stroke
    - Diabetes Mellitus
    - Creatinine level > 2 mg/dL

\*Time interval is based on consensus of eviCore executive cardiology panel.

**Table-2**

Cardiac Risk Stratification List		
High Risk (> 5%)	Intermediate Risk (1-5%)	Low Risk (<1%)
<ul style="list-style-type: none"><li>• Open aortic and other major open vascular surgery</li><li>• Open peripheral vascular surgery</li></ul>	<ul style="list-style-type: none"><li>• Open intraperitoneal and/or intrathoracic surgery</li><li>• Open carotid endarterectomy</li><li>• Head and neck surgery</li><li>• Open orthopedic surgery</li><li>• Open prostate surgery</li></ul>	<ul style="list-style-type: none"><li>• Endoscopic procedures</li><li>• Superficial procedures</li><li>• Cataract surgery</li><li>• Breast surgery</li><li>• Ambulatory surgery</li><li>• Laparoscopic and endovascular procedures that are unlikely to require further extensive surgical intervention</li></ul>

# Transplant (CD-1.6)

CD.ST.0001.6.A

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- Stress Testing in individuals for Non-Cardiac Transplant
  - Individuals who are candidates for any type of organ, bone marrow, or stem cell transplant can undergo imaging stress testing every year (stress echo, SPECT MPI, stress MRI, or stress cardiac PET perfusion per the transplant center's protocol) prior to transplant. See **Kidney Transplant, Pre-Transplant Imaging Studies (AB-42.5)**.
  - Individuals who have undergone organ transplant are at increased risk for ischemic heart disease secondary to their medication. Risk of vasculopathy is 7% at one year, 32% at five years and 53% at ten years. An imaging stress test can be repeated annually after transplant for at least two years or within one year of a prior cardiac imaging study if there is evidence of progressive vasculopathy.
  - After two consecutive normal imaging stress tests, repeated testing is not supported more often than every other year without evidence for progressive vasculopathy or new symptoms.
  - Stress testing after five years may proceed according to normal patterns of consideration.
- Post-Cardiac transplant assessment of transplant CAD:
  - One of the following imaging studies may be performed annually:
    - SPECT MPI
    - Stress ECHO
    - Stress MRI
    - Cardiac PET perfusion

# References (CD-1)

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1. Fihn SD, Gardin JM, Abrams J, et al. 2012 ACCF/AHA/ACP/AATS/PCNA/SCAI/STS Guideline for the Diagnosis and Management of Patients with Stable Ischemic Heart Disease: Executive Summary. *Circulation*. 2012;126(25):3097-3137. doi:10.1161/cir.0b013e3182776f83.
2. Qaseem A, Fihn SD, Williams S, et al. A. Diagnosis of Stable Ischemic Heart Disease: Summary of a Clinical Practice Guideline from the American College of Physicians/American College of Cardiology Foundation/American Heart Association/American Association for Thoracic Surgery/Preventive Cardiovascular Nurses Association/Society of Thoracic Surgeons. *Annals of Internal Medicine*. 2012;157(10):729. doi:10.7326/0003-4819-157-10-201211200-00010.
3. Rybicki FJ, Udelson JE, Peacock WF, et al. 2015 ACR/ACC/AHA/AATS/ACEP/ASNC/NASCI/SAEM/SCCT/SCMR/SCPC/SNMMI/STR/STS Appropriate Utilization of Cardiovascular Imaging in Emergency Department Patients with Chest Pain. *J Am Coll Cardiol*. 2016;67(7):853-879. doi:10.1016/j.jacc.2015.09.011.
4. Fleisher LA, Fleischmann KE, Auerbach AD, et al. 2014 ACC/AHA Guideline on Perioperative Cardiovascular Evaluation and Management of Patients Undergoing Noncardiac Surgery. *J Am Coll Cardiol*. 2014;64(22):e77-e137. doi:10.1016/j.jacc.2014.07.944.
5. Ho PM, Rumsfeld JS, Peterson PN. Chest pain on exercise treadmill test predicts future cardiac hospitalizations. *Clin Cardiol* 2007; 30:505-510. doi:10.1002/clc.20139.
6. Sechtem U. Do heart transplant recipients need annual coronary angiography? *European Heart Journal* 2001; 22:895–897. doi:10.1053/ehj.2001.2660.
7. Tavel ME. Stress testing in cardiac evaluation: Current concepts with emphasis on the ECG. *Chest* 2001; 119:907-925. doi:10.1378/chest.119.3.907.
8. Wolk MJ, Bailey SR, Doherty JU, et al. ACCF/AHA/ASE/ASNC/HFSA/HRS/SCAI/SCCT/SCMR/STS. 2013 Multi-modality appropriate use criteria for the detection and risk assessment of stable ischemic heart disease: a report of the American College of Cardiology Foundation, Appropriate Use Criteria Task Force, American Heart Association, American Society of Echocardiography, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, Society for Cardiovascular Magnetic Resonance, and Society of Thoracic Surgeons. *J Am Coll Cardiol* 2014; 63: forthcoming. doi:10.1016/j.jacc.2013.11.009.
9. Blank P, Scheopf UJ, Leipsic JA. CT in transcatheter aortic valve replacement. *Radiology*, 2013; 269(3). doi:10.1148/radiol.13120696.
10. Leipsic JA, Blanke P, Hanley M, et al. ACR Appropriateness Criteria® Imaging for Transcatheter Aortic Valve Replacement. *Journal of the American College of Radiology*. 2017;14(11). doi:10.1016/j.jacr.2017.08.046.
11. Mieres JH, Gulati M, Bairey Merz N, et al. American Heart Association Cardiac Imaging Committee of the Council on Clinical Cardiology, Cardiovascular Imaging and Intervention Committee of the Council on Cardiovascular Radiology. Role of Noninvasive Testing in the Clinical Evaluation of Women with Suspected Ischemic Heart Disease A Consensus Statement from the American Heart Association. *Circulation*. 2014; 130(4):350. doi:10.1161/CIR.0000000000000061.
12. American College of Cardiology Foundation Appropriate Use Criteria Task Force, American Society of Echocardiography, American Heart Association, et al. ACCF/ASE/AHA/ASNC/HFSA/HRS/SCAI/SCCM/SCCT/SCMR 2011 Appropriate Use Criteria for Echocardiography. A Report of the American College of Cardiology Foundation Appropriate Use Criteria Task Force, American Society of Echocardiography, American Heart Association, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, Society of Critical Care Medicine, Society of Cardiovascular Computed Tomography, and Society for Cardiovascular Magnetic Resonance Endorsed by the American College of Chest Physicians. *J Am Coll Cardiol* 2011; 57:1126. doi:10.1016/j.echo.2010.12.008.
13. Melon CC, Eshtiahi P, Luksun WJ, et al. Validated questionnaire vs physicians' judgment to estimate preoperative exercise capacity. *JAMA Intern Med* 2014; 174:1507. doi:10.1001/jamainternmed.2014.2914.
14. Taqueti V, Dorbala S, Wolinsky D. Myocardial perfusion imaging in women for the evaluation of stable ischemic heart disease—state-of-the-evidence and clinical recommendations. *Journal of Nuclear Cardiology*. June 2017. doi.org/10.1007/s12350-017-0926-8.
15. Chamberlain JJ, Johnson EL, Leal S, et al. Cardiovascular Disease and Risk Management: Review of the American Diabetes Association Standards of Medical Care in Diabetes 2018. *Annals of Internal Medicine*. 2018;168(9):640. doi:10.7326/m18-0222.
16. Bateman TM, Dilsizian V, Beanlands RS, Depuey EG, Heller GV, Wolinsky DA. American Society of Nuclear Cardiology and Society of Nuclear Medicine and Molecular Imaging Joint Position Statement on



- the Clinical Indications for Myocardial Perfusion PET. *Journal of Nuclear Medicine*. 2016;57(10):1654-1656. doi:10.2967/jnumed.116.180448.
17. U.S. Food and Drug Administration. PROLEUKIN® (aldesleukin) for injection, for intravenous infusion. U.S. Food and Drug Administration Website. <https://www.accessdata.fda.gov>.
  18. Gulati M, Levy PD, Mukherjee D, et al. 2021 AHA/ACC/ASE/CHEST/SAEM/SCCT/SCMR Guideline for the Evaluation and Diagnosis of Chest Pain: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines [published correction appears in *Circulation*. 2021 Nov 30;144(22):e455]. *Circulation*. 2021;144(22):e368-e454. doi:10.1161/CIR.0000000000001029.
  19. Kidney Disease: Improving Global Outcomes (KDIGO) Kidney Transplant Candidate Work Group. KDIGO Clinical Practice Guideline on the Evaluation and Management of Candidates for Kidney Transplantation. 2020;104: S1 – S103.
  20. Wenger NK, Lloyd-Jones DM, Elkind MSV, et al. Call to Action for Cardiovascular Disease in Women: Epidemiology, Awareness, Access, and Delivery of Equitable Health Care: A Presidential Advisory From the American Heart Association. *Circulation*. 2022;145(23):e1059-e1071. doi:10.1161/CIR.0000000000001071

# Echocardiography (ECHO)

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# Transthoracic Echocardiogram (TTE) - Coding (CD-2.1)

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## Transthoracic Echocardiography (TTE) - Coding

### Transthoracic Echocardiography

Description	CPT®
TTE for congenital cardiac anomalies, complete	93303
TTE for congenital cardiac anomalies, follow-up or limited	93304
TTE with 2-D, M-mode, Doppler and color flow, complete	93306
TTE with 2-D, M-mode, without Doppler or color flow	93307
TTE with 2-D, M-mode, follow-up or limited	93308

### 3D Echocardiography

Description	CPT®
3D echocardiographic imaging and postprocessing during transesophageal echocardiography, or during transthoracic echocardiography for congenital cardiac anomalies, for the assessment of cardiac structure(s) (eg, cardiac chambers and valves, left atrial appendage, interatrial septum, interventricular septum) and function, when performed (List separately in addition to code for echocardiographic imaging) Code with (93303-93304, 93312, 93314, 93315, 93317, 93350-93351)	+93319

### Doppler Echocardiography

Description	CPT®
Doppler echo, pulsed wave and/or spectral display	+93320
Doppler echo, pulsed wave and/or spectral display, follow-up or limited study	+93321

Description	CPT®
Doppler echo, color flow velocity mapping	+93325
CPT® 93320 and CPT® 93321 should not be requested or billed together	

C codes are unique temporary codes established by CMS. C codes were established for contrast echocardiography. Each echocardiography C code corresponds to a standard echo code (Class I CPT® code) The C code and the matching CPT code should not both be approved.

C Code	Transthoracic Echocardiography	CPT®
C8921	TTE for congenital cardiac anomalies, complete	93303
C8922	TTE for congenital cardiac anomalies, follow-up or limited	93304
C8929	TTE with 2-D, M-mode, Doppler and color flow, complete	93306
C8923	TTE with 2-D, M-mode, without Doppler or color flow	93307
C8924	TTE with 2-D, M-mode, follow-up or limited	93308

#### Myocardial strain imaging

Description	CPT®
Myocardial strain imaging using speckle tracking-derived assessment of myocardial mechanics (List separately in addition to codes for echocardiography imaging)	+93356

#### Investigational codes

Description	CPT®
Myocardial contrast perfusion echocardiography, at rest or with stress, for assessment of myocardial ischemia or viability	0439T

#### Transthoracic Echocardiography (TTE) – Coding - General Information (CD-2.1.1)

- Complete transthoracic echocardiogram with spectral and color flow Doppler (CPT® 93306).
  - 93306 includes the Doppler exams, so CPT® codes 93320-93325 should not be assigned together with CPT® 93306.
  - Doppler codes (CPT® 93320, CPT® 93321, and CPT® 93325) are 'add-on codes' (as denoted by the + sign) and are assigned in addition to code for the primary procedure.
- For a 2D transthoracic echocardiogram without Doppler, report CPT® 93307.
- Limited transthoracic echocardiogram should be billed if the report does not "evaluate or document the attempt to evaluate" all of the required structures.

- A limited transthoracic echocardiogram is reported with CPT® 93308.
- CPT® 93321 (not CPT® 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 limited congenital transthoracic echocardiogram is reported with CPT® 93304.
- Doppler echo may be used for evaluation of the following:
  - Shortness of breath
  - Known or suspected valvular disease
  - Known or suspected hypertrophic obstructive cardiomyopathy
  - Shunt detection

### ***Background and Supporting Information***

- Providers performing echo on a pediatric individual, may not know what procedure codes they will be reporting until the initial study is completed.
- If a congenital issue 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 all 4 codes (CPT® 93303, CPT® 93320, CPT® 93325, and CPT® 93306).
- CPT® 76376 and CPT® 76377 are not unique to 3D Echo. These codes also apply to 3D rendering of MRI and CT studies, see **3D Echocardiography – Coding (CD-2.9)**
- CPT® 93325 may also be used with fetal echocardiography
- CPT® 93319 3D echo imaging postprocessing of TEE or TTE to evaluate congenital cardiac abnormalities. see **3D Echocardiography – Coding (CD-2.9)**

### **Myocardial contrast perfusion echocardiography (CPT® 0439T) (CD-2.11)**

- Investigational see **Transthoracic Echocardiography (TTE) – Coding (CD-2.1)**

# Transthoracic Echocardiography (TTE) – Indications/initial evaluation (CD-2.2)

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## Asymptomatic Individuals

- TTE can be approved for screening of an individual when there is documentation of any of the following:
  - First-degree relative with an inherited cardiomyopathy-an initial screening echocardiogram can be approved at the time an inherited cardiomyopathy is diagnosed in a first degree relative
  - First-degree relative with bicuspid aortic valve
  - First degree relative with known thoracic aortic aneurysm or dissection (may repeat every two years if negative). See **Thoracic Aortic Aneurysm (PVD-6.2)**, **Aortic Dissection and Other Aortic Conditions (PVD-6.7)**, **Screening for TAA in individuals with bicuspid aortic valves (PVD-2.3)** in the Peripheral Vascular Disease Imaging Guideline
  - Pulmonary hypertension see **Pulmonary Hypertension (PH) (CD-8.1)**
- TTE can be approved for the initial evaluation of an individual for any of the following documented conditions:
  - Known or suspected connective tissue disease or a genetic condition that predisposes to an aortic aneurysm or dissection to evaluate the ascending aorta (may repeat every two years if negative). See **Screening for Vascular related genetic connective tissue Disorders (PVD-2.2)** in the Peripheral Vascular Disease imaging guidelines
  - Genotype positive individual with inherited cardiomyopathy including any of the following:
    - HCM
    - Non compaction cardiomyopathy
    - Familial Dilated Cardiomyopathy
    - Arrhythmogenic Cardiomyopathy (e.g., ARVC)
  - Prior to solid organ transplant or hematopoietic stem cell transplant (HSCT)
  - Prior to exposure to medications or radiation that could result in cardiotoxicity/heart failure. See **Cardiotoxic agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1)**
  - Suspected pulmonary hypertension (PH) in an individual with documented high risk for developing PH including any of the following conditions:

- Scleroderma
- Lupus
- Mixed connective tissue disease

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**Note** See **Pulmonary Hypertension (PH) (CD-8.1)**

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- Cardiac mass, suspected tumor, or thrombus seen on other imaging (i.e., CT Chest, MRI Chest, CXR) when further assessment is needed for alteration in treatment or therapy
- Newly diagnosed or strongly suspected cerebral ischemia or peripheral embolic event- initial evaluation
- Suspected cardiac injury due to blunt chest trauma
- Post myocardial infarction (MI) can be approved once in follow-up  $\geq 6$  weeks after the MI
- Suspected hypertensive heart disease (initial evaluation)
- Evaluation of adult congenital heart disease see **Adult Congenital Heart Disease (CD-11)** and **Congenital Heart Disease (PEDCD-2)** in the Pediatric Cardiology imaging guidelines

### Symptomatic Individuals

- TTE can be approved to evaluate an individual when there is documentation of any of the following new or worsening clinical signs and symptoms of heart disease:
  - Chest pain
  - New or changing heart murmur
  - Newly diagnosed RBBB or LBBB
  - Frequent VPCs without other evidence of heart disease (Frequent VPCs is defined as Ventricular premature contractions occurring more frequently than 30 times per hour or occurring in a pattern of bigeminy, trigeminy, or runs of ventricular tachycardia)
  - Non sustained or sustained ventricular tachycardia (VT)
  - Ventricular fibrillation (VF)
  - Newly diagnosed atrial fibrillation/flutter
  - Palpitations
  - Dependent lower extremity edema
  - Presyncope/Syncope
  - Dyspnea/shortness of breath, or hypoxemia
  - Suspected endocarditis when there is documentation of **any**:
    - Fever
    - Positive blood cultures indicating bacteremia
    - A new murmur

# Frequency of Echocardiography Testing (CD-2.3)

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## Repeat TTE

Repeat routine echocardiograms are not supported (annually or otherwise) for evaluation of clinically stable syndromes.

## Every three years

A repeat echo is allowed **every three years**, without a change in clinical status, when there is a documented history of:

- Bicuspid aortic valve
- Mild aortic or mitral stenosis
- Prosthetic heart valve
- Aortic sclerosis without stenosis
- A first degree relative with a diagnosis of Hypertrophic Cardiomyopathy
- A first degree relative with a diagnosis of Familial Dilated Cardiomyopathy or Idiopathic cardiomyopathy
- Genotype positive for Familial Dilated Cardiomyopathy

## Every two years

First degree relative with known thoracic aortic aneurysm or dissection a repeat echo is allowed every two years when **both**:

- Prior aortic imaging (echo, CT, MR) is negative
- Last aortic imaging was  $\geq 2$  years

## Once a year

A repeat echo is allowed **once** a year (when no change in clinical status), when there a history of:

- Significant valve dysfunction either:
  - Moderate or severe regurgitation
  - Moderate or severe stenosis
- Significant valve deformity (regardless of extent of regurgitation or stenosis) when there is documentation of **either**:
  - Thickened myxomatous valve
  - Bileaflet prolapse



- Hypertrophic cardiomyopathy- see also: **Transthoracic Echocardiography (TTE) – Indications (CD-2.2), Stress Echocardiography – Indications, other than ruling out CAD (CD-2.7)**
- Chronic pericardial effusions when findings would potentially alter therapy
- Left ventricular systolic dysfunction to evaluate the effectiveness of on-going therapy
- Aortic root dilatation that has not yet been repaired, see also **Congenital Valvular Aortic Stenosis (CD-11.2.9)** and for post-repair see **Post Aortic Endovascular/Open Surgery Surveillance Studies (PVD-6.8)** in the Peripheral Vascular Disease Imaging Guideline
- Systemic Sclerosis or Scleroderma

### **Every 6 Months**

A repeat echocardiogram is allowed every six months for asymptomatic, severe mitral regurgitation

### **Valve Surgery**

- If valve surgery is being considered can have TTE **twice** a year
- Post-surgery (repair or prosthetic valve implantation):
  - 6 weeks post surgery to establish baseline
  - One routine study (surveillance) every 3 years after valve repair or replacement.
- TAVR follow-up:
  - A baseline post-op TTE is usually performed within one week after surgery. This baseline study may also be approved as an outpatient if not performed in the hospital prior to discharge
  - 1 month post procedure
  - 1 year post-procedure
  - Annually thereafter
  - See also **Transcatheter Aortic Valve Replacement (TAVR) (CD-4.8)**
- Mitral valve clip:
  - 1 month post procedure
  - 6 months post-procedure
  - 1 year post-procedure
  - See also **Percutaneous Mitral Valve Repair (mitral valve clip) (CD-13.5)**

### **PFO closure**

- Preoperative evaluation for closure of PFO
- Postprocedural evaluation of PFO repair
- 6 month follow-up post PFO repair
- Annually if there is a residual shunt

(for ASD closure see **ASD-Atrial septal defects (CD 11.2.1)**)

## Left Atrial Appendage Occlusion

TTE with 3D imaging can be approved as part of the preprocedural evaluation

## Pulmonary hypertension

Follow-up echocardiogram (TTE) on patients with PH

- Every 6 months for surveillance of stable patients
- Prior to planned intubation (e.g., for elective surgery)
- Prior to planned pregnancy
- During pregnancy as often as requested by provider
- Anytime, without regard for the number or timing of previous ECHO studies to evaluate either:
  - Change in therapy
  - Change in clinical findings or symptoms
- Echocardiogram at baseline then every 3 months with therapy changes in stable individuals.

## Obstructive Hypertrophic Cardiomyopathy (HCM)

Repeat TTE (CPT® 93306) is indicated in individuals with Obstructive Hypertrophic Cardiomyopathy (HCM) for the following:

### ***Mavacamten for obstructive hypertrophic cardiomyopathy***

See **Mavacamten for Obstructive Hypertrophic Cardiomyopathy (HCM) (CD-12.3)**

Initiation of treatment

- Baseline-at the beginning of treatment
- 4 weeks after treatment initiation
- 8 weeks after treatment initiation
- 12 weeks after treatment initiation
- Then every 12 weeks while on mavacamten

Changes in treatment

- 4 weeks after any interruption of treatment (any missed dose)
- After any dosage change (including restart of treatment):
  - 4 weeks after dosage change
  - 12 weeks after dosage change
- After initiating a weak CYP2C19 inhibitor (e.g., omeprazole) or moderate CYP2A4 inhibitor (e.g., ciprofloxacin):
  - 4 weeks after start of medication
  - 12 weeks after start of medication

- At any time regardless of timing of prior echo when there are new cardiac signs or symptoms, or worsening of clinical status

### ***Post- Septal Reduction Therapy (SRT)***

TTE is indicated within 3 to 6 months after SRT (surgical myectomy or alcohol septal ablation) in individuals with hypertrophic cardiomyopathy to evaluate the procedural results

### **Cardiac device therapy**

- Re-evaluation is indicated three months after revascularization or maximally tolerated optimal medical therapy to determine either:
  - Candidacy for device therapy
  - Optimal choice of device
- Evaluation prior to ICD/CRT placement, while establishing 3 months of optimal medical therapy

### **Anytime**

Repeat echocardiogram is indicated **anytime** (without regard for the number or timing of previous ECHO studies) if there is a **change** in clinical status, or **new signs and symptoms** with documentation of **any** of the following:

- Cardiac murmurs
- Myocardial infarction or acute coronary syndrome
- Congestive heart failure (new or worsening):
  - New symptoms of dyspnea
  - Orthopnea
  - Paroxysmal nocturnal dyspnea
  - Elevated BNP
- Pericardial disease
- Stroke/transient ischemic attack
- Decompression illness
- Prosthetic valve dysfunction or thrombosis
- Cardiac transplant
- Individuals with Left Ventricular Assist Device (LVAD)

### **Cardiac Transplant**

Anytime (without regard for the number or timing of previous ECHO studies) when there is a history of cardiac transplant, per transplant center protocol

### **Cardiotoxic agents**

For re-evaluation in an individual previously or currently undergoing therapy with cardiotoxic agents or radiation therapy follow **Cardiotoxic agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1)**

# Transesophageal Echocardiography (TEE) (CD-2.4) (CD-2.5)

CD.EC.0002.4.A

v2.0.2023

## Guideline

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## Transesophageal echocardiography (TEE) – coding (CD-2.4)

## TEE coding

Transesophageal Echocardiography	CPT®
TEE with 2-D, M-mode, probe placement, image acquisition, interpretation and report	93312
TEE probe placement only	93313
TEE image acquisition, interpretation, and report only	93314
TEE for congenital anomalies with 2-D, M-mode, probe placement, image acquisition, interpretation and report	93315
TEE for congenital anomalies, probe placement only	93316
TEE for congenital anomalies, image acquisition, interpretation and report only	93317
TEE for monitoring purposes, ongoing assessment of cardiac pumping function on an immediate time basis	93318

## Doppler Echocardiography

Description	CPT®
Doppler echo, pulsed wave and/or spectral display	+93320
Doppler echo, pulsed wave and/or spectral display, follow-up or limited study	+93321
Doppler echo, color flow velocity mapping	+93325

Description	CPT®
Doppler echo, if performed, may be reported separately in addition to the primary TEE codes: CPT® 93312, CPT® 93314, CPT® 93315, and CPT® 93317	

## C codes

HCPCS	Description	CPT®
C8925	TEE with 2-D, M-mode, probe placement, image acquisition, interpretation and report	93312
C8926	TEE for congenital anomalies with 2-D, M-mode, probe placement, image acquisition, interpretation and report	93315
C8927	TEE for monitoring purposes, ongoing assessment of cardiac pumping function on an immediate time basis	93318

- The complete transesophageal echocardiogram service, including both (1) probe (transducer) placement and (2) image acquisition/interpretation, is reported with CPT® 93312.
  - Probe placement only is reported with CPT® 93313.
  - The image acquisition/interpretation only is reported with CPT® 93314.
- Physicians assign codes CPT® 93312, CPT® 93313, and/or CPT® 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
  - CPT® 93313 and CPT® 93314 should never be used together. If both services are provided, CPT® 93312 is reported.
- Hospitals should report TEE procedures using CPT® 93312 (the complete service). CPT® 93313 and CPT® 93314 are not used for hospital billing.
- Monitoring of patients undergoing cardiac surgery is CPT® 93318.

**Transesophageal echocardiography (TEE) – indications (CD-2.5)**

- Limited transthoracic echo window when further information is needed to guide management (e.g. suspected or confirmed endocarditis, suspected intracardiac mass, etc.)

- Assessing valvular dysfunction, especially mitral regurgitation, when TTE is inadequate and intervention is being considered to repair/replace valve.
- Evaluation of cardiac mass, suspected tumor or thrombus
- Preprocedural assessment of PFO/ASD
- 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 vegetation, tumor
- Embolic events when there is an abnormal TTE or a history of atrial fibrillation
  - Clarify atria/atrial appendage, aorta, mitral/aortic valve beyond the information that other imaging studies have provided
- Cardiac valve dysfunction
  - Differentiation of tricuspid from bicuspid aortic valve in setting of aortic enlargement or significant stenosis or significant regurgitation
  - Congenital abnormalities
- Assessing for left atrial thrombus prior to cardioversion of atrial fibrillation or atrial flutter.
- Assessing for left atrial thrombus prior to planned atrial fibrillation ablation/pulmonary vein isolation procedure.
- For initial imaging of ascending and descending thoracic aortic aneurysms.
- For repeat imaging or established thoracic aneurysms, TEE is indicated **only** when imaging with CT or MR is contraindicated.
- Left atrial appendage (LAA) Closure device (e.g., WATCHMAN®)
  - Preprocedural evaluation with or without 3D imaging
  - Repeat TEE 45 days post procedure
  - 1 year post procedure
  - See also **Percutaneous Mitral Valve Repair (mitral valve clip) (CD-13.5)**

# Stress echocardiography (stress echo) (CD-2.6) (CD-2.7)

CD.EC.0002.7.A

v2.0.2023

## Stress echo – coding (CD-2.6)

### Associated codes

Stress Echocardiography	CPT®
Echo, transthoracic, with (2D), includes M-mode, during rest and exercise stress test and/or pharmacologically induced stress, with report; <sup>1</sup>	93350
Echo, transthoracic, with (2D), includes M-mode, during rest and exercise stress test and/or pharmacologically induced stress, with report: including performance of continuous electrocardiographic monitoring, with physician supervision <sup>2</sup>	93351
Doppler Echocardiography	
Doppler echo, pulsed wave and/or spectral display <sup>3</sup>	+93320
Doppler echo, pulsed wave and/or spectral display, follow-up/limited study	+93321
Doppler echo, color flow velocity mapping <sup>4</sup>	+93325

### Associated HCPCS codes

CPT®	Stress Echocardiography	HCPCS
93350	Echo, transthoracic, with (2D), includes M-mode, during rest and exercise stress test and/or pharmacologically induced stress, with report; <sup>5</sup>	C8928
93351	Echo, transthoracic, with (2D), includes M-mode, during rest and exercise stress test and/or pharmacologically induced stress, with report: including performance of continuous electrocardiographic monitoring, with physician supervision <sup>6</sup>	C8930

<sup>1</sup> CPT® 93350 and CPT® 93351 do not include Doppler studies

<sup>2</sup> CPT® 93350 and CPT® 93351 do not include Doppler studies

<sup>3</sup> Doppler echo (CPT® +93320 and CPT® +93325), if performed, may be reported separately in addition to the primary SE codes: CPT® 93350 or CPT® 93351.

<sup>4</sup> Doppler echo (CPT® +93320 and CPT® +93325), if performed, may be reported separately in addition to the primary SE codes: CPT® 93350 or CPT® 93351.

<sup>5</sup> CPT® 93350 and CPT® 93351 do not include Doppler studies

<sup>6</sup> CPT® 93350 and CPT® 93351 do not include Doppler studies

**Stress echo—indications other than ruling out CAD (CD-2.7)**

- See: **Stress Testing with Imaging – Indications (CD-1.4)**
- In addition to the evaluation of CAD, stress echo can be used to evaluate the following conditions:
  - Dyspnea on exertion (specifically to evaluate pulmonary hypertension)
  - Right heart dysfunction
  - Valvular heart disease, especially when the outcome would affect a therapeutic or interventional decision
  - Pulmonary hypertension, when the outcome will measure response to therapy and/or prognostic information
  - Hypertrophic cardiomyopathy (as defined in **Obstructive Hypertrophic Cardiomyopathy (HCM) (CD-12.3)**) for **either** of the following:
    - Exercise stress echo (CPT® 93351 or 93350) is indicated for the detection and quantification of dynamic left ventricular outflow tract obstruction in symptomatic individuals with HCM who do **not** have a resting or provokable outflow tract gradient  $\geq 50$  mm Hg on TTE.
    - Stress echo can be repeated in 1 to 2 years in an individual with a documented history of HCM previously evaluated with a stress echo when there is documentation of **either** of the following:
      - Worsening symptoms
      - There has been a therapeutic change (i.e., change in medication, surgical procedure performed).
- In general spectral Doppler (CPT® 93320 or 93321) and color-flow Doppler (CPT® 93325) are necessary in the evaluation of the above conditions and can be added to the stress echo code.



# 3D Echocardiography (CD-2.8)(CD-2.9)

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## 3D echocardiography – coding (CD-2.8)

- CPT® 93319 with one of the following (CPT® 93303, 93304, 93312, 93314, 93315, or 93317) for congenital cardiac abnormalities
- The procedure codes used to report 3D rendering for echocardiography are not unique to echocardiography and are the same codes used to report the 3D post-processing work for CT, MRI, ultrasound, and other tomographic modalities.
  - **CPT® 76376**, not requiring image post-processing on an independent workstation, is the most common code used for 3D rendering done with echocardiography
  - **CPT® 76377** requires the use of an independent workstation

## 3D echocardiography – indications (CD-2.9)

Echocardiography with 3-dimensional (3D) rendering is becoming universally available, yet its utility remains limited based on the current literature.

- 3D Echo may be indicated when a primary echocardiogram is approved and **one** of the following is needed:
  - Left ventricular volume and ejection fraction assessment when measurements are needed for treatment decision (e.g., implantation of ICD, alteration in cardiotoxic chemotherapy)
  - Mitral valve anatomy specifically related to mitral valve stenosis
  - Preprocedural evaluation of left atrial appendage occlusion (e.g., WATCHMAN®)
  - Guidance of transcatheter procedures such as:
    - Mitral valve clipping
    - TAVR
    - Left atrial appendage closure device (e.g., WATCHMAN®)

# Myocardial strain imaging (CPT® 93356) (CD-2.12)

CD.EC.0002.12.A

v2.0.2023

- Myocardial strain imaging (CPT® 93356, speckle tracking longitudinal strain) is indicated for the initial evaluation of LVH, in addition to the primary echocardiogram, when there is documentation of **both**:
  - Unclear etiology
  - Concern for infiltrative cardiomyopathy
- Myocardial strain imaging (CPT® 93356) in addition to the primary echocardiogram in individuals receiving therapy with cardiotoxic agents for ANY of the following:
  - Initial evaluation-prior to treatment with EITHER:
    - Medications that could result in cardiotoxicity/heart failure
    - Radiation that could result in cardiotoxicity/heart failure
  - Re-evaluation of an individual previously or currently undergoing therapy as per echocardiogram parameters. See **Cardiotoxic agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1)**
  - Re-evaluation of an individual undergoing therapy with worsening symptoms

# References (CD-2)

CD.EC.0002.A

v2.0.2023

1. Douglas PS, Khandheria B, Stainback RF, et al. ACCF/AHA/ACEP/AHA/ASNC/SCAI/SCCT/SCMR 2008 appropriateness criteria for stress echocardiography: a report of the American College of Cardiology Foundation Appropriateness Criteria Task Force, American Society of Echocardiography, American College of Emergency Physicians, American Heart Association, American Society of Nuclear Cardiology, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, and Society for Cardiovascular Magnetic Resonance: endorsed by the Heart Rhythm Society and the Society of Critical Care Medicine. *Circulation*. 2008;117(11):1478-1497. doi:10.1161/CIRCULATIONAHA.107.189097.
2. Pellikka PA, Nagueh SF, Elhendy AA, Kuehl CA, Sawada SG. American Society of Echocardiography Recommendations for Performance, Interpretation, and Application of Stress Echocardiography. *J Am Soc Echocardiogr*. 2007;20(9):1021-1041. doi:10.1016/j.echo.2007.07.003.
3. Holmes DR, Mack MJ, Kaul S, et al. 2012 ACCF/AATS/SCAI/STS Expert Consensus Document on Transcatheter Aortic Valve Replacement. *The Annals of Thoracic Surgery*. 2012;93(4):1340-1395. doi:10.1016/j.athoracsur.2012.01.084.
4. Wolk MJ, Bailey SR, Doherty JU, et al. ACCF/AHA/ASE/ASNC/HFSA/HRS/SCAI/SCCT/SCMR/STS 2013 Multimodality Appropriate Use Criteria for the Detection and Risk Assessment of Stable Ischemic Heart Disease. *J Am Coll Cardiol*. 2014;63(4):380-406. doi:10.1016/j.jacc.2013.11.009.
5. Doherty JU, Kort S, Mehran R, Schoenhagen P, Soman P. ACC/AATS/AHA/ASE/ASNC/HRS/SCAI/SCCT/SCMR/STS 2017 Appropriate Use Criteria for Multimodality Imaging in Valvular Heart Disease. *J Am Coll Cardiol* 2017;70(13):1647-1672. doi:10.1016/j.jacc.2017.07.732.
6. Khanna D, Gladue H, Channick R, et al. Recommendations for Screening and Detection of Connective Tissue Disease-Associated Pulmonary Arterial Hypertension. *Arthritis & Rheumatism*. 2013;65(12):3194-3201. doi:10.1002/art.38172.
7. Simonneau G, Gatzoulis MA, Adatia I, et al. Updated Clinical Classification of Pulmonary Hypertension. *J Am Coll Cardiol*. 2013;62(25). doi:10.1016/j.jacc.2013.10.029.
8. Tolle JJ, Waxman AB, Horn TLV, Pappagianopoulos PP, Systrom DM. Exercise-Induced Pulmonary Arterial Hypertension. *Circulation*. 2008;118(21):2183-2189. doi:10.1161/circulationaha.108.787101.
9. Vainrib AF, Harb SC, Jaber W, et al. Left Atrial Appendage Occlusion/Exclusion: Procedural Image Guidance with Transesophageal Echocardiography. *J Am Soc Echocardiogr*. 2018;31(4):454-474. doi:10.1016/j.echo.2017.09.014.
10. Lentine KL, Costa SP, Weir MR, et al. Cardiac Disease Evaluation and Management Among Kidney and Liver Transplantation Candidates. *Circulation*. 2012;126(5):617-663. doi:10.1161/cir.0b013e31823eb07a.
11. Stainback RF, Estep JD, Agler DA, et al. Echocardiography in the Management of Patients with Left Ventricular Assist Devices: Recommendations from the American Society of Echocardiography. *J Am Soc Echocardiogr*. 2015;28(8):853-909. doi:10.1016/j.echo.2015.05.008.
12. Sachdeva R, Valente AM, Armstrong AK, et al. ACC/AHA/ASE/HRS/ISACHD/SCAI/SCCT/SCMR/SOPE 2020 Appropriate Use Criteria for Multimodality Imaging During the Follow-Up Care of Patients with Congenital Heart Disease. *J Am Coll Cardiol*. 2020;75(6):657-703. doi:10.1016/j.jacc.2019.10.002.
13. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Circulation*. 2019;139(14). doi:10.1161/cir.0000000000000603.
14. Otto CM, Nishimura RA, Bonow RO, et al. 2020 ACC/AHA Guideline for the Management of Patients with Valvular Heart Disease: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation*. 2021;143(5). doi:10.1161/cir.0000000000000923.
15. Hiratzka LF, Bakris GL, Beckman JA, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the diagnosis and management of patients with thoracic aortic disease. A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine [published correction appears in J Am Coll Cardiol. 2013 Sep 10;62(11):1039-40]. *J Am Coll Cardiol*. 2010;55(14):e27-e129. doi:10.1016/j.jacc.2010.02.015.
16. Ge Y, Gupta S, Fentanes E, et al. Role of Cardiac CT in Pre-Procedure Planning for Transcatheter Mitral Valve Replacement. *JACC: Cardiovasc Imag*. 2021. doi:10.1016/j.jcmg.2020.12.018.
17. Blanke P, Weir-McCall JR, Achenbach S, et al. Computed Tomography Imaging in the Context of Transcatheter Aortic Valve Implantation (TAVI)/Transcatheter Aortic Valve Replacement (TAVR). *JACC: Cardiovasc Imag*. 2019;12(1):1-24. doi:10.1016/j.jcmg.2018.12.003.

18. Doherty JU, Kort S, Mehran R, et al. ACC/AATS/AHA/ASE/ASNC/HRS/SCAI/SCCT/SCMR/STS 2019 Appropriate Use Criteria for Multimodality Imaging in the Assessment of Cardiac Structure and Function in Nonvalvular Heart Disease: A Report of the American College of Cardiology Appropriate Use Criteria Task Force, American Association for Thoracic Surgery, American Heart Association, American Society of Echocardiography, American Society of Nuclear Cardiology, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, Society for Cardiovascular Magnetic Resonance, and the Society of Thoracic Surgeons. *J Am Coll Cardiol*. 2019;73(4):488-516. doi:10.1016/j.jacc.2018.10.038.
19. Behr ER, Dalageorgou C, Christiansen M, et al. Sudden arrhythmic death syndrome: familial evaluation identifies inheritable heart disease in the majority of families. *Eur Heart J*. 2008;29(13):1670-1680. doi:10.1093/eurheartj/ehn219.
20. Emery MS, Kovacs RJ. Sudden Cardiac Death in Athletes. *JACC Heart Fail*. 2018;6(1):30-40. doi:10.1016/j.jchf.2017.07.014.
21. Ommen SR, Mital S, Burke MA, et al. 2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients With Hypertrophic Cardiomyopathy: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2020;76(25):e159-e240. doi:10.1016/j.jacc.2020.08.045.
22. Sweet M, Taylor MR, Mestroni L. Diagnosis, prevalence, and screening of familial dilated cardiomyopathy. *Expert Opin Orphan Drugs*. 2015;3(8):869-876. doi:10.1517/21678707.2015.1057498.
23. TeRiele, Anneline, James, Cynthia, Approach to family screening in arrhythmogenic right ventricular dysplasia/ Cardiomyopathy. *Eur Heart J*. (2016) 37, 755-763 doi:10.1093/eurheartj/ehv387.
24. Tanaka H. Efficacy of echocardiography for differential diagnosis of left ventricular hypertrophy: special focus on speckle-tracking longitudinal strain. *J Echocardiogr*. 2021;19(2):71-79. doi:10.1007/s12574-020-00508-3.

# Nuclear Cardiac Imaging

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# Myocardial Perfusion Imaging (MPI) (CD-3.1)(CD-3.2)

CD.NC.0003.1.A

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## Myocardial Perfusion Imaging (MPI) – Coding (CD-3.1)

Nuclear Cardiac Imaging Procedure Codes	
Myocardial Perfusion Imaging (MPI)	CPT®
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)	<b>78451</b>
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	<b>78452</b>
Absolute quantitation of myocardial blood flow (AQMBF), single-photon emission computed tomography (SPECT), with exercise or pharmacologic stress, and at rest, when performed (List separately in addition to code for primary procedure)	<b>+0742T</b>

- The most commonly performed myocardial perfusion imaging are single (at rest or stress, CPT® 78451) and multiple (at rest and stress, CPT® 78452) SPECT studies.
  - Evaluation of the individual's left ventricular wall motion and ejection fraction are routinely performed during MPI and are included in the code's definition.
  - First pass studies, (CPT® 78481 and CPT® 78483), MUGA, (CPT® 78472 and CPT® 78473) and SPECT MUGA (CPT® 78494) should not be reported in conjunction with MPI codes.
  - Attenuation correction, when performed, is included in the MPI service by code definition. No additional code should be assigned for the billing of attenuation correction.

- **Multi-day Studies:** In the absence of written payer guidelines to the contrary, it is not appropriate to bill separately for the rest and stress segments of MPI even if performed on separate calendar dates. A single code is assigned to define the entire procedure on the date all portions of the study are completed.

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**Note** 3D rendering should not be billed in conjunction with MPI.

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## MPI – Indications (CD-3.2)

See: **Stress Testing with Imaging – Indications (CD-1.4)**

### **Absolute quantitation of myocardial blood flow (AQMBF)(SPECT)**

- AQMBF obtained by CZT-SPECT is considered experimental, investigational, or unproven at this time.

## References

1. Bailly M, Thibault F, Courtehoux M, Metrard G, Ribeiro MJ. Impact of attenuation correction for CZT-SPECT measurement of myocardial blood flow. J Nucl Cardiol. 2021;28(6):2560-2568. doi:10.1007/s12350-020-02075-7.
2. Acampa W, Zampella E, Assante R, et al. Quantification of myocardial perfusion reserve by CZT-SPECT: A head to head comparison with 82Rubidium PET imaging. J Nucl Cardiol. 2021;28(6):2827-2839. doi:10.1007/s12350-020-02129-w.
3. Wang L, Zheng Y, Zhang J, et al. Diagnostic value of quantitative myocardial blood flow assessment by NaI(Tl) SPECT in detecting significant stenosis: a prospective, multi-center study [published online ahead of print, 2022 Aug 15]. J Nucl Cardiol. 2022;10.1007/s12350-022-03085-3. doi:10.1007/s12350-022-03085-3.
4. Pang Z, Wang J, Li S, Chen Y, Wang X, Li J. Diagnostic analysis of new quantitative parameters of low-dose dynamic myocardial perfusion imaging with CZT SPECT in the detection of suspected or known coronary artery disease. Int J Cardiovasc Imaging. 2021 Jan;37(1):367-378. doi: 10.1007/s10554-020-01962-x. Epub 2020 Sep 10. PMID: 32914404; PMCID: PMC7878253.



# MUGA – Coding (CD-3.3)

CD.NC.0003.3.A

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Cardiac blood pool imaging, or radionuclide ventriculography, can be used to evaluate ventricular function. Cardiac blood pool imaging includes first pass studies (CPT® 78481 and 78483) as well as gated equilibrium studies (CPT® 78472, 78473, 78494, and +78496).

Gated equilibrium studies can also be referred to as multi-gated acquisition (MUGA) scan or equilibrium radionuclide angiography (ERNA). Imaging for gated equilibrium studies can be planar or three-dimensional (single photon emission computed tomography, SPECT).

Of note, all cardiac blood pool imaging is synchronized with electrographic RR interval (EKG-gated); thus, regular rhythm is required for accurate LV assessment.

Gated Equilibrium Studies – Planar	CPT®
Cardiac blood pool imaging, gated equilibrium; planar, <b>single study</b> at rest or stress, wall motion study plus ejection fraction, with or without quantitative processing	78472
Cardiac blood pool imaging, gated equilibrium; planar, <b>multiple</b> studies, wall motion study plus ejection fraction, <b>at rest and stress</b> , with or without additional quantification	78473
Gated Equilibrium Studies - SPECT	CPT®
Cardiac blood pool imaging, gated equilibrium, SPECT, at rest, wall motion study plus ejection fraction, with or without quantitative processing	78494
First Pass studies	CPT®
Cardiac blood pool imaging (planar), first pass technique; <b>single</b> 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; <b>multiple</b> studies, at rest and with stress (exercise and/or pharmacologic), wall motion study plus ejection fraction, with or without quantification	78483
Cardiac blood pool imaging, gated equilibrium, <b>single study</b> , at rest, with <b>right ventricular ejection fraction</b> by first pass technique (List separately in addition to code for primary procedure) This CPT code is an add-on code to 78472.	+78496

- The technique employed for a MUGA service guides the code assignment.
  - CPT® 78472 is used for a planar MUGA scan at rest or stress
  - CPT® 78473 for planar MUGA scans, multiple studies at rest and stress.



- Planar MUGA studies (CPT® 78472 and CPT® 78473) should not be reported in conjunction with:
  - SPECT MPI (CPT® 78451 - CPT® 78454)
  - First pass studies (CPT® 78481- CPT® 78483)
  - SPECT MUGA (CPT® 78494).
- CPT® +78496 is assigned only in conjunction with CPT® 78472.

# MUGA Study – Cardiac Indications (CD-3.4)

CD.NC.0003.4.A

v2.0.2023

## MUGA (Multi Gated Acquisition) – Blood Pool Imaging Indications

- Echocardiography is the preferred method of following left ventricular systolic function.
- MUGA may be appropriate when a recent ECHO, as indicated in **Transthoracic Echocardiography (TTE) – Indications (CD-2.2)** and/or **Frequency of Echocardiography Testing (CD 2.3)**, was technically limited and prevented accurate assessment of left ventricular function.
- MUGA may be appropriate when there is a significant discrepancy between LVEF assessment by ECHO and another modality (i.e., one study reports normal LVEF and the other, a reduced LVEF) AND there is clear documentation as to how quantitative measurement of LVEF will affect individual management (e.g., implantation of an ICD, alteration in cardiotoxic chemotherapy, etc.).
- MUGA may be performed in place of an ECHO in the following circumstances:
  - To determine candidacy for ICD/CRT and/or to determine optimal choice of device in individuals who meet criteria for ICD based on ejection fraction and other criteria.
  - When previously or currently undergoing therapy with potentially cardiotoxic agents, including chemotherapy and radiation, AND a history of previous low LV ejection fraction (LVEF <50%). See **Cardiotoxic agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1)**
- MUGA is **not** indicated when requested simply to compare LVEF by the same modality, a prior MUGA is not a reason to approve another MUGA.

## Right ventricular first pass study

- (CPT® 78472 and 78496) may be performed when ECHO is technically limited and prevents accurate assessment of RV function AND when further information about RV function is needed to guide management (e.g. established/diagnosed pulmonary hypertension, suspected or confirmed pulmonary embolus).

## First pass studies

- First pass studies (CPT® 78481 and CPT® 78483) may be approved in place of MUGA when indications are met for MUGA and/or there is need for information that cannot be obtained by MUGA.
- First pass studies, (CPT® 78481 and CPT® 78483), MUGA (CPT® 78472 and CPT® 78473) and SPECT MUGA (CPT® 78494) should not be reported in conjunction with MPI codes.

**MUGA Study – Oncologic Indications for Cancer Therapeutics-Related Cardiac Dysfunction (CTRCD) (CD-3.5)**

- See **Cardiotoxic agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1)**

# Myocardial Sympathetic Innervation Imaging in Heart Failure (CD-3.6)

CD.CS.0009.A

v2.0.2023

- Nuclear imaging using I-123-meta-iodobenzylguanidine (I-123-mIBG) in an attempt to image increased myocardial sympathetic activity is considered to be experimental and investigational.
- The AMA has established the following set of Category III codes to report these studies:
  - **0331T** - Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment
  - **0332T** - Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment; with tomographic SPECT.

## ***Background and supporting information***

In heart failure, the sympathetic nervous system is activated in order to compensate for the decreased myocardial function. Initially, this is beneficial, however, long-term this compensatory mechanism is detrimental and causes further damage.

Markers have been developed, using radioactive iodine, in an attempt to image this increased myocardial sympathetic activity. Currently, AdreView™ (Iodine-123 meta-iodobenzylguanidine), is the only FDA-approved imaging agent available for this purpose.

# Myocardial Tc-99m Pyrophosphate Imaging (CD-3.7)

CD.NC.0003.7.A

v2.0.2023

Myocardial Tc-99m Pyrophosphate Imaging	
<b>MUGA (Multi Gated Acquisition) – Blood Pool Imaging</b>	<b>CPT®</b>
Myocardial Imaging, infarct avid, planar, qualitative or quantitative	<b>78466</b>
Myocardial Imaging, infarct avid, planar, qualitative or quantitative with ejection fraction by first pass technique	<b>78468</b>
Myocardial Imaging, infarct avid, planar, qualitative or quantitative tomographic SPECT with or without quantification	<b>78469</b>
Radiopharmaceutical Localization Imaging Limited area	<b>78800</b>
Radiopharmaceutical Localization Imaging SPECT Note: When reporting CPT® 78803, planar imaging of a limited area or multiple areas should be included with the SPECT	<b>78803</b>
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 (e.g., head, neck, chest, pelvis), single day imaging	<b>78830</b>

# Cardiac Amyloidosis (CD-3.8)

CD.NC.0003.8.A

v2.0.2023

- Tc-99m pyrophosphate imaging (CPT® 78803 or 78830) may be used to identify cardiac amyloidosis. Chest SPECT and planar imaging may be used, as well as whole-body imaging for identification of systemic ATTR (transthyretin) amyloidosis. See **Myocardial Tc-99m Pyrophosphate Imaging (CD-3.7)** for coding information
- For a single planar imaging session alone (without a SPECT study), report CPT® 78800 Radiopharmaceutical Localization Imaging Limited area
- Tc-99m pyrophosphate imaging can be pursued for diagnosis of ATTR amyloidosis in the presence of known systemic amyloidosis if Cardiac MRI (CMR) is either contraindicated or indeterminate in individuals undergoing evaluation for kidney transplant. See **Kidney Transplant, Pre-Transplant Imaging Studies (AB-42.5)**.
- Tc-99m pyrophosphate imaging can be pursued for diagnosis of ATTR amyloidosis after screening for presence of a monoclonal light chain to exclude AL amyloidosis:
  - Serum kappa/lambda free light chain ratio (not SPEP)
    - Abnormal if ratio is <0.26 or >1.65
  - Serum and urine immunofixation electrophoresis (IFE)
    - Abnormal if monoclonal protein detected
- Tc-99m pyrophosphate imaging may also be used for the following:
  - Diagnosis of cardiac ATTR in individuals with cardiac MRI or echocardiography findings consistent with or suggestive of cardiac amyloidosis.
  - Individuals with suspected cardiac ATTR amyloidosis and contraindications to CMR such as renal insufficiency or an implantable cardiac device.
  - Individuals with systemic amyloidosis who are being evaluated for kidney transplant if CMR is either contraindicated or indeterminate. See **Kidney Transplant, Pre-Transplant Imaging Studies (AB-42.5)**.

## ***Background and supporting information***

- The following conditions would raise high index of suspicion:
  - Left ventricular hypertrophy but low voltage on ECG
  - Heart failure with preserved ejection fraction and an increase in left ventricular wall thickness.
  - Unexplained heart failure with preserved ejection fraction and concomitant right heart failure in an individual over the age of 60
  - Individuals, especially elderly males, with signs/symptoms of heart failure and any of the following:
    - Lumbar spinal stenosis
    - Spontaneous biceps tendon rupture
    - Bilateral carpal tunnel syndrome

- Atrial arrhythmias in the absence of usual risk factors
  - Known or suspected familial amyloidosis.
  - Low flow, low gradient aortic stenosis

# Non-imaging Heart Function and Cardiac Shunt Imaging (CD-1.7)

CD.GG.0001.7.C

v2.0.2023

- Echocardiogram is the preferred method for cardiac shunt detection, rather than the cardiac shunt imaging study described by CPT® 78428.
- Ejection fraction can be obtained by echocardiogram, SPECT MPI, MUGA study, cardiac MRI, cardiac CT, or cardiac PET depending on the clinical situation, rather than by the non-imaging heart function study described by CPT® 78414.

## ***Background and supporting information***

Procedures reported with CPT® 78414 and CPT® 78428 are essentially obsolete and should not be performed in lieu of other preferred modalities



## References (CD-3)

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1. American Association of Physicists in Medicine (AAPM) Report 96, January 2008. Report of AAPM Task Group 23, "The measurement, reporting and management of radiation dose in CT." [https://www.aapm.org/pubs/reports/RPT\\_96.pdf](https://www.aapm.org/pubs/reports/RPT_96.pdf).
2. Boden WE, O'Rourke RA, Teo KK, et al. Impact of optimal medical therapy with or without percutaneous coronary intervention on long-term cardiovascular end points in patients with stable coronary artery disease (from the COURAGE trial). *Am J Cardiol*. 2009 July; 104(1):1-4. doi.org/10.1016/j.amjcard.2009.02.059.
3. Guarneri V, Lenihan DJ, Valero V, et al. Long-term cardiac tolerability of trastuzumab in metastatic breast cancer: the M.D. Anderson Cancer Center experience. *J Clinical Oncology* 2006 Sept; 24:4107-4115. doi:10.1200/JCO.2005.04.9551.
4. Hendel RC, Berman DS, Carli MFD, et al. ACCF/ASNC/ACR/AHA/ASE/SCCT/SCMR/SNM 2009 Appropriate Use Criteria for Cardiac Radionuclide Imaging. *J Am Coll Cardiol*. 2009;53(23):2201-2229. doi:10.1016/j.jacc.2009.02.013.
5. Highlights of Prescribing Information HERCEPTIN® (trastuzumab) for injection, for intravenous use Initial U.S. Approval: 1998. Revised: April 2017. <http://www.gene.com/gene/products/information/pdf/herceptin-prescribing.pdf>.
6. Sciammarella MG, Gerson M, Buxton AE, et al. ASNC/SNMMI Model Coverage Policy: Myocardial sympathetic innervation imaging: Iodine-123 meta-iodobenzylguanidine ((123)I-mIBG). *J Nucl Cardiol*. 2015;22(4):804-811. doi:10.1007/s12350-015-0202-8.
7. Bokhari S, Castano A, Pozniakoff T, et al. (99m)Tc-pyrophosphate scintigraphy for differentiating light-chain cardiac amyloidosis from the transthyretin-related familial and senile cardiac amyloidosis. *Circ Cardiovasc Imaging* 2013; 6:195. doi:10.1161/CIRCIMAGING.112.000132.
8. Plana JC, Galderisi M, Barac A, et al. Expert consensus for multimodality imaging evaluation of adult patients during and after cancer therapy: a report from the American Society of Echocardiography and the European Association of Cardiovascular Imaging. *European Heart Journal - Cardiovascular Imaging*. 2014;15(10):1063-1093. doi:10.1093/ehjci/jeu192.
9. Dorbala S, Bokhari S, Miller E, et al. 99mTechnetium-Pyrophosphate Imaging for Transthyretin Cardiac Amyloidosis. *ASNC PRACTICE POINTS 2016*. <https://www.asnc.org/Files/Practice%20Resources/Practice%20Points/ASNC%20Practice%20Point-99mTechnetiumPyrophosphateImaging2016.pdf>.
10. Rapezzi C, Quarta CC, Guidalotti PL, et al. Role of 99mTc-DPD Scintigraphy in Diagnosis and Prognosis of Hereditary Transthyretin-Related Cardiac Amyloidosis. *JACC: Cardiovascular Imaging*. 2011;4(6):659-670. doi:10.1016/j.jcmg.2011.03.016.
11. Dorbala S, Ananthasubramaniam K, Armstrong IS, et al. Single Photon Emission Computed Tomography (SPECT) Myocardial Perfusion Imaging Guidelines: Instrumentation, Acquisition, Processing, and Interpretation. *J Nuc Cardiol*. 2018. doi:10.1007/s12350-018-1283-y.
12. Doherty JU, Kort S, Mehran R, et al. ACC/AATS/AHA/ASE/ASNC/HRS/SCAI/SCCT/SCMR/STS 2019 Appropriate Use Criteria for Multimodality Imaging in the Assessment of Cardiac Structure and Function in Nonvalvular Heart Disease. *J Am Coll Cardiol*. 2019;73(4):488-516. doi:10.1016/j.jacc.2018.10.038.
13. Witteles RM, Liedtke M. AL Amyloidosis for the Cardiologist and Oncologist. *JACC: CardioOncology*. 2019;1(1):117-130. doi:10.1016/j.jacc.2019.08.002.
14. Ruberg FL, Grogan M, Hanna M, Kelly JW, Maurer MS. Transthyretin Amyloid Cardiomyopathy. *J Am Coll Cardiol*. 2019;73(22):2872-2891. doi:10.1016/j.jacc.2019.04.003.
15. Dorbala S, Cuddy S, Falk RH. How to Image Cardiac Amyloidosis. *JACC: Cardiovascular Imaging*. 2020;13(6):1368-1383. doi:10.1016/j.jcmg.2019.07.015.
16. Jitendra M. MUGA scan (CPT code 78472, 78473, 78494) Coding Tips. Medical Coding Guide. <https://www.americanmedicalcoding.com/muga-scan-cpt-code/>. Published November 10, 2020.
17. Kittleson MM, Maurer MS, Ambardekar AV, et al. Cardiac Amyloidosis: Evolving Diagnosis and Management: A Scientific Statement From the American Heart Association [published correction appears in *Circulation*. 2021 Jul 6;144(1):e10] [published correction appears in *Circulation*. 2021 Jul 6;144(1):e11]. *Circulation*. 2020;142(1):e7-e22. doi:10.1161/CIR.0000000000000792.
18. Kidney Disease: Improving Global Outcomes (KDIGO) Kidney Transplant Candidate Work Group. KDIGO Clinical Practice Guideline on the Evaluation and Management of Candidates for Kidney Transplantation. 2020;104: S1 – S103.

# Cardiac CT

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# Cardiac CT and CTA - General information and coding (CD-4.1)

CD.CT.0004.1.A

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## Associated Codes

### Cardiac Imaging Procedure Codes

Cardiac CT and CCTA	CPT®
<p>CT, heart, without contrast, with quantitative evaluation of coronary calcium</p> <ul style="list-style-type: none"> <li>The code set for Cardiac CT and CCTA (CPT® 75572-CPT® 75574), include quantitative and functional assessment (for example, calcium scoring) if performed</li> <li>CPT® 75571 describes a non-contrast CT of the heart with calcium scoring and should be reported only when calcium scoring is performed as a stand-alone procedure. <ul style="list-style-type: none"> <li>Can be used to report a preliminary non-contrast scan which indicates an excessive amount of calcium such that the original scheduled study must be discontinued.</li> <li>CPT® 75571 should not be reported in conjunction with any of the contrast CT/CTA codes (CPT® 75572- CPT® 75574).</li> </ul> </li> </ul>	75571
CT, heart, with contrast, for evaluation of cardiac structure and morphology (including 3D image post-processing, 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 left ventricular [LV] cardiac function, right ventricular [RV] structure and function and evaluation of vascular structures, if performed).	75573
CTA, heart, coronary arteries and bypass grafts (when present), with contrast, including 3D image post-processing (including 3D image post-processing, assessment of cardiac function, and evaluation of venous structures, if performed).	75574

Cardiac CT and CCTA	CPT®
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

### Cardiac Imaging Procedure Codes

Description	CPT®
Automated quantification and characterization of coronary atherosclerotic plaque to assess severity of coronary disease, using data from coronary computed tomographic angiography; data preparation and transmission, computerized analysis of data, with review of computerized analysis output to reconcile discordant data, interpretation and report	0623T
Automated quantification and characterization of coronary atherosclerotic plaque to assess severity of coronary disease, using data from coronary computed tomographic angiography; data preparation and transmission	0624T

Description	CPT®
Automated quantification and characterization of coronary atherosclerotic plaque to assess severity of coronary disease, using data from coronary computed tomographic angiography; computerized analysis of data from coronary computed tomographic angiography	0625T
Automated quantification and characterization of coronary atherosclerotic plaque to assess severity of coronary disease, using data from coronary computed tomographic angiography; review of computerized analysis output to reconcile discordant data, interpretation and report	0626T

### Cardiac CT and CTA - General information (CD-4.1)

- Only one code from the set: CPT® 75572 - CPT® 75574 can be reported per encounter.
- CPT® 75574 includes evaluation of cardiac structure and morphology when performed; therefore, additional code/s should not be assigned.
- Automated quantification and characterization of coronary atherosclerotic plaque (CPT® 0623T, 0624T, 0625T, 0626T) is a service in which coronary computed tomographic angiography (CCTA) data are analyzed using computerized algorithms to assess the extent and severity of coronary artery disease. The use of automated quantification and characterization of coronary atherosclerotic plaque is considered investigational and experimental at this time.

#### **Background and Supporting Information**

The high negative predictive value (98%-99%) of CCTA in ruling out significant coronary artery disease has been confirmed in multiple studies.

3D rendering should not be billed in conjunction with Cardiac CT and CCTA.

# CT for coronary calcium scoring (CPT® 75571) (CD-4.2)

CD.CT.0004.2.A

v2.0.2023

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## CT Calcium Scoring-Asymptomatic and for CAD Screening (CD-4.2.1)

- Coronary calcium scoring is not indicated in someone with known CAD.
- Coronary artery calcium score (CPT® 75571) can be approved when there is documentation of **all** of the following:
  - The results will impact risk-based decisions for preventive interventions
  - The individual is an adult age 40-75
  - The 10-year ASCVD risk including pooled cohort equation is between 5.0% to 19.9%
  - There is no documented CAD
  - Individual is not currently on a statin
  - Individual is not a smoker
  - There is no history of diabetes
  - There is no family history of premature CAD
  - There has been no calcium score performed in the previous 5 years
  - There has been no prior calcium score > 0

## Background and Supporting Information

### State Mandates

#### ***Texas Heart Attack Preventive Screening Law (HR 1290)***

Texas Heart Attack Preventive Screening Law mandates that insurers in Texas cover either a calcium scoring study (CPT® 75571 or HCPCS S8092) or a carotid intima-media thickness study (ultrasound—Category III code 0126T) every five years for certain populations.

- To qualify, the following must apply:
  - Must be a Texas resident.
  - Must be a member of a fully-insured Texas health plan.
  - Must be a male age 45 to 75 or a female age 55 to 75.
  - Must have either diabetes or a Framingham cardiac risk score of intermediate or higher (10% or higher).

- Must not have had a calcium scoring study or a carotid intima-media thickness study within the past 5 years

### ***New Mexico House Bill 126***

New Mexico House Bill 126 Coverage for Health Artery Calcium Scan:

- Coverage may apply per state mandate as stated in House Bill 126. See <https://www.nmlegis.gov> for guidance on specific application.
- Coronary calcium scan can be approved every 5 years to be used as a clinical management tool when all the following apply:
  - Prior CT calcium was >5 years ago
  - Prior CT calcium scan had a calcium score of zero
  - The individual is between the ages of 45 and 65
- The individual has an intermediate risk of developing CAD determined by a health care provider based on a 10 year risk algorithm including pooled cohort equation.

## **CT Calcium Scoring Indications-Symptomatic (CD-4.2.2)**

### **Symptoms concerning for cardiac ischemia**

- Individuals with new, recurrent or worsening symptoms concerning for cardiac ischemia, who have a 'very low', or 'low' pretest probability of CAD\*, see **Table 1** in **General guidelines (CD-1.0)** for definitions of very low, low, intermediate, and high pretest probability of CAD

### **Low gradient aortic stenosis**

- Coronary artery calcium score (CPT® 75571) can be approved in low gradient aortic stenosis when symptomatic, severe aortic stenosis is suspected. Low gradient aortic stenosis is defined as an AVA <1 and a mean gradient <40mmHg.



# CCTA – Indications for CCTA (CPT® 75574) (CD-4.3)

CD.CT.0004.3.A

v2.0.2023

- New, recurrent or worsening symptoms concerning for cardiac ischemia in individuals who have:
  - An 'intermediate' or 'intermediate-high' pretest probability of CAD\*, see **Table-1** in **General guidelines (CD-1.0)**
  - Persistent symptoms in individuals with a 'low', 'intermediate', or 'intermediate-high' pre-test probability of coronary disease after a normal stress test
  - Equivocal, borderline, abnormal or discordant prior noninvasive evaluation where obstructive coronary artery disease remains a concern (<90 days)
  - Abnormal rest ECG findings, such as a new LBBB, or T-wave inversions, when ischemia is a concern
  - A prior CABG when **only** graft patency is a concern
- Evaluation of an individual under the age of 40 for suspected anomalous coronary artery(ies) or for treatment planning when there is a history of one or more of the following:
  - Syncopal episodes during strenuous activities
  - Persistent chest pain brought on by exertion or emotional stress, and normal stress test
  - Full sibling(s) with history of sudden death syndrome before age 40 or with documented anomalous coronary artery
  - Resuscitated sudden death and contraindications for conventional coronary angiography
  - Prior nondiagnostic coronary angiography in determining the course of the anomalous coronary artery in relation to the great vessels, origin of a coronary artery or bypass graft location (any):
    - Anomalies of origin:
      - LCA or the RCA arising from the pulmonary artery;
      - Interarterial course between the pulmonary artery and the aorta of either the RCA arising from the left sinus of Valsalva or the LCA arising from the right sinus of Valsalva
    - Anomalies of course:
      - Myocardial bridging
    - Anomalies of termination:
      - Coronary artery fistula
- Initial imaging study in individuals with hypertrophic cardiomyopathy and stable anginal symptoms.



- Chest discomfort is common in individuals with hypertrophic cardiomyopathy. The incidence of false positive myocardial perfusion imaging abnormalities is higher in these individuals, whereas the incidence of severe coronary artery stenosis is low.
- Individuals who have recovered from unexplained sudden cardiac arrest in lieu of invasive coronary angiography (**both**):
  - Confirm the presence or absence of ischemic heart disease
  - Exclude the presence of an anomalous coronary artery.

## CCTA – Regardless of symptoms (CPT® 75574) (CD-4.4)

CD.CT.0004.4.A

v2.0.2023

- Evaluation of newly diagnosed congestive heart failure or cardiomyopathy (all):
  - No prior history of coronary artery disease, the ejection fraction is less than 50 percent, and low or intermediate risk on the pre-test probability assessment, and
  - No contraindications to cardiac CT angiography.
  - No cardiac catheterization, SPECT, cardiac PET, or stress echocardiogram has been performed since the diagnosis of congestive heart failure or cardiomyopathy.
- Unclear coronary artery anatomy despite conventional cardiac catheterization
- Re-do CABG
  - Assess bypass graft patency
  - Evaluate the location of the left internal mammary artery (LIMA) and or right internal mammary artery (RIMA) prior to repeat bypass surgery
- Follow-up Left main stent one time at 6-12 months
- Evaluate coronary artery anomalies and other complex congenital heart disease of cardiac chambers or great vessels:
  - Report CPT® 75574 for evaluating coronary artery anomalies.
  - Report CPT® 75573 for congenital heart disease.
  - To evaluate the great vessels, CTA Chest (CPT® 71275) can be performed instead of CCTA or in addition to CCTA.
  - For anomalous pulmonary venous return, can add CT Abdomen and Pelvis with contrast (CPT® 74177).
- When CCTA will replace conventional invasive coronary angiography for any of the following:
  - Ventricular tachycardia (6 beat runs or greater)
  - Delayed presentation or retrospective evaluation of suspected Takotsubo syndrome (stress cardiomyopathy)
  - Preoperative assessment of the coronary arteries in planned surgery for any of the following:
    - Aortic dissection
    - Aortic aneurysm
    - Valvular surgery
  - To assess for coronary involvement in individuals with systemic vasculitis (e.g. Giant Cell Arteritis, Takayasu's, Kawasaki's disease) when there are clinical features suggestive of underlying vasculitis including:

- Unexplained elevated cardiac markers (erythrocyte sedimentation rate, C-reactive protein)
- Constitutional symptoms (fever, chills, night sweats, weight loss)
- Multiple visceral infarcts in the absence of embolic etiology
- **Cardiac Trauma:** CTA Chest (CPT® 71275) and CCTA (CPT® 75574) are useful in detecting aortic and coronary injury and can help in the evaluation of myocardial and pericardial injury see **Cardiac Trauma – Imaging (CD-10.1)**

# Fractional Flow Reserve by Computed Tomography (CD-4.5)

**CD.CT.0004.5.A****v2.0.2023**

Fractional flow reserve (FFR) is typically measured using invasive techniques. FFR can be obtained noninvasively from coronary computed tomography angiography data (FFR-CT).

- Indications for FFR-CT:
  - To further assess CAD seen on a recent CCTA that is of uncertain physiologic significance

# CT Heart – Indications (CPT® 75572) (CD-4.6)

CD.CT.0004.6.A

v2.0.2023

## CT heart – indications (CPT® 75572) (CD-4.6)

- Cardiac vein identification for lead placement in individuals needing left ventricular pacing.
- Pulmonary vein isolation procedure (ablation) for atrial fibrillation:
  - MRI Cardiac (CPT® 75557 or CPT® 75561), MRV Chest (CPT® 71555), CTV Chest (CPT® 71275), or CT Cardiac (CPT® 75572) can be performed to evaluate the anatomy of the pulmonary veins prior to an ablation procedure performed for atrial fibrillation.
  - Study may be repeated post-procedure between 3-6 months after ablation because of a 1%-2% incidence of asymptomatic pulmonary vein stenosis
  - See **Pulmonary Vein Imaging – Indications (CD-8.2)**
- If echocardiogram is inconclusive for:
  - Cardiac or pericardial tumor or mass
  - Cardiac thrombus
  - Pericarditis/constrictive pericarditis
  - Complications of cardiac surgery
- In place of MRI when there is clinical suspicion of arrhythmogenic right ventricular dysplasia or arrhythmogenic cardiomyopathy (ARVD/ARVC) if the clinical suspicion is supported by established criteria for ARVD-see **Cardiac MRI – Indications (excluding Stress MRI) (CD-5.2)**
- Recurrent laryngeal nerve palsy due to cardiac chamber enlargement.
- CT Cardiac (CPT® 75572) can be performed instead of TEE for assessment of left atrial appendage (LAA) occlusion device or to assess for thrombus, see: **Transesophageal Echocardiography (TEE) – Indications (CD-2.5)**
- Coronary imaging is not included in the code definition for CPT® 71275.
  - The AMA definition for CPT® 71275 reads: "CTA Chest (non-coronary), with contrast material(s), including non-contrast images, if performed, and image post-processing."

# CT Heart for Congenital Heart Disease (CD-4.7)

CD.CT.0004.7.A

v2.0.2023

## CT Heart for Congenital Heart Disease (CPT® 75573) (CD-4.7)

- Coronary artery anomaly evaluation
  - A cardiac catheterization was performed, and not all coronary arteries were identified.
- Thoracic arteriovenous anomaly evaluation
  - A MRI Cardiac or CT angiogram Chest was performed and suggested congenital heart disease.
- Complex adult congenital heart disease evaluation
  - No CT Cardiac or MRI Cardiac has been performed, and there is a contraindication to MRI Cardiac.
  - A CT Cardiac or MRI Cardiac was performed one year ago or more.
- See also section **Adult Congenital Heart Disease (CD-11)**

# Transcatheter aortic valve replacement (TAVR) (CD-4.8)

CD.CT.0004.8.A

v1.0.2023

## Pre-surgical aortic valve replacement

- Once the decision has been made for aortic valve replacement, the following may be used to determine if an individual is a candidate for TAVR:
  - CTA Chest (CPT® 71275), Abdomen and Pelvis (combination code CPT® 74174) are considered appropriate, and
  - CT Cardiac (CPT® 75572) may be considered to measure the aortic annulus or
  - Coronary CTA (CCTA CPT® 75574) may be considered to both measure the aortic annulus and assess the coronary arteries in lieu of heart catheterization.
- A repeat diagnostic left heart catheterization is **not** medically necessary when the individual is undergoing a transcatheter aortic valve replacement (TAVR).

### ***Transfemoral access not feasible***

Alternative imaging can be obtained to evaluate vascular access for TAVR in individuals for whom it is documented either via the office note or prior imaging that transfemoral access would not be feasible due to **any** of the following exclusion criteria:

- Small vessels
- Highly calcified vessels
- Stenosed or occluded vessels
- Prior aortoiliac vascular intervention

Imaging is indicated based on the documented intended access site (transaxillary or transcarotid) and should be of the involved body areas. The following studies are indicated based on the documented planned access site:

- CTA of the Head (CPT® 70496) and/or Neck (CPT® 70498) for transcarotid access
- CTA of the Chest (CPT® 71275) and/or Upper extremity (CPT® 73206) for transaxillary access

## Post TAVR

- TTE follow-up is indicated at:
  - A baseline post-op TTE is indicated within one week after surgery if not performed in the hospital prior to discharge.
  - 1 month
  - One year post-procedure
  - Then annually thereafter.

# References (CD-4)

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1. Taylor AJ, Cerqueira M, Hodgson JM, et al. ACCF/SCCT/ACR/AHA/ASE/ASNC/ NASCI/SCAI/SCMR 2010 appropriate use criteria for cardiac computed tomography: a report of the American College of Cardiology Foundation Appropriate Use Criteria Task Force, the Society of Cardiovascular Computed Tomography, the American College of Echocardiography, the American Society of Nuclear Cardiology, the North American Society for Cardiovascular Imaging, the Society for Cardiovascular Angiography and Interventions, and the Society for Cardiovascular Magnetic Resonance. *J Am Coll Cardiol* 2010; 56:1864-1894. doi:10.1016/j.jacc.2010.07.005.
2. Curry SJ, Krist AH, Owens DK, et al. Risk Assessment for Cardiovascular Disease with Nontraditional Risk Factors. *Jama*. 2018;320(3):272-280. doi:10.1001/jama.2018.8359.
3. Boden WE, O'Rourke RA, Teo KK, et al. Impact of Optimal Medical Therapy With or Without Percutaneous Coronary Intervention on Long-Term Cardiovascular End Points in Patients With Stable Coronary Artery Disease (from the COURAGE Trial). *J Am Coll Cardiol*. 2009;104(1):1-4. doi:10.1016/j.amjcard.2009.02.059.
4. Einstein AJ, Henzlova MJ, Rajagopalan S. Estimating Risk of Cancer Associated with Radiation Exposure From 64-Slice Computed Tomography Coronary Angiography. *Jama*. 2007;298(3):317. doi:10.1001/jama.298.3.317.
5. Schlosser T, Konorza T, Hunold P, et al. Noninvasive visualization of coronary artery bypass grafts using 16-detector row computed tomography. *J Am Coll Cardiol*, 2004; 44:1224-1229. doi:10.1016/j.jacc.2003.09.075.
6. Douglas PS, DeBruyne B, Pontone G, Patel MR, et al. 1-year outcomes of FFRct-guided care in patients with suspected coronary disease: The PLATFORM Study. *J Am Coll Cardiol*, 2016; 68:435-45. doi:10.1016/j.jacc.2016.05.056.
7. Norgaard B, Leipsic J, Gaur S, et al. Diagnostic performance of noninvasive fractional flow reserve derived from coronary computed tomography angiography in suspected coronary artery disease. *J Am Coll Cardiol*, 2014; 63:1145-55. doi:10.1016/j.jacc.2013.11.043.
8. Ko BS, Cameron JD, Munnur RK, Wong DTL, et al. Cardiac CT: atherosclerosis to acute coronary syndrome. *J Am Coll Cardiol*. December 2016;4(6). doi:10.3978/j.issn.2223-3652.2014.11.03.
9. Holmes D Jr, Mack M, Kaul S, et al. 2012 ACCF/AATS/SCAI/STS expert consensus document on transcatheter aortic valve replacement. *J Am Coll Cardiol*, 2012; 59:1200. doi:10.1016/j.jacc.2012.01.001.
10. NICE medical technology advisory committee. Overview: HeartFlow FFRCT for estimating fractional flow reserve from coronary CT angiography: Guidance. NICE: National Institute for health and care excellence. <https://www.nice.org.uk/guidance/mtg32>. Published February 2017.
11. American College of Cardiology Foundation Task Force on Expert Consensus Documents, Mark DB, Berman DS, et al. ACCF/ACR/AHA/NASCI/SAIP/SCAI/SCCT 2010 expert consensus document on coronary computed tomographic angiography: a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents. *J Am Coll Cardiol*. 2010; 55:2663. doi:10.1161/CIR.0b013e3181d4b618.
12. Doherty JU, Kort S, Mehran R, Schoenhagen P, Soman P. ACC/AATS/AHA/ASE/ASNC/HRS/SCAI/SCCT/SCMR/STS 2017 Appropriate Use Criteria for Multimodality Imaging in Valvular Heart Disease. *Journal of Nuclear Cardiology*. 2017;24(6):2043-2063. doi:10.1007/s12350-017-1070-1.
13. The Medicare Learning Network®. MEDICARE PREVENTIVE SERVICES. Preventive Services Chart Medicare Learning Network®. ICN MLN006559. [https://www.cms.gov/Medicare/Prevention/PrevntnGenInfo/medicare-preventive-services/MPS-QuickReferenceChart-1.html#CARDIO\\_DIS](https://www.cms.gov/Medicare/Prevention/PrevntnGenInfo/medicare-preventive-services/MPS-QuickReferenceChart-1.html#CARDIO_DIS). Published June 2019.
14. Grundy SM, Stone NJ, Bailey AL, et al. 2018 AHA/ACC/AACVPR/AAPA/ABC/ACPM/ADA/AGS/APhA/ASPC/NLA/PCNA Guideline on the Management of Blood Cholesterol: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Circulation*. 2019;139(25). doi:10.1161/cir.0000000000000625.
15. Arnett DK, Blumenthal RS, Albert MA, et al. 2019 ACC/AHA Guideline on the Primary Prevention of Cardiovascular Disease. *Circulation*. 2019;140(11):e596-e646. doi:10.1161/cir.0000000000000678.
16. Korsholm K, Berti S, Iriart X, et al. Expert Recommendations on Cardiac Computed Tomography for Planning Transcatheter Left Atrial Appendage Occlusion. *JACC: Cardiovascular Interventions*. 2020;13(3):277-292. doi:10.1016/j.jcin.2019.08.054.
17. Koster MJ, Warrington KJ. Vasculitis of the Coronary Arteries. *American College of Cardiology Latest in Cardiology*. <https://www.acc.org/latest-in-cardiology/articles/2019/03/13/06/50/vasculitis-of-the-coronary-arteries>. Published March 13, 2019. Accessed July 29, 2020.



18. Opolski MP, Staruch AD, Jakubczyk M, et al. CT Angiography for the Detection of Coronary Artery Stenoses in Patients Referred for Cardiac Valve Surgery. *JACC: Cardiovascular Imaging*. 2016;9(9):1059-1070. doi:10.1016/j.jcmg.2015.09.028.
19. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Circulation*. 2019;139(14). doi:10.1161/cir.0000000000000603.
20. Al-Khatib SM, Stevenson WG, Ackerman MJ, et al. 2017 AHA/ACC/HRS Guideline for Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death. *Circulation*. 2018;138(13):e272-e391. doi:10.1161/cir.0000000000000549.
21. Gräni C, Buechel RR, Kaufmann PA, Kwong RY. Multimodality Imaging in Individuals with Anomalous Coronary Arteries. *JACC: Cardiovascular Imaging*. 2017;10(4):471-481. doi:10.1016/j.jcmg.2017.02.004.
22. Kim SY, Seo JB, Do K-H, et al. Coronary Artery Anomalies: Classification and ECG-gated Multi-Detector Row CT Findings with Angiographic Correlation. *RadioGraphics*. 2006;26(2):317-333. doi:10.1148/rg.262055068.
23. Ghadri JR, Kazakauskaitė E, Braunschweig S, et al. Congenital coronary anomalies detected by coronary computed tomography compared to invasive coronary angiography. *BMC Cardiovascular Disorders*. 2014;14(1). doi:10.1186/1471-2261-14-81.
24. Shariat M, Thavendiranathan P, Nguyen E, et al. Utility of coronary CT angiography in outpatients with hypertrophic cardiomyopathy presenting with angina symptoms. *J Cardiovasc Comput Tomogr*. 2014;8(6):429-437. doi:10.1016/j.jcct.2014.09.007.
25. Lyon AR, Bossone E, Schneider B, et al. Current state of knowledge on Takotsubo syndrome: a Position Statement from the Taskforce on Takotsubo Syndrome of the Heart Failure Association of the European Society of Cardiology. *European Journal of Heart Failure*. 2015;18(1):8-27. doi:10.1002/ehf.424.
26. Levine A, Hecht HS. Cardiac CT Angiography in Congestive Heart Failure. *Journal of Nuclear Medicine*. 2015;56(Supplement\_4). doi:10.2967/jnumed.114.150441.
27. Hecht H, Blaha MJ, Berman DS, et al. Clinical indications for coronary artery calcium scoring in asymptomatic patients: Expert consensus statement from the Society of Cardiovascular Computed Tomography. *J Cardiovasc Comput Tomogr*. 2017;11(2):157-168. doi:10.1016/j.jcct.2017.02.010.
28. Williams MC, Kwiecinski J, Doris M, et al. Low-Attenuation Noncalcified Plaque on Coronary Computed Tomography Angiography Predicts Myocardial Infarction. *Circulation*. 2020;141(18):1452-1462. doi:10.1161/circulationaha.119.044720.
29. Daghem M, Bing R, Fayad ZA, Dweck MR. Noninvasive Imaging to Assess Atherosclerotic Plaque Composition and Disease Activity. *JACC: Cardiovascular Imaging*. 2020;13(4):1055-1068. doi:10.1016/j.jcmg.2019.03.033.
30. Shaw LJ, Blankstein R, Bax JJ, et al. Society of Cardiovascular Computed Tomography / North American Society of Cardiovascular Imaging – Expert Consensus Document on Coronary CT Imaging of Atherosclerotic Plaque. *J Cardiovasc Comput Tomogr*. 2020. doi:10.1016/j.jcct.2020.11.002.
31. Writing Committee Members, Otto CM, Nishimura RA, et al. 2020 ACC/AHA Guideline for the Management of Patients With Valvular Heart Disease: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines [published correction appears in *J Am Coll Cardiol*. 2021 Feb 2;77(4):509] [published correction appears in *J Am Coll Cardiol*. 2021 Mar 9;77(9):1275]. *J Am Coll Cardiol*. 2021;77(4):e25-e197. doi:10.1016/j.jacc.2020.11.018

# Cardiac MRI

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# Cardiac MRI – Coding (CD-5.1)

CD.MRI.0005.1.A

v2.0.2023

Cardiac Imaging Procedure Codes	
Cardiac MRI	CPT®/HCPCS
Cardiac magnetic resonance imaging for morphology and function without contrast	<b>75557</b>
Cardiac magnetic resonance imaging for morphology and function without contrast; with stress imaging	<b>75559</b>
Cardiac magnetic resonance imaging for morphology and function without and with contrast and further sequences	<b>75561</b>
Cardiac magnetic resonance imaging for morphology and function without and with contrast and further sequences; with stress imaging	<b>75563</b>
Cardiac magnetic resonance imaging for velocity flow mapping (List separately in addition to code for primary procedure)	<b>+75565</b>
Cardiac magnetic resonance imaging for morphology and function, quantification of segmental dysfunction; with strain imaging	<b>C9762</b>
Cardiac magnetic resonance imaging for morphology and function, quantification of segmental dysfunction; with stress imaging	<b>C9763</b>

- Only one procedure code from the set (CPT® 75557- CPT® 75563) should be reported per session.
- Only one flow velocity measurement (CPT® +75565) should be reported per session when indicated.
- C9762--Cardiac magnetic resonance imaging for morphology and function, quantification of segmental dysfunction; with strain imaging. The use of CMR strain imaging for the quantification of segmental dysfunction is considered investigational and experimental at this time.
- C9763--Cardiac magnetic resonance imaging for morphology and function, quantification of segmental dysfunction; with stress imaging. The use of stress CMR for the quantification of segmental dysfunction is considered investigational and experimental at this time.

# Cardiac MRI and MRA Chest – Indications (excluding Stress MRI) (CD-5.2)

CD.MRI.0005.2.A

v2.0.2023

- Assess myocardial viability (to differentiate hibernating myocardium from scar) when necessary to determine if revascularization should be performed (CPT® 75561)
- Assessment of global ventricular function, myocardial composition, and mass if a specific clinical question is left unanswered by a recent echocardiogram and results will affect individual management (CPT® 75557 or CPT® 75561). Particularly useful in evaluating:
  - Cardiomyopathy (ischemic, diabetic, hypertrophic, or muscular dystrophy)
  - Noncompaction
  - Infiltrative heart disease such as amyloid, iron overload cardiomyopathy (hemosiderosis, hemochromatosis)
  - Post cardiac transplant
  - Hypertrophic cardiomyopathy
  - Suspected acute myocarditis, cardiac aneurysm, trauma, and contusions
  - Monitoring cancer chemotherapy effect on the heart (especially if an accurate assessment of right ventricular function is documented as necessary).
- Pre and postoperative congenital heart disease assessment see **Adult Congenital Heart Disease (CD-11)** for defect specific indications (CPT® 75557 or CPT® 75561).
  - MRA Chest (CPT® 71555) may be added if the aorta or pulmonary artery need to be visualized beyond the root.
  - May add CPT® +75565 in conjunction with CPT® 75557 or CPT® 75561, only if there is a need to clarify findings on a recent echocardiogram and cardiac Doppler study when there is documentation of **either** of the following:
    - Significant valvular disease that may require intervention
    - Intracardiac flow disturbances (e.g., ASD, VSD)
- MRA Chest (CPT® 71555) may be indicated for the following:
  - Thoracic aortic dissection see **Aortic Dissection and Other Aortic Conditions (PVD-6.7)** in the Peripheral Vascular Disease Imaging Guidelines
  - Coarctation of the aorta see:
    - **Coarctation of the Aorta (CD-11.3.2)** for adults
    - **Aortic Coarctation and IAA (interrupted aortic arch) (PEDCD-2.4.11)** for infants and children in the Pediatric Cardiac Imaging Guideline
  - Thoracic aortic aneurysm see **Thoracic Aortic Aneurysm (TAA) (PVD-6.2)** in the Peripheral Vascular Disease Imaging Guidelines.

- Coarctation of the aorta
  - Follow-up (surveillance) imaging after repair of coarctation:
    - Adults: see **Coarctation of the Aorta (CD-11.3.2)**
    - Infants and children: see **Aortic Coarctation and IAA (interrupted aortic arch) (PEDCD-2.4.11)** in the Pediatric Cardiac Imaging Guideline
- Arrhythmogenic right ventricular dysplasia or arrhythmogenic right ventricular cardiomyopathy (ARVD/ARVC) suspicion (CPT® 75557 or CPT® 75561)—must have one of the following:
  - Nonsustained or sustained VT of LBBB morphology OR >500 PVC's over 24 hours on event recorder or Holter monitor.
  - ARVD/ARVC confirmed in a first degree relative either by criteria, autopsy, pathogenic genetic mutation or sudden death <35 years of age with suspected ARVD/ARVC.
  - Inverted T waves in right precordial leads (V1, V2 and V3) or beyond in individuals >14 years of age in the absence of complete RBBB
  - Right ventricular akinesis, dyskinesis or aneurysm noted on echo or RV angiography.
- Differentiate constrictive pericarditis from restrictive cardiomyopathy (CPT® 75561).
- Evaluate cardiac tumor or mass when echocardiogram is inconclusive.
- Evaluate valvular heart disease when echocardiogram is inconclusive:
  - CPT® 75557 **or** CPT® 75561
  - May add CPT® 75565 when there is documentation of either of the following:
    - Significant valvular disease that may require intervention
    - Intracardiac flow disturbances (e.g., ASD, VSD)
- MRI Cardiac (CPT® 75557 or CPT® 75561) *or* chest MRV (CPT® 71555), but not both, for pulmonary vein anatomy for planned ablation procedures in individuals with atrial fibrillation. See **Pulmonary Vein Imaging – Indications (CD-8.2)** for guidelines on follow-up imaging after ablation procedure.
- Suspected cardiac thrombus when echocardiogram is inconclusive (CPT® 75557).
- Right ventricular function evaluation (CPT® 75557 in conjunction with CPT® +75565) if a recent ECHO has been done, and there is documented need to perform Cardiac MRI in order to resolve an unanswered question about flow dynamics.
- Shunting through a VSD (CPT® 75557 in conjunction with CPT® +75565) 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 about flow dynamics.
- Conditions that would **not** require an echo prior to an MRI:
  - Anomalous coronary arteries: Cardiac MRI (CPT® 75561) or CCTA (CPT® 75574) is much better at detecting this than conventional angiography.
  - Assess coronary arteries in Kawasaki's disease.
  - Fabry disease

- Late enhancement MRI may predict the effect of enzyme replacement therapy on myocardial changes that occur with this disease (CPT® 75561).
- Initial evaluation for cardiac sarcoidosis.

# Cardiac MRI – Indications for Stress MRI (CD-5.3)

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CD.MRI.0005.3.A

v2.0.2023

- For indications for Stress MRI see **Stress Testing with Imaging – Indications (CD-1.4)**.
- If a nuclear perfusion (MPI) stress test was performed and was equivocal, a stress MRI is appropriate.

# Cardiac MRI – Aortic Root and Proximal Ascending Aorta (CD-5.4)

CD.MRI.0005.4.A

v2.0.2023

- See- Thoracic Aortic Aneurysm (TAA) (PVD-6.2) in the Peripheral Vascular Disease imaging guidelines



# Cardiac MRI – Evaluation of Pericardial Effusion or Diagnosis of Pericardial Tamponade (CD-5.5)

CD.MRI.0005.5.A

v2.0.2023

- Contrast-enhanced cardiac MRI (CPT® 75561) is useful for evaluating pericarditis, neoplastic and other effusion, tamponade or myocardial infiltration if a specific clinical question is left unanswered by echocardiogram or another recent imaging study.

# Cardiac MRI – Myocarditis (CD-5.6)

CD.MRI.0005.6.A

v2.0.2023

## Clinical evaluation of suspected myocarditis

Initial testing for suspected myocarditis should consist of an electrocardiogram, measurement of cardiac troponin, and an echocardiogram.

Cardiac MRI is indicated for suspected myocarditis in the presence of **all** of the following:

- New onset or persisting symptoms suggestive of myocarditis documented by **any** of the following:
  - Dyspnea
  - Chest pain
  - Palpitations
  - Syncope
  - Effort intolerance
- Evidence for recent or ongoing myocardial injury documented by **any** of the following results on initial screening:
  - Ventricular dysfunction noted on any cardiac imaging study, or
  - New or persisting ECG abnormalities suspicious for myocarditis
    - New ST changes, T wave changes, Q waves, or
    - New conduction abnormalities, such as LBBB or AV block, or
    - VT or VF
  - Elevated troponin
- Strong suspicion for viral etiology of myocardial injury with documentation of **both**:
  - Recent systemic viral disease, recent mRNA COVID-19 vaccination, or prior myocarditis
  - No evidence of coronary ischemia as documented by **any** of the following:
    - Lack of risk factors for CAD
    - Age under 35 years
    - Negative cardiac imaging study, such as MPI, CCTA, cath

## Return to Play Screening for athletes at risk for myocarditis

Cardiac MRI is indicated for **Return to Play Screening** for athletes when there is documentation of **both** of the following:

- Individual has a history of a clinical condition associated with myocarditis (i.e., COVID-19 infection or recent mRNA COVID-19 vaccination)
- Initial screening has been performed with documentation of **either** of the following:
  - Initial screening showed evidence for recent or ongoing myocardial injury (as defined above in Clinical Evaluation of Suspected Myocarditis ) with ongoing symptoms concerning for myocarditis (dyspnea, chest pain, palpitations, syncope, or effort intolerance).
  - Normal results of initial screening with persistent or new onset symptoms concerning for myocarditis.

### ***Background and Supporting Information***

As noted in the "2022 ACC expert consensus decision pathway on cardiovascular sequelae of COVID-19 in adults" and the 2017 "Sports cardiology: core curriculum for providing cardiovascular care to competitive athletes and highly active people", an athlete is defined as an individual who places a high premium on exercise training, competition, and sports achievement.

# Cardiac MRI – Duchenne Muscular Dystrophy (DMD) (CD-5.7)

**CD.MRI.0005.7.A****v2.0.2023**

Cardiac MRI (CPT® 75557 or 75561-does not include 75565 or 71555 unless otherwise indicated)

- Asymptomatic individual with documented DMD can have annual surveillance cardiac MRI starting at 6 years old (yearly echo is recommended prior to age 6)
- Asymptomatic, documented carrier of DMD can have cardiac MRI every 3 years starting at 18

## References (CD-5)

v2.0.2023

1. Hamdan A, Charalampos K, Roettgen R, et al. Magnetic resonance imaging versus computed tomography for characterization of pulmonary vein morphology before radiofrequency catheter ablation of atrial fibrillation. *Am J Cardiol*. 2009; 104:1540-1546. doi:10.1016/j.amjcard.2009.07.029.
2. Hendel RC, Kramer CM, Patel MR, et al. ACCF/ACR/SCCT/SCMR/ ASNC/NASCI/SCAI/SIR 2006 Appropriateness Criteria for computed tomography and cardiac magnetic resonance imaging. *J Am Coll Cardiol*. 2006; 48(7):1475-1497. Accessed November 30, 2017. doi:10.1016/j.jacc.2006.07.003.
3. Doherty JU, Kort S, Mehran R, et al. ACC/AATS/AHA/ASE/ASNC/HRS/SCAI/SCCT/SCMR/STS 2019 Appropriate Use Criteria for Multimodality Imaging in the Assessment of Cardiac Structure and Function in Nonvalvular Heart Disease. *J Am Coll Cardiol*. 2019;73(4):488-516. doi:10.1016/j.jacc.2018.10.038.
4. Hundley WG, Bluemke DA, Finn JP, et al. ACCF/ACR/AHA/NASCI/SCMR 2010 Expert Consensus Document on Cardiovascular Magnetic Resonance. *J Am Coll Cardiol*. 2010;55(23):2614-2662. doi:10.1016/j.jacc.2009.11.011.
5. Friedrich MG, Sechtem U, Schulz-Menger J, et al. Cardiovascular Magnetic Resonance in Myocarditis: A JACC White Paper. *J Am Coll Cardiol*. 2009;53(17):1475-1487. doi:10.1016/j.jacc.2009.02.007.
6. Riele AST, Tandri H, Sanborn DM, Bluemke DA. Noninvasive Multimodality Imaging in ARVD/C. *JACC: Cardiovasc Imaging*. 2015;8(5):597-611. doi:10.1016/j.jcmg.2015.02.007.
7. Verhaert D, Richards K, Rafael-Fortney JA, Raman SV. Cardiac involvement in patients with muscular dystrophies: magnetic resonance imaging phenotype and genotypic considerations. *Circ Cardiovasc Imaging*. 2011;4(1):67-76. doi:10.1161/CIRCIMAGING.110.960740.
8. Feingold B, Mahle WT, Auerbach S, et al. Management of Cardiac Involvement Associated With Neuromuscular Diseases: A Scientific Statement From the American Heart Association. *Circulation*. 2017;136(13). doi:10.1161/cir.0000000000000526
9. Mah ML, Cripe L, Slawinski MK, et al. Duchenne and Becker muscular dystrophy carriers: Evidence of cardiomyopathy by exercise and cardiac MRI testing. *International Journal of Cardiology*. 2020;316:257-265. doi:10.1016/j.ijcard.2020.05.052
10. Power LC, O'Grady GL, Hornung TS, Jefferies C, Gusso S, Hofman PL. Imaging the heart to detect cardiomyopathy in Duchenne muscular dystrophy: A review. *Neuromuscular Disorders*. 2018;28(9):717-730. doi:10.1016/j.nmd.2018.05.011
11. Hor KN, Mah ML, Johnston P, Cripe TP, Cripe LH. Advances in the diagnosis and management of cardiomyopathy in Duchenne muscular dystrophy. *Neuromuscular Disorders*. 2018;28(9):711-716. doi:10.1016/j.nmd.2018.06.014.
12. Birnkrant DJ, Bushby K, Bann CM, et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management. *The Lancet Neurology*. 2018;17(4):347-361. doi:10.1016/s1474-4422(18)30025-5.
13. Baggish AL, Battle RW, Beckerman JG, et al. Sports Cardiology: Core Curriculum for Providing Cardiovascular Care to Competitive Athletes and Highly Active People. *J Am Coll Cardiol*. 2017;70(15):1902-1918. doi:10.1016/j.jacc.2017.08.055.
14. Ammirati E, Frigerio M, Adler ED, et al. Management of Acute Myocarditis and Chronic Inflammatory Cardiomyopathy: An Expert Consensus Document. *Circ Heart Fail*. 2020;13(11):e007405. doi:10.1161/CIRCHEARTFAILURE.120.007405.
15. Gluckman TJ, Bhavne NM, et al. 2022 ACC Expert Consensus Decision Pathway on Cardiovascular Sequelae of COVID-19 in Adults: Myocarditis and Other Myocardial Involvement, Post-Acute Sequelae of SARS-CoV-2 Infection, and Return to Play: A Report of the American College of Cardiology Solution Set Oversight Committee. *J Am Coll Cardiol*. 2022;79(17):1717-1756. doi:10.1016/j.jacc.2022.02.003.

# Cardiac PET

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## Cardiac PET – Coding (CD-6.1)

CD.PET.0006.1.A

v2.0.2023

Cardiac Imaging Procedure Codes	
Cardiac PET	CPT®
Myocardial imaging, positron emission tomography (PET), metabolic evaluation study (including ventricular wall motion[s] and/or ejection fraction[s], when performed), single study	78459
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)	78491
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)	78492
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

Cardiac Imaging Procedure Codes	
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	<b>78431</b>
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 (e.g., myocardial viability);	<b>78432</b>
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 (e.g., myocardial viability); with concurrently acquired computed tomography transmission scan	<b>78433</b>
Absolute quantitation of myocardial blood flow (AQMBF), positron emission tomography (PET), rest and pharmacologic stress (List separately in addition to code for primary procedure)	<b>+78434</b>
Positron emission tomography (PET) with concurrently acquired computed tomography (CT) for attenuation correction and anatomical localization imaging; skull base to mid-thigh	<b>78815</b>

- 3D rendering should not be billed in conjunction with PET.
- Separate codes for such related services as treadmill testing (CPT® 93015-CPT® 93018) and radiopharmaceuticals should be assigned in addition to perfusion PET. These services are paid according to each individual payer.



# Cardiac PET – Perfusion – Indications (CD-6.2)

CD.PET.0006.2.A

v2.0.2023

CPT® 78430, CPT® 78431, CPT® 78491 and CPT® 78492

- Meets all of the criteria for an imaging stress test in **Stress Testing with Imaging – Indications (CD-1.4)** and additionally any one of the following:
  - Individual is severely obese (for example BMI >40 kg/m<sup>2</sup>) or
  - Individual has large breasts or implants
  - Individual incapable of exercise due to physical (musculoskeletal or neurological) inability to achieve target heart rate. Target heart rate is calculated as 85% of the maximum age predicted heart rate (MPHR). MPHR is estimated as 220 minus the individual's age
- Equivocal nuclear perfusion (SPECT MPI) stress test
- Routine use in post heart transplant assessment of transplant CAD

# Cardiac PET – Absolute Quantitation of Myocardial Blood Flow (AQMBF) (CD-6.3)

CD.PET.0006.3.A  
v2.0.2023

## CPT® 78434\*

Quantitation of myocardial blood flow at rest and with stress in ml/g/min and the calculation of myocardial perfusion reserve (the ratio of stress to rest flow) can be used for diagnosis and prognosis of coronary artery disease and cardiac endothelial dysfunction that can be seen in diabetes, left ventricular hypertrophy, heart transplantation vasculopathy among other conditions.

- AQMBF with PET (CPT 78434) is an add-on procedure that is indicated when one of the following apply:
  - Primary study Myocardial PET rest/stress perfusion (CPT® 78492 or 78431 only) has been approved
  - Primary study Myocardial PET rest/stress perfusion (CPT® 78492 or 78431 only) has been ordered and is being approved at the same time (see- **Cardiac PET - Perfusion - Indications (CD-6.2)** or **Stress Testing with Imaging -Indications (CD-1.4)**)

## Background and supporting information

Despite its utility, AQMBF is a technically challenging measurement. Variables include:

- Different tracers (N-13 ammonia vs Rb-82 Cl) give different values
- Different mathematical models used (static vs dynamic)
- Different stressors are used that give different hyperemic flow results (adenosine vs dipyridamole vs regadenoson)
- Data can be collected in 2D vs 3D modes. Saturation of crystals is more problematic in 3D.
- Cardiac, respiratory and patient motion can degrade measurement accuracy .
- Different vendor software is used by different reading labs.
- Resting blood flow can be elevated due to pain, anxiety, lack of vagal tone, hypertension, etc. and can be normalized by using the rate pressure product (RPP) for calculation of myocardial perfusion reserve (MBF) the ratio of myocardial hyperemic flow/rest flow.

eviCore along with the American Society of Nuclear Medicine, the American College of Cardiology, and the Society of Nuclear Medicine and Metabolic Imaging agree that to minimize the above listed variables, AQMBF should only be approved when performed by **(all)**:

- Laboratories that are Intersocietal Accreditation Commission (IAC), American College of Radiology (ACR), or Joint Commission cardiac PET accredited.
- Interpreting physician(s) must be Board certified in Nuclear Cardiology (CBNC), Nuclear Medicine (ABNM), or Radiology (ABR) and have additional training in measuring AQMBF.
- Individual laboratories should have a standard protocol (same tracer, camera, software, stressor, model etc.) for use for all patients.
- Reports should contain rest myocardial blood flow (MBF) and stress MBF in ml/g/min, and myocardial blood flow reserve (MBFR) reported as the ratio of stress to rest MBF (with normal limits).
- Laboratories should have the ability to perform rate-pressure-product (RPP) correction of resting MBF when resting MBF is elevated due to elevated resting RPP and include mention of the true measured resting MBF and MBFR as well as the RPP-corrected resting MBF and RPP-corrected MBFR in the conclusions of the report.
- Health plans will be responsible for verifying requirements.

# Cardiac PET – Metabolic – Indications (CD-6.4)

CD.PET.0006.4.A

v2.0.2023

- Cardiac PET Metabolic (CPT® 78459 or CPT® 78429)
  - To determine myocardial viability when a previous study has shown significant left ventricular dysfunction when under consideration for revascularization
  - To diagnose strongly suspected cardiac sarcoid or monitor response to therapy for established cardiac sarcoid. See **Cardiac Sarcoidosis (CD-3.9)**
- Cardiac PET Metabolic and Perfusion (MPI SPECT CPT® 78451 and CPT® 78459, or CPT® 78432, or CPT® 78433)
  - To diagnose strongly suspected cardiac sarcoid or monitor response to therapy for established cardiac sarcoid. See **Cardiac Sarcoidosis (CD-3.9)**
- Full body PET/CT (CPT® 78815) is not indicated for the diagnosis or monitoring response to therapy of cardiac sarcoid. It may be considered to assist in diagnosis and/or treatment options in some instances of pulmonary sarcoid. See **Sarcoid (CH-15.1)** in the Chest Imaging Guidelines

## FDG PET/CT for infections (CD-6.5)

CD.PET.0006.5.A

v2.0.2023

- FDG PET/CT (CPT® 78815 or CPT® 78429) is indicated in the assessment of suspected prosthetic heart valve endocarditis when there is documentation of **both** of the following:
  - TTE and/or TEE are equivocal or non-diagnostic
  - Suspicion for prosthetic heart valve endocarditis remains high (all):
    - C-reactive protein  $\geq 40$  mg/L
    - No evidence of prolonged antibiotic therapy
    - The implantation was  $\geq 3$  months ago and there is no evidence of surgical adhesives used during the valve implantation
- FDG PET/CT for LVAD driveline infection (CPT® 78815 or 78429)
  - Early infection detection for LVAD drivelines is desirable, since once the infection extends to the cannula and pump pocket, eradication becomes difficult. CT findings are nonspecific and metal device artifacts of the driveline itself affects sensitivity.
  - FDG PET/CT can be approved for suspected LVAD infection if other studies and examination remain inconclusive.

## References (CD-6)

**v2.0.2023**

1. Einstein AJ, Moser KW, Thompson RC, et al. Radiation Dose to Patients from Cardiac Diagnostic Imaging. *Circulation*. 2007;116(11):1290-1305. doi:10.1161/circulationaha.107.688101.
2. Youssef G, Mylonas I, Leung E, et al. The Use of 18F-FDG PET in the Diagnosis of Cardiac Sarcoidosis: A Systematic Review and Metaanalysis Including the Ontario Experience. *Journal of Nuclear Medicine*. <http://jnm.snmjournals.org/content/53/2/241.long>. Published February 1, 2012.
3. Blankstein R, Osborne M, Naya M, et al. Cardiac Positron Emission Tomography Enhances Prognostic Assessments of Patients with Suspected Cardiac Sarcoidosis. *Journal of the American College of Cardiology*. 2014;63(4):329-336. doi:10.1016/j.jacc.2013.09.022.
4. Habib G, Lancellotti P, Antunes MJ, et al. 2015 ESC Guidelines for the management of infective endocarditis. *European Heart Journal*. 2015;36(44):3075-3128. doi:10.1093/eurheartj/ehv319.
5. Swart LE, Gomes A, Scholtens AM, et al. Improving the Diagnostic Performance of 18 F-Fluorodeoxyglucose Positron-Emission Tomography/Computed Tomography in Prosthetic Heart Valve Endocarditis. *Circulation*. 2018;138(14):1412-1427. doi:10.1161/circulationaha.118.035032.
6. Kim J, Feller ED, Chen W, Liang Y, Dilsizian V. FDG PET/CT for Early Detection and Localization of Left Ventricular Assist Device Infection. *JACC: Cardiovascular Imaging*. 2019;12(4):722-729. doi:10.1016/j.jcmg.2018.01.024.
7. Tam MC, Patel VN, Weinberg RL, et al. Diagnostic Accuracy of FDG PET/CT in Suspected LVAD Infections. *JACC: Cardiovascular Imaging*. 2020;13(5):1191-1202. doi:10.1016/j.jcmg.2019.04.024.
8. Harnett DT, Hazra S, Maze R, et al. Clinical performance of Rb-82 myocardial perfusion PET and Tc-99m-based SPECT in patients with extreme obesity. *J Nucl Cardiol*. 2017;26(1):275-283. doi:10.1007/s12350-017-0855-6.
9. Defining Adult Overweight and Obesity. Centers for Disease Control and Prevention. <https://www.cdc.gov/obesity/adult/defining.html>. Published March 3, 2021.
10. Gulati M, Levy PD, Mukherjee D, et al. 2021 AHA/ACC/ASE/CHEST/SAEM/SCCT/SCMR Guideline for the Evaluation and Diagnosis of Chest Pain: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines [published correction appears in *Circulation*. 2021 Nov 30;144(22):e455]. *Circulation*. 2021;144(22):e368-e454. doi:10.1161/CIR.0000000000001029

# Diagnostic Heart Catheterization

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# Diagnostic Heart Catheterization – Coding (CD-7.1) (CD-7.2)

CD.DHC.0007.1.A

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## Diagnostic Heart Catheterization – Code Sets (CD-7.1)

### Cardiac Catheterization Procedure Codes

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



Cardiac Cath Procedure	CPT®
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® 93593 to 93597 are appropriate for invasive evaluation of congenital heart disease. See specific conditions in **Adult Congenital Heart Disease (CD-11)**

### Diagnostic Heart Catheterization – Coding Notes (CD-7.2)

- Cardiac catheterization (CPT® 93451-CPT® 93461) includes all "road mapping" angiography necessary to place the catheters, including any injections and imaging supervision, interpretation, and report.
- Cardiac catheterization (CPT® 93452-CPT® 93461) (for all conditions other than congenital heart disease) includes contrast injections, imaging supervision, interpretation, and report for imaging typically performed.
- Catheter placements in native coronaries or bypass grafts (CPT® 93454-CPT® 93461) include intraprocedural injections for bypass graft angiography, imaging supervision, and interpretation.
- Injection codes CPT® 93563-CPT® 93565 should not be used in conjunction with CPT® 93452-CPT® 93461.
- Codes CPT® 93451-CPT® 93461 do not include contrast injections and imaging supervision, interpretation, and report for imaging that is separately identified by the following specific procedure codes: CPT® 93566, CPT® 93567 and CPT® 93568.
- Separate diagnostic cardiac catheterization codes should only be assigned in conjunction with interventional procedures in the following circumstances:
  - No prior or recent diagnostic catheterization is available to guide therapy
  - Individual's condition has significantly changed since the last diagnostic cath
  - The treatment plan may be affected

- Other vessels may be identified for treatment
- Further establishment of a diagnosis from a non-invasive study is necessary

# LHC – Unstable/Active Coronary Artery Syndromes (CD-7.3.1)

CD.DHC.0008.A

v2.0.2023

Diagnostic Left Heart Catheterization (LHC) is indicated for individuals in acute settings or with **active** unstable angina and should be handled as medical emergencies.

- LHC may be indicated for new onset, accelerating, or worsening ischemic symptoms suggestive of acute coronary syndrome (ACS) occurring at rest, or with minimal exertion resolving with rest, including:
  - Cardiac chest pain (typical angina) with or without new onset, evolving ischemic EKG changes
  - Symptoms consistent with the known angina pattern in an individual with a history of CABG or PCI
- Left and right heart cath may be indicated in place of a left heart cath if the above criteria has been met and there is documentation of any of the following:
  - The major component of the individual's symptoms is dyspnea
  - Newly diagnosed or worsening cardiomyopathy
  - For surgical planning prior to valve surgery or congenital heart defect repair

# Diagnostic Left Heart Catheterization (LHC) (CD-7.3)

CD.DHC.0007.3.A

v2.0.2023

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## LHC – Stable Established CAD Post Revascularization with CABG or PCI (CD-7.3.2)

These guidelines apply to individuals with stable conditions and who are not in the acute setting (acute coronary syndrome or unstable angina).

- Diagnostic Left Heart Catheterization (LHC) is indicated in patients with established Coronary Artery Disease (CAD) post revascularization by percutaneous coronary intervention (PCI) or coronary artery bypass graft surgery (CABG) when there is documentation of any **one** of the following:
  - New, recurrent, or worsening ischemic symptoms with documentation of **one** of the following:
    - Symptoms occur despite current treatment with **at least two** classes of anti-anginal medications (including beta blockers, calcium channel blockers, long-acting nitrates, ranolazine).
    - Intermediate or high risk findings on non-invasive stress testing as documented by **one** of the following:
      - Cardiac chest pain induced by exercise treadmill testing or dobutamine stress testing
      - Exercise treadmill testing inducing one of the following:
        - At least 1 mm downsloping ST-depression
        - 2 mm horizontal ST-depression
        - At least 1 mm ST-elevation in two leads
        - Ventricular tachycardia of at least 3 consecutive beats
      - Myocardial perfusion imaging (SPECT or PET) with ≥5% reversible ischemic burden
      - Stress echo with at least 2 segments of inducible ischemia
      - Severe stress-induced LV dysfunction (drop in LVEF with stress >10%)

- New or worsened left ventricular dysfunction
- New or worsened congestive heart failure
- Ventricular fibrillation
- Sustained ventricular tachycardia
- Unheralded syncope (not near syncope)
- Left and right heart cath may be indicated in place of a left heart cath if the above criteria has been met and there is documentation of **any** of the following:
  - The major component of the individual's symptoms is dyspnea
  - Newly diagnosed or worsening cardiomyopathy
  - For surgical planning prior to valve surgery or congenital heart defect repair

### **Stable Symptomatic Suspected or Established Coronary Artery Disease (CD-7.3.3)**

- Diagnostic left heart catheterization to screen for coronary artery disease (CAD) in asymptomatic individuals who are not anticipating other cardiac procedures is **not** medically necessary
- LHC with coronary arteriography is indicated when there is documentation of one of the following:
  - New onset, persistent, or worsening of cardiac chest pain (typical angina) and either:
    - Symptomatic failure of a 12 week trial of OMT including as tolerated all of the following:
      - Anti-platelet therapy
      - Statin and/or other lipid-lowering therapy
      - Anti-anginal therapy implemented to pursue a goal heart rate of 60 beats per minute or less
      - Anti-hypertensive therapy as may be indicated to pursue a goal systolic blood pressure (sbp) of less than 140 mmHg and a goal diastolic blood pressure (DBP) of less than 90 mmHg
    - Worsening of cardiac chest pain (typical angina) during 12 week trial of OMT
  - New onset, persistent, or worsening of cardiac chest pain (typical angina) and documentation of **both** of the following:
    - Intermediate/high pretest probability of CAD see **General Guidelines (CD-1.0)**, **Pre-Test Probability Grid (Table 1)** or **established CAD per CD-1.0**, and
    - Cardiac chest pain (typical angina) at a low level of exercise or at rest despite optimal medical therapy
  - LHC may be indicated irrespective of OMT for symptomatic individuals with any pre-test probability for coronary artery disease (CAD) who also have high-risk

findings on Coronary CT Angiography See **CCTA – Indications for CCTA (CPT® 75574) (CD-4.3)**, to include any of the following:

- Left main coronary artery stenosis  $\geq 40\%$
- Proximal or mid left anterior descending coronary artery stenosis  $\geq 70\%$
- Proximal or mid double-vessel coronary artery stenosis  $\geq 60\%$
- Proximal or mid triple-vessel coronary artery stenosis  $\geq 50\%$
- CT-FFR measured to be  $\leq 0.8$  in the proximal or mid segment of any coronary artery irrespective of degree of stenosis
- LHC may be indicated irrespective of OMT for symptomatic individuals who have BOTH high pretest probability of CAD see **General Issues (CD-1.0)**, Pre-Test Probability Grid (Table 1) and high-risk findings on non-invasive stress testing including any of the following:
  - Cardiac chest pain induced by exercise treadmill testing or dobutamine stress testing
  - Myocardial perfusion imaging with  $\geq 10\%$  reversible ischemic burden
  - Stress echo with at least 3 segments of inducible ischemia
  - Exercise treadmill testing inducing at least 2.5 mm downsloping ST-depression or 3 mm horizontal ST-depression in two leads
  - Ventricular tachycardia of at least 3 consecutive beats induced by an exercise treadmill test
- Left and right heart cath may be indicated in place of a left heart cath if the above criteria has been met and there is documentation of any of the following:
  - The major component of the individual's symptoms is dyspnea
  - Newly diagnosed or worsening cardiomyopathy
  - For surgical planning prior to valve surgery or congenital heart defect repair

### ***Background and supporting information***

In addition to OMT, physician-guided behavioral modification therapy (BMT) is recommended including all of the following:

- Mediterranean diet
- Moderate intensity physical activity for at least thirty minutes per day at least five times per week as possible
- Attempts at smoking cessation to include at least one of the following:
  - Cognitive behavioral therapy
  - Nicotine withdrawal replacement therapy

### **Exclusion of Significant Coronary Artery Disease Involvement in other Cardiac Pathology (CD-7.3.4)**

- LHC may be indicated when the etiology is unclear for **any** of the following:

- New or worsened left ventricular dysfunction or congestive heart failure if coronary artery disease is suspected
- Ventricular fibrillation or sustained ventricular tachycardia
- Unheralded syncope (not near syncope)
- Suspected myocarditis
- Left and right heart cath may be indicated in place of a left heart cath if the above criteria has been met and there is documentation of any of the following:
  - The major component of the individual's symptoms is dyspnea
  - Newly diagnosed or worsening cardiomyopathy
  - For surgical planning prior to valve surgery or congenital heart defect repair

### Evaluation of structural heart disease (CD-7.3.5)

- Evaluation prior to planned surgery
  - Ruling out coronary artery disease prior to planned non-coronary cardiac or great vessel surgery (i.e., cardiac valve surgery, aortic dissection, aortic aneurysm, congenital disease repair such as atrial septal defect, etc.).
  - Pre-organ transplant (non-cardiac) - in place of stress imaging for initial pre-transplant evaluation (per the transplant center's protocol) **or** if stress imaging is positive for ischemia. Repeat periodic screening while on a transplant waiting list (in the absence of other clinical indications) is not supported. See **Kidney Transplant, Pre-Transplant Imaging Studies (AB-42.5)**.
- Valvular heart disease when either:
  - There is a discrepancy between the clinical findings (history, physical exam, and non-invasive test results)
  - Valvular surgery is being considered.
- Suspected pericardial disease.
- Previous cardiac transplant:
  - Per transplant center protocol
  - To assess for accelerated coronary artery disease associated with cardiac transplantation.
- Left and right heart cath may be indicated in place of a left heart cath if the above criteria has been met and there is documentation of any of the following:
  - The major component of the individual's symptoms is dyspnea
  - Newly diagnosed or worsening cardiomyopathy
  - For surgical planning prior to valve surgery or congenital heart defect repair

# Right Heart Catheterization (RHC) (CD-7.4)

CD.DHC.0007.4.A  
v2.0.2023

## Diagnostic Right Heart Catheterization – Indications (CD-7.4.2)

- Diagnostic Right heart cath is indicated when results will impact the diagnosis and management of **any** of the following:
  - Atrial septal defect (ASD) including shunt detection and quantification
  - Ventricular septal defect (VSD) including shunt detection and quantification
  - Patent foramen ovale (PFO)
  - Anomalous pulmonary venous return
  - Congenital defects including persistent left vena cava
  - Pulmonary hypertension
  - Pericardial diseases (constrictive or restrictive pericarditis)
  - Valvular disease
  - Right heart failure
  - Left heart failure
  - Newly diagnosed or worsening cardiomyopathy
  - Preoperative evaluation for valve surgery
  - During a left heart cath where the etiology of the symptoms remains unclear
  - Pre-lung transplant to assess pulmonary pressures
  - Uncertain intravascular volume status with an unclear etiology
  - Assessment post-cardiac transplant
    - For routine endomyocardial biopsy
    - Assess for rejection
    - Assess pulmonary artery pressure
    - Can be done per the institution protocol or anytime organ rejection is suspected and biopsy is needed for assessment
  - Evaluation of right ventricular morphology.
  - Suspected arrhythmogenic right ventricular dysplasia.



***Background and supporting information*****General information RHC (CPT®93451) (CD-7.4.1)**

- It is performed most commonly from the femoral vein, less often through the subclavian, brachial, or internal jugular vein and inter-atrial septal puncture approach.
- It includes a full oximetry for detection and quantification of shunts.
- Cardiac outputs are calculated by several techniques including the Fick thermodilution

# Combined Right and Left Heart Catheterization Indications (CD-7.5)

CD.DHC.0007.5.A

v2.0.2023

- Preoperative evaluation for valve surgery
- The indications for **Diagnostic Left Heart Catheterization (LHC) (CD-7.3)** are met and **any** of the following are present:
  - The major component of the individual's symptoms is dyspnea
  - The indications are met according to **Right Heart Catheterization (RHC) (CD-7.4)**
  - Newly diagnosed or worsening cardiomyopathy

# Planned (Staged) Coronary Interventions (CD-7.6)

CD.DHC.0007.6.A

v2.0.2023

- The CPT® codes for percutaneous coronary interventions (PCI) include the following imaging services necessary for the procedure(s):
  - Contrast injection, angiography, 'road-mapping', and fluoroscopic guidance
  - Vessel measurement
  - Angiography following coronary angioplasty, stent placement, and atherectomy
- Separate codes for these services should not be assigned in addition to the PCI code/s because the services are already included.
- A repeat diagnostic left heart catheterization is not medically necessary when the individual is undergoing a planned staged percutaneous coronary intervention.

# Evaluation of Conditions other than Coronary Artery Disease (CD-7.7)

CD.DHC.0077.A

v2.0.2023

- Right and left heart catheterization (CPT® 93453) is indicated for any of the following:
  - Preoperative assessment prior to planned valvular surgery
  - Evaluation of pulmonary hypertension out of proportion to or unexplained by the severity of valvular disease documented by other non-invasive imaging modalities (i.e., echo, CMR)
  - Left ventricular dysfunction out of proportion to the severity of valvular disease documented by other non-invasive imaging modalities
  - Suspected pericardial tamponade as documented by clinical findings or other non-invasive imaging modalities
  - Suspected, or clinical uncertainty, between constrictive pericarditis vs. restrictive cardiomyopathy physiology when there are questions left unanswered by other cardiac non-invasive imaging modalities
  - Known or suspected cardiomyopathy with or without heart failure documented by prior advanced imaging
  - Re-evaluation of known cardiomyopathy for any of the following:
    - Change in clinical status
    - Change in cardiac exam
    - When required to guide therapy
  - Hypertrophic Cardiomyopathy
  - Subvalvular aortic stenosis
- Right and left heart catheterization (CPT® 93453) is indicated when there is uncertainty between clinical impression and other non-invasive imaging modalities to evaluate the following valvular diseases:
  - Mitral stenosis
  - Mitral regurgitation
  - Aortic stenosis
  - Aortic regurgitation
- Left heart catheterization (CPT® 93452) for hemodynamic evaluation of the left ventricle and aorta is indicated to evaluate aortic stenosis when there is uncertainty between the clinical impression and non-invasive imaging modality findings.

# References (CD-7)

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1. Olade RB. Cardiac Catheterization of Left Heart: Background, Indications, Contraindications. TheHeart.org. <https://emedicine.medscape.com>. Published January 7, 2017.
2. Amsterdam EA, Wenger NK, Brindis RG, et al. 2014 AHA/ACC Guideline for the Management of Patients with Non-ST-Elevation Acute Coronary Syndromes: Executive Summary: A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *Circulation*. 2014;130(25):2354-2394. doi:10.1161/cir.000000000000133.
3. Levine GN, Bates ER, Bittl JA, et al. 2016 ACC/AHA Guideline Focused Update on Duration of Dual Antiplatelet Therapy in Patients With Coronary Artery Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines: An Update of the 2011 ACCF/AHA/SCAI Guideline for Percutaneous Coronary Intervention, 2011 ACCF/AHA Guideline for Coronary Artery Bypass Graft Surgery, 2012 ACC/AHA/ACP/AATS/PCNA/SCAI/STS Guideline for the Diagnosis and Management of Patients with Stable Ischemic Heart Disease, 2013 ACCF/AHA Guideline for the Management of ST-Elevation Myocardial Infarction, 2014 AHA/ACC Guideline for the Management of Patients With Non-ST-Elevation Acute Coronary Syndromes, and 2014 ACC/AHA Guideline on Perioperative Cardiovascular Evaluation and Management of Patients Undergoing Noncardiac Surgery. *Circulation*. 2016;134(10). doi:10.1161/cir.0000000000000404.
4. Fihn SD, Gardin JM, Abrams J, et al. 2012 ACCF/AHA/ACP/AATS/PCNA/SCAI/STS Guideline for the Diagnosis and Management of Patients with Stable Ischemic Heart Disease. *J Am Coll Cardiol*. 2012;60(24). doi:10.1016/j.jacc.2012.07.013.
5. Fihn SD, Blankenship JC, Alexander KP, et al. 2014 ACC/AHA/AATS/PCNA/SCAI/STS Focused Update of the Guideline for the Diagnosis and Management of Patients with Stable Ischemic Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines, and the American Association for Thoracic Surgery, Preventive Cardiovascular Nurses Association, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. *Circulation*. 2014;130(19):1749-1767. doi:10.1161/cir.0000000000000095.
6. Boden WE, O'Rourke RA, Teo KK, et al. Impact of Optimal Medical Therapy With or Without Percutaneous Coronary Intervention on Long-Term Cardiovascular End Points in Patients With Stable Coronary Artery Disease (from the COURAGE Trial). *Am J Cardiol*. 2009;104(1):1-4. doi:10.1016/j.amjcard.2009.02.059.
7. Whelton PK, Carey RM, Aronow WS, et al. 2017 ACC/AHA/AAPA/ABC/ACPM/AGS/APhA/ASH/ASPC/NMA/PCNA guideline for the prevention, detection, evaluation, and management of high blood pressure in adults: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2018;71:e127-248. doi:10.1016/j.jacc.2017.11.00.
8. Maron DJ, Hochman JS, Reynolds HR, et al. Initial Invasive or Conservative Strategy for Stable Coronary Disease. *N Engl J Med*. 2020;382(15):1395-1407. doi:10.1056/nejmoa1915922.
9. Neumann FJ, Sousa-Uva M, Ahlsson A, et al. 2018 ESC/EACTS Guidelines on myocardial revascularization. *European Heart Journal*. 2018;40(2):87-165. doi:10.1093/eurheartj/ehy394.
10. Gulati M, Levy PD, Mukherjee D, et al. 2021 AHA/ACC/ASE/CHEST/SAEM/SCCT/SCMR Guideline for the Evaluation and Diagnosis of Chest Pain: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines [published correction appears in *Circulation*. 2021 Nov 30;144(22):e455]. *Circulation*. 2021;144(22):e368-e454. doi:10.1161/CIR.0000000000001029.
11. Patel MR, Bailey SR, Bonow RO, et al. ACCF/SCAI/AATS/AHA/ASE/ASNC/HFSA/HRS/SCCM/SCCT/SCMR/STS 2012 appropriate use criteria for diagnostic catheterization: American College of Cardiology Foundation Appropriate Use Criteria Task Force Society for Cardiovascular Angiography and Interventions American Association for Thoracic Surgery American Heart Association, American Society of Echocardiography American Society of Nuclear Cardiology Heart Failure Society of America Heart Rhythm Society, Society of Critical Care Medicine Society of Cardiovascular Computed Tomography Society for Cardiovascular Magnetic Resonance Society of Thoracic Surgeons. *Catheter Cardiovasc Interv*. 2012;80(3):E50-E81. doi:10.1002/ccd.24467.
12. Nishimura RA, Carabello BA. Hemodynamics in the cardiac catheterization laboratory of the 21st century. *Circulation*. 2012;125(17):2138-2150. doi:10.1161/CIRCULATIONAHA.111.060319.
13. Patel MR, Calhoun JH, Dehmer GJ, et al. ACC/AATS/AHA/ASE/ASNC/SCAI/SCCT/STS 2017 Appropriate Use Criteria for Coronary Revascularization in Patients With Stable Ischemic Heart Disease: A Report of the American College of Cardiology Appropriate Use Criteria Task Force, American Association for Thoracic Surgery, American Heart Association, American Society of Echocardiography, American Society of Nuclear Cardiology, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular

- Computed Tomography, and Society of Thoracic Surgeons [published correction appears in *J Am Coll Cardiol*. 2018 Apr 13;:]. *J Am Coll Cardiol*. 2017;69(17):2212-2241. doi:10.1016/j.jacc.2017.02.001.
14. Kidney Disease: Improving Global Outcomes (KDIGO) Kidney Transplant Candidate Work Group. KDIGO Clinical Practice Guideline on the Evaluation and Management of Candidates for Kidney Transplantation. 2020;104: S1 – S103.

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# Congenital heart disease – General Information (CD-11.1)

CD.CHD.0011.1.A

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- This section covers adult congenital heart disease (CHD), for other associated disorders please see the condition specific sections
  - Marfan Syndrome
  - Hypertrophic cardiomyopathy (HCM)
  - Bicuspid aortic valve (BAV)

## Definitions (CD-11.1.1)

- Physiological stages (A, B, C, D)
  - Each congenital heart lesion is divided into 4 physiological stages (A, B, C, D)

Characteristics	Physiological stage			
	A	B	C	D
<b>NYHA functional class</b>	<b>I</b>	<b>II</b>	<b>III</b>	<b>IV</b>
Hemodynamic or anatomic sequelae	None	Mild ventricular enlargement of dysfunction, small shunt	Moderate or greater, ventricular dysfunction. Any venous or arterial stenosis	Moderate or greater, ventricular dysfunction. Any venous or arterial stenosis
Valvular	None	Mild	Moderate or greater	Moderate or greater
Aortic enlargement	None	Mild	Moderate	Severe
Exercise capacity limitation	Normal	Abnormal objective cardiac limitation	Moderate	Severe



Characteristics	Physiological stage			
	A	B	C	D
Renal hepatic pulmonary dysfunction	None		Mild but responsive to medication	Refractory to treatment
Cyanosis/hypoxemia	None		Mild	Severe
Arrhythmias	None	Arrhythmia not requiring treatment	Needs rx	Refractory to rx
Pulmonary hypertension	None		Mild to moderate	Severe or Eisenmenger

- CHD Anatomic classification
  - Class I-Simple
    - Native disease
      - Isolated small ASD
      - Isolated small VSD
      - Mild isolated pulmonic stenosis
    - Repaired conditions
      - Previously ligated or occluded ductus arteriosus
      - Repaired secundum ASD or sinus venosus defect without significant residual shunt or chamber enlargement
      - Repaired VSD without significant residual shunt or chamber enlargement
  - Class II-Moderate Complexity
    - Repaired or unrepaired conditions
      - Aorto-left ventricular fistula
      - Anomalous pulmonary venous connection, partial or total
      - Anomalous coronary artery arising from the pulmonary artery
      - Anomalous aortic origin of a coronary artery from the opposite sinus
      - AVSD (partial or complete, including primum ASD)
      - Congenital aortic valve disease
      - Congenital mitral valve disease
      - Coarctation of the aorta
      - Ebstein anomaly (disease spectrum includes mild, moderate, and severe variations)
      - Infundibular right ventricular outflow obstruction
      - Ostium primum ASD
      - Moderate and large unrepaired secundum ASD

- Moderate and large persistently patent ductus arteriosus
- Pulmonary valve regurgitation (moderate or greater)
- Pulmonary valve stenosis (moderate or greater)
- Peripheral pulmonary stenosis
- Sinus of Valsalva fistula/aneurysm
- Sinus venosus defect
- Subvalvular aortic stenosis (excluding HCM; HCM not addressed in these guidelines)
- Supravalvular aortic stenosis
- Straddling atrioventricular valve
- Repaired tetralogy of Fallot
- VSD with associated abnormality and/or moderate or greater shunt
- Class III- Great Complexity (or Complex)
  - Cyanotic congenital heart defect (unrepaired or palliated, all forms)
  - Double-outlet ventricle
  - Fontan procedure
  - Interrupted aortic arch
  - Mitral atresia
  - Single ventricle (including double inlet left ventricle, tricuspid atresia, hypoplastic left heart, any other anatomic abnormality with a functionally single ventricle)
  - Pulmonary atresia (all forms)
  - TGA (classic or d-TGA; CCTGA or l-TGA)
  - Truncus arteriosus
  - Other abnormalities of atrioventricular and ventriculoarterial connection (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

### Modalities (CD-11.1.2)

- Echocardiogram- transthoracic (TTE) or transesophageal (TEE)
  - Transthoracic echocardiography (TTE) is an indispensable tool in the initial and serial follow-up evaluation to identify abnormalities and changes that commonly influence management decisions.
- Cardiac MRI (CMR)
  - CMR plays a valuable role in assessment of RV size and function, because it provides data that are reproducible and more reliable than data obtained with alternative imaging techniques
  - For intracardiac congenital heart disease, CMR will typically include flow velocity mapping for shunts and flow assessment.
  - Imaging that only requires aortic arch imaging, does not require intracardiac CMR, only MRA Chest.

- Cardiac Computed Tomography (CCT) and Cardiac Computed Tomography Angiography (CCTA)
  - The most important disadvantage of CCT (including CT angiography) as an imaging technique is the associated exposure to ionizing radiation.
- Cardiac catheterization
  - (hemodynamic and/or angiographic) in individuals with adult CHD AP classification II and III, or interventional cardiac catheterization in individuals with adult CHD AP classification I to III should be performed by, or in collaboration with, cardiologists with expertise in adult CHD
- Exercise Testing
  - Exercise test does not imply stress imaging
- Stress Imaging
  - Includes-MPI, stress echo, stress MRI
  - PET stress may be included as per **Cardiac PET (CD-6)**
- Circumstances where CMR, CCT, TEE, and/or Cardiac Catheterization may be Superior to TTE
  - Assessment of RV size and function in repaired Tetralogy of Fallot (TOF), systemic right ventricles, and other conditions associated with right ventricular (RV) volume and pressure overload
  - Identification of anomalous pulmonary venous connections
  - Serial assessment of thoracic aortic aneurysms, especially when the dilation might extend beyond the echocardiographic windows
  - Accurate assessment of pulmonary artery (PA) pressure and pulmonary vascular resistance
  - Assessment for re-coarctation of the aorta
  - Sinus venosus defects
  - Vascular rings
  - Evaluation of coronary anomalies
  - Quantification of valvular regurgitation

### Coding (CD-11.1.3)

Modality	
Echocardiogram	
Transthoracic echocardiogram (TTE)	CPT®
TTE for congenital cardiac anomalies; complete	93303
TTE for congenital cardiac anomalies; limited study	93304

Modality	
TTE (2D) m-mode recording, complete, with spectral and color flow doppler echocardiography	93306
TTE (2D) with or without m-mode recording; complete	93307
TTE (2D) with or without m-mode recording; limited study	93308
Transesophageal echocardiogram (TEE)	
TEE (2D) including probe placement, imaging, interpretation, and report	93312
TEE for congenital cardiac anomalies; including probe placement, imaging, interpretation, and report	93315
<b>MRI</b>	
cardiac (CMR)	CPT®
Cardiac MRI for morphology and function without contrast	75557
Cardiac MRI for morphology and function without and with contrast	75561
MRI Chest	
MRI Chest without contrast	71550
MRI Chest with contrast	71551
MRI Chest with & without contrast	71552
MRI Angiography (MRA) MRA Chest	
MRA Chest (excluding myocardium) with or without contrast	71555
<b>CT</b>	
Cardiac (CCT)	CPT®
CT, Heart, with contrast material, for evaluation of cardiac structure and morphology	75572
CT, Heart, with contrast material, for evaluation of cardiac structure and morphology in the setting of congenital heart disease	75573
CT Angiography-cardiac (CCTA)	CPT®

Modality	
CTA Heart, coronary arteries and bypass grafts (when present), with contrast, including 3D image post processing	75574
CT-Chest	CPT®
CT Thorax without contrast	71250
CT Thorax with contrast	71260
CT Thorax without & with contrast	71270
CT Angiography-Chest (CTA Chest)	CPT®
CTA Chest without and with contrast	71275
<b>Stress Imaging (echo, MRI, MPI)</b>	
Stress echo	CPT®
Echocardiography (TTE), (2D), with or without m-mode, during rest and cardiovascular stress, with interpretation and report	93350
Echocardiography (TTE), (2D), m-mode, during rest and cardiovascular stress test using treadmill, bicycle exercise and/or pharmacologically induced stress, with interpretation	93351
Stress MRI	CPT®
Cardiac MRI for morphology and function without contrast, with stress imaging	75559
Cardiac MRI for morphology and function without and with contrast, with stress imaging	75563
Myocardial perfusion imaging (MPI)	CPT®
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)	78451

Modality	
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	78452
Pulmonary perfusion imaging	CPT®
Pulmonary perfusion imaging (e.g., particulate)	78580
Pulmonary ventilation (e.g., aerosol or gas) and perfusion imaging	78582
Quantitative differential pulmonary perfusion, including imaging when performed	78597
Quantitative differential pulmonary perfusion and ventilation (e.g., aerosol or gas), including imaging when performed	78598

# Congenital Heart Disease Imaging Indications (CD-11.2)

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- The following sections are based on the congenital heart lesion. Requests for imaging based on other cardiac conditions, such as CAD, HCM, acquired valvular lesions, should follow the adult cardiac guidelines for those conditions.

# ASD-Atrial septal defects (CD-11.2.1)

CD.CHD.0011.2.1.A

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## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram at time of diagnosis
  - CMR, CCT (CPT® 75573), and/or TEE are useful if echo (TTE) is suboptimal and either:
    - ASD is suspected
    - To evaluate pulmonary venous connections in known ASD
  - MRA Chest or CTA Chest may be indicated if echo shows pulmonary venous anomalies
    - If normal, repeat pulmonary vein imaging is not required.
- Transesophageal echocardiogram (TEE) is recommended to guide percutaneous ASD closure
- Diagnostic cath is indicated when there is either:
  - Evidence of pulmonary hypertension
  - Unanswered questions on CMR/CCT for venous drainage.

## Post procedure imaging

- TTE is indicated post ASD device placement:
  - 6 months to evaluate for erosion
  - 1 week (if amplatzer)
  - 1 month
  - 6 months
  - 12 months
  - then every 1-2 years
- Due to low risk of erosion in PFO devices- PFO device closure requires follow-up at 6-12 months. No additional evaluation unless PFO not closed

Stress imaging and coronary artery imaging would be based on **Stress Testing with Imaging – Indications (CD-1.4)**



**Follow-up ASD if surgically closed or if no interventions**

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
TTE	36	24	12	12

# Anomalous Pulmonary Venous Connections (CD-11.2.2)

CD.CHD.0011.2.2.A

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## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram at time of diagnosis
  - CMR and/or MRA Chest, or CT Cardiac and/or CTA Chest at time of diagnosis if any issues with pulmonary veins or RV volume.
  - Cardiac Cath at time of diagnosis for hemodynamic data and issues not answered on other imaging
- Routine stress imaging or coronary artery imaging not required.
- Echo, CMR, CT, per cardiology request for clinical changes
  - Diagnostic heart catheterization if questions unanswered on imaging

## Follow-up Anomalous Pulmonary Venous Connections

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
Echo (TTE)	36	24	12	12

# Ventricular Septal Defect (VSD) (CD-11.2.3)

CD.CHD.0011.2.3.A

v2.0.2023

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echo (TTE) at time of diagnosis
  - CMR or CCT can be performed if questions are unanswered on echo
  - Catheterization at time of diagnosis for hemodynamics if pulmonary hypertension (PHT) or shunt size is a question

## Long term follow-up VSD

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
Echo (TTE)	36	24	12	12

# Atrioventricular Septal Defect (AV Canal, AVSD, endocardial cushion defect) (CD-11.2.4)

CD.CHD.0011.2.4.A  
v2.0.2023

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echo (TTE) at time of diagnosis
  - CMR or CT Cardiac at time of diagnosis if there are unanswered questions on echo
  - Cardiac cath at time of diagnosis when CMR and TTE leave questions unanswered that affect individual management
- Stress imaging per **Stress Testing with Imaging – Indications (CD-1.4)**

## Long term follow-up -AVSD

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
Echo (TTE)	24	24	12	12

# Patent Ductus Arteriosus (PDA) (CD-11.2.5)

CD.CHD.0011.2.5.A

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## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echo at time of diagnosis
  - MR Chest or CT Chest if there are questions left unanswered by echo
  - Cardiac Cath for hemodynamics (if planned device closure, diagnostic cardiac cath is not indicated as it is included in the procedure code)
- Stress imaging per **Stress Testing with Imaging – Indications (CD-1.4)**

## Long term follow-up PDA

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
Echo (TTE)	36	24	12	12

## Cor Triatriatum (CD-11.2.6)

CD.CHD.0011.2.6.A

v2.0.2023

### Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
  - CMR and/or MRA Chest or CT Cardiac and/or CTA Chest may be approved
  - Diagnostic cath may be approved if additional information is required for medical management

### Long term follow-up

- Stress imaging per **Stress Testing with Imaging – Indications (CD-1.4)**

# Congenital Mitral Stenosis (CD-11.2.7)

CD.CHD.0011.2.7.A

v2.0.2023

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis

## Long term follow-up congenital mitral stenosis

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
Echo (TTE)	24	24	12	12

# Subaortic Stenosis (SAS) (CD-11.2.8)

CD.CHD.0011.2.8.A

v2.0.2023

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
- Stress imaging (stress echo or stress MRI) for any of the following:
  - Once at the time of diagnosis
  - New or changed signs or symptoms of ischemia
  - Changes in cardiac function
  - If cardiac intervention is being considered
  - Any signs or symptoms allowed in **Stress Testing with Imaging – Indications (CD-1.4)**

## Long term follow-up SAS

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
Echo (TTE)	24	24	12	12
Stress imaging		24	24	12



# Congenital Valvular Aortic Stenosis (CD-11.2.9)

CD.CHD.0011.2.9.A

v2.0.2023

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
- TEE may be required if TTE limited or equivocal
- MRA Chest or CTA Chest if one of the following:
  - Suspicion of Coarctation based on exam and echocardiogram
  - Proximal ascending aorta not well visualized on TTE

## Routine follow-up Congenital Valvular Aortic Stenosis

Modality	Physiological stage / intervals for routine imaging			
Stage (valvular AS)	Progressive (stage B) Mild Vmax 2.0-2.9 m/s	Progressive (stage B) Moderate Vmax 3.0-3.9 m/s	Severe (stage C) $\geq 4.0$ m/s	Aortic root dilation $>4.5$ cm
echo (TTE)	3 years	1 years	6 months	12 months
MRA Chest or CTA				if ascending allowed yearly

Degree of aortic stenosis (AS) severity			
	Mild AS	Moderate AS	Severe AS
Vmax (m/s) <sup>a</sup> maximum Doppler velocity	2.0-2.9	3.0-3.9	$\geq 4.0$
Mean gradient (mmHg) <sup>a</sup>	$<30$	30-49	$\geq 50$
AVA (cm <sup>2</sup> ) aortic valve area	$>1.5$	1.0-1.5	$<1.0$

Degree of aortic stenosis (AS) severity			
	Mild AS	Moderate AS	Severe AS
AVA <sub>i</sub> (cm <sup>2</sup> /m <sup>2</sup> BSA) indexed aortic valve area	≥1.0	0.6-0.9	<0.6
<sup>a</sup> At normal transvalvular flow, BSA= body surface area			

Adapted from: ESC Guidelines for the management of grown-up congenital heart disease (new version 2010): The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC).

# Aortic disease in Turner Syndrome (CD-11.2.10)

CD.CHD.0011.2.10.A

v2.0.2023

Dissection more common for a given aortic diameter. Mid-ascending aortic disease more common and may not be reliably seen on echocardiogram

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
- MRA Chest or CTA Chest to rule out mid ascending aortic aneurysm if mid aorta was not seen on echocardiogram.

## Surveillance

- Echocardiogram (TTE) yearly
  - MRA Chest or CTA if mid ascending aorta not visualized
- For documented thoracic aortic aneurysm (TAA)  $\leq 4$ cm
  - Routine MRA Chest or CTA yearly
- For documented thoracic aortic aneurysm (TAA)  $> 4$ cm
  - MRA Chest or CTA every 6 months.

## Aortopathies with CHD (CD-11.3)

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**v2.0.2023**

- Dilated aortic arches are not uncommon with several congenital heart diseases and postoperative procedures including- Aortic stenosis, Ross repair, Tetralogy of Fallot, Transposition of the great arteries (TGA), Pulmonary atresia, hypoplastic left heart syndrome (HLHS), Truncus Arteriosis, single ventricle.

# Supravalvular Aortic Stenosis (CD-11.3.1)

CD.CHD.0011.3.1.A

v2.0.2023

Supravalvular aortic stenosis is a relatively rare condition overall but is seen commonly in individuals with Williams syndrome or homozygous familial hypercholesterolemia.

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
- MRA Chest or CTA Chest
- Cardiac MRI or CTA Cardiac to assess coronary ostia
- Cardiac cath for any individuals pre cardiac intervention for coronary arteries
- New cardiac symptoms-any of the following:
  - CT Cardiac or cardiac MR
  - CTA Chest or MRA Chest
  - Stress imaging as per **Stress Testing with Imaging – Indications (CD-1.4)**

## Routine follow-up supravalvular AS

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
TTE	24	24	12	12
CMR or CCT	36	36	36	36

# Coarctation of the Aorta (CD-11.3.2)

CD.CHD.0011.3.2.A

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Coarctation is suspected based on clinical findings:

- BP higher in upper extremities than in the lower extremities
- Absent femoral pulses
- Continuous murmur
- Abdominal bruit
- Berry aneurysm with hemorrhage
- Rib notching on x-ray
- Abnormal thoracic aortic imaging and blood pressures

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
  - No further imaging is required if echocardiogram (TTE), blood pressure, and exam rule out Coarctation.
  - If echo and exam are equivocal or positive one of the following is indicated:
    - CTA Chest
    - MRA Chest
  - Individuals with Coarctation of the aorta do not require intracardiac MR unless issue cannot be resolved on echocardiogram.
  - Screening for intracranial aneurysm by MRA or CTA of head is allowed
- ETT for diagnosis of exercise induced hypertension does not require imaging
- Cardiac MR not required unless issues unresolved by echo for intracardiac anatomy
- Diagnostic cath can be approved prior to stenting of the coarctation
- Stress imaging, TEE, Cardiac MR or CT, Coronary imaging not routine

## Symptomatic

- Individuals with Coarctation are at risk for dissection. When individual has new or worsening symptoms any of the following:
  - Echocardiogram (TTE)
  - MRA Chest or CTA.
- For exertional symptoms, one of the following:
  - Stress imaging-per **Stress Testing with Imaging – Indications (CD-1.4)**
  - Cardiac MRI or CT Cardiac

**Routine follow-up Coarctation of the Aorta**

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
TTE	24	24	12	12
MRA Chest or CTA Chest	36	36	12	12

# Valvular Pulmonary Stenosis (CD-11.3.3)

CD.CHD.0011.3.3.A

v2.0.2023

## Overview Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
- For issues affecting management not well visualized on TTE
  - Cardiac MRI or CT Cardiac
  - MRA Chest or CTA Chest

## Valvular PS routine follow-up and testing.

- Echocardiogram-stages
  - Mild PS – peak gradient <36 mmHg (peak velocity < 3m/s)
  - Moderate PS- peak gradient 36-64 mmHg (peak velocity 3-4 m/s)
  - Severe PS- peak gradient >64 mmHg (peak velocity > 4 m/s); or mean gradient >35 mmHg.
- Routine stress imaging is not required
- Routine chest or cardiac or ischemia workup not required.

## Valvular PS routine imaging

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
TTE	36	24	12	12

## Isolated Pulmonary regurgitation after PS repair-Echo and CMR at same interval as TOF

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
TTE	24	12	12	12
CMR	36	24	12	12



# Branch and Peripheral pulmonary stenosis (CD-11.3.4)

CD.CHD.0011.3.4.A

v2.0.2023

## Overview

- Can be seen in newborns as a normal variant in the first 6 months of life
- Can be seen in surgeries of right ventricular outflow (TOF)
  - Noonan
  - Alagille
  - Williams
  - Maternal rubella exposure
  - Keutel syndrome

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
- Baseline MRA Chest or CTA Chest
- Cath may be considered if other advanced imaging is not adequate for management
- VQ scan or MRA Chest for differential blood flow

## Routine follow-up branch and peripheral pulmonary stenosis

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
TTE	24	24	12	12
Cardiac MRI or CT Cardiac	36	36	24	24
MRA Chest or CTA Chest	36	36	24	24

## Double chambered RV (CD-11.3.5)

CD.CHD.0011.3.5.A

v2.0.2023

### Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis

### Routine follow-up double chambered right ventricle (RV)

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
Echo (TTE)	24	24	12	12

# Ebstein Anomaly (CD-11.3.6)

CD.CHD.0011.3.6.A

v2.0.2023

## Overview Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
- TEE if either:
  - TTE is not adequate
  - If surgery/intervention planned
- Cardiac MRI or CT Cardiac at time of Diagnosis

## Routine follow-up Ebstein Anomaly

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
Echo (TTE)	12	12	12	12
Cardiac MRI or CT Cardiac	60	36	24	12

# Tetralogy of Fallot (TOF, VSD with PS) (CD-11.3.7)

CD.CHD.0011.3.7.A

v2.0.2023

Includes TOF with pulmonary atresia, VSD PA

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
- Cardiac MR or CTA Cardiac at time of diagnosis
- MRA Chest or CTA Chest at time of diagnosis
- Cardiac catheterization if other advanced imaging leaves unanswered questions

## Prior to cardiac intervention or surgery

- Repeat imaging Echo/MR/CT
- Cath prior to surgery or intervention
  - If planned Catheter Pulmonary Valve replacement, procedure includes diagnostic cath and hemodynamics and diagnostic cath is not billed separately

## New or worsening symptoms

- Repeat advanced imaging
  - New or worsening symptoms
  - New EKG changes
- Stress imaging (stress echo, stress MRI, or MPI) allowed for typical chest pain, even if intermediate pretest probability at atypical symptoms in individuals with known or undefined coronary artery (CA) anatomy or CA pathology
- VQ scan or MRA chest for left/right perfusion abnormality

## Routine Follow-up Tetralogy of Fallot (TOF)

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
TTE	24	12	12	12
Cardiac MRI or CCTA	36	24	12	12

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
CTA Chest or MRA	36	24	12	12

# Right Ventricle-to-Pulmonary Artery Conduit (CD-11.3.8)

CD.CHD.0011.3.8.A

v2.0.2023

**Initial studies-Diagnosis, clinical changes, consideration of surgery. Surgical repair for many lesions such as TOF/ Truncus /Pulmonary atresia**

- Echocardiogram (TTE) at time of diagnosis
- Cardiac MRI or CTA Cardiac
- MRA Chest or CTA Chest
- Prior to interventions or surgery may repeat any of the above imaging
- Cath allowed for new symptoms or with new imaging findings as needed for management
- Stress imaging (stress echo, stress MRI or MPI) as requested for symptoms

## Routine follow-up Right Ventricle-to-Pulmonary Artery Conduit

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
TTE	12	12	12	12
CMR or CCTA	36	36	12	12
MRA Chest or CTA Chest	36	36	12	12

# Transposition of the great arteries (TGA) (CD-11.3.9)

CD.CHD.0011.3.9.A

v2.0.2023

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
- Baseline Cardiac MRI or CCTA
- Baseline MRA Chest or CTA
- Stress imaging as requested for symptoms or signs of ischemia
- V/Q scan for left to right PA perfusion or MRA Chest
- Symptomatic individuals should be offered stress physiological imaging and repeat anatomic imaging considered if symptoms are suggestive of coronary ischemia (regardless of diamond forester pretest probability category)
- Cath right and left heart when issues not elucidated on advanced imaging

## Routine follow-up TGA

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
TTE	12	12	12	12
CMR or CCTA	36	24	12	12
MRA Chest or CTA Chest	36	24	12	12

# Congenitally corrected TGA (CD-11.3.10)

CD.CHD.0011.3.10.A

v2.0.2023

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis
- Baseline CMR and MRA Chest
- CMR and/or Echo for changes in clinical status

## Routine follow-up congenitally corrected TGA

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
Echo (TTE)	12	12	12	12
CMR or CCTA	36	36	12	12
CTA Chest or MRA Chest	36	36	12	12



# Fontan Palliation of Single Ventricle Physiology (CD-11.3.11)

CD.CHD.0011.3.11.A

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Including Tricuspid Atresia and Double Inlet Left Ventricle, HLHS, HRHS, PA, Mitral atresia, AVC unbalanced, single ventricle, DIRV, pulmonary atresia, HLHS, Glen procedure, TA, double outlet right ventricle (DORV), and single ventricle physiology

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE) at time of diagnosis and with any new Symptoms
- CMR or CCTA can be done annually (vs. based on below chart) on individuals who have prior issues that were equivocal on echo, and the data is required (i.e. very poor windows)
  - Cardiac catheterization prior to surgical interventions
- Echo/CMR or CCTA/MRA Chest or CTA Chest/cath with any new signs or symptoms
- V/Q scan or MRA for lung perfusion left vs. right

## Routine follow-up Fontan Palliation of Single Ventricle Physiology

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
Echo (TTE)	12	12	12	12
CMR or CT Cardiac	36	24	24	24
CTA Chest or MRA	36	24	24	24

# Severe Pulmonary artery hypertension (PHT) and Eisenmenger syndrome (CD-11.3.12)

CD.CHD.0011.3.12.A

v2.0.2023

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echo (TTE)
  - Initial diagnosis
  - With new signs or symptoms
- Cardiac cath
  - Echo (TTE) results suggest PHT
  - New signs or symptoms with PHT

## Long term follow-up Severe Pulmonary artery hypertension (PHT) and Eisenmenger syndrome

Modality	Physiological stage / intervals for routine imaging (months)			
Physiological stage	A	B	C	D
TTE			6	6
CMR or CCT			As needed	As needed
MRA Chest or CTA Chest			As needed	As needed
Cath			As needed	As needed

# Coronary artery anomalies (CD-11.3.13)

CD.CHD.0011.3.13.A

v2.0.2023

## Initial studies-Diagnosis, clinical changes, consideration of surgery

- Echocardiogram (TTE)
  - At baseline
  - Any signs or symptoms
- Coronary CT/MR/Cath for initial evaluation
- CA from wrong sinus-baseline stress imaging regardless of symptoms
- Stress imaging for any cardiac signs or symptoms
- For Kawasaki GL regarding echo, Stress imaging, coronary imaging, see pediatric GL **Kawasaki Disease (PEDCD-6)**

# Pregnancy – Maternal Imaging (CD-11.4)

CD.DHC.0011.4.A

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- Overview
  - World Health Organization (WHO) classification:
    - WHO classification I: no detectable increased risk of maternal mortality and no/mild increase in morbidity.
      - Uncomplicated small or mild pulmonary stenosis
      - Patent Ductus Arteriosus (PDA)
      - Mitral valve prolapse
      - Successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous connection)
    - WHO classification II: small increase in maternal risk mortality or moderate increase in morbidity.
      - Unrepaired atrial or ventricular septal defect
      - Repaired tetralogy of Fallot
    - WHO classification II–III (depending on individual)
      - Mild left ventricular impairment
      - Native or tissue valvular heart disease not considered WHO I or IV
      - Marfan syndrome without aortic dilation
      - Aorta <45 mm in association with bicuspid aortic valve disease
      - Repaired coarctation
    - WHO classification III: significantly increased risk of maternal mortality or severe morbidity. Expert counseling required. If pregnancy is decided upon, intensive specialist cardiac and obstetric monitoring needed throughout pregnancy, childbirth and the puerperium.
      - Mechanical valve
      - Systemic right ventricle
      - Fontan circulation
      - Unrepaired cyanotic heart disease
      - Other complex congenital heart disease
      - Aortic dilation 40–45 mm in Marfan syndrome
      - Aortic dilation 45–50 mm in bicuspid aortic valve disease
    - WHO classification IV: extremely high risk of maternal mortality or severe morbidity; pregnancy contraindicated. If pregnancy occurs, termination should be discussed. If pregnancy continues, care as for WHO class III.
      - Pulmonary arterial hypertension from any cause

- Severe systemic ventricular dysfunction (LVEF <30%, NYHA functional class III–IV)
- Severe mitral stenosis; severe symptomatic aortic stenosis
- Marfan syndrome with aorta dilated >45 mm
- Aortic dilation >50 mm in aortic disease associated with bicuspid aortic valve
- Native severe coarctation of the aorta

Adapted from: Elkayam U, Goland S, Pieper PG, Silversides CK. High-Risk Cardiac Disease in Pregnancy. Journal of the American College of Cardiology..

- Congenital heart disease imaging in pregnancy
  - Echocardiogram (TTE) when planning pregnancy
  - TEE if TTE equivocal
  - CMR can be performed prior to planning pregnancy in those lesions where CMR would be routinely performed at some later date
  - CTA Chest or MRA Chest of arch if known disease with aortic involvement or if known dilation
  - Repeat echocardiogram and MR (can be without gad) can be performed based on the II, III, IV, or other risk factors
  - Severe complex CHD, may require echo monthly, or even weekly (every two weeks) (major physiological changes)-may be best as often as needed (Pulmonary hypertension, changes in function, can guide delivery after 24 weeks)
  - Echo can be performed if new signs or Symptoms during pregnancy
  - Postpartum first year can have more frequent imaging
  - Stress imaging pre/during pregnancy for individuals with known Coronary artery anomaly, pulmonary hypertension, LVOT obstruction, cardiac dysfunction, single ventricle.
  - WHO II, III, IV, can have echo/MR/CT/stress imaging prior to pregnancy
  - WHO I- one echocardiogram during pregnancy
  - WHO II- one echocardiogram per trimester during pregnancy
  - WHO II/III- echocardiogram every 2 months during pregnancy
  - WHO III/IV- echocardiogram monthly during pregnancy
    - Individuals may require more (even weekly) if treatment decision, delivery is considered.
- Syndromes that allow cardiac imaging at the time of diagnosis if not previously done. This list is not exhaustive
  - DiGeorge/velocardiofacial (22q11.2)
  - Down syndrome (trisomy 21)
  - Holt Oram (TBX5)
  - Klinefelter syndrome (47 XXY)

- Noonan (PTPN11, KRAS, SOS1 RAF1, NRAS, BRAF, MAP2K1)
- Turner (45X)
- Williams (7q11.23 deletion)
- Any syndrome associated with congenital heart disease.
- Echocardiogram at time of Diagnosis (either genetic testing or clinical features)
- CMR or CCTA if arch involved in disease.
- See **Maternal Imaging in Cardiovascular Disease (CD-15)**

# References (CD-11)

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1. Mcelhinney DB, Quartermain MD, Kenny D, Alboliras E, Amin Z. Relative Risk Factors for Cardiac Erosion Following Transcatheter Closure of Atrial Septal Defects. *Circulation*. 2016;133(18):1738-1746. doi:10.1161/circulationaha.115.019987.
2. Center for Devices and Radiological Health. 2018 Meeting Materials of the Circulatory System Devices Panel. U.S. Food and Drug Administration. <https://www.fda.gov/advisory-committees/circulatory-system-devices-panel/2018-meeting-materials-circulatory-system-devices-panel>. Published December 3, 2018.
3. Doherty JU, Kort S, Mehran R, Schoenhagen P, Soman P. ACC/AATS/AHA/ASE/ASNC/HRS/SCAI/SCCT/SCMR/STS 2019 Appropriate Use Criteria for Multimodality Imaging in the Assessment of Cardiac Structure and Function in Nonvalvular Heart Disease. *J Nucl Cardiol*. 2019;26(4):1392-1413. doi:10.1007/s12350-019-01751-7.
4. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Circulation*. 2019;139(14). doi:10.1161/cir.0000000000000603
5. Silvestry FE, Cohen MS, Arnsby LB, et al. Guidelines for the Echocardiographic Assessment of Atrial Septal Defect and Patent Foramen Ovale: From the American Society of Echocardiography and Society for Cardiac Angiography and Interventions. *J Am Soc Echocardiogr*. 2015;28(8):910-958. doi:10.1016/j.echo.2015.05.015.
6. El-Said HG, Bratincsak A, Foerster SR, et al. Safety of Percutaneous Patent Ductus Arteriosus Closure: An Unselected Multicenter Population Experience. *Journal of the American Heart Association*. 2013;2(6). doi:10.1161/jaha.113.000424.
7. Franklin RCG, Béland MJ, Colan SD, et al. Nomenclature for congenital and paediatric cardiac disease: the International Paediatric and Congenital Cardiac Code (IPCCC) and the Eleventh Iteration of the International Classification of Diseases (ICD-11). *Cardiology in the Young*. 2017;27(10):1872-1938. doi:10.1017/s1047951117002244.
8. Whelton PK, Carey RM, Aronow WS, et al. 2017 ACC/AHA/AAPA/ABC/ACPM/AGS/APhA/ASH/ASPC/NMA/PCNA Guideline for the Prevention, Detection, Evaluation, and Management of High Blood Pressure in Adults: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Hypertension*. 2018;71(6). doi:10.1161/hyp.0000000000000065.
9. Shen W-K, Sheldon RS, Benditt DG, et al. 2017 ACC/AHA/HRS Guideline for the Evaluation and Management of Patients with Syncope: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *Circulation*. 2017;136(5). doi:10.1161/cir.0000000000000499.
10. Page RL, Joglar JA, Caldwell MA, et al. 2015 ACC/AHA/HRS Guideline for the Management of Adult Patients with Supraventricular Tachycardia. *Circulation*. 2016;133(14). doi:10.1161/cir.0000000000000311.
11. Chaikof EL, Dalman RL, Eskandari MK, et al. The Society for Vascular Surgery practice guidelines on the care of patients with an abdominal aortic aneurysm. *Journal of Vascular Surgery*. 2018;67(1). doi:10.1016/j.jvs.2017.10.044.
12. AMPLATZER PFO Occluder: PFO Closure Device. Abbott Cardiovascular. <https://www.cardiovascular.abbott/us/en/hcp/products/structural-heart/amplatzer-pfo.html>.
13. Madhbour R, Wahl A, Praz F, Meier B. Amplatzer patent foramen ovale occluder: safety and efficacy. *Expert Review of Medical Devices*. 2019;16(3):173-182. doi:10.1080/17434440.2019.1581060.
14. Drummond A. AMPLATZER Patent Foramen Ovale (PFO) Occluder: FDA Review of P120021 Office of Device Evaluation Center for Devices and Radiological Health (CDRH) Food and Drug Administration May 24, 2016. <https://www.fda.gov/media/98643/download>.
15. Updates to Instructions for Use (IFU) concerning Erosion with the Amplatzer Atrial Septal Occluder (ASO). Last updated on 06 Jul 2014. [https://www.hsa.gov.sg/content/hsa/en/Health\\_Products\\_Regulation/Safety\\_Information\\_and\\_Product\\_Recalls/Dear\\_Healthcare\\_Professional\\_Letters/2013/Updates\\_to\\_Instructions\\_for\\_Use\\_IFU\\_concerning\\_Erosion\\_with\\_the\\_Amplatzer\\_Atrial\\_Septal\\_Occluder\\_AS.html](https://www.hsa.gov.sg/content/hsa/en/Health_Products_Regulation/Safety_Information_and_Product_Recalls/Dear_Healthcare_Professional_Letters/2013/Updates_to_Instructions_for_Use_IFU_concerning_Erosion_with_the_Amplatzer_Atrial_Septal_Occluder_AS.html).
16. Amsterdam EA, Wenger NK, Brindis RG, et al. 2014 AHA/ACC Guideline for the Management of Patients with Non-ST-Elevation Acute Coronary Syndromes. *Circulation*. 2014;130(25). doi:10.1161/cir.0000000000000134.
17. Priori SG, Wilde AA, Horie M, et al. Executive summary: HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes. *EP Europace*. 2013;15(10):1389-1406. doi:10.1093/europace/eut272.

18. Daniels CJ, Bradley EA, Landzberg MJ, et al. Fontan-Associated Liver Disease. *J Am Coll Cardiol*. 2017;70(25):3173-3194. doi:10.1016/j.jacc.2017.10.045.
19. Collado FMS, Poulin MF, Murphy JJ, Jneid H, Kavinsky CJ. Patent Foramen Ovale Closure for Stroke Prevention and Other Disorders. *Journal of the American Heart Association*. 2018;7(12). doi:10.1161/jaha.117.007146.
20. Khairy P, Hare GFV, Balaji S, et al. PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease. *Canadian Journal of Cardiology*. 2014;30(10). doi:10.1016/j.cjca.2014.09.002.
21. Kusumoto FM, Schoenfeld MH, Barrett C, et al. 2018 ACC/AHA/HRS Guideline on the Evaluation and Management of Patients with Bradycardia and Cardiac Conduction Delay. *J Am Coll Cardiol*. 2018. doi:10.1016/j.jacc.2018.10.044.
22. Fleisher LA, Fleischmann KE, Auerbach AD, et al. 2014 ACC/AHA Guideline on Perioperative Cardiovascular Evaluation and Management of Patients Undergoing Noncardiac Surgery. *J Am Coll Cardiol*. 2014;64(22). doi:10.1016/j.jacc.2014.07.944.
23. Galiè N, Humbert M, Vachiery J-L, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *European Respiratory Journal*. 2015;46(4):903-975. doi:10.1183/13993003.01032-2015.
24. Otto CM, Kumbhani DJ, Alexander KP, et al. 2017 ACC Expert Consensus Decision Pathway for Transcatheter Aortic Valve Replacement in the Management of Adults with Aortic Stenosis. *J Am Coll Cardiol*. 2017;69(10):1313-1346. doi:10.1016/j.jacc.2016.12.006.
25. O'gara PT, Kushner FG, Ascheim DD, et al. 2013 ACCF/AHA Guideline for the Management of ST-Elevation Myocardial Infarction. *J Am Coll Cardiol*. 2013;61(4). doi:10.1016/j.jacc.2012.11.019.
26. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease. *J Am Coll Cardiol*. 2018;73(12). doi:10.1016/j.jacc.2018.08.1029.
27. Elkayam U, Goland S, Pieper PG, Silversides CK. High-Risk Cardiac Disease in Pregnancy. *J Am Coll Cardiol*. 2016;68(4):396-410. doi:10.1016/j.jacc.2016.05.048.
28. Sachdeva R, Valente AM, Armstrong AK, et al. ACC/AHA/ASE/HRS/ISACHD/SCAI/SCCT/SCMR/SOPE 2020 Appropriate Use Criteria for Multimodality Imaging During the Follow-Up Care of Patients with Congenital Heart Disease. *J Am Coll Cardiol*. 2020;75(6):657-703. doi:10.1016/j.jacc.2019.10.002.



# Maternal Imaging in Cardiovascular Disease

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# Diagnostic Cardiovascular Imaging Pre-Pregnancy to Post-Partum (CD-15.1)

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## Maternal imaging in cardiovascular disease

Ultrasound and magnetic resonance are the preferred imaging modalities to minimize radiation exposure in pregnancy. When imaging using ionizing radiation is necessary, radiation exposure should follow the ALARA principle (As Low As Reasonably Achievable). Shared decision making regarding diagnostic testing should occur in consultation with the individual, cardiologist, and obstetrical team when weighing the risk of fetal exposure to radiation against the need to diagnose or plan treatment for serious illness.

## Peripartum Red Flag Signs and Symptoms

Imaging as requested, as listed within **Imaging Modalities in Maternal Imaging** (Echocardiogram, Exercise stress echo, Coronary angiography, Cardiac MRI), is indicated for peripartum individuals for any of the following **red flag** signs or symptoms:

- Chest pain
- Dyspnea
- Orthopnea
- Cough
- Lower extremity edema when there is a concern for heart disease
- Tachycardia
- Unheralded syncope
- Headache
- Acute visual changes
- New onset hypotension
- Hypertension

## Imaging Modalities in Maternal Imaging

### Transthoracic echocardiography (TTE)

- TTE (CPT® 93306) is the primary cardiac imaging modality in pregnancy. Baseline and surveillance echocardiography is indicated for several conditions as noted in **Maternal imaging in cardiovascular disease (CD-15.2)** Table 1.
- A repeat echocardiogram is indicated when there are new or worsening cardiovascular signs or symptoms, as described in **Peripartum Red Flags, Transthoracic Echocardiography (TTE)– Indications/initial evaluation (CD-2.2)** and **Frequency of Echocardiography Testing (CD-2.3)**, during and after pregnancy.

### Exercise Stress Echo

- Exercise stress echo (CPT® 93350, 93351) is indicated pre-conception to assist with risk stratification in individuals with a documented history of **any** of the following:
  - Current left ventricular dysfunction
  - Previous history of left ventricular dysfunction
  - Valvular heart disease of any severity
  - There is a concern for myocardial reserve
- See Stress echocardiogram in **Stress Testing with Imaging – Indications (CD-1.4)** and **Stress echo–indications other than ruling out CAD (CD-2.7)**
- See **Hypertrophic Cardiomyopathy (CD-14)**

### Coronary Angiography

- Fetal risk from ionizing radiation is highest before 20 weeks gestational age. When coronary angiography is medically necessary, the ALARA principle should be followed. Invasive management of acute coronary syndrome is associated with lower in-hospital mortality and should be considered. See **Diagnostic Heart Catheterization**

### Cardiac MRI

- Cardiac MRI (CPT® 75557) is utilized in pregnant individuals to measure aortic dimensions, wall motion and ventricular function when the echocardiogram is non-diagnostic. Gadolinium based contrast agents are not necessary in aortic imaging or most other indications in pregnancy. **See Cardiac MRI**

### Background and Supporting Information

- Cardiovascular disease (CVD) in pregnancy has become increasingly prevalent in recent years.
- The increase in plasma volume during pregnancy requires significant physiological adaptation.
- Maternal mortality has increased in the last two decades with CVD accounting for 33% of all deaths.

- Invasive management of myocardial infarction (MI) is associated with lower in-hospital mortality.
- Research has underrepresented females of childbearing age leading to significant deficits in our knowledge of cardiovascular care of these individuals.
- **Cardiac Imaging using ionizing radiation**
  - Multiple imaging modalities expose the pregnant individual and fetus to ionizing radiation.
  - This exposure causes concern for an elevated risk of childhood cancer.
  - Shared decision-making should be employed when weighing the fetal exposure to radiation against the need to diagnose serious illness

## References

1. Park K, Bairey Merz CN, Bello NA, et al. Management of Women With Acquired Cardiovascular Disease From Pre-Conception Through Pregnancy and Postpartum: JACC Focus Seminar 3/5. *J Am Coll Cardiol.* 2021;77(14):1799-1812. doi:10.1016/j.jacc.2021.01.057.
2. Bello NA, Bairey Merz CN, Brown H, et al. Diagnostic Cardiovascular Imaging and Therapeutic Strategies in Pregnancy: JACC Focus Seminar 4/5. *J Am Coll Cardiol.* 2021;77(14):1813-1822. doi:10.1016/j.jacc.2021.01.056.

# Maternal imaging in cardiovascular disease (CD-15.2)

CD.MI.0015.2.A

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Table 1: Suggested frequency of echo monitoring during pregnancy

Cardiovascular disease	Monitoring frequency
Hypertensive disorders of pregnancy (BP $\geq 130/90$ )	<ul style="list-style-type: none"> <li>An echocardiogram (TTE) (CPT® 93303, 93304, 93306, 93307, 93308) is indicated once during pregnancy in all hypertensive disorders of pregnancy.</li> <li>A repeat TTE is indicated when there are new or worsening cardiovascular signs or symptoms</li> </ul>
Valvular disorders/Native and Prosthetic	<ul style="list-style-type: none"> <li>One TTE is indicated during the first trimester (weeks 1-12 of pregnancy) for individuals with known or suspected valvular heart disease.</li> <li>A repeat TTE is indicated when there are new or worsening cardiovascular signs or symptoms</li> </ul>
Severe Aortic stenosis (AS)	<ul style="list-style-type: none"> <li>A repeat TTE is indicated every 1-2 months or when there are new or worsening cardiovascular signs or symptoms</li> </ul>
Mitral stenosis (MS)	<ul style="list-style-type: none"> <li>TTE is indicated each trimester (12 weeks) and prior to delivery in individuals with mild MS.</li> <li>TTE is indicated every 1–2 months in individuals with moderate to severe MS.</li> <li>A repeat TTE is indicated when there are new or worsening cardiovascular signs or symptoms</li> </ul>
Dilated cardiomyopathy	<ul style="list-style-type: none"> <li>TTE is indicated during each trimester (12 weeks)</li> <li>A repeat TTE is indicated when there are new or worsening cardiovascular signs or symptoms</li> </ul>
Hypertrophic cardiomyopathy	<ul style="list-style-type: none"> <li>TTE is indicated in <b>asymptomatic</b> individuals each trimester</li> <li>TTE is indicated in <b>symptomatic</b> individuals every 1-2 months.</li> <li>A repeat TTE is indicated when there are new or worsening cardiovascular signs or symptoms</li> </ul>

Cardiovascular disease	Monitoring frequency
Peripartum cardiomyopathy	<p>TTE is indicated in individuals with signs and symptoms of heart failure.</p> <ul style="list-style-type: none"> <li>• A repeat TTE is indicated when there are new or worsening cardiovascular signs or symptoms</li> <li>• TTE is indicated in subsequent pregnancies: <ul style="list-style-type: none"> <li>• At the time of the first prenatal visit</li> <li>• At the end of the first and second trimesters</li> <li>• One month prior to delivery</li> <li>• After delivery prior to discharge</li> <li>• One month postpartum</li> <li>• At any time when there are worsening signs or symptoms of heart failure</li> </ul> </li> </ul>
Pulmonary hypertension	<ul style="list-style-type: none"> <li>• TTE is indicated in individuals with signs and symptoms of pulmonary hypertension</li> <li>• A repeat TTE is indicated at the discretion of the health care provider.</li> </ul>

## References

1. O'Kelly AC, Sharma G, Vaught AJ, Zakaria S. The Use of Echocardiography and Advanced Cardiac Ultrasonography During Pregnancy. *Curr Treat Options Cardiovasc Med*. 2019;21(11):71. Published 2019 Nov 21. doi:10.1007/s11936-019-0785-5.
2. Bello NA, Bairey Merz CN, Brown H, et al. Diagnostic Cardiovascular Imaging and Therapeutic Strategies in Pregnancy: JACC Focus Seminar 4/5. *J Am Coll Cardiol*. 2021;77(14):1813-1822. doi:10.1016/j.jacc.2021.01.056.
3. Hemnes AR, Kiely DG, Cockrill BA, et al. Statement on pregnancy in pulmonary hypertension from the Pulmonary Vascular Research Institute. *Pulm Circ*. 2015;5(3):435-465. doi:10.1086/682230.

# Maternal Imaging in Individuals with Aortopathy (CD-15.3)

CD.MI.0015.3.A  
v2.0.2023

## Pre-pregnancy imaging

Individuals at risk for aortic aneurysms (**Table 15-3-1**) should be evaluated with echocardiogram (TTE) **and** Computed Tomography (CT)/ Magnetic Resonance Imaging (MRI) of the Chest/Abdomen/Pelvis (**Table 15-3-2**) within 1 year prior to conception to evaluate for aortic valve disease and aortic dimensions.

Table 15-3-1

Individuals at risk for aortic aneurysm, aortic dissection, limb-threatening ischemia
Bicuspid Aortic Valve
Turner Syndrome
Coarctation of the Aorta
Marfan Syndrome
Type IV Ehlers-Danlos
Loeys-Dietz
Familial Thoracic Aortic Disease and Aortic Dissection (defined as first degree relative with history of aortic dissection or thoracic aortic aneurysm or two second degree relatives with history of aortic dissection or thoracic aortic aneurysm)

Table 15-3-2

Imaging for Aortic conditions	CPT®
CT Chest and/or Abdomen and/or Pelvis	71260
	74177
	74160
	72193

Imaging for Aortic conditions	CPT®
CTA Chest and/or Abdomen and/or Pelvis	71275 74175 72191 74174
MRA Chest and/or Abdomen and/or Pelvis	71555 74185 72198
Echocardiogram (TTE)	93303 93304 93306 93307 93308
Echocardiogram (TEE)	93312 93313 93314 93315 93316 93317
Doppler echocardiography- is indicated as add-on codes for TEE	+93320 +93321 +93325

### Surveillance imaging during pregnancy and postpartum

Follow-up imaging with Echocardiogram (TTE) and CTA/MRA. TEE can be substituted if TTE is equivocal.

**Table 15-3-3**

Suggested Frequency of Aortic Imaging during pregnancy and postpartum in individuals known to be high risk for aortic aneurysm



Condition	WHO Class	Imaging frequency	Postpartum (up to 42 days after birth)
Turner Syndrome	<ul style="list-style-type: none"> <li>II-III: Aortic root &lt;20mm/m<sup>2</sup> with associated risk factors or &lt;25 mm/m without associated risk factors</li> </ul>	<ul style="list-style-type: none"> <li>Once during pregnancy if normal aortic dimension, or every two months if repaired coarctation</li> </ul>	<ul style="list-style-type: none"> <li>Once during the postpartum period</li> </ul>
	<ul style="list-style-type: none"> <li>IV: Aortic root ≥20mm/m<sup>2</sup> with associated risk factors or ≥25 mm/m without associated risk factors</li> </ul>	<ul style="list-style-type: none"> <li>Every 6 weeks if aorta diameter dilated &gt; 30mm</li> </ul>	<ul style="list-style-type: none"> <li>Once during the postpartum period</li> </ul>
	<ul style="list-style-type: none"> <li>Any patient with Turner who has severe coarctation</li> </ul>	<ul style="list-style-type: none"> <li>At discretion of provider</li> </ul>	<ul style="list-style-type: none"> <li>Once during the postpartum period</li> </ul>
Marfan Syndrome	<ul style="list-style-type: none"> <li>III: Aortic root &lt;45mm, mod-severe Aortic Insufficiency</li> </ul>	<ul style="list-style-type: none"> <li>Every trimester if &lt;40mm</li> </ul>	<ul style="list-style-type: none"> <li>Once during the postpartum period</li> </ul>
	<ul style="list-style-type: none"> <li>IV: Aortic root ≥45mm, history of dissection</li> </ul>	<ul style="list-style-type: none"> <li>Every 6 weeks if aorta is ≥40mm</li> </ul>	<ul style="list-style-type: none"> <li>Once during the postpartum period</li> </ul>
Vascular Ehlers-Danlos	<ul style="list-style-type: none"> <li>Type IV</li> </ul>	<ul style="list-style-type: none"> <li>Every 6 weeks</li> </ul>	<ul style="list-style-type: none"> <li>Once during the postpartum period</li> </ul>
Loeys-Dietz	<ul style="list-style-type: none"> <li>III: Aortic diameter &lt;40mm</li> </ul>	<ul style="list-style-type: none"> <li>Every 6 weeks</li> </ul>	<ul style="list-style-type: none"> <li>Once during the postpartum period</li> </ul>
	<ul style="list-style-type: none"> <li>IV: Aortic diameter ≥40mm</li> </ul>	<ul style="list-style-type: none"> <li>Every 6 weeks</li> </ul>	<ul style="list-style-type: none"> <li>Once during the postpartum period</li> </ul>
Familial thoracic aortic aneurysms and	<ul style="list-style-type: none"> <li>III: Aortic diameter &lt;40mm</li> </ul>	<ul style="list-style-type: none"> <li>Every trimester if &lt;40mm aortic diameter</li> </ul>	<ul style="list-style-type: none"> <li>Once during the postpartum period</li> </ul>

Condition	WHO Class	Imaging frequency	Postpartum (up to 42 days after birth)
dissections	<ul style="list-style-type: none"><li>IV: Aortic diameter <math>\geq 40</math>mm</li></ul>	<ul style="list-style-type: none"><li>Every 6 weeks if <math>\geq 40</math>mm aortic diameter</li></ul>	<ul style="list-style-type: none"><li>Once during the postpartum period</li></ul>

### ***Background and supporting information***

First degree relative (sibling, parent, child).

Second degree relative (aunt/uncles, grandparent, niece, nephew, cousin, or half-sibling of an individual)

### **References**

1. Lindley KJ, Bairey Merz CN, Asgar AW, et al. Management of Women With Congenital or Inherited Cardiovascular Disease From Pre-Conception Through Pregnancy and Postpartum: JACC Focus Seminar 2/5. *J Am Coll Cardiol.* 2021;77(14):1778-1798. doi:10.1016/j.jacc.2021.02.026.

# Imaging in Pregnancy with Congenital Heart Disease (CHD) (CD-15.4)

CD.MI.0015.4.A

v2.0.2023

**Pre-pregnancy imaging based on the World Health Organization (WHO) chart for imaging in pregnancy with CHD**

Imaging modality and indication	CPT®
Echo (TTE) when planning pregnancy	93306
TEE if TTE equivocal	93312
CMR (cardiac MRI) can be performed prior to pregnancy in those lesions where CMR would be routinely performed at some later date	75557
CTA or MRA of chest if known aortic disease, Pulmonary artery disease, anomalous pulmonary veins, anomalous systemic veins. (also see peripartum aortopathy table.)	71275, 71555
Echo with new signs or symptoms	93303, 93304, 93306, 93308
Postpartum imaging per provider requested frequency	imaging as noted above
Stress imaging pre/during pregnancy when known Coronary artery anomaly, pulmonary hypertension, LVOT obstruction, cardiac dysfunction, single	93350, 93351, 93320, 93325
WHO II, III, IV, can have echo/MR/CT/stress imaging prior to pregnancy	imaging as noted above

## Surveillance Imaging

Surveillance imaging after baseline studies.

TTE frequency after initial imaging, (Individuals who also have aortopathy can have Chest MRA in addition to echo at same frequency.) Individuals with known poor/inadequate imaging on echo, can have CMR in lieu of echocardiogram.

## The World Health Organization modified classification of maternal cardiovascular risk

The World Health Organization established a modified classification of maternal cardiovascular risk used as a tool to evaluate risk status for pregnant individuals with various cardiovascular conditions. See **Pregnancy-Maternal Imaging (CD-11.4)**

Class	Risk	Sample Lesions
WHO Class I	No detectable increased risk of maternal mortality and no or mild increase in morbidity.	<ul style="list-style-type: none"> <li>Mild Pulmonary stenosis</li> <li>Small PDA</li> <li>Mild MVP</li> <li>Successfully repaired simple lesions (ASD, VSD, PDA, anomalous pulmonary venous drainage)</li> <li>Isolated PACs or PVCs</li> </ul>
WHO Class II	Small increased risk of maternal mortality or moderate increase in morbidity.	<ul style="list-style-type: none"> <li>Unoperated ASD or VSD</li> <li>Repaired TOF (uncomplicated)</li> <li>Most arrhythmias</li> </ul>
WHO Class II-III	Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity.	<ul style="list-style-type: none"> <li>Mild left ventricular impairment</li> <li>Hypertrophic cardiomyopathy</li> <li>Native or tissue valvular heart disease not considered WHO I or IV</li> <li>Aorta &lt;45 mm in aortic disease associated with bicuspid aortic valve</li> <li>Repaired Coarctation</li> </ul>
WHO Class III	Significantly increased risk of maternal mortality or severe morbidity. <ul style="list-style-type: none"> <li>Expert counseling required.</li> <li>If pregnancy is decided upon, intensive specialist cardiac and obstetric monitoring needed throughout pregnancy, childbirth and the postpartum period.</li> </ul>	<ul style="list-style-type: none"> <li>Mechanical valve</li> <li>Systemic right ventricle</li> <li>Fontan circulation</li> <li>Unrepaired cyanotic heart disease</li> <li>Other complex congenital heart disease</li> </ul>

Class	Risk	Sample Lesions
WHO Class IV	<p>Extremely high risk of maternal mortality or severe morbidity.</p> <ul style="list-style-type: none"><li>• Pregnancy contraindicated.</li><li>• If pregnancy occurs, termination should be discussed.</li><li>• If pregnancy continues, care as for WHO class III.</li></ul>	<ul style="list-style-type: none"><li>• Pulmonary arterial hypertension from any cause</li><li>• Severe systemic ventricular dysfunction (LVEF &lt;30%, NYHA functional class III-IV)</li><li>• Severe mitral stenosis; severe symptomatic aortic stenosis</li><li>• Aortic dilation <math>\geq 50</math> mm in aortic disease associated with bicuspid aortic valve</li><li>• Native severe coarctation of the aorta</li></ul>

## References

1. Steiner JM, Lokken E, Bayley E, et al. Cardiac and Pregnancy Outcomes of Pregnant Patients With Congenital Heart Disease According to Risk Classification System. Am J Cardiol. 2021;161:95-101. doi:10.1016/j.amjcard.2021.08.037.
2. Canobbio MM, Warnes CA, Aboulhosn J, et al. Management of Pregnancy in Patients With Complex Congenital Heart Disease: A Scientific Statement for Healthcare Professionals From the American Heart Association. Circulation. 2017;135(8):e50-e87. doi:10.1161/CIR.0000000000000458.

# Condition Specific Imaging

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# Cardiotoxic Agent-Related Cardiac Dysfunction (CD-12)

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## Cardiotoxic agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1)

- Echocardiogram to determine LV function in individuals on cardiotoxic chemotherapeutic drugs:
  - The time frame should be determined by the provider, but no more often than baseline and at every 6 weeks.
  - May repeat every 4 weeks if cardiotoxic chemotherapeutic drug is withheld for significant left ventricular cardiac dysfunction
  - If the LVEF is <50% on echocardiogram follow up can be done with MUGA at appropriate intervals.
- Echocardiography vs. MUGA for Determining Left Ventricular Ejection Fraction (LVEF) in Individuals on Cardiotoxic Chemotherapy Drugs:
  - eviCore guidelines support using **echocardiography rather than MUGA** for the determination of LVEF and/or wall motion EXCEPT in one of the circumstances described previously in **MUGA Study – Cardiac Indications (CD-3.4)**.
- Echocardiogram is recommended for cancer survivors with a history of chest radiotherapy or anthracycline exposure who are pregnant or planning to become pregnant as follows:
  - Baseline exam
  - Once in the first trimester
  - Once in the third trimester
  - Study can be repeated for any symptoms at any other time as needed during or immediately following pregnancy
- Adults who received anthracyclines in childhood see **PEDONC-19.2**

**Background and supporting information**

- Advantages of Echocardiography in comparison to MUGA in individuals on cardiotoxic chemotherapy:
  - No ionizing radiation
  - No IV access required when echo contrast is not used
  - Allows view of the pericardium to look for effusion
  - Allows estimate of pulmonary pressure
  - May allow visualization of a clot or tumor in the Inferior Vena Cava (IVC) and/or the right heart

**Myocardial Strain Imaging (CD-12.2)**

- Myocardial strain imaging (CPT® 93356) in addition to the primary echocardiogram in individuals receiving therapy with cardiotoxic agents for ANY of the following:
  - Initial evaluation-prior to treatment with EITHER:
    - Medications that could result in cardiotoxicity/heart failure
    - Radiation that could result in cardiotoxicity/heart failure
  - Re-evaluation of an individual previously or currently undergoing therapy as per echocardiogram parameters. See **Cardiotoxic agent/Cancer Therapeutics-Related Cardiac Dysfunction (CD-12.1)**
  - Re-evaluation of an individual undergoing therapy with worsening symptoms

**Mavacamten for Obstructive Hypertrophic Cardiomyopathy (HCM) (CD-12.3)**

Echocardiogram (CPT® 93306) is indicated for individuals treated with mavacamten for class II-III obstructive HCM as follows:

**Initiation of treatment**

- Baseline-at the beginning of treatment
- 4 weeks after treatment initiation
- 8 weeks after treatment initiation
- 12 weeks after treatment initiation
- Then every 12 weeks while on mavacamten

**Changes in treatment**

- 4 weeks after any interruption of treatment (any missed dose)
- After any dosage change (including restart of treatment):
  - 4 weeks after dosage change
  - 12 weeks after dosage change



- After initiating a weak CYP2C19 inhibitor (e.g., omeprazole) or moderate CYP2A4 inhibitor (e.g., ciprofloxacin):
  - 4 weeks after start of medication
  - 12 weeks after start of medication
- At any time regardless of timing of prior echo when there are new cardiac signs or symptoms, or worsening of clinical status

### **Background and supporting information**

Hypertrophic Cardiomyopathy (HCM) is a clinical diagnosis, established by imaging with 2D echocardiography or cardiovascular magnetic resonance (CMR) showing a maximal end-diastolic wall thickness of  $\geq 15$  mm anywhere in the left ventricle, in the absence of another cause of hypertrophy in adults. More limited hypertrophy (13–14 mm) can be diagnostic, particularly when present in family members of a patient with HCM or in conjunction with a positive genetic test, and/or associated with typical dynamic outflow obstruction, or distinctly abnormal ECG patterns.

### **References (CD-12)**

1. Hendel RC, Berman DS, Carli MFD, et al. ACCF/ASNC/ACR/AHA/ASE/SCCT/SCMR/SNM 2009 Appropriate Use Criteria for Cardiac Radionuclide Imaging. *Circulation*. 2009;119(22). doi:10.1161/circulationaha.109.192519.
2. Genentech: Herceptin® (trastuzumab) - Information for Healthcare Providers. Genentech: Herceptin® (trastuzumab) - Information for Healthcare Providers. <https://www.gene.com/medical-professionals/medicines/herceptin>.
3. Virizuela JA, García AM, Peñas RDL, et al. SEOM clinical guidelines on cardiovascular toxicity (2018). *Clinical and Translational Oncology*. 2019;21(1):94-105. doi:10.1007/s12094-018-02017-3.
4. Friedman DL, Hudson MM. Health Link: Heart Health. Long-Term Follow-Up Guidelines for Survivors of Childhood, Adolescent, and Young Adult Cancers. <http://www.survivorshipguidelines.org/>. Published October 2018.
5. Marian AJ, Braunwald E. Hypertrophic Cardiomyopathy: Genetics, Pathogenesis, Clinical Manifestations, Diagnosis, and Therapy. *Circ Res*. 2017;121(7):749-770. doi:10.1161/CIRCRESAHA.117.311059.
6. CAMZYOS. Highlights of Prescribing Information. Bristol Myers Squibb. April 2022. [https://packageinserts.bms.com/pi/pi\\_camzyos.pdf](https://packageinserts.bms.com/pi/pi_camzyos.pdf).

# Cardiac Sarcoidosis (CD-3.9)

CD.CS.0003.9.A

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## Suspected cardiac sarcoidosis (see Background and Supporting Information)

- MRI imaging of the heart with gadolinium (CPT® 75561). Initial imaging for identification of suspected cardiac sarcoid should be cardiac MRI with late gadolinium enhancement (LGE) protocol unless there is a contraindication to MRI imaging (non MRI safe pacemaker, renal failure). Absence of LGE is a strong negative predictor for low rates of cardiac morbidity and mortality from cardiac sarcoid and further testing is not usually indicated.
- PET – Metabolic imaging with F-18 FDG for diagnosis if there is a contraindication to MRI and cardiac sarcoid is suspected. Requires PET with F-18 FDG metabolic study combined with a PET perfusion study (CPT® 78432 or CPT® 78433) OR PET metabolic study (CPT® 78459 or CPT® 78429) and SPECT perfusion image (CPT® 78451).
  - For equivocal MRI
  - To confirm diagnosis if suggested by MRI
  - Prior to treatment of cardiac sarcoid

## Monitoring of treatment of established cardiac sarcoidosis

- PET - Cardiac PET metabolic is indicated to monitor therapy in cardiac sarcoidosis. Requires PET with F-18 FDG metabolic study combined with a PET perfusion study (CPT® 78432 or CPT® 78433) OR PET metabolic study (CPT® 78459 or CPT® 78429) and SPECT perfusion image (CPT® 78451).
  - A pretreatment PET is indicated.
  - PET (heart FDG metabolic with perfusion study as above) can be repeated at 3-6 month intervals if there is active disease or to make therapeutic decisions.

## Background and supporting information

- Cardiac imaging is reasonable to detect cardiac sarcoid in the following:
  - Any patient with extra cardiac sarcoid even if no cardiac symptoms
  - Echo with basal thinning of the intraventricular septum, depressed EF (<50) or regional wall motion abnormality not associated with CAD
  - Young patients with unexplained ventricular tachycardia, especially monomorphic VT
  - Patients with unexplained cardiomyopathy or heart failure (i.e., CAD has been ruled out)
  - Patients with unexplained arrhythmia especially advanced AV block or VT

## References

1. Birnie DH, Sauer WH, Bogun F, et al. HRS expert consensus statement on the diagnosis and management of arrhythmias associated with cardiac sarcoidosis. *Heart Rhythm*. 2014;11(7):1305-1323. doi:10.1016/j.hrthm.2014.03.043.
2. Blankstein R, Waller AH. Evaluation of Known or Suspected Cardiac Sarcoidosis. *Circ Cardiovasc Imaging*. 2016;9(3):e000867. doi:10.1161/CIRCIMAGING.113.000867.
3. Bravo PE, Singh A, Di Carli MF, Blankstein R. Advanced cardiovascular imaging for the evaluation of cardiac sarcoidosis. *J Nucl Cardiol*. 2019;26(1):188-199. doi:10.1007/s12350-018-01488-9.
4. Kim SJ, Pak K, Kim K. Diagnostic performance of F-18 FDG PET for detection of cardiac sarcoidosis; A systematic review and meta-analysis. *J Nucl Cardiol*. 2020;27(6):2103-2115. doi:10.1007/s12350-018-01582-y.
5. Manabe O, Oyama-Manabe N, Aikawa T, et al. Advances in Diagnostic Imaging for Cardiac Sarcoidosis. *J Clin Med*. 2021;10(24):5808. doi:10.3390/jcm10245808.
6. Ramirez R, Trivieri M, Fayad ZA, et al. Advanced Imaging in Cardiac Sarcoidosis. *J Nucl Med*. 2019;60(7):892-898. doi:10.2967/jnumed.119.228130
7. Writing group; Document reading group; EACVI Reviewers: This document was reviewed by members of the EACVI Scientific Documents Committee for 2014–2016 and 2016–2018. A joint procedural position statement on imaging in cardiac sarcoidosis: from the Cardiovascular and Inflammation & Infection Committees of the European Association of Nuclear Medicine, the European Association of Cardiovascular Imaging, and the American Society of Nuclear Cardiology. *Eur Heart J Cardiovasc Imaging*. 2017;18(10):1073-1089. doi:10.1093/ehjci/jex146.
8. Terasaki F, Azuma A, Anzai T, et al. JCS 2016 Guideline on Diagnosis and Treatment of Cardiac Sarcoidosis - Digest Version. *Circ J*. 2019;83(11):2329-2388. doi:10.1253/circj.CJ-19-0508.
9. Trivieri MG, Spagnolo P, Birnie D, et al. Challenges in Cardiac and Pulmonary Sarcoidosis: JACC State-of-the-Art Review. *J Am Coll Cardiol*. 2020;76(16):1878-1901. doi:10.1016/j.jacc.2020.08.042.
10. Ungprasert P, Carmona EM, Utz JP, et al. Epidemiology of Sarcoidosis 1946-2013: A Population-Based Study. *Mayo Clin Proc*. 2016;91(2):183-188. doi:10.1016/j.mayocp.2015.10.024.

# Cardiac Trauma Imaging (CD-10.1)

CD.CS.0010.1.A

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- Any of the following can be used to evaluate cardiac or aortic trauma:
  - Echocardiogram (TTE, TEE)
  - Cardiac MRI Cardiac (CPT® 75557, CPT® 75561, and CPT® 75565)
  - Cardiac CT Cardiac (CPT® 75572)
  - CCTA (CPT® 75574)
  - Chest CTA Chest (CPT® 71275)
  - Chest CT Chest (CPT® 71260, CPT® 71270)

## References (CD-10)

1. Conn A. Chest trauma. In: Trauma: A Comprehensive Emergency Medicine Approach. New York, NY: Cambridge University Press; 2011:190-212.
2. Stojanovska J, Hurwitz Kowek LM, Chung JH, et al. ACR Appropriateness Criteria® Blunt Chest Trauma-Suspected Cardiac Injury. Revised 2020. Am Coll Radiol (ACR). Available at <https://acsearch.acr.org/docs/3082590/Narrative/>.

# Congestive Heart Failure (CD-9)

CD.CS.0009.A

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## CHF – Imaging (CD-9.1)

- Congestive heart failure (CHF), including post-cardiac transplant failure:
  - Echocardiogram is the first study after the clinical evaluation for suspected CHF.
  - MUGA, cardiac MRI or cardiac CT may be indicated if the ECHO is limited or does not completely answer the question.
  - Stress test to assess for CAD may be indicated. Follow stress testing guideline: **Stress Testing with Imaging – Indications (CD-1.4)**
- Arteriovenous fistula with “high output” heart failure:
  - CT Chest with contrast (CPT® 71260) and/or CT Abdomen and/or CT Pelvis with contrast (CPT® 74160 or CPT® 72193 or CPT® 74177) **OR**
  - CTA Chest (CPT® 71275) and/or CTA Abdomen and/or CTA Pelvis (CPT® 74175 or CPT® 72191 or CPT® 74174) **OR**
  - MRI Chest and/or MRI Abdomen and/or MRI Pelvis without and with contrast (CPT® 71552 and/or CPT® 74183 and/or CPT® 72197) **OR**
  - MRA Chest and/or MRA Abdomen and/or MRA Pelvis (CPT® 71555 and/or CPT® 74185 and/or CPT® 72198)
- Right-sided congestive heart failure can be a manifestation of pulmonary hypertension or serious lung disease.
  - CT Chest (CPT® 71260) or CTA Chest (CPT® 71275) to evaluate for recurrent pulmonary embolism

## Myocardial Sympathetic Innervation Imaging in Heart Failure (CD-3.6)

- Nuclear imaging using I-123-meta-iodobenzylguanidine (I-123-mIBG) in an attempt to image increased myocardial sympathetic activity is considered to be experimental and investigational.
- The AMA has established the following set of Category III codes to report these studies:
  - **0331T** - Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment
  - **0332T** - Myocardial sympathetic innervation imaging, planar qualitative and quantitative assessment; with tomographic SPECT.

### **Background and supporting information**

In heart failure, the sympathetic nervous system is activated in order to compensate for the decreased myocardial function. Initially, this is beneficial, however, long-term this compensatory mechanism is detrimental and causes further damage.

Markers have been developed, using radioactive iodine, in an attempt to image this increased myocardial sympathetic activity. Currently, AdreView™ (Iodine-123 meta-iodobenzylguanidine), is the only FDA-approved imaging agent available for this purpose.

### **Left ventricular assist devices (LVAD) (CD-9.4)**

Left ventricular assist devices (LVAD) are implantable devices used in individuals with advanced heart failure refractory to medical therapy, often as a bridge to transplantation.

- Echocardiograms (TTE) are obtained frequently for surveillance following implantation:
  - Post implant-generally at 2 weeks
  - Then as follows at:
    - One month
    - Three months
    - Six months
    - Twelve months
    - Every 6 months thereafter

### **References (CD-9)**

1. Yancy CW, Jessup M, Bozkurt B, et al. 2013 ACCF/AHA Guideline for the Management of Heart Failure: Executive Summary: A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. *Circulation*. 2013;128(16):1810-1852. doi:10.1161/cir.0b013e31829e8807.
2. Yancy CW, Jessup M, Bozkurt B, et al. 2016 ACC/AHA/HFSA Focused Update on New Pharmacological Therapy for Heart Failure: An Update of the 2013 ACCF/AHA Guideline for the Management of Heart Failure. *Journal of Cardiac Failure*. 2016;22(9):659-669. doi:10.1016/j.cardfail.2016.07.001.
3. Ponikowski P, Voors AA, Anker SD, et al. 2016 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure. *European Heart Journal*. 2016;37(27):2129-2200. doi:10.1093/eurheartj/ehw128.
4. Nakata T, Nakajima K, Yamashina S, et al. A Pooled Analysis of Multicenter Cohort Studies of 123I-mIBG Imaging of Sympathetic Innervation for Assessment of Long-Term Prognosis in Heart Failure. *JACC: Cardiovascular Imaging*. 2013;6(7):772-784. doi:10.1016/j.jcmg.2013.02.007.
5. Stainback RF, Estep JD, Agler DA, et al. Echocardiography in the Management of Patients with Left Ventricular Assist Devices: Recommendations from the American Society of Echocardiography. *Journal of the American Society of Echocardiography*. 2015;28(8):853-909. doi:10.1016/j.echo.2015.05.008.
6. Slaughter MS, Pagani FD, Rogers JG, et al. Clinical management of continuous-flow left ventricular assist devices in advanced heart failure. *The Journal of Heart and Lung Transplantation*. 2010;29(4). doi:10.1016/j.healun.2010.01.011.

# Pre-Surgical Cardiac Testing (CD-13)

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## Pre-Surgical Cardiac Testing – General Information (CD-13.1)

- It is important to differentiate requests for preoperative CT imaging before cardiac surgery according to type of procedure planned:
  - Primary cardiac operation—individuals who have not had prior heart surgery
  - Redo procedures—individuals who have had a prior procedure (it is important to determine the type of procedure as this may impact which modality is most appropriate for the pre-operative assessment)
  - Minimally invasive procedures, such as minimally invasive aortic valve operations, minimally invasive or robotic mitral operations, TAVR, MitraClip™ or other percutaneous valve procedures (such as valve in valve aortic or mitral, percutaneous tricuspid and TMVR which will be increasing in the future)
- In re-operative cardiac surgery, the benefit of preoperative CT is to assess for aortic calcifications, to evaluate the anatomic relationships in the mediastinum, such as the location of the various cardiac chambers and great vessels and proximity to the sternum, and to assess for the location of prior bypass grafts. Information can then be used to change the operative strategy including non-midline approach, peripheral vascular exposure, and alternative cannulation sites and for establishing cardiopulmonary bypass before re-sternotomy. These techniques can result in decreased incidence of intraoperative injury to heart, great vessels and prior bypass grafts and lower rates of postoperative stroke. IV contrast is necessary with these studies to delineate the anatomic structures. However, in individuals with renal insufficiency, the provider might choose to forgo the contrast if does not want to contrast load the individual prior to placing them on the heart-lung machine.
- Aortic atherosclerosis is recognized as the single most important determinant of postoperative stroke. There is evidence to support that preoperative CT is associated with lower postoperative stroke rates and mortality after primary cardiac surgery.
  - CT Chest without contrast (CPT® 71250) can be performed pre-operatively to allow the surgeon to:
    - Visualize the extent and location of aortic atherosclerosis



- Change the operative strategy such as those problematic areas are avoided

### **Primary Cardiac Surgery – No Previous Cardiac Surgery (CD-13.2)**

- CT Chest without contrast (CPT® 71250) to evaluate for the presence of ascending aortic calcifications may be indicated prior to primary cardiac surgery when there is documented high risk for aortic calcification including any of the following:
  - Aortic calcification on chest x-ray or other diagnostic test (TEE, fluoroscopy, etc.)
  - Calcific aortic stenosis
  - End stage renal disease (dialysis)

### **Re-operative cardiac surgery (CD-13.3)**

- Individuals undergoing re-operative cardiac surgery may undergo **one** of the following tests for preoperative assessment:
  - CT Chest with contrast
  - CTA Chest
  - CCTA only if prior CABG (this might be in addition to CT with contrast as CCTA will not show the extent of the thoracic aorta that needs to be visualized)
  - CT Heart usually does not provide the necessary information, and should not be approved routinely.

### **Minimally Invasive Valve Surgery (CD-13.4)**

- See **Transcatheter Aortic Valve Replacement (TAVR) (CD-4.8)**
- For an individual undergoing minimally invasive aortic valve surgery and minimally invasive or robotic mitral valve surgery, **ONE** of the following for preoperative assessment of an individual's suitability for the approach and for subsequent procedure planning:
  - CTA Chest, CTA Abdomen and Pelvis
  - CT Chest and CT Abdomen and Pelvis with contrast

### **Percutaneous Mitral Valve Repair (mitral valve clip) (CD-13.5)**

- Percutaneous treatment of mitral regurgitation can be accomplished using venous access to apply a clip device (e.g., MitraClip™ currently FDA approved) to provide edge-to-edge mitral leaflet coaptation, approximating opposing sections of the anterior and posterior mitral valve leaflets. FDA approved indications include treatment for individuals with symptomatic, moderate to severe or severe primary mitral regurgitation whose surgical risks are prohibitive, as well as symptomatic moderate to severe or severe secondary mitral regurgitation who have failed optimal medical therapy. This therapy should include, if indicated, cardiac resynchronization therapy.
- The following imaging may be used to determine if an individual is eligible for the procedure:



- Transthoracic echo with or without 3D rendering
- Transesophageal echo with or without 3D rendering
- Heart catheterization, including right heart cath if requested
- Because this is a venous approach, CTA of Abdomen, Chest, and/or Pelvis **is not** indicated.
- Post procedure transthoracic echo (TTE) can be performed at the following intervals:
  - One month
  - Six months
  - One year

### References (CD-13)

1. Cantinotti M. The importance and ways of exploring the entire chest before and after cardiac surgery: Chest radiography, lung ultrasonography, and computed tomography. *The Journal of Thoracic and Cardiovascular Surgery*. 2018;155(5):2041-2042. doi:10.1016/j.jtcvs.2018.01.032.
2. Merlo A, Chen K, Deo S, Markowitz A. Does routine preoperative computed tomography imaging provide clinical utility in patients undergoing primary cardiac surgery? *Interactive CardioVascular and Thoracic Surgery*. 2017;25(4):659-662. doi:10.1093/icvts/ivx098.
3. Erthal F, Inacio JR, Hazra S, Chan V, Chow BJW. Cardiac Computed Tomography Before and After Cardiac Surgery. *J Thorac Imaging*. 2018 May;33(3):156-167. doi:10.1097/RTI.0000000000000295.
4. Moodley S, Schoenhagen P, Gillinov AM, et al. Preoperative multidetector computed tomography angiography for planning of minimally invasive robotic mitral valve surgery impact on decision making. *J Thorac Cardiovasc Surg*. 2013 Aug;146(2):262-8. doi:10.1016/j.jtcvs.2012.06.052.
5. den Harder AM, de Heer LM, Meijer RC, et al. Effect of computed tomography before cardiac surgery on surgical strategy, mortality and stroke. *Eur J Radiol*. 2016 Apr;85(4):744-50. doi:10.1016/j.ejrad.2016.01.003.
6. Dass C, Simpson SA, Steiner RM, Guy TS. Preprocedural Computed Tomography Evaluation for Minimally Invasive Mitral Valve Surgery. *Journal of Thoracic Imaging*. 2015;30(6):386-396. doi:10.1097/rti.0000000000000170.
7. Adler Y, Fisman EZ, Shemesh J, et al. Spiral computed tomography evidence of close correlation between coronary and thoracic aorta calcifications. *Atherosclerosis*. 2004 Sep;176(1):133-8. doi: 10.1016/j.atherosclerosis.2004.03.027.
8. van der Linden J, Hadjinikolaou L, Bergman P, et al. Postoperative stroke in cardiac surgery is related to the location and extent of atherosclerotic disease in the ascending aorta. *J Am Coll Cardiol*. 2001 Jul;38(1):131-5. doi:10.1016/s0735-1097(01)01328-6.
9. Lapa DJ, Ailawadi G, Irvine JN Jr, et al. Preoperative computed tomography is associated with lower risk of perioperative stroke in reoperative cardiac surgery. *Interact Cardiovasc Thorac Surg*. 2011 Jun;12(6):919-23. doi:10.1510/icvts.2010.265165.
10. Nishi H, Mitsuno M, Tanaka H, Ryomoto M, Fukui S, Miyamoto Y. Who needs preoperative routine chest computed tomography for prevention of stroke in cardiac surgery? *Interact Cardiovasc Thorac Surg*. 2010 Jul;11(1):30-3. doi:10.1510/icvts.2009.231761.
11. Akhtar NJ, Markowitz AH, Gilkeson RC. Multidetector computed tomography in the preoperative assessment of cardiac surgery patients. *Radiol Clin North Am*. 2010 Jan;48(1):117-39. doi:10.1016/j.rcl.2009.09.002.
12. Khan NU, Yonan N. Does preoperative computed tomography reduce the risks associated with re-do cardiac surgery? *Interact Cardiovasc Thorac Surg*. 2009 Jul;9(1):119-23. doi:10.1510/icvts.2008.189506.
13. Bergman P, Linden JVD, Forsberg K, Ohman M. Preoperative Computed Tomography or Intraoperative Epiaortic Ultrasound for the Diagnosis of Atherosclerosis of the Ascending Aorta? *The Heart Surgery Forum*. 2004;7(3). doi:10.1532/hfs98.20033009.
14. Lee R, Matsutani N, Polimenakos AC, et al. Preoperative noncontrast chest computed tomography identifies potential aortic emboli. *Ann Thorac Surg*. 2007 Jul;84(1):38-41; discussion 42.
15. Nishi H, Mitsuno M, Ryomoto M, Miyamoto Y. Comprehensive approach for clamping severely calcified ascending aorta using computed tomography. *Interactive CardioVascular and Thoracic Surgery*. 2010;10(1):18-20. doi:10.1510/icvts.2009.216242.

16. Aviram G, Sharony R, Kramer A, et al. Modification of Surgical Planning Based on Cardiac Multidetector Computed Tomography in Reoperative Heart Surgery. *The Annals of Thoracic Surgery*. 2005;79(2):589-595. doi:10.1016/j.athoracsur.2004.07.012.
17. Harder AMD, Heer LMD, Maurovich-Horvat P, et al. Ultra low-dose chest CT with iterative reconstructions as an alternative to conventional chest x-ray prior to heart surgery (CRICKET study): Rationale and design of a multicenter randomized trial. *Journal of Cardiovascular Computed Tomography*. 2016;10(3):242-245. doi:10.1016/j.jcct.2016.01.016.
18. O'gara PT, Grayburn PA, Badhwar V, et al. 2017 ACC Expert Consensus Decision Pathway on the Management of Mitral Regurgitation. *J Am Coll Cardiol*. 2017;70(19):2421-2449. doi:10.1016/j.jacc.2017.09.019.
19. Doherty JU, Kort S, Mehran R, Schoenhagen P, Soman P. ACC/AATS/AHA/ASE/ASNC/HRS/SCAI/SCCT/SCMR/STS 2017 appropriate use criteria for multimodality imaging in valvular heart disease: a report of the American College of Cardiology Appropriate Use Criteria Task Force, American Association for Thoracic Surgery, American Heart Association, American Society of Echocardiography, American Society of Nuclear Cardiology, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, Society for Cardiovascular Magnetic Resonance, and Society of Thoracic Surgeons. *J Am Coll Cardiol*. 2017;70:1647-72. doi:10.1007/s12350-017-1070-1.
20. Nishi H, Mitsuno M, Tanaka H, Ryomoto M, Fukui S, Miyamoto Y. Who needs preoperative routine chest computed tomography for prevention of stroke in cardiac surgery? *Interactive CardioVascular and Thoracic Surgery*. 2010;11(1):30-33. doi:10.1510/icvts.2009.231761.

# Pulmonary Hypertension (PH) (CD-8.1)

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v2.0.2023

## Pulmonary Hypertension - Imaging indications

Transthoracic echocardiogram (TTE) (CPT® 93306) should be performed initially as it can help determine the probability of pulmonary hypertension.

## Screening

A screening echocardiogram (TTE) for PH is indicated in individuals with documented history of **any** of the following:

- Individuals preoperatively for planned liver transplant.
- Individuals evaluated for transjugular portosystemic shunt
- Portal Hypertension
- Liver disease with signs and symptoms of PH
- Bronchopulmonary dysplasia

Annual screening echo (TTE) is indicated in individuals with a documented history of any of the following:

- Systemic sclerosis (SSc)
- Individuals with PH mutations (e.g., BMPR2)
- First degree relatives of individuals with PH
- Connective tissue disorder with symptoms consistent with PH
- Individuals with TR velocity  $\geq 2.8$  m/s, with no other findings on additional testing
- Individuals being treated with medications associated with PH
- Individuals who have a concern documented for PH, who had a negative echocardiogram, but still show signs or symptoms of pulmonary hypertension

Follow-up testing is not indicated in individuals with TR velocity  $< 2.8$  m/s and no other signs, symptoms, or risk factors of PH

## Initial imaging

**Transthoracic echocardiogram (TTE) (CPT® 93306)** is indicated for symptoms and signs of pulmonary hypertension (PH) including **any** of the following:

- Notes documenting clinical concern for pulmonary hypertension

- EKG findings concerning for PH such as **any** of the following:
  - Right ventricular hypertrophy (RVH)
  - Right axis deviation
  - Right atrial enlargement
- Right ventricular hypertrophy or pulmonary artery dilation on other images
- History of pulmonary embolism with persistent or new onset DOE, or exercise limitation
- Suspected PH in individuals with lung disease
- DOE in individuals with connective tissue disorder, HIV, portal hypertension, SSc.
- Symptoms of PH (**any** of the following):
  - Breathlessness
  - SOB
  - Decreased exercise tolerance
  - Fatigue and rapid exhaustion
  - Palpitations
  - Dyspnea on bending forward
  - Hemoptysis
  - Exercise induced abdominal distention and nausea
  - Weight gain due to fluid retention
  - Syncope during or shortly after physical activity
  - Exertional chest pain

**Stress Echo (CPT® 93350 or 93351)** is indicated for **any** of the following:

- To assess for treatment in the setting of concomitant valvular disease
- As indicated by **Stress Testing with Imaging - Indications (CD-1.4)** or **Stress Echo-Indications Other than Ruling out CAD (CD-2.7)**
- There is documented concern for chronic thrombo-embolic pulmonary hypertension

**Cardiac MRI (CPT® 75557)** is indicated when there is documentation of **any** of the following:

- An echocardiogram that is equivocal or unclear (e.g., for RV function) and the information is needed for management
- MRI and echocardiogram may both be required for individuals who need RV pressure and function assessed, and prior RV function cannot be assessed by echocardiogram
- If the issue that makes the imaging by echo unclear is likely to be seen in future echocardiograms, MRI can replace echocardiogram

**Other advanced imaging** is indicated after TTE for the following:

- High-resolution CT Chest (CPT® 71250) is indicated in the setting of hypoxemia to rule out restrictive lung disorders such as pulmonary fibrosis

- CTA Chest (CPT® 71275) or MRA Chest (CPT® 71555) is indicated to evaluate for suspected acute and/or chronic pulmonary embolism
- V/Q scan (CPT® 78580-Pulmonary Perfusion Imaging or CPT® 78582- Pulmonary Ventilation (e.g., Aerosol or Gas) and Perfusion Imaging) is indicated to evaluate for any of the following:
  - Suspected acute pulmonary embolism
  - To evaluate for chronic thromboembolic pulmonary hypertension at 3 to 6 months post pulmonary embolism if both of the following apply:
    - Persistent or new onset dyspnea on exertion or exercise limitation
    - Evidence of pulmonary hypertension on follow up echo
  - To evaluate for chronic thromboembolic pulmonary hypertension in individuals with pulmonary hypertension of uncertain etiology
- SPECT imaging (CPT® 78803) or SPECT/CT imaging (CPT® 78830) can be added to V/Q scan if requested
- Transesophageal (TEE) contrast echocardiography or other imaging techniques (e.g., CT angiography, cardiac MRI) may be indicated, in addition to 2D Doppler and contrast examinations, to identify CHD to detect or exclude **any** of the following:
  - Sinus venosus
  - Atrial septal defects
  - Patent ductus arteriosus
  - Anomalous pulmonary venous connections

### Indications for initial Catheterization

Right heart catheterization (RHC) is indicated for **any** of the following:

- Echo findings
  - TR gradient  $\geq 3.4$  m/second
  - TR gradient  $\geq 2.9$  m/second and presence of other PH signs on echo or other testing, or risk factors or associated indications
- Individuals with SSc where breathlessness remains unexplained (RHC is recommended despite normal echocardiogram).
- Individuals with connective tissue disorder who have symptoms or concerns for PH with a negative or equivocal echocardiogram.
- When recommended to determine if shunt closure is recommended due to congenital heart disease
- RHC if **moderate** to **severe** PH seen based on echocardiogram.
- RHC is indicated prior to starting PH medical therapy.
- Individuals with **low** risk profile only need cath if indicated for another reason or equivocal studies.
- RHC allowed when LHC indicated for separate indication.
- RHC preoperative for surgical intervention treating the cause of PH (MV, TV, AV, PV).

- Eisenmenger syndrome – RHC is indicated when requested by provider.

**Left heart catheterization (LHC) or Right and left heart catheterization** as per the following guidelines:

- **Evaluation of Conditions Other than Coronary Artery Disease (CD-7.7)**
- **Diagnostic Left Heart Catheterization (LHC) (CD-7.3)**

## Repeat Testing

### **Follow-up echocardiogram (TTE) on patients with PH**

- Every 6 months for surveillance of stable patients
- Prior to planned intubation (e.g., for elective surgery)
- Prior to planned pregnancy
- During pregnancy as often as requested by provider
- Anytime, without regard for the number or timing of previous ECHO studies to evaluate either:
  - Change in therapy
  - Change in clinical findings or symptoms
- Echocardiogram at baseline then every 3 months with therapy changes in stable patients

### **Pulmonary embolism (PE)**

- TTE is indicated 3 to 6 months post pulmonary embolism if **any** of the following apply:
  - Persistent or new onset dyspnea on exertion, or exercise limitation
  - Pulmonary hypertension or right ventricular dysfunction on initial echo at PE diagnosis
  - History of recurrent pulmonary embolism

**RHC** is indicated for known PH as follows:

- At baseline
- Then every 6 months
- Anytime for clinical changes or with treatment changes

## Other related sections

- **Frequency of Echocardiography Testing (CD-2.3)** in the Cardiac Imaging Guidelines
- **Right Heart Catheterization (RHC) (CD-7.4)** in the Cardiac Imaging Guidelines
- **Pulmonary hypertension (PHT) and Eisenmenger syndrome (CD-11.3.12)** in the Cardiac Imaging Guidelines

- Congenital Heart Disease Modality Considerations (PEDCD-2.3) in the Pediatric Cardiac Imaging Guidelines
- Pediatric Pulmonary Hypertension - General (PEDCD-7) in the Pediatric Cardiac Imaging Guidelines
- Pulmonary Embolism (PE) (CH-25) in the Chest Imaging Guidelines

### ***Background and supporting information***

Pulmonary hypertension (PH) is a complex, chronic disease with multiple etiologies, that requires extensive evaluation, including ECG (right ventricular hypertrophy with/without strain, right atrial dilatation); chest x-ray; arterial blood gas, pulmonary function testing, CT angiography based on the etiology.

PH can be subdivided into the following five groups based on the underlying cause:

- Pulmonary arterial hypertension (PAH)
- PH due to left heart disease
- PH due to lung disease
- Chronic thromboembolic PH (CTEPH)
- PH with unclear and/or multifactorial mechanisms

Probability of PH is assessed at initial evaluation:

- **High** probability:
  - TR velocity  $\geq 3.4$  m/s
  - TR velocity between 2.9 to 3.4 m/s and **one** of the following:
    - Right ventricle or pulmonary artery enlargement
    - Interventricular septum flattening
    - Right ventricular systolic dysfunction
- **Intermediate** probability:
  - TR velocity between 2.9 to 3.4 m/s in the absence of other signs of PH
- **Low** probability:
  - TR velocity  $< 2.8$  m/s

Peak TR velocity  $\geq 2.8$  m/s may suggest PH; however, the presence or absence of PH cannot be reliably determined by TR velocity alone

In addition to the tricuspid regurgitation velocity, other findings on echo can increase the probability of PH, examples of these findings include:

- Abnormal tricuspid annular plane systolic excursion (TAPSE)
- Abnormal RV fractional area change (RV-FAC)
- Abnormal RV free-wall strain
- Abnormal tricuspid annulus velocity (S' wave) derived from tissue Doppler imaging
- Abnormal RV ejection fraction (RVEF) derived from 3D echocardiography



MRI can be a useful test especially with respect to RV function

Right heart cath is the gold standard for diagnosing PH

See **Severe Pulmonary Artery Hypertension (PH) and Eisenmenger Syndrome (CD-11.3.12)** for additional information regarding Eisenmenger Syndrome

## References

1. Abman SH, Hansmann G, Archer SL, et al. Pediatric Pulmonary Hypertension: Guidelines From the American Heart Association and American Thoracic Society [published correction appears in *Circulation*. 2016 Jan 26;133(4):e368]. *Circulation*. 2015;132(21):2037-2099. doi:10.1161/CIR.0000000000000329.
2. Alabed S, Shahin Y, Garg P, et al. Cardiac-MRI Predicts Clinical Worsening and Mortality in Pulmonary Arterial Hypertension: A Systematic Review and Meta-Analysis [published correction appears in *JACC Cardiovasc Imaging*. 2021 Apr;14(4):884]. *JACC Cardiovasc Imaging*. 2021;14(5):931-942. doi:10.1016/j.jcmg.2020.08.013.
3. Alabed S, Uthoff J, Zhou S, et al. Machine learning cardiac-MRI features predict mortality in newly diagnosed pulmonary arterial hypertension. *European Heart Journal - Digital Health*. 2022;3(2):265-275. doi:10.1093/ehjdh/ztac022.
4. Bossone E, Dellegrottaglie S, Patel S, et al. Multimodality imaging in pulmonary hypertension. *Can J Cardiol*. 2015;31(4):440-459. doi:10.1016/j.cjca.2015.02.012.
5. Broncano J, Bhalla S, Gutierrez FR, et al. Cardiac MRI in Pulmonary Hypertension: From Magnet to Bedside. *Radiographics*. 2020;40(4):982-1002. doi:10.1148/rg.2020190179.
6. Dong Y, Pan Z, Wang D, et al. Prognostic Value of Cardiac Magnetic Resonance-Derived Right Ventricular Remodeling Parameters in Pulmonary Hypertension: A Systematic Review and Meta-Analysis. *Circ Cardiovasc Imaging*. 2020;13(7):e010568. doi:10.1161/CIRCIMAGING.120.010568.
7. Expert Panel on Thoracic Imaging, Sirajuddin A, Mirmomen SM, et al. ACR Appropriateness Criteria® Suspected Pulmonary Hypertension: 2022 Update. *J Am Coll Radiol*. 2022;19(11S):S502-S512. doi:10.1016/j.jacr.2022.09.018.
8. Goh ZM, Balasubramanian N, Alabed S, et al. Right ventricular remodelling in pulmonary arterial hypertension predicts treatment response. *Heart*. 2022;108(17):1392-1400. Published 2022 Aug 11. doi:10.1136/heartjnl-2021-320733.
9. Hulten EA, Bradley AJ. Cardiac Magnetic Resonance Evaluation of Pulmonary Arterial Hypertension: Transforming From Supplementary to Primary Imaging Modality? *JACC Cardiovasc Imaging*. 2021;14(5):943-946. doi:10.1016/j.jcmg.2020.11.022.
10. Humbert M, Kovacs G, Hoeper MM, et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Heart J*. 2022;43(38):3618-3731. doi:10.1093/eurheartj/ehac237.
11. Kaemmerer H, Apitz C, Brockmeier K, et al. Pulmonary hypertension in adults with congenital heart disease: Updated recommendations from the Cologne Consensus Conference 2018. *Int J Cardiol*. 2018;272S:79-88. doi:10.1016/j.ijcard.2018.08.078.
12. Lewis MJ, Van Dissel A, Kochav J, et al. Cardiac MRI predictors of adverse outcomes in adults with a systemic right ventricle. *ESC Heart Fail*. 2022;9(2):834-841. doi:10.1002/ehf2.13745.
13. Mazurek A, Dziuk M, Witkowska-Patena E, Piszczek S, Gizewska A. The Utility of Hybrid SPECT/CT Lung Perfusion Scintigraphy in Pulmonary Embolism Diagnosis. *Respiration*. 2015;90(5):393-401. doi:10.1159/000439543.
14. McLaughlin VV, Archer SL, Badesch DB, et al. ACCF/AHA 2009 expert consensus document on pulmonary hypertension a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association developed in collaboration with the American College of Chest Physicians; American Thoracic Society, Inc.; and the Pulmonary Hypertension Association. *J Am Coll Cardiol*. 2009;53(17):1573-1619. doi:10.1016/j.jacc.2009.01.004.
15. Moledina S, Pandya B, Bartsota M, et al. Prognostic significance of cardiac magnetic resonance imaging in children with pulmonary hypertension. *Circ Cardiovasc Imaging*. 2013;6(3):407-414. doi:10.1161/CIRCIMAGING.112.000082.
16. Ostenfeld E, Kjellström B. The Conundrum of Right Ventricular Remodeling and Outcome in Pulmonary Hypertension. *Circ Cardiovasc Imaging*. 2020;13(7):e011208. doi:10.1161/CIRCIMAGING.120.011208.
17. Reddy YNV, Carter RE, Obokata M, Redfield MM, Borlaug BA. A Simple, Evidence-Based Approach to Help Guide Diagnosis of Heart Failure with Preserved Ejection Fraction. *Circulation*. 2018;138(9):861-870. doi:10.1161/CIRCULATIONAHA.118.034646.
18. Remy-Jardin M, Ryerson CJ, Schiebler ML, et al. Imaging of Pulmonary Hypertension in Adults: A Position Paper from the Fleischner Society. *Radiology*. 2021;298(3):531-549. doi:10.1148/radiol.2020203108.



19. Runser LA, Gauer R, Houser A. Syncope: Evaluation and Differential Diagnosis. *AAFP Home*. <https://www.aafp.org/afp/2017/0301/p303.html>. Published March 1, 2017.
20. Sato T, Ambale-Venkatesh B, Zimmerman SL, et al. Right ventricular function as assessed by cardiac magnetic resonance imaging-derived strain parameters compared to high-fidelity micromanometer catheter measurements. *Pulm Circ*. 2021;11(4):20458940211032529. Published 2021 Sep 24. doi:10.1177/20458940211032529.
21. Thenappan T, Ormiston ML, Ryan JJ, Archer SL. Pulmonary arterial hypertension: pathogenesis and clinical management. *BMJ*. 2018;360:j5492. Published 2018 Mar 14. doi:10.1136/bmj.j5492.
22. van der Bruggen CE, Handoko ML, Bogaard HJ, et al. The Value of Hemodynamic Measurements or Cardiac MRI in the Follow-up of Patients with Idiopathic Pulmonary Arterial Hypertension. *Chest*. 2021;159(4):1575-1585. doi:10.1016/j.chest.2020.10.077.
23. van Wolferen SA, Marcus JT, Boonstra A, et al. Prognostic value of right ventricular mass, volume, and function in idiopathic pulmonary arterial hypertension. *Eur Heart J*. 2007;28(10):1250-1257. doi:10.1093/eurheartj/ehl477.
24. Xue L, Yang Y, Sun B, Liu B, Zeng Q, Xiong C. Mildly Elevated Pulmonary Arterial Pressure Is Associated with a High Risk of Progression to Pulmonary Hypertension and Increased Mortality: A Systematic Review and Meta-Analysis. *J Am Heart Assoc*. 2021;10(7):e018374.
25. Yaghi S, Novikov A, Trandafirescu T. Clinical update on pulmonary hypertension. *J Investig Med*. 2020;68(4):821-827. doi:10.1136/jim-2020-001291.

# Pulmonary Vein Imaging – Indications (CD-8.2)

CD.CS.0008.2.A

v2.0.2023

- MRI Cardiac (CPT® 75557 or CPT® 75561), MRV Chest (CPT® 71555), CTV Chest (CPT® 71275), or CT Cardiac (CPT® 75572) to evaluate anatomy of the pulmonary veins:
  - Prior to planned atrial fibrillation ablation/pulmonary vein isolation procedure
  - Post-procedure between 3-6 months after ablation because of a 1% to 2% incidence of asymptomatic pulmonary vein stenosis
    - If no pulmonary vein stenosis is present, no further follow-up imaging is required
    - If pulmonary vein stenosis is present on imaging following ablation and symptoms of pulmonary vein stenosis (usually shortness of breath) are present, can be imaged at 1, 3, 6, and 12 months

## Background and Supporting Information

The majority (81%) of pulmonary vein stenosis remain stable over 1 year. Progression occurs in 8.8% and regression occurs in a small percentage.

## References

1. Abman SH, Hansmann G, Archer SL, et al. Pediatric Pulmonary Hypertension: Guidelines From the American Heart Association and American Thoracic Society [published correction appears in *Circulation*. 2016 Jan 26;133(4):e368]. *Circulation*. 2015;132(21):2037-2099. doi:10.1161/CIR.0000000000000329.
2. Alabed S, Shahin Y, Garg P, et al. Cardiac-MRI Predicts Clinical Worsening and Mortality in Pulmonary Arterial Hypertension: A Systematic Review and Meta-Analysis [published correction appears in *JACC Cardiovasc Imaging*. 2021 Apr;14(4):884]. *JACC Cardiovasc Imaging*. 2021;14(5):931-942. doi:10.1016/j.jcmg.2020.08.013.
3. Alabed S, Uthoff J, Zhou S, et al. Machine learning cardiac-MRI features predict mortality in newly diagnosed pulmonary arterial hypertension. *European Heart Journal - Digital Health*. 2022;3(2):265-275. doi:10.1093/ehjdh/ztac022.
4. Bossone E, Dellegrottaglie S, Patel S, et al. Multimodality imaging in pulmonary hypertension. *Can J Cardiol*. 2015;31(4):440-459. doi:10.1016/j.cjca.2015.02.012.
5. Broncano J, Bhalla S, Gutierrez FR, et al. Cardiac MRI in Pulmonary Hypertension: From Magnet to Bedside. *Radiographics*. 2020;40(4):982-1002. doi:10.1148/rg.2020190179.
6. Dong Y, Pan Z, Wang D, et al. Prognostic Value of Cardiac Magnetic Resonance-Derived Right Ventricular Remodeling Parameters in Pulmonary Hypertension: A Systematic Review and Meta-Analysis. *Circ Cardiovasc Imaging*. 2020;13(7):e010568. doi:10.1161/CIRCIMAGING.120.010568.
7. Expert Panel on Thoracic Imaging, Sirajuddin A, Mirmomen SM, et al. ACR Appropriateness Criteria® Suspected Pulmonary Hypertension: 2022 Update. *J Am Coll Radiol*. 2022;19(11S):S502-S512. doi:10.1016/j.jacr.2022.09.018.
8. Goh ZM, Balasubramanian N, Alabed S, et al. Right ventricular remodelling in pulmonary arterial hypertension predicts treatment response. *Heart*. 2022;108(17):1392-1400. Published 2022 Aug 11. doi:10.1136/heartjnl-2021-320733.

9. Hulten EA, Bradley AJ. Cardiac Magnetic Resonance Evaluation of Pulmonary Arterial Hypertension: Transforming From Supplementary to Primary Imaging Modality? *JACC Cardiovasc Imaging*. 2021;14(5):943-946. doi:10.1016/j.jcmg.2020.11.022.
10. Humbert M, Kovacs G, Hoeper MM, et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Heart J*. 2022;43(38):3618-3731. doi:10.1093/eurheartj/ehac237.
11. Kaemmerer H, Apitz C, Brockmeier K, et al. Pulmonary hypertension in adults with congenital heart disease: Updated recommendations from the Cologne Consensus Conference 2018. *Int J Cardiol*. 2018;272S:79-88. doi:10.1016/j.ijcard.2018.08.078.
12. Lewis MJ, Van Dissel A, Kochav J, et al. Cardiac MRI predictors of adverse outcomes in adults with a systemic right ventricle. *ESC Heart Fail*. 2022;9(2):834-841. doi:10.1002/ehf2.13745.
13. Mazurek A, Dziuk M, Witkowska-Patena E, Piszczek S, Gizewska A. The Utility of Hybrid SPECT/CT Lung Perfusion Scintigraphy in Pulmonary Embolism Diagnosis. *Respiration*. 2015;90(5):393-401. doi:10.1159/000439543.
14. McLaughlin VV, Archer SL, Badesch DB, et al. ACCF/AHA 2009 expert consensus document on pulmonary hypertension a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association developed in collaboration with the American College of Chest Physicians; American Thoracic Society, Inc.; and the Pulmonary Hypertension Association. *J Am Coll Cardiol*. 2009;53(17):1573-1619. doi:10.1016/j.jacc.2009.01.004.
15. Moledina S, Pandya B, Bartsota M, et al. Prognostic significance of cardiac magnetic resonance imaging in children with pulmonary hypertension. *Circ Cardiovasc Imaging*. 2013;6(3):407-414. doi:10.1161/CIRCIMAGING.112.000082.
16. Ostenfeld E, Kjellström B. The Conundrum of Right Ventricular Remodeling and Outcome in Pulmonary Hypertension. *Circ Cardiovasc Imaging*. 2020;13(7):e011208. doi:10.1161/CIRCIMAGING.120.011208.
17. Reddy YNV, Carter RE, Obokata M, Redfield MM, Borlaug BA. A Simple, Evidence-Based Approach to Help Guide Diagnosis of Heart Failure with Preserved Ejection Fraction. *Circulation*. 2018;138(9):861-870. doi:10.1161/CIRCULATIONAHA.118.034646.
18. Remy-Jardin M, Ryerson CJ, Schiebler ML, et al. Imaging of Pulmonary Hypertension in Adults: A Position Paper from the Fleischner Society. *Radiology*. 2021;298(3):531-549. doi:10.1148/radiol.2020203108.
19. Runser LA, Gauer R, Houser A. Syncope: Evaluation and Differential Diagnosis. *AAFP Home*. <https://www.aafp.org/afp/2017/0301/p303.html>. Published March 1, 2017.
20. Sato T, Ambale-Venkatesh B, Zimmerman SL, et al. Right ventricular function as assessed by cardiac magnetic resonance imaging-derived strain parameters compared to high-fidelity micromanometer catheter measurements. *Pulm Circ*. 2021;11(4):20458940211032529. Published 2021 Sep 24. doi:10.1177/20458940211032529.
21. Thenappan T, Ormiston ML, Ryan JJ, Archer SL. Pulmonary arterial hypertension: pathogenesis and clinical management. *BMJ*. 2018;360:j5492. Published 2018 Mar 14. doi:10.1136/bmj.j5492.
22. van der Bruggen CE, Handoko ML, Bogaard HJ, et al. The Value of Hemodynamic Measurements or Cardiac MRI in the Follow-up of Patients with Idiopathic Pulmonary Arterial Hypertension. *Chest*. 2021;159(4):1575-1585. doi:10.1016/j.chest.2020.10.077.
23. van Wolferen SA, Marcus JT, Boonstra A, et al. Prognostic value of right ventricular mass, volume, and function in idiopathic pulmonary arterial hypertension. *Eur Heart J*. 2007;28(10):1250-1257. doi:10.1093/eurheartj/ehl477.
24. Xue L, Yang Y, Sun B, Liu B, Zeng Q, Xiong C. Mildly Elevated Pulmonary Arterial Pressure Is Associated with a High Risk of Progression to Pulmonary Hypertension and Increased Mortality: A Systematic Review and Meta-Analysis. *J Am Heart Assoc*. 2021;10(7):e018374.
25. Yaghi S, Novikov A, Trandafirescu T. Clinical update on pulmonary hypertension. *J Investig Med*. 2020;68(4):821-827. doi:10.1136/jim-2020-001291.

# Hypertrophic Cardiomyopathy (HCM) (CD-14)

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Hypertrophic Cardiomyopathy (HCM) is a clinical diagnosis, established by imaging with 2D echocardiography or cardiovascular magnetic resonance (CMR) showing a maximal end-diastolic wall thickness of  $\geq 15$  mm anywhere in the left ventricle, in the absence of another cause of hypertrophy in adults. More limited hypertrophy (13–14 mm) can be diagnostic, particularly when present in family members of a patient with HCM or in conjunction with a positive genetic test, and/or associated with typical dynamic outflow obstruction, or distinctly abnormal ECG patterns.

## Screening

- Screening for inherited hypertrophic cardiomyopathy see **Transthoracic Echocardiography (TTE) – Indications (CD-2.2)** and **Frequency of Echocardiography Testing (CD-2.3)**

## Initial imaging, new or changed symptoms

### TTE

- TTE is indicated for the initial evaluation of a genotype positive individual with inherited hypertrophic cardiomyopathy

### Stress echocardiogram

- Exercise stress echo (CPT® 93351 or 93350) is indicated for the detection and quantification of dynamic left ventricular outflow tract obstruction in symptomatic individuals with HCM who do **not** have a resting or provokable outflow tract gradient  $\geq 50$  mm Hg on TTE.
- Stress echo can be repeated in 1 to 2 years in an individual with a documented history of HCM previously evaluated with a stress echo when there is documentation of **either** of the following:
  - Worsening symptoms
  - There has been a therapeutic change (i.e., change in medication, surgical procedure performed).

### CCTA (CPT® 75574)

- Initial imaging study in individuals with hypertrophic cardiomyopathy and stable anginal symptoms.
  - Chest discomfort is common in individuals with hypertrophic cardiomyopathy. The incidence of false positive myocardial perfusion imaging abnormalities is

higher in these individuals, whereas the incidence of severe coronary artery stenosis is low.

#### Cardiac MRI (CMR)

- Cardiac MRI (CPT® 75557 or CPT® 75561) for assessment of global ventricular function, myocardial composition, and mass if a specific clinical question is left unanswered by a recent echocardiogram and results will affect patient management.

#### Surveillance imaging

- TTE is indicated every three years when there is no change in clinical status or treatment

#### Monitoring treatment

Repeat TTE (CPT® 93306) is indicated in individuals with Obstructive Hypertrophic Cardiomyopathy (HCM) for the following:

#### Mavacamten for obstructive hypertrophic cardiomyopathy

##### Initiation of treatment

- Baseline-at the beginning of treatment
- 4 weeks after treatment initiation
- 8 weeks after treatment initiation
- 12 weeks after treatment initiation
- Then every 12 weeks while on mavacamten

##### Changes in treatment

- 4 weeks after any interruption of treatment (any missed dose)
- After any dosage change (including restart of treatment):
  - 4 weeks after dosage change
  - 12 weeks after dosage change
- After initiating a weak CYP2C19 inhibitor (e.g., omeprazole) or moderate CYP2A4 inhibitor (e.g., ciprofloxacin):
  - 4 weeks after start of medication
  - 12 weeks after start of medication
- At any time regardless of timing of prior echo when there are new cardiac signs or symptoms, or worsening of clinical status

**Post- Septal Reduction Therapy (SRT)**

TTE is indicated within 3 to 6 months after SRT (surgical myectomy or alcohol septal ablation) to evaluate the procedural results in individuals with hypertrophic cardiomyopathy