Adult Congenital Heart Disease

Clinical Guidelines for Medical Necessity Review

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Important Notices

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Other Notices:

Guideline Information:

Disease Area: Cardiology
Care Path Group: General Cardiology
Care Path Name: Adult Congenital Heart Disease
Type: [**X**] Adult (18+ yo) | [_] Pediatric (0-17 yo)

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Care Path Overview

Care Path Clinical Discussion

Congenital heart disease (CHD) refers to congenital malformations of the heart. These include heart valves, atrial and ventricular septal defects, stenosis, and heart muscle abnormalities. Approximately 90% of children diagnosed with severe CHD survive to adulthood.[1] Even following successful pediatric treatment, almost all adult congenital heart disease (ACHD) patients will have sequelae of either their original defect or surgical repair or palliation. [1] There is a wide variety in the anatomy, physiology, and surgical repair or palliation of ACHD patients.

A bicuspid aortic valve (BAV) is the most prevalent congenital cardiac abnormality in adults, with an estimated prevalence of 4.6 per 1,000 live births. [1] Isolated VSDs are the most common form of CHD in the pediatric population, which can lead to long-term implications throughout adult life. [1] Valvular pulmonic stenosis is also one of the most common congenital heart defects after BAV, occurring in approximately 7% of children born with CHD. [1] Atrial septal defects are common in adults with congenital heart disease, accounting for approximately 10% to 15% of all cardiac defects. Tetralogy of Fallot is the most common form of cyanotic congenital heart disease in children and adults.

Congenital heart disease may be associated with underlying genetic syndromes. Adults with Trisomy 21 often have a history of repaired and unrepaired CHDs. Prior childhood genetic workup may not be available, potentially causing missed diagnosis of genetic syndromes in patients with ACHD. Up to 5% of children with CHD have a chromosomal disorder called DiGeorge Syndrome (22q11.2 deletion), which often presents with conotruncal defects. DiGeorge Syndrome is an autosomal dominant condition, so genetic testing is reasonable for patients with conotruncal defects.[1]It is also indicated that female patients who are pregnant with a previous history of CHD undergo genetic testing to risk stratify the recurrence of CHDs in offspring.

Late surviving adults with CHD may experience significant cardiac events, such as:

- Myocardial infarction.
- Stroke.
- Heart failure.
- Infective endocarditis.
- Malignant arrhythmias.[1]

- Sudden cardiac death.[1]

Heart failure is common in patients with ACHD. However, many causes may be reversible, including valve dysfunction, shunts, arrhythmias, venous obstruction, conduit dysfunction, and ventricular dysfunction. Because clinical trials often exclude patients with ACHD, there is limited data to support treatment decisions in these patients.[1] Treatment decisions must be individualized to the patient's anatomy, surgical repair, and physiology. [1]

Congenital heart disease can affect patients in other ways, including:

- Depression and anxiety are prevalent in this population. [1]
- ACDH patients who received a blood transfusion before 1992 are at higher risk of hepatitis C and should undergo testing.[1]
- Women with ACHD should receive pre-pregnancy counseling to evaluate maternal cardiac, obstetrical, and fetal risks and potential long-term risks to the mother. [1]

The information contained herein gives a general overview of the pathway of this specific diagnosis, beginning with the initial presentation, recommended assessments, and treatment options as supported by the medical literature and existing guidelines. It should be noted that the care of patients can be complex. The information below is meant to support clinical decision-making in adult patients. It is not necessarily applicable to every case, as the entire clinical picture (including comorbidities, history, etc.) should be considered.

Key Information

- ➢ Physicians may occasionally see adults with previously undiagnosed congenital heart disease. These patients may present with shortness of breath, palpitations, fatigue, or hypertension. Physicians may encounter congenital heart disease in adults who have fallen out of care after being treated as children, with complications of CHD or treatment, or previously undiagnosed CHD.
- ➢ Because of the success of pediatric cardiology and congenital cardiac surgery, the prevalence of ACHD is increasing, with survival to age 18 years now expected for 90% of children diagnosed with severe CHD.[1] Patients with ACHD are a heterogeneous population in underlying anatomy, physiology, and surgical repair or palliation.[1]
- ➢ Transthoracic echocardiography (TTE) is the most common imaging technique used to evaluate congenital heart lesions. Transesophageal echocardiography, MRI, nuclear cardiology, and cardiac catheterization may be appropriate when TTE does not provide sufficient information for anatomical or functional assessment. [2]
- ➢ Genetic testing for DiGeorge syndrome is reasonable for patients with conotruncal defects. [1]
- ➢ Late surviving adults with CHD may acquire other cardiac problems, such as myocardial infarction, coronary artery disease, heart failure, infective endocarditis, malignant arrhythmia, sudden cardiac death, and need for a pacemaker or implantable cardioverter-defibrillator (ICD). [1]

Definitions

- **Atrial Septal Defect (ASD):** A hole in the wall between the two upper chambers of the heart caused by incomplete development in utero. The most common type of ASD is a secundum septal defect; primum septal defects are commonly associated with Trisomy 21 (Down Syndrome).
- **Ventricular Septal Defect (VSD):** A hole in the wall between the two lower chambers of the heart; these can have variable locations within the septum. Residual VSDs are often components of repaired complex ACHD.
- **Tetralogy of Fallot (TOF):** The most common cyanotic heart lesion, with 4 components: VSD, pulmonary stenosis, aortic override of the VSD, and right ventricular hypertrophy. Repaired TOF patients are at-risk for

ventricular arrhythmias and sudden death due to scarring from incisions of the right ventricle and often need frequent pulmonary valve replacements.[1,3,4]

- **Dextro-transposition of the great arteries (d-TGA):** A conotruncal abnormality where the relative position of the great arteries is reversed, causing severe cyanosis before repair. Modern repair switches the great arteries to their regular positions with reimplantation of the coronary arteries. However, early techniques involved diverting blood flow within the atria (see Mustard/Senning procedures).[1]
- **Mustard/Senning Procedure:** A procedure that creates a baffle system within the atria to divert systemic and pulmonary venous return to the appropriate outflow chamber. Patients with this anatomy have a significant risk of baffle leaks, sinus node dysfunction, bradycardia, and atrial arrhythmia due to extensive suture lines in the atrial tissue.
- **Levo-Transposition of the great arteries (L-TGA):** Otherwise known as ventricular inversion, this is a condition where the great arteries are in the proper anatomic location but connected with the incorrect ventricle. Patients with this lesion have few options for repair and are at high-risk for heart failure and progressive AV block.
- **Ebstein's Anomaly:** A developmental anomaly of the Tricuspid valve caused by lack of separation of the septal leaflet from right ventricular tissue. This causes a displacement of the tricuspid valve towards the right ventricular apex, creating a smaller ventricular chamber. There is often associated tricuspid valve insufficiency, atrial communication, and thinning atrialization of part of the right ventricle. Accessory electrical connections causing supraventricular tachycardia are prevalent.[3]
- **Fontan Operation:** A procedure that completes a conduit connecting all systemic venous return to the pulmonary arteries, bypassing the right heart. This is used to palliate many CHD lesions where only one ventricle is viable. These patients are at-risk for sinus node dysfunction, atrial arrhythmias, venous thrombosis, pulmonary embolism, and heart failure.[1,5]
- **Orthodeoxia:** A condition where a patient's oxygen saturation drops when in the upright position but improves when lying down.
- **Platypnea:** A condition where shortness of breath worsens in an upright position but improves when lying down.
- **Pretest Probability (of CAD):** The pretest probability of CAD is the likelihood that the patient has CAD, calculated before the test result is

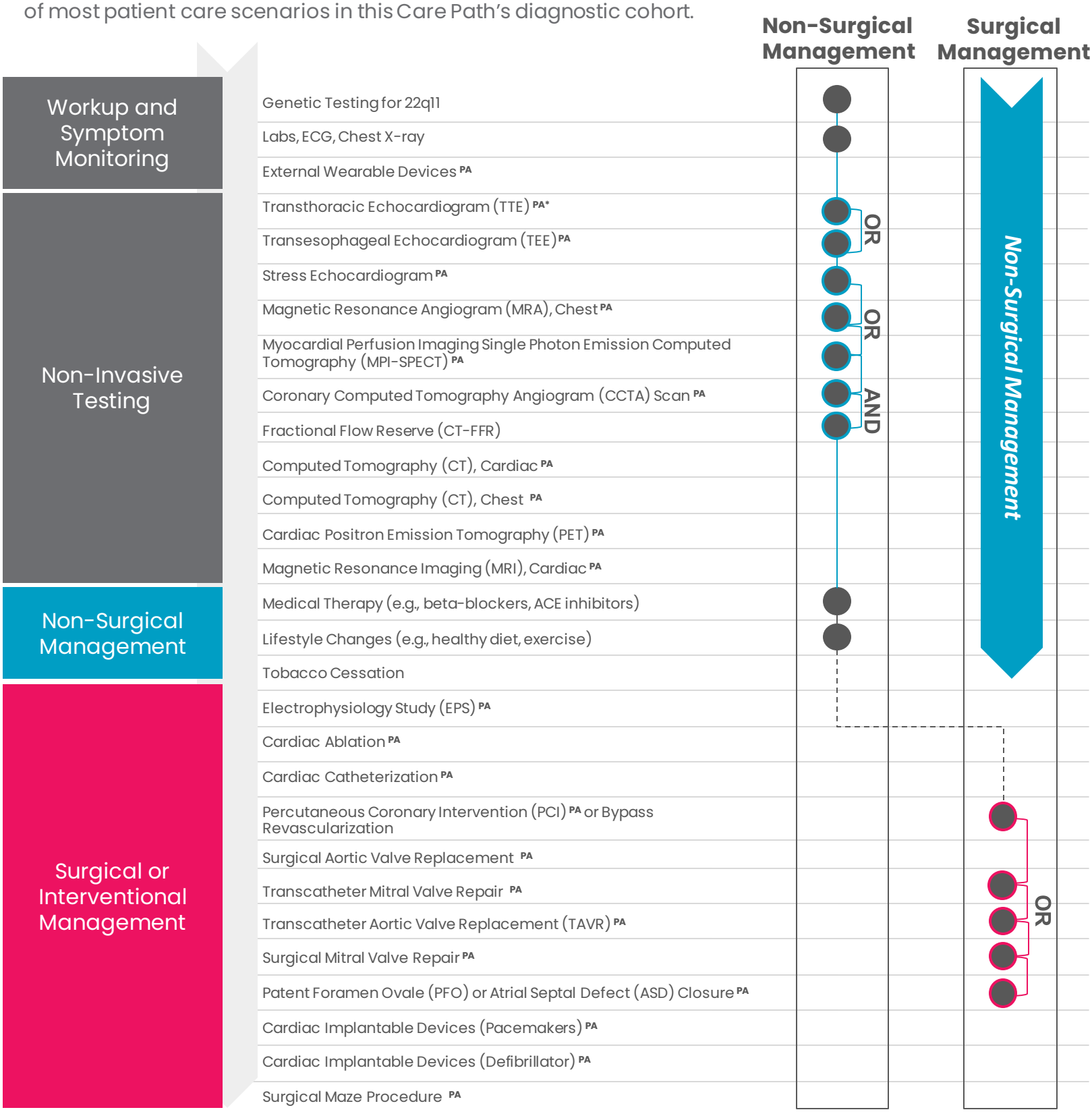
known. These guidelines reference the 2019 European Society of Cardiology (ESC) Guidelines for the diagnosis and management of chronic coronary syndromes model to calculate the pretest probability based on age, sex, and type of chest pain.[6,7,8]

- **Canadian Cardiovascular Society grading of Angina Pectoris**:
    - Grade I: Ordinary physical activity does not cause angina, such as walking and climbing stairs. Angina with strenuous or rapid or prolonged exertion at work or recreation.
    - Grade II: Slight limitation of ordinary activity. Walking or climbing stairs rapidly, walking uphill, walking or stair climbing after meals, or in the cold, or in the wind, or under emotional stress, or only during the few hours after awakening. Walking more than two blocks on the level and climbing more than one flight of ordinary stairs at a normal pace and in normal conditions.
    - Grade III: Marked limitation of ordinary physical activity. Walking one or two blocks on the level and climbing one flight of stairs in normal conditions and at normal pace.
    - Grade IV: Inability to carry on any physical activity without discomfort, anginal syndrome may be present at rest.
- **New York Heart Association (NYHA) Classification:** A common measure of heart failure.
    - Class I: The patient has a cardiac disease that does not limit ordinary physical activity.
    - Class II (Mild): The patient has cardiac disease, causing slight limitations in physical activity. Ordinary physical activity results in fatigue, palpitation, dyspnea, or anginal pain. Comfortable at rest.
    - Class III (Moderate): The patient has a cardiac disease that noticeably limits physical activity. Less than ordinary activity causes fatigue, palpitation, dyspnea, or anginal pain. Comfortable at rest.
    - Class IV (Severe): The patient has a cardiac disease that prevents them from physical activity. Experiences symptoms and discomfort at rest.

Adult Congenital Heart Disease

What is a "Cohere Care Path"?

These Care Paths organize the services typically considered most clinically optimal and likely to be automatically approved. These service recommendations also include the suggested sequencing and quantity or frequency determined clinically appropriate and medically necessary for the management of most patient care scenarios in this Care Path's diagnostic cohort.



Key

PA = Service may require prior authorization
★ = Denotes preferred service
AND = Services completed concurrently
OR = Services generally mutually exclusive

= Non-surgical management prior authorization group of services
= Surgical management prior authorization group of services
= Subsequent service
= Management path moves to a different management path

Care Path Diagnostic Criteria

Disease Classification

Congenital Heart Disease (Heart disease from birth).

ICD-10 Codes Associated with Classification

ICD-10 Code	Code Description/Definition
I23.2	Ventricular septal defect as current complication following acute myocardial infarction
I25.41	Coronary artery aneurysm
I27.83	Eisenmenger's syndrome
I28.9	Disease of pulmonary vessels, unspecified
I28.0	Arteriovenous fistula of pulmonary vessels
I36	Nonrheumatic tricuspid valve disorders
I37.0	Nonrheumatic pulmonary valve stenosis
I51.0	Cardiac septal defect, acquired
I77.0	Arteriovenous fistula, acquired
I77.810	Thoracic aortic ectasia
I77.812	Thoracoabdominal aortic ectasia
I77.819	Aortic ectasia, unspecified site
I82.210	Acute embolism and thrombosis of superior vena cava
I82.220	Acute embolism and thrombosis of inferior vena cava
M31.4	Aortic arch syndrome [Takayasu]
Q20.0	Common Arterial Trunk
Q20.1	Double outlet right ventricle
Q20.3	Dextro-Transposition of the Great Arteries
Q20.4	Double inlet ventricle
Q20.5	Levo-Transposition of the Great Arteries
Q20.8	Other congenital malformations of cardiac chambers and

	connections
Q20.9	Congenital malformation of cardiac chambers and connections, unspecified
Q21.0	Ventricular septal defect
Q21.10	Atrial septal defect, unspecified
Q21.11	Secundum atrial septal defect
Q21.12	Patent foramen ovale
Q21.13	Coronary sinus atrial septal defect
Q21.14	Superior sinus venosus atrial septal defect
Q21.15	Inferior sinus venosus atrial septal defect
Q21.16	Sinus venosus atrial septal defect, unspecified
Q21.19	Other specified atrial septal defect
Q21.20	Atrioventricular septal defect, unspecified as to partial or complete
Q21.21	Partial atrioventricular septal defect
Q21.22	Transitional atrioventricular septal defect
Q21.23	Complete atrioventricular septal defect
Q21.3	Tetralogy of Fallot
Q21.8	Other congenital malformations of cardiac septa
Q21.9	Congenital malformation of cardiac septum, unspecified
Q22.0	Pulmonary valve atresia
Q22.1	Congenital pulmonary valve stenosis
Q22.2	Congenital pulmonary valve insufficiency
Q22.4	Congenital tricuspid stenosis
Q22.5	Ebstein's Anomaly
Q22.8	Other congenital malformations of tricuspid valve
Q23.0	Congenital stenosis of aortic valve
Q23.1	Congenital insufficiency of aortic valve
Q23.2	Congenital mitral stenosis
Q23.3	Congenital mitral insufficiency

Q23.4	Hypoplastic left heart syndrome
Q23.8	Other congenital malformations of aortic and mitral valves
Q23.9	Congenital malformation of aortic and mitral valves, unspecified
Q24.0	Dextrocardia
Q24.1	Levocardia
Q24.4	Congenital subaortic stenosis
Q24.5	Malformation of coronary vessels
Q24.8	Other specified congenital malformations of heart
Q24.9	Congenital malformation of heart, unspecified
Q25.0	Patent ductus arteriosus
Q25.1	Coarctation of aorta
Q25.21	Interruption of aortic arch
Q25.3	Supravalvular aortic stenosis
Q25.40	Congenital malformation of aorta unspecified
Q25.43	Congenital aneurysm of aorta
Q25.44	Congenital dilation of aorta
Q25.45	Double aortic arch
Q25.46	Tortuous aortic arch
Q25.47	Right aortic arch
Q25.5	Atresia of pulmonary artery
Q25.6	Stenosis of pulmonary artery
Q25.72	Congenital pulmonary arteriovenous malformation
Q25.79	Other congenital malformations of pulmonary artery
Q25.8	Other congenital malformations of other great arteries
Q26.1	Persistent left superior vena cava
Q26.2	Total anomalous pulmonary venous connection
Q26.3	Partial anomalous pulmonary venous connection
Q26.4	Anomalous pulmonary venous connection, unspecified
Q26.8	Other congenital malformations of great veins

Q87.410	Marfan's syndrome with aortic dilation
Q87.418	Marfan's syndrome with other cardiovascular manifestations
Q89.3	Situs inversus
T82.01XD	Breakdown (mechanical) of heart valve prosthesis, subsequent encounter
T82.03XA	Leakage of heart valve prosthesis, initial encounter
T82.03XD	Leakage of heart valve prosthesis, subsequent encounter
T82.09XA	Other mechanical complication of heart valve prosthesis, initial encounter
T82.118A	Breakdown (mechanical) of other cardiac electronic device, initial encounter
T82.223A	Leakage of biological heart valve graft, initial encounter
T82.538D	Leakage of other cardiac and vascular devices and implants, subsequent encounter
T82.6XXA	Infection and inflammatory reaction due to cardiac valve prosthesis, initial encounter
T82.817A	Embolism due to cardiac prosthetic devices, implants and grafts, initial encounter
T82.857A	Stenosis of other cardiac prosthetic devices, implants and grafts, initial encounter
T82.897A	Other specified complication of cardiac prosthetic devices, implants and grafts, initial encounter
T82.897D	Other specified complication of cardiac prosthetic devices, implants and grafts, subsequent encounter
Z36.83	Encounter for fetal screening for congenital cardiac abnormalities
Z45.09	Encounter for adjustment and management of other cardiac device
Z87.74	Personal history of (corrected) congenital malformations of heart and circulatory system
Z95.2	Presence of prosthetic heart valve

Z95.3	Presence of xenogenic heart valve
Z95.4	Presence of other heart-valve replacement
Z95.811	Presence of heart assist device

Presentation and Etiology

Causes and Risk Factors

Risk factors for congenital heart disease include:

- Genetic or chromosomal abnormalities, such as Down syndrome.
- Excessive alcohol consumption during pregnancy.
- The use of certain medications in pregnancy, such as ACE inhibitors, statins, and lithium.
- Maternal viral infection.
- Maternal diabetes, type 1 or 2.
- A family history of congenital heart disease.

Clinical Presentation[9]

Physicians may occasionally see adults with previously undiagnosed congenital heart disease. These patients may present with shortness of breath, palpitations, fatigue, or hypertension. Physicians may often encounter congenital heart disease in adults who have fallen out of care after being diagnosed and treated as children. Some adult patients present with complications from their initial surgeries, such as pulmonary regurgitation from surgery as an infant that involved enlarging the pulmonary valve and right ventricular outflow tract.

A thorough history of adults with congenital heart disease includes a detailed pediatric history. This includes the prenatal course and details of any interventions. The physician should inquire about the patient's functional abilities and cardiac symptoms, such as shortness of breath, limited ability to exercise, palpitations, and fatigue.

Typical Physical Exam Findings

A detailed physical examination should be performed. A patient with congenital heart disease may have a scar from a previous procedure that leads to further evaluation. Essential exam components include vital signs, as well as a thorough lung and cardiovascular assessment. The physician should look for signs of heart failure, murmurs, and other cardiac comorbidities. Evaluation should be comprehensive, including JVD

assessment, liver examination, and looking for peripheral stigmata of underlying conditions.

Typical Evaluation and Diagnostic Findings

The clinical approach to adults with congenital heart diseases is unique because each patient will be anatomically and physiologically unique—the type of repair, associated defects, long-term risk factors, and complications will vary. Due to improved survival rates of ACHD patients, acquired heart problems such as coronary artery disease contribute to morbidity and mortality. Therefore, the physician should also pay attention to the signs and symptoms of acquired heart disease and perform an appropriate evaluation.

Following a thorough history and physical exam, an ECG and chest x-ray help clarify the diagnosis and determine further evaluation. Transthoracic echocardiography (TTE) is the primary imaging technique for anatomic evaluation. Transesophageal echocardiography (TEE), computed tomography (CT/CTA), magnetic resonance imaging (MRI/MRA), nuclear cardiology, and cardiac catheterization may be appropriate when TTE does not provide sufficient information for anatomical or functional assessment. [2] The American College of Cardiology recommends limiting radiation exposure for patients with ACHD whenever appropriate. [1]

Patients with ACHD require periodic follow-up. The frequency of repeat cardiac testing depends on the clinical scenario, including the type and severity of the defect. In addition to routine monitoring, a repeat TTE may also be appropriate if new symptoms arise or there is a change in the physical examination.

Care Path Services & Medical Necessity Criteria

Workup and Symptom Monitoring

Service: Genetic Testing, 22q11

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** Complete testing for a specific genetic disease only once unless new capabilities for detecting additional mutations develop.
- **Recommended Clinical Approach[10]:** Genetic testing for 22q11 deletions is reasonable for patients with conotruncal cardiac defects. [!] This genetic syndrome is also associated with characteristic facial features and behavioral disorders such as autistic spectrum disorder, ADHD, or schizophrenia. The mainstay of genetic testing for 22q11 deletions is fluorescence in situ hybridization (FISH), which accounts for about 78% of all mutations in the gene segment.
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **Genetic testing (*22q11*)** is considered appropriate if **ALL** of the following is **TRUE**[10]:

- ◆ The patient has **ANY** of the following:
  - Truncus arteriosus.
  - Tetralogy of Fallot.
  - An interrupted aortic arch.
  - A double outlet right or left ventricle.
  - Transposition of the great arteries.
  - An aortopulmonary septal defect.
  - Subarterial VSD with right aortic arch.
  - Pregnant or considering pregnancy with previously treated or untreated CHD.
- ◆ The patient has not had prior genetic testing for 22q11.

Non-Indications

- ➔ **Genetic testing (*22q11*)** is not considered appropriate if **ANY** of the following is **TRUE**:
  - ◆ Genetic testing was already completed.

Site of Service Criteria

Outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
81422	Genomic Sequencing Procedures and Other Molecular Multianalyte Assays

Service: Genetic Testing, Cytogenomic Constitutional (genome-wide) Microarray Analysis (CMA)

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** Complete testing for a specific genetic disease only once unless new capabilities for detecting additional mutations develop.
- **Recommended Clinical Approach:** The majority of all congenital heart defects (CHDs) are isolated or non-syndromic; however, up to 20-30% may be part of well-known chromosomal and single-gene syndromes (e.g., trisomy 21, trisomy 18, and Noonan syndrome).[11] Published guidelines have emphasized testing for chromosomal abnormalities by G-banded karyotyping and testing for single-gene disorders. However, other techniques, such as testing using Cytogenomic constitutional (genome-wide) microarray analysis (CMA), including interrogation of genomic regions for copy number and single nucleotide polymorphism (SNP) variants for chromosomal abnormalities, are now commonly ordered and provide improved diagnostic yield.[12]
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **Genetic testing (CMA)** is considered appropriate if **ALL** of the following are **TRUE**:
- ◆ The patient has **ALL** of the following[13]:
  - Multiple congenital anomalies
  - Findings not specific to a well-delineated genetic syndrome.
- ◆ The patient has not had prior CMA genetic testing.

Non-Indications

➔ **Genetic testing (CMA)**is not considered appropriate if **ANY** of the following is **TRUE**:
- ◆ CMA testing was already completed.

Site of Service Criteria

Outpatient.

HCPCS Code	Code Description/Definition
81229	Cytogenomic constitutional (genome-wide) microarray analysis; interrogation of genomic regions for copy number and single nucleotide polymorphism (SNP) variants for chromosomal abnormalities

Service: External Wearable Devices

General Guidelines

- **Units, Frequency, & Duration:** When medically necessary, use external wearable devices for 24 hours to 30 days, depending on symptom frequency.
- **Criteria for Subsequent Requests:** Subsequent requests may be appropriate when the device malfunctions, has a high burden of poor quality data/artifact, or cannot/did not capture a recording of patient symptoms (e.g., the initial monitor chosen was of shorter duration or was solely dependent on patient-activated data.) In certain high-risk clinical conditions, routine annual or biannual testing may be appropriate.
- **Recommended Clinical Approach:** Ambulatory electrocardiographic monitoring is appropriate for patients with congenital heart disease (CHD) who are at-risk of tachyarrhythmia, bradyarrhythmia, or heart block, or when the patient has symptoms suggesting arrhythmia.[1] The external wearable monitor should be selected based on the patient's symptom frequency and duration of the episodes. A 24-48 hour Holter monitor may address daily symptoms or ongoing rhythm abnormalities (e.g., frequent premature ventricular contractions). Less frequent episodic palpitations are more likely to be captured with more extended monitoring, either a 30-day loop recorder, cardiac mobile telemetry, or an extended-wear patch device. Consideration of the patient's ability to trigger a device effectively may also guide device selection in favor of those with more passive event recording capability.[14]
- **Exclusions:** 2 types of monitors cannot be ordered simultaneously. A long-term event monitoring device can be preceded by a short-term monitoring device (holter) for 24-72 hours. If the holter does not catch the arrhythmias, an event monitor could be placed.

Medical Necessity Criteria

Indications

➔ **External wearable devices** are considered appropriate if **ALL** of the following are **TRUE**[14]:

- ◆ The patient has known or suspected congenital heart disease and **ANY** of the following:
  - Palpitations.

- Syncope.
- d-TGA with atrial switch or Fontan operation if treated with beta-blockers or other rate-slowing agents.[1]
- Atrial switch or Fontan operation with progressive bradycardia or lack of AV synchrony. [5]
- Repaired Tetralogy of Fallot with QRS duration of greater than or equal to 180 msec, or spontaneous ventricular ectopy
- Significant bradycardia or conduction abnormalities a 12-lead ECG

◆ Arrhythmia should reasonably be expected to occur every 21 days
◆ The patient does not have superseding symptoms of a more urgent cardiac condition that ambulatory cardiac monitoring would delay.
◆ The patient does not have an implantable cardiac device that would acquire similar clinical information regarding the patient's symptoms.
◆ In considering mobile cardiac telemetry, the patient has worn an event monitor for at least 21 days without any diagnostic findings.
◆ If the patient has had 3 or more external wearable devices in the last six months, consider an internal loop recorder.

Non-Indications

➔ **External wearable devices** are not considered appropriate if **ANY** of the following is **TRUE**:
◆ Palpitations are associated with symptoms suggestive of angina or clinically significant coronary artery obstruction, and monitoring would delay other needed testing or intervention.
◆ The patient has an implantable cardiac device capable of acquiring clinical data of a similar or equivalent quality to an external cardiac monitor.
◆ For mobile cardiac telemetry, the patient has not yet worn an event monitor or has not completed at least 21 days of monitoring.

Site of Service Criteria

Outpatient, in-office.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
93224	External electrocardiographic recording up to 48 hours by

(Holter monitoring)	continuous rhythm recording and storage; includes recording, scanning analysis with report, review and interpretation by a physician or other qualified healthcare professional
93225	External electrocardiographic recording up to 48 hours by continuous rhythm recording and storage; recording (includes connection, recording, and disconnection)
93226	External electrocardiographic recording up to 48 hours by continuous rhythm recording and storage; scanning analysis with report
93227	External electrocardiographic recording up to 48 hours by continuous rhythm recording and storage; review and interpretation by a physician or other qualified healthcare professional
0295T (Patch monitors)	External electrocardiographic recording for more than 48 hours up to 21 days by continuous rhythm recording and storage; includes recording, scanning analysis with report, review and interpretation
0296T	External electrocardiographic recording for more than 48 hours up to 21 days by continuous rhythm recording and storage; recording (includes connection and initial recording)
0297T	External electrocardiographic recording for more than 48 hours up to 21 days by continuous rhythm recording and storage; scanning analysis with report
0298T	External electrocardiographic recording for more than 48 hours up to 21 days by continuous rhythm recording and storage; review and interpretation
93268 (Event monitoring)	External patient and, when performed, auto activated electrocardiographic rhythm derived event recording with symptom-related memory loop with remote download capability up to 30 days, 24-hour attended monitoring; includes transmission, review and interpretation by a physician or other qualified healthcare professional
93270	External patient and, when performed, auto activated electrocardiographic rhythm derived event recording with symptom-related memory loop with remote download

	capability up to 30 days, 24-hour attended monitoring; recording (includes connection, recording, and disconnection)
93271	External patient and, when performed, auto activated electrocardiographic rhythm derived event recording with symptom-related memory loop with remote download capability up to 30 days, 24-hour attended monitoring; transmission and analysis
93272	External patient and, when performed, auto activated electrocardiographic rhythm derived event recording with symptom-related memory loop with remote download capability up to 30 days, 24-hour attended monitoring; review and interpretation by a physician or other qualified healthcare professional
93228 (Mobile cardiac telemetry)	External mobile cardiovascular telemetry with electrocardiographic recording, concurrent computerized real time data analysis and greater than 24 hours of accessible ECG data storage (retrievable with query) with ECG triggered and patient selected events transmitted to a remote attended surveillance center for up to 30 days; review and interpretation with report by a physician or other qualified healthcare professional
93229	External mobile cardiovascular telemetry with electrocardiographic recording, concurrent computerized real time data analysis and greater than 24 hours of accessible ECG data storage (retrievable with query) with ECG triggered and patient selected events transmitted to a remote attended surveillance center for up to 30 days; technical support for connection and patient instructions for use, attended surveillance, analysis and transmission of daily and emergent data reports as prescribed by a physician or other qualified healthcare professional

Non-Invasive Testing

Service: Computed Tomography , Cardiac

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** None.
- **Recommended Clinical Approach:** Cardiac CTA is a non-invasive imaging modality that can evaluate the anatomy and pathology of the pericardium, cardiac chambers, central great vessels, and heart function, including the cardiac valves.[15] It is beneficial in defining vascular structures and allowing for precise measurements, which may influence decisions for intervention. Image quality may be affected by high levels of calcification in the heart, which may be a consideration in patients with existing conduits or bioprosthetic valve replacements.
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **Cardiac CTA** is considered appropriate if **ANY** of the following is **TRUE**:

- Known or suspected congenital heart disease[1,3,16-17] and **ALL** of the following:
  - Echocardiography that was inconclusive or discordant with clinical findings.
  - As pre-procedural planning prior to a repair or valve replacement.
- In cases where the diameter of the aortic sinuses or ascending aorta is greater than or equal to 4.0 cm by echocardiography.[18]
- A known or suspected partial anomalous pulmonary venous connection.[1]
- Patients with no cardiac CT or MRI in the last 1-3 years if serial studies are required (intervals between scans depend on the risk of the condition and the expected rate of change).[15]
- Prosthetic valve replacement or prior valve repair and suspected valve dysfunction.[18]
- Known or suspected paravalvular infections when echocardiography is inconclusive or discordant with clinical findings [18]
- Suspected low-flow, low-gradient severe AS with normal or reduced LVEF.[18]
- Known or suspected pulmonary outflow tract obstruction.

- Known or suspected cardiac mass, tumor, thrombus, or potential cardiac source of emboli.

Non-Indications

➔ **Cardiac CTA** may not be considered appropriate if **ANY** of the following is **TRUE**[19]:

- The patient has non-rate controlled atrial fibrillation.
- The patient has contrast dye hypersensitivity.
- In pregnant patients.
- The patient has impaired renal function because angiographic contrast is utilized for the study.
- The patient uses metformin.[20]

Site of Service Criteria

Outpatient or inpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
75572	Computed tomography (CT) of heart with contrast material for evaluation of cardiac structure and morphology, including 3-dimensional (3D) image postprocessing, assessment of cardiac function, and evaluation of venous structures
75573	Computed tomography (CT) of heart with contrast material for evaluation of cardiac structure and morphology in congenital heart disease

Service: Computed Tomography Angiogram (CTA), Chest

General Guidelines

- **Units, Frequency, & Duration:** Single request based on medical necessity criteria.
- **Criteria for Subsequent Requests:** New indication or follow-up after an intervention.
- **Recommended Clinical Approach:** Chest CT provides anatomical information but little functional data and exposes the patient to radiation. It is best reserved for those for whom CMR is not an option.
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **Chest CTA** is considered appropriate if **ANY** of the following is **TRUE**:

- The patient has a known or suspected aortic coarctation, including patients with a prior surgical or catheter intervention.[1]
- There is known or suspected distal pulmonary artery obstruction.
- The diameter of the aortic sinuses or ascending aorta is greater than or equal to 4.0 cm, as measured by echocardiography.[18]
- Echocardiography has not adequately imaged the cardiac chambers valves, great vessels, and pericardium.
- The patient has another distal vascular anomaly in the chest (e.g., Williams Syndrome, Tetralogy of Fallot).

Non-Indications

➔ **Chest CTA** may not be considered appropriate if **ANY** of the following is **TRUE**:

- Non-rate-controlled (i.e., rapid) atrial fibrillation or other tachyarrhythmias.
- Renal failure if angiographic contrast is needed.
- Contrast dye allergy.
- In a patient unable to cooperate with breath-holding.
- The patient takes metformin.
- In pregnant patients.[20]

Site of Service Criteria

Outpatient service.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition

71275	Computed tomographic angiography (CTA) of chest with contrast material and image postprocessing

Service: Coronary Computed Tomography Angiography (CCTA)

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** Clinical reason and judgment according to college and clinical practice guidelines and usage of the ACR Appropriateness Criteria.
- **Recommended Clinical Approach:** Coronary CTA is a non-invasive test that helps clarify whether a patient has coronary artery disease (CAD).
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **CCTA** is considered appropriate if **ANY** of the following is **TRUE**:

- The patient has symptoms of coronary artery disease (CAD) and known or suspected Congenital Heart Disease (CHD).
- Suspected abnormal anatomy of the coronary arteries.
- The patient requires an evaluation of coronary artery anatomy and patency after d-TGA is repaired with an arterial switch procedure.[1]
- CCTA is needed for a preoperative assessment of the coronary arteries before valve surgery if the patient has a low or intermediate risk of CAD.[18]
- CCTA is needed for a preoperative assessment for transcatheter aortic valve implantation (TAVR).
- CCTA is needed for evaluating ostial narrowing in a patient who has Williams syndrome or is suspected of having supravalvular aortic stenosis and is symptomatic for coronary ischemia.[1]

Non-Indications

➔ **CCTA** may not be considered appropriate if **ANY** of the following is **TRUE**:

- Non-rate controlled atrial fibrillation.
- Known or suspected allergy to contrast media.
- Known or suspected renal insufficiency.
- The patient is pregnant.
- The patient takes metformin.[20]

Site of Service Criteria

None.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
75574	Computed tomographic angiography (CTA) of coronary arteries and bypass grafts, with contrast material and 3-dimensional (3D) image postprocessing

Service: Fractional Flow Reserve (CT-FFR)

General Guidelines

- **Units, Frequency, & Duration:** Single instance, must be ordered in conjunction with Coronary CTA imaging.
- **Criteria for Subsequent Requests:** For pre-intervention surveillance of coronary artery lesions or new clinical indications.
- **Recommended Clinical Approach:** Following a positive CCTA, non-invasive fractional flow reserve (FFR) may be medically necessary to guide decisions about invasive coronary angiography in patients with intermediate or high-risk coronary lesions on imaging.[21,16] CT-FFR is not recommended in patients with complex congenital heart disease.
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **FFR** is considered appropriate if **ANY** of the following is **TRUE**[22]:
- ◆ For functional evaluation of coronary CTA lesions which are 40-90% stenosed in a proximal to a middle coronary segment on CCTA.[23,24]
- ◆ For evaluating multivessel disease and identifying culprit lesions causing symptoms.
- ◆ For evaluating the physiologic severity of multiple lesions in a single vessel

***FFR can only be requested with a Coronary CTA, or after a recently performed Coronary CTA

Non-Indications

➔ **FFR** is not appropriate if **ANY** of the following conditions is **TRUE**[25]:
- ◆ The original CCTA was of suboptimal quality.
- ◆ The patient is not a candidate for revascularization.
- ◆ The patient is post coronary artery bypass surgery.
- ◆ The patient has a metal intracoronary stent in the vessel to be studied.[23]
- ◆ Coronary obstruction that is low risk (less than 30% stenosis).
- ◆ The patient has complex congenital heart disease.

Site of Service Criteria

Outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
0501T	non-invasive 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.
0502T	non-invasive 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
0503T	non-invasive 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
0504T	Non-invasive 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
0523T	Intraprocedural coronary fractional flow reserve (FFR) with 3D functional mapping of color-coded FFR values for the coronary tree, derived from coronary angiogram data, for real-time review and interpretation of possible atherosclerotic stenosis(es) intervention (List separately in addition to code for primary procedure)

Service: Cardiac Positron Emission Tomography (PET)

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** None.
- **Recommended Clinical Approach:** Positron emission tomography (PET) is a minimally invasive diagnostic imaging procedure used to evaluate metabolism in normal tissues and diseased tissues in conditions such as cancer, ischemic heart disease, and some neurologic disorders. The benefits of PET scans include greater accuracy for patients who cannot adequately exercise and less radiation exposure than SPECT. It is particularly beneficial in obese patients and others prone to SPECT attenuation artifact, in younger patients (men less than 40, women less than 50), and following equivocal or nondiagnostic testing.
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **Cardiac PET** is considered appropriate if **ALL** of the following are **TRUE**:
- ◆ The patient has symptoms of coronary artery disease (CAD) and intermediate or high pretest probability of CAD.[6-8] If a patient has ongoing or unstable symptoms, stress testing should not be performed before symptoms relieve.
- ◆ One or more of the following:
  - Likely to experience attenuation artifact with SPECT imaging due to factors such as morbid obesity, large breasts, breast implants, previous mastectomy, chest wall deformity, pleural/pericardial effusion.[26]
  - Previous inadequate SPECT/MPI imaging due to inadequate findings, technical difficulties with interpretation, or discordant results with previous clinical data.

Non-Indications

➔ **Cardiac PET** may not be considered appropriate if **ANY** of the following is **TRUE**:
- ◆ The patient is pregnant.
- ◆ The patient has allergic reactions or intolerance to radiotracers.[26]
- ◆ Normal coronary angiogram or CCTA with no stenosis or plaque within the last two years
- ◆ Normal stress test (given adequate stress) within the last year

Site of Service Criteria

Outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
78429	Single positron emission tomography (PET) myocardial imaging study for metabolic evaluation with concurrently acquired computed tomography (CT) transmission scan
78430	Single positron emission tomography (PET) myocardial perfusion imaging study with evaluation of ejection fraction, at rest, with concurrently acquired computed tomography (CT) transmission scan
78431	Multiple positron emission tomography (PET) myocardial perfusion imaging studies with evaluation of ejection fraction, at rest, with concurrently acquired computed tomography (CT) transmission scan
78432	Positron emission tomography (PET) combined myocardial perfusion imaging study and metabolic evaluation study using dual radiotracer
78433	Positron emission tomography (PET) combined myocardial perfusion imaging and metabolic evaluation study using dual radiotracer, with concurrently acquired computed tomography (CT) transmission scan
78459	Single positron emission tomography (PET) myocardial imaging study for metabolic evaluation
78491	Single positron emission tomography (PET) myocardial perfusion imaging study with evaluation of ejection fraction, at rest
78492	Multiple positron emission tomography (PET) myocardial perfusion imaging studies with evaluation of ejection fraction, at rest and with exercise stress
78811	Positron emission tomography (PET) imaging of chest
G0235	Pet not otherwise specified
G0252	Pet imaging initial dx

Service: Magnetic Resonance Angiogram (MRA), Chest

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** Considerations of additional phase, dynamic sequences, positioning of the patient, and marker use are at the discretion of the protocoling radiologist.
- **Recommended Clinical Approach:** Chest magnetic resonance angiogram (MRA) helps assess the aorta, its branch vessels, and the pulmonary vasculature.[27]
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **MRA** is considered appropriate if **ANY** of the following is **TRUE**:

- Known or suspected congenital heart disease when echo is inconclusive or discordant with clinical findings.[28]
- Before repair or revision of ACHD for surgical planning.
- Known or suspected complications following repair of vascular structures.
- To assess shunt and conduits
- Clinically suspected abnormality of the thoracic aorta.[28]
- Known or suspected Pulmonary hypertension.[1]
- Vascular-related thoracic outlet syndrome.[17,28]
- Known or suspected pulmonary arteriovenous malformation or pulmonary vascular abnormality.[1,28]
- Known or suspected anomalous coronary artery.[1]

Non-Indications

➔ **MRA** is not considered appropriate if **ANY** of the following is **TRUE**:

- The patient has non-compatible implanted devices.[27]
- The patient has metallic intraocular foreign bodies.
- The patient is severely claustrophobic.
- There is a potential for adverse reactions to contrast media.[20]
- If the patient has renal insufficiency (eGFR less than 30 mL/min per 1.73 m^2) and if gadolinium contrast is requested, an MRI/MRA may not be considered appropriate.[29]

Site of Service Criteria

Outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
71555	Magnetic resonance angiography (MRA) of chest with contrast material
C8909	Mra w/cont, chest
C8911	Mra w/o fol w/cont, chest
C8910	Mra w/o cont, chest

Service: Magnetic Resonance Imaging (MRI), Cardiac

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** Considerations of additional phase, dynamic sequences, positioning of the patient, and marker use are at the discretion of the protocoling radiologist.
- **Recommended Clinical Approach:** Cardiac MRI is effective for the complete depiction of the pathologic anatomy of CHD, can accurately assess cardiac chamber volume and function, and has the benefit of avoiding radiation.[30]
- **Exclusions:** Exclusions include contraindications of MRI (e.g., retained metal, incompatible width to bore size, claustrophobia), incompatibility with following directions (i.e., breath-hold), and renal insufficiency (eGFR less than 30 mL/min per 1.73 m^2) if gadolinium is requested.

Medical Necessity Criteria

Indications

➔ **Cardiac MRI** is considered appropriate if **ANY** of the following is **TRUE:**

- ◆ MRI is needed before intervention for ACHD for surgical planning.
- ◆ Known or suspected pulmonary hypertension.
- ◆ The patient has ACHD and is at-risk of developing right ventricular enlargement and dysfunction (e.g., Tetralogy of Fallot with severe pulmonary insufficiency).[1]
- ◆ Patients with an atrial septal defect (ASD) when prior echocardiography (i.e., TTE or TEE) was inconclusive or discordant with clinical findings.[29]
- ◆ Known or suspected coarctation of the aorta.[29]
- ◆ Patients with no cardiac CT or MRI in the last 1-3 years if serial studies are required (intervals between scans depend on the risk of the condition and the expected rate of change).[1,31]

Non-Indications

➔ **Cardiac MRI** is not considered appropriate if **ANY** of the following is **TRUE**:

- ◆ Non-compatible implanted devices.
- ◆ Metallic intraocular foreign bodies.
- ◆ Severely claustrophobic.
- ◆ There is a potential for adverse reactions to contrast media.[20]

- A cardiac CTA was requested for the same indications within 3 months of an MRI/MRA request.
- If the patient has renal insufficiency (eGFR less than 30 mL/min per 1.73 m^2) and if gadolinium contrast is requested, an MRI/MRA may not be considered appropriate.

Site of Service Criteria

Outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
75557	Cardiac magnetic resonance imaging (MRI) without contrast material, for evaluation of morphology and function
75559	Cardiac magnetic resonance imaging (MRI) with stress imaging, without contrast material, for evaluation of morphology and function
75561	Cardiac magnetic resonance imaging (MRI) without contrast material, followed by contrast material and further sequences, for evaluation of morphology and function
75563	Cardiac magnetic resonance imaging (MRI) with stress imaging, without contrast material, followed by contrast material and further sequences, for evaluation of morphology and function
C9762	Cardiac magnetic resonance imaging for morphology and function, quantification of segmental dysfunction; with strain imaging
C9763	Cardiac magnetic resonance imaging for morphology and function, quantification of segmental dysfunction; with stress imaging
S8042	MRI Low Field

Service: Myocardial Perfusion Imaging Single Photon Emission Computed Tomography (MPI-SPECT)

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** None.
- **Recommended Clinical Approach:** MPI-SPECT is typically appropriate for patients with adult congenital heart disease and an intermediate or high pretest probability of coronary artery disease (CAD).[6-8]
    - If the patient is unable to exercise or has ECG abnormalities that interfere with the interpretation of an ECG during exercise, MPI-SPECT or stress echo should be considered.
    - Limitations of MPI-SPECT include cost and radiation. In addition, an interpretation of MPI-SPECT can be affected by attenuation artifacts related to soft tissue overlying the heart or extracardiac radioisotope (e.g., liver or gastrointestinal uptake that may be adjacent to the heart).
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **MPI-SPECT** is considered appropriate if **ANY** of the following is **TRUE:**

- ◆ The patient has symptoms that suggest coronary artery disease (CAD) and intermediate or high pretest probability of CAD.[6-8]
- ◆ The patient has chest pain or shortness of breath (SOB) and **ANY** of the following ECG abnormalities that prevent the ECG diagnosis of ischemia:
    - Inability to achieve the target heart rate with a standard exercise treadmill test (greater than or equal to 85% of age-predicted maximal heart rate).
    - Ventricular preexcitation (Wolff-Parkinson-White pattern).
    - Ventricular paced rhythm.
    - Left bundle branch block (LBBB).
    - Greater than 1 mm ST depression at rest.
    - Digoxin use.
    - Left ventricular hypertrophy with ST-T abnormalities.
- ◆ The patient has chest pain or shortness of breath and stress test or stress echocardiography was performed with inconclusive results, technical difficulties with interpretation, or results were discordant with previous clinical data.
- ◆ The patient has chest pain or SOB and **ANY** of the following:
    - Severe breathing difficulty that prevents test completion.

- Technical limitations making echocardiography inappropriate (e.g., the patient cannot be in an appropriate position).
- Segmental wall motion abnormalities at rest due to cardiomyopathy, recent MI, or pulmonary hypertension.
- Inability to exercise requiring a pharmacological stress test.

Non-Indications

➔ **MPI-SPECT** may not be considered appropriate if **ANY** of the following is **TRUE**:

- The patient is pregnant.
- Normal coronary angiogram or CCTA with no stenosis or plaque within the last two years.
- Normal stress test (given adequate stress) within the last year.
- An active cardiac condition that has not been stabilized (e.g., uncontrolled hypertension, uncontrolled arrhythmias, undiagnosed chest pain).
- An active cardiac condition that has not been stabilized (e.g., difficulty breathing, the possibility of pulmonary embolism).
- Vasodilators (i.e., adenosine, regadenoson, and dipyridamole) are contraindicated in patients with hypotension, sinus node dysfunction, high-degree atrioventricular (AV) block (in the absence of back up pacemaker capability), and reactive airway disease.

Site of Service Criteria

Outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
78451	Single-photon emission computed tomography (SPECT) myocardial perfusion imaging study with stress
78452	Multiple single-photon emission computed tomography (SPECT) myocardial perfusion imaging studies with stress
78494	Cardiac blood pool single photon emission computed tomography (SPECT) imaging, gated equilibrium study, at rest, with wall motion study plus ejection fraction

Service: Stress Echocardiogram

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** None.
- **Recommended Clinical Approach:** Stress echocardiography is an option to help clarify the diagnosis of coronary artery disease (CAD) or detect complications of ACHD. It can be accomplished using either exercise or pharmacologic agents (i.e., dobutamine) as the stress mechanism.[32] This test results in no radiation exposure and is typically lower cost than myocardial perfusion imaging (MPI-SPECT). Other advantages of stress echo compared to MPI-SPECT include shorter patient time commitment, and additional information on cardiac structures (valves, ascending aorta, pericardial space). The test is less technically demanding than MPI-SPECT.[33]
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **Stress echo** is considered appropriate if **ANY** of the following is **TRUE**:

- The patient has left ventricular outflow tract (LVOT) obstruction and otherwise equivocal indications for intervention. [1]
- The patient has coarctation of the aorta, and a stress echo is needed to assess for exercise-induced hypertension and left ventricular (LV) function.[1]
- The patient has anatomically severe aortic stenosis (AS, less than 1.0 cm^2) and left ventricular ejection fraction (LVEF) less than 50% with a low transvalvular velocity and pressure gradient at rest (i.e., velocity less than 4 m/s or mean gradient less than 40 mmHg). Stress echo is needed to distinguish between severe AS with LV systolic dysfunction and moderate AS.[18]
- The patient has asymptomatic moderate or severe chronic primary mitral regurgitation.[18]
- The patient has asymptomatic but severe valve disease and is considering pregnancy.[18]
- The patient has moderate or asymptomatic severe AS (stages B and C) for measurement of changes in valve hemodynamics with exercise or pharmacological stress.[34]
- The patient has mitral valve disease, and there is a discrepancy between clinical symptoms and resting echo findings.[34]
- The patient has moderate or severe aortic regurgitation for assessment of symptoms and functional capacity.[34]

- Patients who have anginal symptoms of chest pain or exertional dyspnea (ischemic equivalent) and intermediate or high pretest probability of CAD [6-8] and **ANY** of the following that would interfere with the specificity and diagnosis of ECG interpretation alone:
    - An inability to achieve the target heart rate with a standard exercise treadmill test (greater than or equal to 85% of age-predicted maximal HR).
    - An ECG with ventricular preexcitation (Wolff-Parkinson-White pattern).
    - Ventricular-paced rhythm.
    - Left bundle branch block (LBBB).
    - An ECG with greater than 1 mm ST depression at rest.
    - Left ventricular hypertrophy with ST-T abnormalities.
    - The patient takes digoxin.
    - Unable to exercise requiring pharmacological stress test.

Non-Indications

- **Stress echo** may not be considered appropriate if **ANY** of the following is **TRUE**[35]:
    - Normal coronary angiogram or CCTA with no stenosis or plaque within the last two years (if the assessment is for the presence of coronary artery disease).
    - Normal stress test (given adequate stress) within the last year (if the assessment is for cardiac ischemia).
    - The patient has a high-priority clinical illness to address before stress testing, such as **ANY** of the following[32]:
        - Acute coronary syndrome (STEMI, NSTEMI, unstable angina).
        - Acute pericarditis/Myocarditis.
        - Severe aortic stenosis.
        - Uncontrolled arrhythmias.
        - Symptomatic congestive heart failure.
        - Left main coronary artery stenosis.
        - Severe hypertension (greater than 180/100mm Hg).
        - Inability to exercise sufficiently or tolerate pharmacologic agents to simulate exercise.

Site of Service Criteria

Outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
93350	Real time transthoracic echocardiography with

	2-dimensional (2D) image documentation during rest and cardiovascular stress test using treadmill and pharmacologically induced stress, with interpretation and report
93351	Real time transthoracic echocardiography with 2-dimensional (2D) image documentation during rest and cardiovascular stress test using treadmill, bicycle exercise and pharmacologically induced stress, with interpretation and report, including performance of continuous electrocardiographic monitoring, with physician supervision
C8928	Tte w or w/o fol w/con,stres
C8930	Tte w or w/o contr, cont ecg

Service: Transesophageal Echocardiogram (TEE)

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** None.
- **Recommended Clinical Approach:** Transthoracic echocardiography (TEE) can be useful for patients with congenital heart disease when transthoracic echocardiography results are discordant with history and physical exam. It is also more accurate imaging than TTE for visualization of tissue rims around an atrial septal defect, which is key for safe transcatheter device closure. The posterior structures of the heart are also better seen, which can be important when planning mitral valve surgery or detecting cardiac masses or vegetations.
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **TEE** is considered appropriate if **ANY** of the following is **TRUE**:

- ◆ TEE is needed for intraprocedural guidance during surgery.[1]
- ◆ TEE is needed for surgical planning for **ANY** of the following:
  - Ebstein's anomaly if TTE images are inadequate to evaluate tricuspid valve morphology and function. [1]
  - Mitral valve intervention.[18,28,34]
  - Transcatheter aortic valve replacement (TAVR).[34]
- ◆ TTE provided insufficient or discordant information.[28]
- ◆ There is a concern for a baffle leak in a patient with d-TGA with an atrial switch, and the TTE was inadequate to confirm.[1]
- ◆ The patient has an atrial septal defect (ASD) to evaluate pulmonary venous connections.[1]
- ◆ Williams syndrome or patients suspected of having supravalvular aortic stenosis.[1]
- ◆ In patients with a prior valve replacement or valve repair and clinical symptoms or signs that suggest prosthetic valve dysfunction, even if TTE does not show valve dysfunction.[18]
- ◆ In patients with known or suspected infectious endocarditis (IE) and **ANY** of the following:
  - Nondiagnostic TTE results.
  - Intracardiac device leads are present.[18]
  - The patient is being considered for an early change to oral antibiotic therapy for the treatment of stable IE; perform a baseline TEE before switching to oral therapy and a repeat

TEE 1 to 3 days before completing the oral antibiotic regimen.[18]
- The patient has a prosthetic valve and persistent fever but does not have bacteremia or a new murmur.[18]

◆ There is suspicion of a cardiac mass, tumor, thrombus, or cardiac source of embolus.[34]
◆ The patient is pregnant and has a mechanical prosthetic valve and either 1) prosthetic valve obstruction; or 2) experienced an embolic event.[18]

Non-Indications

➔ **TEE** may not be considered appropriate if **ANY** of the following is **TRUE**:
◆ Another imaging modality (e.g., CT, MRI) is requested simultaneously to evaluate for intracardiac thrombus.
◆ The patient has a history of esophageal stricture, malignancy, recent surgery of the esophagus, active GI bleeding, esophageal varices (relative), or prior surgery (relative).
◆ The patient has a history of undiagnosed dysphagia.[36]

Site of Service Criteria

Outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
93312	Real time transesophageal echocardiography with 2-dimensional (2D) image documentation, M-mode recording, probe placement, image acquisition, interpretation, and report
93313	Real time transesophageal echocardiography with 2-dimensional (2D) image documentation and placement of transesophageal probe only
93314	Interpretation and report only of real time transesophageal echocardiography with 2-dimensional (2D) image documentation and image acquisition
93315	Transesophageal echocardiography (TEE) with probe placement, image acquisition, interpretation, and report
93316	Transesophageal echocardiography (TEE) for placement of transesophageal probe only
93317	Interpretation and report only of transesophageal echocardiography (TEE) with image acquisition
93318	Real time transesophageal echocardiography (TEE) with

	probe placement, 2-dimensional (2D) image acquisition and interpretation
93355	Transesophageal echocardiography (TEE) for guidance of transcatheter closure of left atrial appendage, with quantitative measurements, probe manipulation, interpretation and report
C8925	2d tee w or w/o fol w/con,in
C8926	Tee w or w/o fol w/cont,cong
C8927	Tee w or w/o fol w/cont, mon

Service: Transthoracic Echocardiogram (TTE)

General Guidelines

- **Units, Frequency, & Duration:** Perform single procedures as needed per defined criteria.
- **Criteria for Subsequent Requests:**
  - Single repeat TTEs are appropriate for:
    - Evaluating significant changes in signs/symptoms since the patient's last TTE.
    - Providing objective evidence of value for patients undergoing medical treatment to improve left ventricular (LV) function.
  - A repeat TTE is appropriate for patients at various intervals with a history of:
    - Significant valve disfunction or disformity (e.g., severe stenosis, mitral valve prolapse) with the frequency of repeat echocardiograms based on the type and severity of the valve lesion, the known rate of progression of the specific valve lesion, and the effect of the valve lesion on the affected ventricle(s).[1]
    - Pulmonary hypertension.[1]
- **Recommended Clinical Approach:** Transthoracic echocardiography is the standard diagnostic test in the evaluation of patients with known or suspected congenital heart disease, with the timing of serial assessment based on heart defect severity and clinical status.[1]
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **TTE** is considered appropriate if **ANY** of the following is **TRUE**:

- The patient has a suspected congenital heart defect.
- Known ACHD with change in clinical status
- Re-Evaluation to guide therapy in know ACHD
- Routine surviellance (>1 year) of ACHD following incomplete/palliative repair
  - With residual structural or hemodynamic abnormality
  - Without a change in clinical status or cardiac exam
- The patient has a first-degree relative with a bicuspid aortic valve or an enlargement of the thoracic aorta.[28]

- There is suspected cardiac vegetation, tumor, thrombus, or a cardiac source of embolus.[28]
- The patient has known congestive heart failure and a change in clinical status or cardiac examination.
- A TTE is needed before a planned intervention.
- A TTE is needed for the initial follow-up after a valve intervention.[18]
- For routine surveillance of ANY congenital heart disease, repaired or unrepaired, that could potentially deteriorate since the last imaging study (e.g., worsening native or prosthetic valve insufficiency or stenosis, heart function, associated hemodynamic effects).[18]
    - Mild severity or minimally progressive congenital heart disease: a TTE is appropriate every 3 years.
    - Moderate severity or moderately progressive congenital heart disease: a TTE is appropriate every 2 years.
    - Severe or rapidly progressive congenital heart disease: a TTE is appropriate every year (i.e., annually.)
- A TTE is needed for following up on any congenital heart lesion with new symptoms, evaluating the effect of medical therapy, and after a patient was hospitalized for cardiac reasons.
- Williams syndrome or patients suspected of having supravalvular aortic stenosis.[1]
- Assessing for baffle leak in patients with d-TGA with atrial switch.[1]

Non-Indications

➔ **TTE** is not considered appropriate if **ANY** of the following is **TRUE**:

- Echocardiography has no contraindications. Transthoracic echocardiography is of limited benefit in assessing anomalous pulmonary venous connections in adults with atrial septal defect (ASD) and the superior and posterior atrial septum.[15,34]

Site of Service Criteria

Inpatient, outpatient, or observation status.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
93303	Complete transthoracic echocardiography for congenital cardiac anomalies
93304	Follow-up transthoracic echocardiography for congenital cardiac anomalies
93306	Real time transthoracic echocardiography with 2-dimensional (2D) image documentation, M-mode

	recording with spectral Doppler echocardiography, and color flow Doppler echocardiography
93307	Complete real time transthoracic echocardiography with 2-dimensional (2D) image documentation
93308	Follow-up real time transthoracic echocardiography with 2-dimensional (2D) image documentation
C8921	Tte w or w/o fol w/cont, com
C8922	Tte w or w/o fol w/cont, f/u
C8923	2d tte w or w/o fol w/con,co
C8924	2d tte w or w/o fol w/con,fu
C8929	Tte w or wo fol wcon,doppler

Non-Surgical Management

Service: Cardiac Rehabilitation

General Guidelines

- **Units, Frequency, & Duration:** Cardiac rehabilitation is generally appropriate for 36 sessions, 60 minutes each, typically over 12 - 18 weeks. Additional sessions can be requested.[37]
- **Criteria for Subsequent Requests:** Current guidelines do not support the need for repeat cardiac rehabilitation in the absence of a new cardiac event.
- **Recommended Clinical Approach:** Cardiac rehabilitation (CR) is an evidence-based intervention that uses patient education, health behavior modification, and exercise training to improve secondary prevention outcomes and is recognized as an integral component of care for patients with cardiovascular disease.[37,41] Referral to CR is recommended within 12 months after a myocardial infarction (MI), percutaneous coronary intervention, or coronary artery bypass graft surgery or in the setting of stable angina or symptomatic peripheral arterial disease (i.e., intermittent claudication).[37] Referral to CR is also recommended after heart valve surgery, cardiac transplantation, or in the setting of chronic heart failure (NYHA Class I-III) with reduced ejection fraction (HFrEF).[37] The effects of cardiac rehabilitation on mortality, cardiovascular events, hospitalizations, or health-related quality of life are less certain in patients with atrial fibrillation, adult congenital heart disease, and after permanent pacemaker/ICD implantation. However, these are described as useful by various national and international specialty societies.[38-40] Medicare coverage may not be available for these diagnoses.
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **Cardiac rehabilitation** is considered appropriate if **ANY** of the following are **TRUE** (within a one year period)[39,40,42]:

- Acute myocardial infarction
- Acute coronary artery syndrome
- Chronic stable angina
- Chronic congestive heart failure (NYHA Class I-III, including with LV assist devices)
- After coronary artery bypass surgery

- After a percutaneous coronary intervention
- After valvular surgery
- Cardiac transplantation
- Symptomatic peripheral arterial disease
- Atrial fibrillation
- Adult congenital heart disease
- After permanent pacemaker/ICD implantation

Non-Indications

➔ **Cardiac rehabilitation** may not be considered appropriate if **ANY** of the following are present[41]:

- Active unstable angina
- Decompensated cardiac failure
- Active dangerous or complex arrhythmias
- Dissecting aneurysm
- Myocarditis
- Acute pericarditis
- Severe obstruction of the left ventricular outflow tract
- Severe hypertension
- Exertional hypotension or syncope
- Severe orthopedic limitations
- Recent systemic or pulmonary embolus)
- Severe or symptomatic aortic stenosis
- Previous cardiac rehabilitation in the absence of a new cardiac event.

Site of Service Criteria

Outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
S9472	Cardiac rehabilitation program, nonphysician provider, per diem
93798	Physician or other qualified healthcare professional services for outpatient cardiac rehabilitation; with continuous ECG monitoring (per session)

Surgical or Interventional Management

Service: Electrophysiology Study (EPS)

General Guidelines

- **Units, Frequency, & Duration:** One instance, as indicated by clinical guidelines.
- **Criteria for Subsequent Requests:** None.
- **Recommended Clinical Approach:** Electrophysiology studies have a role in many adults with congenital heart disease. These include a preoperative screening of patients with Ebstein's anomaly to determine the presence of accessory pathways and those with Tetralogy of Fallot for inducible ventricular arrhythmias. Some of these procedures can lead to ablation of the arrhythmogenic substrate (or cause of the arrhythmia). Others can lead to a decision to implant a defibrillator, especially in patients with hemodynamic risk factors that could increase the risk of sudden cardiac death.[1]
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **EPS** is considered appropriate if **ANY** of the following is **TRUE**:
- ◆ The patient has congenital heart disease and **ALL** of the following are TRUE:
  - All evaluations were inconclusive, including comprehensive ECG monitoring and stress testing
  - The patient has **ANY** of the following symptoms of significant rhythm abnormalities:
    - Palpitations
    - Syncope
    - Chest pain
    - Shortness of breath
- ◆ EPS is needed before surgical intervention on the tricuspid valve in a patient with Ebstein anomaly.[1,42]
- ◆ The patient has Tetralogy of Fallot and either syncope or nonsustained ventricular tachycardia on non-invasive monitoring.
- ◆ The patient has complex congenital heart disease, unexplained syncope, AND a high-risk substrate for ventricular arrhythmias (e.g., ventriculotomy).[4]

Non-Indications

- ➔ **EPS** is not considered appropriate if **ANY** of the following is **TRUE**:
  - ◆ Other outpatient testing revealed symptomatic sinus bradycardia/arrest that correlate with patient symptoms.

Site of Service Criteria

Outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
93600	Bundle of His recording
93602	Intra-atrial recording
93603	Right ventricular recording
93610	Intra-atrial pacing
93612	Intraventricular pacing
93618	Induction of arrhythmia by electrical pacing
93619	Comprehensive electrophysiologic evaluation with insertion and repositioning of multiple electrode catheters, with right atrial pacing and recording, right ventricular pacing and recording, and His bundle recording
93620	Comprehensive electrophysiologic evaluation with insertion and repositioning of multiple electrode catheters, with attempted induction of arrhythmia, with right atrial pacing and recording, right ventricular pacing and recording, and His bundle recording
+93623	
93624	Electrophysiologic follow-up study with pacing and recording to test effectiveness of therapy with attempted induction of arrhythmia
93631	Intra-operative epicardial and endocardial pacing and mapping to localize the site of tachycardia or zone of slow conduction for surgical correction

Service: Cardiac Ablation

General Guidelines

- **Units, Frequency, & Duration:** Single event, no applicable frequency
- **Criteria for Subsequent Requests:** Subsequent requests may be appropriate if the initial procedure was unsuccessful or if symptomatic atrial fibrillation recurred for greater than three months after the previous cardiac ablation.
- **Recommended Clinical Approach:** Catheter ablation can have a role in many adults with congenital heart disease. These include the preoperative screening for accessory pathways in patients with Ebstein's anomaly and screening for inducible ventricular arrhythmias in those with Tetralogy of Fallot.[42-43] Intra-atrial reentrant tachycardia (a form of atrial flutter) has a prevalence of 30-50% in Mustard/Senning repairs for d-TGA. The same applies to Fontan palliations for single ventricle physiology. In addition, late repair of atrial septal defects is associated with increased atrial fibrillation, even when hemodynamics have normalized.
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **Cardiac Ablation** is considered appropriate if **ANY** of the following is **TRUE**:
- ◆ The patient has **ALL** of the following
  - **ANY** of the following is **TRUE**:
    - Paroxysmal or persistent atrial fibrillation has become refractory
    - The patient cannot tolerate treatment with a Class I or III antiarrhythmic
  - There is reasonable access to a pulmonary vein substrate.
- ◆ Ablation is needed for a patient with Ebstein's anomaly for **ANY** of the following[1]:
  - Clinical or inducible reentrant tachycardia
  - high-risk pathway conduction
  - Multiple accessory pathways
- ◆ After a Fontan palliation or a Mustard/Senning operation with **ANY** of the following:
  - Intra-atrial reentrant tachycardia
  - Focal atrial tachycardia.[1]

- For preoperative assessment of Tetralogy of Fallot (TOF) for sustained monomorphic ventricular tachycardia.[43]
- This is a subsequent request, and the patient had greater than three months of symptomatic atrial fibrillation after the last cardiac ablation.

Non-Indications

➔ **Cardiac Ablation** is not considered appropriate if **ANY** of the following is **TRUE:**

- As a routine screening for ventricular arrhythmia in congenital heart disease patients who do not have a high-risk substrate.

Site of Service Criteria

Outpatient or observation status.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
93653	Comprehensive electrophysiologic evaluation with insertion and repositioning of multiple electrode catheters, with attempted induction of arrhythmia, with right atrial pacing and recording, with treatment of supraventricular tachycardia by ablation
93654	Comprehensive electrophysiologic evaluation with insertion and repositioning of multiple electrode catheters, induction or attempted induction of an arrhythmia with right atrial pacing and recording and catheter ablation of arrhythmogenic focus, including intracardiac electrophysiologic 3-dimensional mapping, right ventricular pacing and recording, left atrial pacing and recording from coronary sinus or left atrium, and His bundle recording, when performed; with treatment of ventricular tachycardia or focus of ventricular ectopy including left ventricular pacing and recording, when performed
93656	Comprehensive electrophysiologic evaluation with transseptal catheterization, with insertion and repositioning of multiple electrode catheters, with attempted induction of arrhythmia, with atrial pacing and recording

Service: Cardiac Catheterization

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** None.
- **Recommended Clinical Approach:** If non-invasive testing is inconclusive, a hemodynamic cardiac catheterization with direct intracardiac measurements of transvalvular pressure gradients and cardiac output measurements may provide important clinical information.[18,44] When possible, cardiac catheterization in patients with adult congenital heart disease (ACHD) should be performed by, or in collaboration with, cardiologists with expertise in ACHD.[1]
- **Exclusions:** Perform elective cardiac catheterization at a facility that offers coronary intervention and has the staffing and lab availability for a PCI if indicated. Perform catheterization of patients with complex congenital heart disease at a comprehensive ACHD center.

Medical Necessity Criteria

Indications

➔ **Cardiac catheterization** is considered appropriate if **ANY** of the following is **TRUE**:

- ◆ The patient has Canadian Cardiovascular Society Class II or higher angina and **ANY** of the following are **TRUE**:
  - The patient is on antianginal medications
  - There is documentation on why the patient is not on antianginal medication due to contraindications or adverse effects such as blood pressure is too low
- ◆ The patient has suspected or known congenital heart disease when non-invasive tests are inconclusive or discordant with clinical assessment.
- ◆ The patient has suspected or known pulmonary hypertension.[1]
- ◆ Before valve surgery (e.g., TAVR) if the patient has **ANY** of the following[18]:
  - Symptoms of angina.
  - Objective evidence of ischemia.
  - Decreased ventricular systolic function.
  - Coronary artery disease (CAD).
  - Coronary risk factors (including men greater than 40 years of age and postmenopausal women).
- ◆ Before **ANY** of the following procedures:
  - Assessment before listing the patient for a heart transplant.
  - Assessing any shunt inside the heart or lungs.[1]

        - Initial Fontan surgery or revision of a prior Fontan connection.
    - During transcatheter atrial septal defect (ASD) closure.[1]
    - Partial anomalous pulmonary venous connection to define vascular connections.[1]
    - In patients with repaired Tetralogy of Fallot (TOF) or Right Ventricular-Pulmonary Artery (RV-PA) conduit where non-invasive testing cannot assess the severity of obstruction in the conduit or pulmonary arteries andthe patient has **ANY** of the following:
        - Arrhythmia.
        - Heart Failure.
        - Unexplained ventricular dysfunction.
        - Suspected right ventricular hypertension.
        - Cyanosis.[1]
    - For assessment of atrial baffle function (suspected obstruction or leak) after Mustard/Senning operation.
    - After a Fontan palliation procedure in patients with **ANY** of the following[1]:
        - Creation or closure of a fenestration.
        - Suspected collateral vessels.
        - Baffle obstruction.
        - Protein-losing enteropathy or ascites.
        - New onset or worsening atrial tachyarrhythmias.
        - Symptomatic and non-invasive testing is insufficient to guide therapy.

Non-Indications

- ➔ **Cardiac catheterization** may not be considered appropriate if **ANY** of the following is **TRUE**[44]:
    - The patient is pregnant.
    - Acute or chronic kidney disease.
    - Coagulopathy.
    - Fever or a systemic infection.
    - Uncontrolled arrhythmia.
    - Uncontrolled hypertension.
    - Uncompensated heart failure.
    - Severe contrast agent allergy.[20]

Site of Service Criteria

Inpatient or outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
93451	Right heart catheterization
93452	Left heart catheterization with intraprocedural injection for left ventriculography
93453	Combined right and left heart catheterization with intraprocedural injection for left ventriculography
93454	Catheter placement in coronary artery for coronary angiography, with intraprocedural injection for coronary angiography, imaging supervision, and interpretation
93455	Catheter placement in coronary artery for coronary angiography, with intraprocedural injection for coronary angiography, imaging supervision, and interpretation, with catheter placement in bypass graft, with intraprocedural injections for bypass graft angiography
93456	Catheter placement in coronary artery for coronary angiography, with intraprocedural injection for coronary angiography, imaging supervision, and interpretation, with right heart catheterization
93457	Catheter placement in coronary artery for coronary angiography, with intraprocedural injection for coronary angiography, imaging supervision, and interpretation, with catheter placement in bypass graft, with intraprocedural injection for bypass graft angiography and right heart catheterization
93458	Catheter placement in coronary artery for coronary angiography, with intraprocedural injection for coronary angiography, imaging supervision, and interpretation, with left heart catheterization, with intraprocedural injection for left ventriculography
93459	Catheter placement in coronary artery for coronary angiography, with intraprocedural injection for coronary angiography, imaging supervision and interpretation, with left heart catheterization, catheter placement in bypass

	graft, with bypass graft angiography
93460	Catheter placement in coronary artery for coronary angiography, with intraprocedural injection for coronary angiography, imaging supervision, and interpretation, with right and left heart catheterization
93461	Catheter placement in coronary artery for coronary angiography, with intraprocedural injection for coronary angiography, imaging supervision, and interpretation, with right and left heart catheterization, catheter placement in bypass graft, with bypass graft angiography
93593	Catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone.
93594	Right heart catheterization for congenital heart defect(s) including imaging guidance by the proceduralist to advance the catheter to the target zone; abnormal native connection
93595	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
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); normal native connections
93597	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

Service: Percutaneous Coronary Intervention (PCI)/Angioplasty

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** None.
- **Recommended Clinical Approach:** This procedure is done during a heart catheterization for a symptomatic, significant coronary artery stenosis or blockage refractory to optimal medical therapy.[45]
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **PCI** is considered appropriate if **ANY** of the following is **TRUE**[45]:

- The patient has unstable angina.
- The patient is undergoing TAVR and has significant left main or proximal CAD (with or without angina) documented by CCTA or prior coronary angiography.[18]
- The patient has supravalvular aortic stenosis and coronary ostial stenosis with anginal symptoms.[1]
- The patient is on at least two antianginal medications (or documentation on why the patient is not on antianginal medication due to contraindications or adverse effects such as blood pressure being too low) and has **ANY**of the following:
  - Canadian Cardiovascular Society Class II or higher angina.
  - Anginal equivalent (e.g., dyspnea, arrhythmia, dizziness, or syncope).
  - High-risk stress test findings.

Non-Indications

None.

Site of Service Criteria

Inpatient or outpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
92920	Percutaneous transluminal coronary angioplasty into single major coronary artery
92928	Percutaneous transcatheter insertion of stent into single

	major coronary artery
92937	Percutaneous transluminal revascularization of a single coronary artery bypass graft with angioplasty
92943	Percutaneous transluminal revascularization of chronic total occlusion of a single coronary artery branch with atherectomy, angioplasty, and insertion of stent
C9600	Perc drug-el cor stent sing
C9604	Perc d-e cor revasc t cabg s
C9607	Perc d-e cor revasc chro sin
33990	Insertion of percutaneous arterial ventricular assist device by arterial access only
33991	Insertion of percutaneous arterial ventricular assist device by arterial and venous access, with transseptal puncture, with radiological supervision and interpretation

Service: Surgical Aortic Valve Replacement

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** The same criteria as a first request.
- **Recommended Clinical Approach:** When aortic valve replacement is indicated, a shared decision-making process is recommended to determine the type of valve. This discussion should include the patient's values and preferences, the indications for and risks of anticoagulant therapy, and the potential benefits and risks associated with valve reintervention. This discussion is particularly important for premenopausal women and the risk of anticoagulation during pregnancy.[18] A mechanical valve requires anticoagulation therapy but is more durable than a bioprosthetic valve. A bicuspid aortic valve is also very commonly associated with dilatation and aneurysmal change in the ascending aorta. This change can increase the risk of dissection or rupture. Replacement of this area with a graft may also be performed with or without an aortic valve replacement.
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **Aortic valve replacement** is considered appropriate if **ANY** of the following is **TRUE**:

- ◆ The patient has a bicuspid aortic valve (BAV), and the diameter of the aortic sinuses or the ascending aorta is greater than 5.5 cm.[18]
- ◆ In asymptomatic patients with a BAV, a diameter of the aortic sinuses or ascending aorta of 5.0 to 5.5 cm, and an additional risk factor for dissection (e.g., family history of aortic dissection, aortic growth rate greater than 0.5 cm per year, aortic coarctation)[18]
- ◆ The patient's history or exercise testing revealed severe high-gradient aortic stenosis (AS)and symptoms of **ANY** of the following[18]:
  - Exertional dyspnea
  - Heart failure (HF)
  - Angina
  - Syncope or presyncope.
- ◆ The patient is symptomatic with low-flow, low-gradient severe AS, and **ALL** of the following[18]:
  - Normal left ventricular ejection fraction (LVEF)
  - AS is the most likely cause of symptoms.

- The patient is symptomatic with severe aortic stenosis (AS), regardless of left ventricular (LV) systolic function.[18]
- The patient is asymptomatic with severe AS and **ANY** of the below[18]:
  - The patient's surgical risk is low, and exercise testing demonstrated decreased exercise tolerance (normalized for age and sex) OR a fall in systolic blood pressure of greater than or equal to 10 mm Hg from baseline.
  - LVEF is less than 50%.
  - Very severe AS (aortic velocity greater than or equal to 5 m/s) and low surgical risk.
  - Serum B-type natriuretic peptide (BNP) level is more than 3 times higher than normal, and surgical risk is low.
  - Serial testing shows an increase in aortic velocity greater than or equal to 0.3 m/s per year, and the patient's surgical risk is low.
  - Progressive decrease in LVEF on at least 3 serial imaging studies to less than 60%.
  - Undergoing cardiac surgery for other indications.
  - Low-flow, low-gradient severe AS with reduced LVEF (Stage D2).
  - The patient is considering pregnancy.
- In asymptomatic patients with severe AR and **ANY** of the below[18]:
  - Normal LV systolic function (LVEF greater than 55%), and a severely enlarged left ventricle (lV end-systolic diameter (LVESD) greater than 50 mm or indexed LVESD greater than 25 mm/m^2) (Stage C2).
  - A progressive decline in LVEF on at least 3 serial studies to the low–normal range (LVEF 55% to 60%)
  - A progressive increase in LV dilation into the severe range (LV end-diastolic dimension (LVEDD) greater than 65 mm).
  - Severe prosthetic regurgitation and low operative risk.
  - LV systolic dysfunction (LVEF less than or equal to 55%), aortic valve surgery is indicated if no other cause for systolic dysfunction is identified.
- The patient is undergoing cardiac surgery for another indication and has **ANY** of the below[18]:
  - Moderate AS (Stage B).
  - The patient is asymptomatic with severe AS.
  - The patient has moderate AR (Stage B) or severe AR.
- In patients with intractable hemolysis or HF attributable to prosthetic transvalvular or paravalvular leak, unless the surgical risk is prohibitively high.[18]
- In pregnant women with severe aortic valve disease and **ALL** of the following[18]:

- Hemodynamic deterioration or New York Heart Association (NYHA) class III or IV HF symptoms (for AS) OR NYHA class IV symptoms (for AR).
- Severe, intractable symptoms despite maximal medical therapy.

◆ In patients with significant AS and significant coronary artery disease (CAD) (luminal reduction greater than or equal to 70% diameter, fractional flow reserve (FFR) less than or equal to 0.8, an instantaneous wave-free ratio less than or equal to 0.89) and **ALL** of the following:
- Significant CAD consists of complex bifurcation left main or multivessel CAD with a SYNTAX (Synergy Between Percutaneous Coronary Intervention With Taxus and Cardiac Surgery) score greater than 33.
- SAVR and CABG are reasonable and preferred over TAVR and PCI.[18]

Non-Indications

➔ **Aortic valve replacement** may not be indicated if **ANY** of the following is **TRUE**:
- ◆ The patient is pregnant (unless severe, intractable symptoms develop despite maximal medical therapy).
- ◆ The patient has active bacterial endocarditis or another systemic infection (unless severe, intractable symptoms develop despite maximal medical therapy).
- ◆ The patient is to receive a mechanical valve replacement, but oral anticoagulation medication is not tolerated.

Site of Service Criteria

Inpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
33405	Replacement, aortic valve, with cardiopulmonary bypass; with prosthetic valve other than homograft or stentless valve
33410	Replacement, aortic valve, with cardiopulmonary bypass; with stentless tissue valve
33858	Ascending aorta graft, with cardiopulmonary bypass, includes valve suspension, when performed; for aortic

	dissection
33859	Ascending aorta graft, with cardiopulmonary bypass, includes valve suspension, when performed; for aortic disease other than dissection (e.g., aneurysm)

Service: Transcatheter Mitral Valve Repair

General Guidelines

- **Units, Frequency, & Duration:** Single request.
- **Criteria for Subsequent Requests:** None.
- **Recommended Clinical Approach:** Transcatheter mitral valve repair is an acceptable option for patients with symptomatic moderate or severe mitral valve insufficiency whose surgical risk is high. Devices like the MitraClip are FDA approved to improve mitral insufficiency due to either primary degenerative mitral valve abnormalities or functional mitral valve insufficiency due to diminished left ventricular function.[18,46-47] Transcatheter mitral valve repair has been shown to reduce rehospitalization and all-cause deaths vs. optimal medical therapy in patients with heart failure and moderate-severe mitral insufficiency.
- Percutaneous mitral valve repair includes- Edge to Edge repair (Mitra-clip/Pascal repair), Indirect annuloplasty (Carillon, ARTO, mitral loop cerclage), Chordal repair (NeoChord/Harpoon), Direct annuloplasty (Cardioband, millipede, Mitralign, Accucinch)
- **Exclusions:** The procedure should be performed in a comprehensive cardiac care center.

Medical Necessity Criteria

Indications

➔ **Transcatheter mitral valve repair** is considered appropriate if **ANY** of the following is **TRUE**[18]**:**

- ◆ In severely symptomatic patients (NYHA class III or IV) with primary severe MR and high or prohibitive surgical risk, transcatheter edge-to-edge repair (TEER) is indicated by **ALL** of the following:
  - Mitral valve anatomy is favorable for the repair procedure.
  - Patient life expectancy is at least 1 year.
- ◆ In patients with chronic severe secondary MR related to LV systolic dysfunction (LVEF <50%) and **ALL** of the following:
  - Persistent symptoms (NYHA class II, III, or IV) while on optimal medical therapy.
  - Appropriate anatomy as defined on TEE, and with
  - LVEF between 20% and 50%.
  - LVESD less than or equal to 70 mm.
  - Pulmonary artery systolic pressure less than or equal to 70 mm Hg.

Non-Indications

- ➔ **Transcatheter mitral valve repair** is not considered appropriate if **ANY** of the following is **TRUE**:
  - ◆ Another cardiac surgery is planned where surgical mitral valvuloplasty can be performed effectively.[18]
  - ◆ Mitral valve insufficiency is due to congenital cleft (transitional AV canal).
  - ◆ Mitral valve insufficiency is due to damage from infective endocarditis.
  - ◆ Mitral valve insufficiency is due to rheumatic heart disease.
  - ◆ Any intracardiac or venous (IVC or femoral vein) thrombosis is present.

Site of Service Criteria

Inpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
33418	Transcatheter mitral valve repair with initial prosthetic valve by percutaneous approach
0345T	Transcatheter mitral valve repair

Service: Surgical Mitral Valve Repair

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** If a patient has had a mitral valve repair and it needs further intervention, a mitral valve replacement is typically indicated.
- **Recommended Clinical Approach:** Mitral valve repair has a lower operative mortality rate than mitral valve replacement, and it avoids the complications of prosthetic valves. Mitral valve repair is recommended for patients with severe primary mitral regurgitation (MR) when the anatomic cause of MR is a degenerative disease if repair is possible.[18] TEE can help predict surgical strategy, but the final decision about repair versus replacement is made at the time of surgery.[18] Perform mitral valve repair at a Comprehensive Valve Center.
- **Exclusions:** Exclusions to surgical mitral valve repair include prohibitive surgical risk (predicted risk of death or major morbidity (all-cause) greater than 50% at 1 year, Frailty (2 or more of 7 indices, 3 or more major organ systems not to be improved postoperatively, or Severe procedure-specific impediment)).

Medical Necessity Criteria

Indications

➔ **Surgical mitral valve repair** is considered appropriate if **ANY** of the following is **TRUE**[18]:

- ◆ In symptomatic patients with severe primary mitral regurgitation (MR).
- ◆ In asymptomatic patients with severe primary MR and **ANY** of the following:
  - Left ventricular (LV) systolic dysfunction (LV ejection fraction (LVEF) less than or equal to 60% or LV-end systolic diameter (LVESD) greater than or equal to 40 mm).
  - Normal LV systolic function (LVEF greater than 60% and LVESD less than 40 mm) with a low expected surgical mortality risk and a high likelihood of a successful repair.
  - Normal LV systolic function (LVEF greater than 60% and LVESD less than 40 mm) AND a progressive increase in LV size or decrease in EF on 3 or more serial imaging studies.
- ◆ In patients with severe secondary MR who are undergoing CABG.
- ◆ Severe secondary MR from mitral annular dilation with preserved LV systolic function (LVEF greater than or equal to 50%) who have

severe persistent symptoms (New York Health Association (NYHA) class III or IV) despite optimal therapy for heart failure (HF).
- Severe secondary MR related to LV systolic dysfunction (LVEF less than 50%) in patients with persistent and severe symptoms despite optimal therapy for heart failure (HF).
- In asymptomatic women with severe MR who are considering pregnancy.

Non-Indications

➔ **Surgical mitral valve repair** may not be appropriate if **ANY** of the following is **TRUE**[48]:
- Severe LV dysfunction (LVEF less than 20%).
- Severe emphysema.
- Restrictive lung disease.
- Pulmonary hypertension.

Site of Service Criteria

All procedures must be performed at a comprehensive valve center.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
33426	Valvuloplasty, mitral valve, with cardiopulmonary bypass; with prosthetic ring

Service: Transcatheter Aortic Valve Replacement/Implantation (TAVR)

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** Dysfunction of previous TAVR.
- **Recommended Clinical Approach:** TAVR isn't an effective procedure for the treatment of severe symptomatic AS in adults.[18] Compared to surgical aortic valve replacement (SAVR), TAVR has a lower risk of stroke, major bleeding, and atrial fibrillation (AF), as well as a shorter hospital length of stay and shorter recovery. Compared with SAVR, TAVR has a higher rate of vascular complications, paravalvular regurgitation, permanent pacemaker implantation, and valve intervention, but the advantages of TAVR often outweigh these disadvantages. TAVR valves are durable for at least 5 years.[18] The Society of Thoracic Surgeons estimated surgical risk score provides a useful measure of the extent of patient comorbidities and may help identify which patients will benefit from TAVR.[18]
- **Exclusions:** Life expectancy less than 1 year even with a successful procedure or those with a chance of "survival with benefit" of less than 25% at 2 years.[18]

Medical Necessity Criteria

Indications

➔ **TAVR** is considered appropriate if **ANY** of the following is **TRUE**:
- ◆ In symptomatic patients age 65 or greater with severe AS.
- ◆ In asymptomatic patients age 65 or greater with severe AS and LVEF less than 50%.[18]
- ◆ Valve-in-valve procedures (new artificial valve placed inside the orifice of a previous valve) for failed prior bioprosthetic valves as defined in the guidelines.[18]
- ◆ In patients with an indication for AVR, a high/prohibitive surgical risk, AND a predicted post-TAVR survival greater than 12 months.

Non-Indications

➔ **TAVR** may not be considered appropriate if **ANY** of the following is **TRUE**:
- ◆ The patient is pregnant.
- ◆ Patient life expectancy is less than 12 months due to a non-cardiac cause.
- ◆ The patient had a myocardial infarction within the last 30 days.

- There is a congenital unicuspid, bicuspid, or noncalcified valve.
- The patient has hypertrophic cardiomyopathy.
- There is a short distance between the annulus and coronary ostium.
- There is echocardiographic evidence of intracardiac mass, thrombus, or unhealed vegetation.
- The native aortic annulus is smaller than 18 or larger than 30 mm.
- There is severe mitral regurgitation.
- The patient has active Endocarditis.
- A significant aortoiliac disease that would interfere with delivery and deployment of the stent-valve.[18]

Site of Service Criteria

Inpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
33361	Transcatheter aortic valve replacement (TAVR/TAVI) with prosthetic valve using percutaneous femoral artery approach
33362	Transcatheter aortic valve replacement (TAVR/TAVI) with prosthetic valve by open femoral artery approach
33363	Transcatheter aortic valve replacement (TAVR/TAVI) with prosthetic valve by open axillary artery approach
33364	Transcatheter aortic valve replacement (TAVR/TAVI) with prosthetic valve by open iliac artery approach
33365	Transcatheter aortic valve replacement (TAVR/TAVI) with prosthetic valve by median mediastinotomy
33366	Transcatheter aortic valve replacement (TAVR/TAVI) using prosthetic valve with transapical exposure

Service: Patent Foramen Ovale (PFO) and Atrial Septal Defect (ASD) closure

General Guidelines

- **Units, Frequency, & Duration:** None.
- **Criteria for Subsequent Requests:** None.
- **Recommended Clinical Approach:** Patent foramen ovale (PFO) refers to the nonclosure between septum primum and secundum located at the superior and inferior margin of the foramen ovale. Closure can prevent stroke in appropriate patients. Patients with a hemodynamically important isolated secundum atrial septal defect (ASD) benefit from surgical or transcatheter closure of the secundum ASD. Patients who do not undergo ASD closure may experience atrial arrhythmias, reduced functional capacity, and greater degrees of pulmonary arterial hypertension (PAH).[1] While secundum ASDs can be closed percutaneously, anatomy permitting, the primum, sinus venosus, and coronary sinus ASDs require surgical treatment.
- **Exclusions:** Exclusions to percutaneous device closure may include the presence of an inferior vena cava filter, elevated bleeding risk or coagulopathy, and vascular, cardiac, or PFO anatomy that is unsuitable for device placement.

Medical Necessity Criteria

Indications

➔ **PFO closure** is considered appropriate if **ANY** of the following is **TRUE**[49,50]:
- ◆ The patient age is less than or equal to 60 years with PFO with ALL of the following:
  - Right-to-left shunt.
  - Embolic-appearing ischemic stroke with no other evident source of stroke.
- ◆ The patient's age is less than or equal to 60 years with PFO with unexplained recurrent embolic stroke despite medical therapy (without another identified cause).
- ◆ The patient has orthodeoxia/platypnea syndromes after pulmonary disease has been excluded.

➔ **ASD closure** is considered appropriate if **ANY** of the following is **TRUE**:
- ◆ The patient has an isolated secundum ASD and **ALL** of the following:
  - Impaired functional capacity.
  - RA or RV enlargement.

- Hemodynamically significant net left-to-right shunt (Qp:Qs greater than or equal to 1.5:1).

◆ The patient is asymptomatic and **ALL** of the following:
- Isolated atrial septal defect (ASD).
- RA and RV enlargement.
- Net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs 1.5:1 or greater).

◆ Percutaneous or surgical closure may be considered for adults with **ANY** of the following:
- ASD when net left-to-right shunt (Qp:Qs) is 1.5:1 or greater.
- PA systolic pressure is 50% or more of systemic arterial systolic pressure.
- Pulmonary vascular resistance is greater than one-third of the systemic resistance.

◆ Worsening hypoxia in a patient with a fenestrated Fontan circuit.
◆ Can be considered for documented recurrent paradoxical embolization event on treatment (without another identified cause).[50]
◆ The patient has orthodeoxia/platypnea syndrome.
◆ Unrepaired Ebsteins anamoly with moderate-severe tricuspid regurgitation and impaired exercise tolerance.

Non-Indications

➔ **PFO closure** is not considered appropriate if **ANY** of the following is **TRUE**:
- ◆ The patient has severe pulmonary hypertension.
- ◆ PFO was discovered incidentally without associated symptoms.

➔ **ASD closure** is not considered appropriate if the following is **TRUE**:
- ◆ The patient has severe pulmonary hypertension.[1]
- ◆ Net right to left shunt.

Site of Service Criteria

Inpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
93580	Percutaneous transcatheter closure of congenital interatrial communication (i.e., fontan fenestration, atrial septal defect) with implant.

Service: Cardiac Implantable Device (Pacemakers)

General Guidelines

- **Units, Frequency, & Duration:** One instance, as needed per inclusion criteria.
- **Criteria for Subsequent Requests:** Subsequent requests may be considered for device replacement due to battery end of life (EOL) or elective replacement interval (ERI), replacement after infection, clinical need for different pacing modes, or replacement after manufacturer recall.
- **Recommended Clinical Approach:** Pacing is an important tool in congenital heart disease, as bradyarrhythmias may have more severe consequences in this population. Sinus bradycardia or loss of AV synchrony can have significant hemodynamic effects on patients with complex congenital heart disease. Pacing may assist patients with sick sinus syndrome to relieve clinically symptomatic bradycardia, restore physiologic heart rate response, and control brady-tachy syndrome. Many antiarrhythmics used to control atrial fibrillation can exacerbate sick sinus syndrome, which may require pacing to prevent symptomatic bradycardia if no alternative treatments are feasible.[28] Patients who have cardiac surgeries resulting in high-grade second degree or complete AV block should be paced if cardiac conduction shows no evidence of recovery.
- **Exclusions:** Conduction disease-causing symptomatic bradycardia or unstable rhythm is an appropriate diagnosis for pacemaker requests; additional clinical information is needed for defibrillator implant requests.

Medical Necessity Criteria

Indications

➔ **Pacemakers** are considered appropriate if **ANY** of the following is **TRUE**:
- ◆ Congenital Heart Disease and **ANY** of the following:
  - Sinus node dysfunction and symptoms related to bradycardia[5]
  - Advanced 2nd or 3rd degree atrioventricular (AV) block with symptomatic bradycardia, ventricular dysfunction, or low cardiac output.[3]
  - Syncope with bifascicular block and transient high-grade 2nd degree or 3rd degree AV block.
- ◆ In a patient with symptomatic sinus node dysfunction caused by standard of care treatments (i.e., pharmacologic treatments)

AND there is no alternative management OR alternative management was exhausted.
- Chronotropic incompetence due to bradycardia.
- Brady-tachy syndrome for symptoms attributable to bradycardia.
- Post-operative advanced second or third degree AV block that persists for atleast 7-10 days after cardiac surgery

Non-Indications

➔ **Pacemakers** are not considered appropriate if **ANY** of the following is **TRUE**:
- The patient has asymptomatic sinus bradycardia or sinus pauses due to physiologically elevated parasympathetic (vagal) tone.
- The patient has an asymptomatic bifascicular block in the absence of a transient complete AV block.
- There are isolated sleep-related sinus bradycardia or sinus pauses.
- There is syncope of undetermined etiology.
- In patients with asymptomatic Sinus Node Dysfunction, or in those in whom the symptoms have been documented to occur in the absence of bradycardia or chronotropic incompetence, permanent pacing should not be performed.[5]
- In patients with first-degree atrioventricular block or second-degree Mobitz type I (Wenckebach) or 2:1 atrioventricular block, which is believed to be at the level of the atrioventricular node, with symptoms that do not temporally correspond to the atrioventricular block, permanent pacing should not be performed.[5]
- In asymptomatic patients with first-degree atrioventricular block or second-degree Mobitz type I (Wenckebach) or 2:1 atrioventricular block, which is believed to be at the level of the atrioventricular node, permanent pacing should not be performed.[5]
- In patients who had acute atrioventricular block attributable to a known reversible and nonrecurrent cause, and have had complete resolution of the atrioventricular block with treatment of the underlying cause, permanent pacing should not be performed.[5]

Site of Service Criteria

Outpatient or observation status.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
33206	Insertion of permanent atrial pacemaker with transvenous electrode
33207	Insertion of permanent ventricular pacemaker with transvenous electrode
33208	Insertion of permanent atrial and ventricular pacemaker with transvenous electrode
33210	Insertion of temporary transvenous single chamber cardiac electrode
33211	Insertion of temporary transvenous dual chamber pacing electrodes
33212	Insertion of pacemaker pulse generator with connection to existing single lead
33213	Insertion of pacemaker pulse generator with connection to existing dual leads
33214	Conversion of single chamber implanted pacemaker system to dual chamber system
33216	Insertion of transvenous electrode of permanent pacemaker
33217	Insertion of 2 transvenous electrodes of permanent cardioverter-defibrillator
33221	Insertion of pacemaker pulse generator with existing multiple leads
33224	Transvenous insertion of pacing electrode for left ventricular pacing, with connection to existing pacemaker
33227	Removal and replacement of permanent pacemaker pulse generator in single lead system
33228	Removal and replacement of permanent pacemaker pulse generator in dual lead system
33229	Removal and replacement of permanent pacemaker pulse generator in multiple lead system
33233	Removal of permanent pacemaker pulse generator
33234	Removal of transvenous atrial pacemaker electrode in single lead

	system
33235	Removal of transvenous pacemaker electrode in dual lead system
33274	Transcatheter insertion of permanent leadless right ventricular pacemaker
33275	Transcatheter removal of permanent leadless pacemaker from right ventricle using imaging guidance
C1779	Lead, pmkr, transvenous vdd
C1785	Pmkr, dual, rate-resp
C1786	Pmkr, single, rate-resp
C1898	Lead, pmkr, other than trans
C1900	Lead, coronary venous
C2619	Pmkr, dual, non rate-resp
C2620	Pmkr, single, non rate-resp
C2621	Pmkr, other than sing/dual

Service: Cardiac Implantable Device (Defibrillators)

General Guidelines

- **Units, Frequency, & Duration:** One instance, as needed per inclusion criteria.
- **Criteria for Subsequent Requests:** Subsequent requests may be appropriate for device replacement due to battery end of life or elective replacement interval, replacement after infection, a clinical need for different pacing modes, or replacement after manufacturer recall.
- **Recommended Clinical Approach:** Patients with congenital heart diseases who have undergone multiple surgical interventions are at higher risk for ventricular arrhythmias due to myocardial scarring and remodeling. Repaired Tetralogy of Fallot is a higher risk lesion due to surgeries involving ventriculostomies.[3,43] Other lesions, especially those with a systemic right ventricle, are subject to progressive ventricular remodeling and declining function, which increase the risk of sudden cardiac death (SCD).[4] Implantable cardioverter-defibrillators (ICDs) can extend these patients' lifespans.
- **Exclusions:** In a patient whose expected remaining lifespan is less than 12 months.

Medical Necessity Criteria

Indications

➔ **Implantable cardioverter-defibrillators (ICDs)** are considered appropriate if **ANY** of the following is **TRUE**:

- ◆ The congenital heart disease patient has clinical episodes of sustained ventricular tachycardia (VT), ventricular fibrillation (VF), or aborted cardiac arrest.
- ◆ The patient has a systemic right ventricle (RV) (e.g., HLHS, DORV, L-TGA), advanced RV dysfunction (EF systemic RV less than 35%), and additional risk factors for sudden cardiac death, including **ANY** of the following[28]:
  - Ventricular tachycardia.
  - Severe atrioventricular valve regurgitation.
  - QRS greater than 140 ms AND L-transposition of the great arteries.
- ◆ In a patient with Tetralogy of Fallot and one or more risk factors for sudden cardiac death, including **ANY** of the following [1,4,28]:
  - Left ventricular dysfunction.
  - Sustained, symptomatic ventricular tachycardia.
  - QRS duration is greater than 180 ms.
  - Extensive RV scarring on magnetic resonance imaging.

- Inducible, sustained VT on programmed electrical stimulation.

Non-Indications

➔ **Implantable Cardioverter-Defibrillators (ICDs)** are NOT considered appropriate if **ANY** of the following is **TRUE**[51]:
- ◆ In a patient whose expected lifespan is less than 12 months.
- ◆ VT, which is amenable to catheter ablation.
- ◆ In a patient with incessant, drug-refractory VT or VF.
- ◆ The ventricular arrhythmias are due to reversible circumstances (e.g., electrolyte abnormalities, toxic ingestion.)

Site of Service Criteria

Outpatient or observation status.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
33217	Insertion of 2 transvenous electrodes of permanent cardioverter-defibrillator
33230	Insertion of pacing cardioverter-defibrillator pulse generator with connection to existing dual leads
33231	Insertion of pacing cardioverter-defibrillator pulse generator with connection to existing multiple leads
33240	Insertion of pacing cardioverter-defibrillator pulse generator with connection to existing single lead
33249	Insertion of dual chamber permanent pacing cardioverter-defibrillator system with transvenous lead
33262	Removal and replacement of pacing cardioverter-defibrillator pulse generator in single lead system
33263	Removal and replacement of pacing cardioverter-defibrillator pulse generator in dual lead system
33264	Removal and replacement of pacing cardioverter-defibrillator pulse generator in multiple lead system
33270	Insertion of permanent subcutaneous implantable defibrillator system with subcutaneous electrode
33271	Insertion of subcutaneous implantable defibrillator electrode

93640	Electrophysiologic evaluation of dual chamber pacing cardioverter-defibrillator leads with defibrillation threshold evaluation at time of initial implantation
93641	Electrophysiologic evaluation of dual chamber pacing cardioverter-defibrillator leads with defibrillation threshold evaluation at time of initial implantation with testing of pacing cardioverter-defibrillator pulse generator
93642	Electrophysiologic evaluation of dual chamber pacing cardioverter-defibrillator with defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing and pacing for arrhythmia termination and programming of sensing parameters
93644	Electrophysiologic evaluation of subcutaneous implantable defibrillator with defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination and programming of sensing parameters
0571T	Insertion or replacement of implantable cardioverter-defibrillator system with substernal electrode(s), including all imaging guidance and electrophysiological evaluation (includes defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters), when performed
0572T	Insertion of substernal implantable defibrillator electrode
0577T	Electrophysiologic evaluation of implantable cardioverter-defibrillator system with substernal electrode (includes defibrillation threshold evaluation, induction of arrhythmia, evaluation of sensing for arrhythmia termination, and programming or reprogramming of sensing or therapeutic parameters)
C1721	Aicd, dual chamber
C1722	Aicd, single chamber
C1777	Lead, aicd, endo single coil
C1882	Aicd, other than sing/dual
C1895	Lead, aicd, endo dual coil
C1899	Lead, pmkr/aicd combination

Service: Surgical Maze Procedure

General Guidelines

- **Units, Frequency, & Duration:** Single procedure.
- **Criteria for Subsequent Requests:** None.
- **Recommended Clinical Approach:** Surgical management of atrial fibrillation is typically approached with a maze procedure, electrically segmenting the left atrium with radiofrequency ablation. These procedures are highly recommended for atrial fibrillation associated with mitral valve disease (Class I, Level A) that requires surgical intervention. It should also be considered during 1) aortic valve replacement; 2) coronary artery bypass grafting; or 3) aortic valve replacement and coronary artery bypass grafting in patients with symptomatic atrial fibrillation (Class I, Level B).[18] Standalone procedures are reasonable for patients refractory to Class I/III antiarrhythmics; however these are much less common given recent advances in percutaneous catheter procedures.[52]

  Maze procedures are also appropriate for patients with congenital heart disease and refractory atrial fibrillation that require cardiac surgery to repair or revise their anatomy. These are especially recommended for patients with high-risk physiology (e.g., Ebstein's Anomaly, univentricular hearts.) Prophylactic Maze procedure is also reasonable in high-risk patients.[53]

  Surgical LAA excision or exclusion in conjunction with surgical ablation to prevent thromboembolic complications of atrial fibrillation received a Class IIa (Level of evidence: C; "limited data") recommendation in the 2017 Society of Thoracic Surgeons clinical practice guidelines.[53] Updated guidelines suggest that surgical occlusion of the LAA may be considered in patients with AF undergoing cardiac surgery as a component of an overall heart team approach to the management of AF.[52]
- **Exclusions:** None.

Medical Necessity Criteria

Indications

➔ **Surgical maze procedures** are considered appropriate if **ANY** of the following is **TRUE**[18,53]:

- ◆ In a patient with symptomatic atrial fibrillation at the time of coronary artery bypass graftin or any other open heart surgery in ACHD..

- In a patient who, at the time of congenital heart disease repair or revision, has atrial fibrillation OR is at a high-risk for developing atrial arrhythmias.
- In a patient with symptomatic and refractory atrial arrhythmias where percutaneous catheter ablation is not available or is not likely to be effective.

Non-Indications

➔ **Surgical maze procedures** are not considered appropriate if **ANY** of the following is **TRUE**[18,53]:

- In a patient with symptomatic atrial fibrillation where no cardiac surgery is planned and percutaneous catheter ablation is a feasible treatment option.

Site of Service Criteria

Inpatient.

Procedure Codes (HCPCS/CPT)

HCPCS Code	Code Description/Definition
33254	Operative tissue ablation and reconstruction of atria, limited (e.g., modified Maze procedure)
33255	Operative tissue ablation and reconstruction of atria, extensive (e.g., Maze procedure); without cardiopulmonary bypass
33256	Operative tissue ablation and reconstruction of atria, extensive (e.g., Maze procedure); with cardiopulmonary bypass
33257	Operative tissue ablation and reconstruction of atria, performed at the time of other cardiac procedure(s), limited (e.g., modified Maze procedure)
33258	Operative tissue ablation and reconstruction of atria, performed at the time of other cardiac procedure(s), extensive (e.g., Maze procedure); without cardiopulmonary bypass
33259	Operative tissue ablation and reconstruction of atria, performed at the time of other cardiac procedure(s), extensive (e.g., Maze procedure); with cardiopulmonary bypass

33265	Endoscopy, surgical; operative tissue ablation and reconstruction of atria, limited (e.g., modified Maze procedure); without cardiopulmonary bypass
33266	Endoscopy, surgical; operative tissue ablation and reconstruction of atria, extensive (e.g., Maze procedure); without cardiopulmonary bypass

Surgical Risk Factors

Patient Medical Risk Stratification

Patient Risk Score	Patient Characteristic	Min Range	Max Range	Guidance
1- Very Low Risk	No known medical problems			
2- Low Risk	Hypertension		180/110 mm Hg	
2- Low Risk	Asthma	peak flow >80% of predicted or personal best value		
2- Low Risk	Prior history of alcohol abuse			Screen for liver disease and malnutrition
2- Low Risk	Prior history of tobacco use			
3- Intermediate Risk	Asthma	peak flow <80% of predicted or personal best value		
3- Intermediate Risk	Active alcohol abuse			
3- Intermediate Risk	Age	65	75	
3- Intermediate Risk	History of treated, stable coronary artery disease (CAD)			
3- Intermediate Risk	Stable atrial fibrillation			
3- Intermediate Risk	Diabetes mellitus	HbA1C >7%		
3- Intermediate Risk	Morbid obesity	BMI 30	BMI 40	
3- Intermediate Risk	Anemia	hemoglobin <11 (females), <12 (males)		Workup to identify etiology
3- Intermediate Risk	HIV	CD4 <200 cells/mm3		Get clearance from HIV specialist
3- Intermediate Risk	Rheumatologic disease			Preoperative consultation with rheumatologist re: perioperative medication management
3- Intermediate Risk	Peripheral vascular disease or history of peripheral vascular bypass	ankle-brachial pressure index (ABPI) <0.9		Preoperative consultation with vascular surgeon

3- Intermediate Risk	History of venous thromboembolism (VTE)			
3- Intermediate Risk	Well-controlled obstructive sleep apnea			
3- Intermediate Risk	Malnutrition	transferrin <200 mg/dL albumin <3.5 g/dL prealbumin <22.5 mg/dL total lymphocyte count <1200-1500 cell/mm3 BMI <18		Preoperative consultation with nutritionist
3- Intermediate Risk	Active tobacco Use			Enroll patient in smoking cessation program
3- Intermediate Risk	Known allergy or hypersensitivity to medication needed for procedure			
4- High Risk	Advanced Renal Disease (Creatinine > 2)			
4- High Risk	Diabetes mellitus with complications	HbA1c >8%		
4- High Risk	Age	76	85	
4- High Risk	Oxygen dependent pulmonary disease			
4- High Risk	Sickle cell anemia			
4- High Risk	Obesity	BMI 40		
4- High Risk	Cirrhosis, history of hepatic decompensation or variceal bleeding			
4- High Risk	Impaired cognition; dementia			
4- High Risk	Compensated CHF			
4- High Risk	Cerebrovascular disease			
4- High Risk	Uncontrolled or suspected obstructive sleep apnea (OSA)			
4- High Risk	Renal insufficiency	serum creatinine >1.5 mg/dL or creatinine clearance <100 mL/min		

4- High Risk	Opioid dependence			
5- Very High Risk	Percutaneous Coronary Intervention (PCI) within 1 month			
5- Very High Risk	Cardiovascular: unstable angina, recent myocardial infarction (60 days), uncontrolled atrial fibrillation or other high-grade abnormal rhythm, severe valvular disease, decompensated heart failure			
5- Very High Risk	Primary pulmonary hypertension			Preoperative consultation with pulmonologist warranted
5- Very High Risk	Cirrhosis or severe liver disease, history of hepatic decompensation or variceal bleeding			
5- Very High Risk	Severe frailty, dependence for ADLs, or history of 3 or more falls in last 6 mos			
5- Very High Risk	Obesity		BMI >50	
5- Very High Risk	Age		>85	
5- Very High Risk	History of VTE with CI to anticoagulation, failure of anticoagulation, cessation of anticoagulation therapy secondary to bleeding			Preoperative consultation with hematologist or internist
5- Very High Risk	Renal failure requiring dialysis			
5- Very High Risk	Immunosuppression			
5- Very High Risk	Chronic Pain			

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Clinical Guideline Revision History/Information

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Review History	
February 11, 2022 (V.1)	**Physician author:** Mary Krebs, MD (Primary Care Physician), Giovanni Lorenz, MD (Radiologist) **Peer reviewed by:** Alisa Niksch, MD (Pediatric Cardiologist/ Electrophysiologist), Russell Rotondo, MD FACC (Cardiologist) **Approving Physician:** Russell Rotondo, MD FACC (Cardiologist)
October 31, 2022 (V.2)	**Peer reviewed by:** Pushpa Shivaram, MD (Cardiologist) **Approving Physician:** Russell Rotondo, MD FACC (Cardiologist)