*Evolent	
Clinical guideline **TRANSTHORACIC (TTE) ECHO**	**Original Date: October 2009**
CPT codes: 93303, 93304, 93306, 93307, 93308, +93320, +93321, +93325, +93356	**Last Revised Date: April 2023**
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GENERAL INFORMATION

- *It is an expectation that all patients receive care/services from a licensed clinician. All appropriate supporting documentation, including recent pertinent office visit notes, laboratory data, and results of any special testing must be provided. If applicable: All prior relevant imaging results and the reason that alternative imaging cannot be performed must be included in the documentation submitted.*
- *Where a specific clinical indication is not directly addressed in this guideline, medical necessity determination will be made based on widely accepted standard of care criteria. These criteria are supported by evidence-based or peer-reviewed sources such as medical literature, societal guidelines and state/national recommendations.*

ADULT PATIENTS – INDICATIONS FOR TRANSTHORACIC ECHOCARDIOGRAPHY (TTE)[1]

(Indications for pediatric patients follow this section)

Evaluation of Cardiac Structure and Function

- When initial evaluation including history, physical examination, electrocardiogram (ECG), remote monitor or other testing suggests a cardiac etiology for symptoms, including but not limited to:
  - Chest pain when another study is not planned to evaluate
  - Shortness of breath
  - Palpitations
- Hypotension suggestive of cardiac etiology not due to other causes, such as:
  - Medications, dehydration, or infection
- ECG Abnormalities
  - Previously unevaluated pathological Q waves (in two contiguous leads) defined as the following:
    - 40 ms (1 mm) wide
    - > 2 mm deep
    - > 25% of depth of QRS complex

- New left bundle branch block (as documented in MD notes and on ECG).
    - New isolated RBBB is **not** an indication for TTE.
- Previously unevaluated left ventricular hypertrophy (i.e., concern for hypertrophic cardiomyopathy).

Murmur or Click

- Initial evaluation when there is a reasonable suspicion for valvular or structural heart disease such as:
    - High grade ≥ 3/6: Note that TTE can be approved for documented concern that murmur suggests a **specific valve pathology** (such as "aortic valve sclerosis/stenosis" or "mitral regurgitation") **regardless of grade of murmur**
    - Holosystolic
    - Continuous
    - Diastolic

Arrhythmias

- Frequent premature ventricular contractions (PVCs, greater than 30 per hour on remote monitoring or ≥ 1 PVC on 12 lead ECG)
    - Isolated premature atrial complexes (PACs) are not an indication for TTE.
- Sustained or nonsustained ventricular tachycardia (VT) or ventricular fibrillation (VF), or ventricular bigeminy
- New onset atrial fibrillation (as documented in MD notes and on ECG) which was not evaluated by a prior transthoracic echocardiogram (TTE)

Syncope[2, 3]

- History, physical examination, or electrocardiogram (ECG) consistent with a cardiac diagnosis known to cause presyncope or syncope, including but not limited to, known or suspected:
    - Structural heart disease (including but limited to):
        - Hypertrophic cardiomyopathy
        - Systolic heart failure
    - Exercise-induced syncope

  And not due to other causes such as:
    - Vaso-vagal syncope, neurogenic orthostatic syncope
    - Orthostasis related to medication or dehydration

Perioperative Evaluation[4, 5]

- Preoperative left ventricular function assessment in patients who are candidates for solid organ transplantation (can be done yearly prior to transplant)

Pulmonary Hypertension

- Evaluation of suspected pulmonary hypertension including evaluation of right ventricular function and estimated pulmonary artery pressure
- Re-evaluation of known pulmonary hypertension if there is a change in clinical status or cardiac exam or a need to change medications[6] such as:
    - New chest pain
    - Worsening shortness of breath
    - Syncope
    - Increased murmur
    - Worsening rales on lung examination
- Initial evaluation of patients with pulmonary embolism to risk stratify and initiate appropriate therapy[7]
    - Repeat TTE can be approved for persistent dyspnea 3-6 months after PE[8] to evaluate for possible chronic thromboembolic pulmonary hypertension (CTEPH)
- Annual screening can be performed for pulmonary hypertension in patients with[6, 9]:
    - Scleroderma
    - Portal hypertension (including evaluation prior to TIPS procedure)
    - Carriers of Bone Morphogenic Protein Receptor 2 (BMPR2) mutation
    - Sickle cell disease

Evaluation of Valvular Function[2, 10-12]

- Screening of first-degree relatives of patients with a bicuspid aortic valve

Native Valvular Stenosis

- Routine surveillance (≥ 3 yrs.) of bicuspid aortic valve, aortic sclerosis, or mild valvular stenosis
- Re-evaluation (≥ 1 yr) of moderate stenosis
- Re-evaluation of severe aortic stenosis (AS) every 6 - 12 months
- Re-evaluation after starting medication in patients with low flow/low gradient severe aortic stenosis

Native Valvular Regurgitation[2, 13, 14]

- Re-evaluation (≥ 3 yrs.) of mild valvular regurgitation
- Re-evaluation (≥ 1 yr) of moderate valvular regurgitation
- Re-evaluation of asymptomatic patient every 6 - 12 months with severe valvular regurgitation

Prosthetic Valves/Native Valve Repair

- Initial evaluation of prosthetic valve or native valve repair, for establishment of baseline, typically 6 weeks to 3 months postoperative

- Routine surveillance of surgical bioprosthetic valve: every 3 years after surgery
- Routine surveillance of surgical bioprosthetic and mechanical valve: at 10 years postoperatively and annually thereafter
- Routine surveillance of surgical mitral valve repair: 1-year post-op and then every 2-3 years
- Evaluation of prosthetic valve or native valve repair with suspected dysfunction, with symptoms including but not limited to:
    - Chest pain
    - Shortness of breath
    - New or Increased murmur on heart examination
    - New rales on lung examination
    - Elevated jugular venous pressure on exam

Transcatheter Heart Interventions

Transcatheter Aortic Valve Replacement (TAVR)[2, 12, 15]

- Pre TAVR evaluation
- Post TAVR at 30 days (6 weeks to 3 months also acceptable) and annually
- Assessment post TAVR when there is suspicion of valvular dysfunction, including but not limited to:
    - Chest pain
    - Shortness of breath
    - New or increased murmur on heart examination
- Assessment of stroke post TAVR

Percutaneous Mitral Valve Repair[2, 12, 13]

- Pre-procedure evaluation
- Reassessment for degree of MR and left ventricular function (1, 6 months, and annually)

Closure of PFO or ASD[10]

- Pre-procedure evaluation
- Routine follow-up post procedure for device position and integrity (see Table 2: Adult and Pediatric Congenital Heart Disease Follow-Up)
- Evaluation for clinical concern for infection, malposition, embolization, or persistent shunt
- Routine surveillance of an asymptomatic patient with a PFO is **not** indicated[16]

Left Atrial Appendage (LAA) Occlusion[10]

- Pre-procedure evaluation

Pericardial Disease[7, 10, 17, 18]

- Suspected pericardial effusion
- Re-evaluation of known pericardial effusion when findings would lead to change in management
- Suspected pericardial constriction or reevaluation of status when management would be changed

Evaluation of Cardiac Source of Emboli or Cardiac Mass[2]

- Embolic source in patients with recent transient ischemic attack (TIA), stroke, or peripheral vascular emboli
- Evaluation of intracardiac mass or re-evaluation of known mass[19]

Infective Endocarditis (Native or Prosthetic Valves)[2, 11, 20]

- Initial evaluation of suspected infective endocarditis with positive blood cultures or a new murmur
- Re-evaluation of infective endocarditis with, but not limited to:
    - Changing cardiac murmur
    - Evidence of embolic phenomena such as TIA or CVA
    - New chest pain, shortness of breath, or syncope
    - A need to change medications due to ongoing fever, positive blood cultures, or evidence of new AV block on ECG
- Re-evaluation of patient with infective endocarditis at high risk of progression or complication (extensive infective tissue/large vegetation, or staphylococcal, enterococcal, or fungal infections)
- At completion of antimicrobial therapy and serial examinations at 1, 3, 6, and 12 months during the subsequent year[20]

Thoracic Aortic Disease[21-26]

In the absence of recent computed tomography (CT) or cardiovascular magnetic resonance (CMR), which are preferred for imaging beyond the proximal ascending aorta

- Screening of first-degree relatives of individuals with:
    - Thoracic aortic aneurysm (defined as ≥ 50% above normal) or dissection
    - Bicuspid aortic valve
    - Presence of an aortopathic syndrome (i.e., Marfan's, Ehlers-Danlos, Loeys-Dietz, or Turner's)
- If one or more first-degree relatives of a patient with a known thoracic aortic aneurysm or dissection, have thoracic aortic dilatation, aneurysm, or dissection; then imaging of 2nd degree relatives is reasonable
- Six-month follow-up after initial finding of a dilated thoracic aorta

- Annual follow-up of enlarged thoracic aorta that is above top normal for age, gender, and body surface area
- Biannual (twice/year) follow-up of enlarged aortic root ≥ 4.5 cm or showing growth rate ≥ 0.5 cm/year
- Evaluation of the ascending aorta in known or suspected connective tissue disease or genetic conditions that predispose to aortic aneurysm or dissection (e.g., Marfan syndrome, Ehlers-Danlos or Loeys-Dietz syndromes) at time of diagnosis and 6 months thereafter for growth rate assessment, followed by annual imaging, or biannual (twice yearly) if diameter ≥ 4.5 or expanding ≥ 0.5 cm/yr
- Turner's Syndrome:
    - Baseline evaluation at the time of diagnosis to assess for bicuspid aortic valve, coarctation of the aorta, aortic root and ascending aortic dilatation and other congenital defects.
    - Surveillance imaging (initial imaging normal and no additional risk factors for dissection such as HTN or bicuspid aortic valve):
        - Children: every 5 years
        - Adults: every 10 years
        - Prior to planned pregnancy
        - Annual imaging can be approved if an abnormality is found[27] (such as bicuspid aortic valve)
- Re-evaluation of known ascending aortic dilation or history of aortic dissection with one of the following:
    - New chest pain
    - Shortness of breath
    - Syncope
    - TIA or CVA
    - New or increased aortic valve murmur on clinical examination
    - New rales on lung examination or increased jugular venous pressure
    - **OR** when findings would lead to referral to a procedure or surgery
- Follow-up of aortic disease when there has been no surgical intervention:
    - Acute dissection: 1 month, 6 months, 12 months, then annually
    - Chronic dissection: annually
- Follow-up thoracic aortic aneurysm repair: chest CTA or chest MRA are the recommended surveillance imaging modalities.
- Evaluation of sinus of Valsalva aneurysms and associated shunting secondary to rupture.[25]

Hypertension (HTN) (Adult)[10,27]

- Initial evaluation of suspected hypertensive heart disease including but not limited to the following:
    - Left ventricular hypertrophy on ECG

- Cardiomegaly
- Evidence of clinical heart failure

Hypertension (HTN) (Pediatric)[28]

- Initial evaluation at time of consideration of pharmacologic treatment of HTN
- Re-evaluation at 6–12-month intervals for:
  - Persistent HTN despite treatment
  - Concentric LVH on prior study
  - Reduced LVEF on prior study
- Re-evaluation of patients without LVH on initial evaluation can have TTE annually for:
  - Stage 2 HTN (BP ≥140/90 mm Hg)
  - Secondary HTN
  - Chronic stage 1 HTN (BP between 130/80- and 139/89-mm Hg) incompletely treated, including drug resistance and noncompliance

Heart Failure[10, 29-31]

- Initial evaluation of suspected heart failure (HF) (systolic or diastolic) based on symptoms, signs, or abnormal test result, including but not limited to:
  - Dyspnea
  - Orthopnea
  - Paroxysmal nocturnal dyspnea
  - Worsening edema
  - Elevated BNP
- Re-evaluation of known HF (systolic or diastolic) with a change in clinical status or cardiac exam (as listed above)

Cardiomyopathy[10, 30-34]

- Initial evaluation of suspected inherited or acquired cardiomyopathy, including but not limited to:
  - Restrictive
  - Infiltrative/Depositional (i.e., hemochromatosis/iron overload, mucopolysaccharidoses, mitochondrial or metabolic storage disease (e.g., Danone disease, Fabry disease))
    - Fabry disease: annual surveillance TTE may be approved for patients receiving enzyme replacement[19]
  - Dilated
  - Hypertrophic
  - Re-evaluation of known cardiomyopathy if there is a need to monitor a change in medications or new symptoms, including but not limited to:
    - Chest pain
    - Shortness of breath

        - Palpitations
        - Syncope
- Heart failure (including Takotsubo cardiomyopathy)[19] with recovered left ventricular ejection fraction defined as (must meet all 3 criteria):
    - Documentation of a decreased LVEF <40% at baseline
    - ≥10% absolute improvement in LVEF
    - A second measurement of LVEF >40%[35]:
        - Repeat echocardiogram every 6 months until 12-18 months after recovery of EF, then annually for 2 years, then every 3-5 years
        - Higher risk patient (persistent left bundle branch block, genetic cardiomyopathy, higher biomarker profiles) may have annual follow-up
- Screening evaluation in first-degree relatives of a patient with an inherited cardiomyopathy
- Suspected cardiac sarcoidosis, including as a screening study in patients with biopsy proven extracardiac sarcoidosis[36]
- Suspected cardiac amyloid and to monitor disease progression and/or response to therapy, and to guide initiation and management of anticoagulation (TEE may be preferred)[34]
    - Light chain amyloidosis (AL): TTE may be repeated every 3-6 months
    - Transthyretin amyloidosis (ATTR): TTE may be repeated every 6-12 months[19]

Hypertrophic Cardiomyopathy (HCM)[33]

- Initial evaluation of suspected HCM
- Re-evaluation of patients with HCM with a change in clinical status or a new clinical event
- Evaluation of the result of surgical myomectomy or alcohol septal ablation
- Re-evaluation in patients with no change in clinical status or events every 1 - 2 years to assess degree of myocardial hypertrophy, dynamic obstruction, MR, and myocardial function
- Evaluation of patients with HCM who have undergone septal reduction therapy within 3-6 months after the procedure
    - Screening for patients who are clinically unaffected or (genotype-positive and phenotype-negative):
        - Children and adolescents, every 1-2 years
        - Adults every 3-5 years
    - Screening of first-degree relatives is recommended at the time HCM is diagnosed in the family member and serial follow-up as below:
        - Children and adolescents from genotype-positive families and families with early onset disease every 1-2 years
        - All other children and adolescents every 2-3 years
        - Adults every 3-5 years

- To guide therapy
    - Camzyos (mevacamten): baseline TTE prior to initiation. Repeat TTE during therapy at the discretion of the ordering physician[37].

Imaging Surveillance for Cardiotoxic Medication[38, 39]

- TTE is the method of choice for the evaluation of patients who will receive or have received cardiotoxic medication. TTE may be approved for:
    - Baseline assessment prior to initiation of therapy
    - Monitoring during therapy. The frequency of testing should be left to the discretion of the ordering physician, but in the absence of new abnormal findings, generally no more often than every 6 weeks while on active therapy.
    - Long term surveillance after completion of therapy may be required, especially for those who have been exposed to anthracycline medication. The frequency of testing is generally every 6-12 months, or at the discretion of the provider.

Imaging Surveillance for Previous Radiation Therapy with Cardiac Exposure[40]

- TTE is indicated for long term surveillance, generally at 5 years and at 10 years following radiation exposure. More frequent surveillance may be indicated at the discretion of the provider.

Device Candidacy or Optimization (Pacemaker, ICD, or CRT)

- Initial evaluation or re-evaluation after revascularization (≥ 90 days) and/or myocardial infarction (≥ 40 days) and/or 3 months of guideline-directed medical therapy when ICD is planned[41]
- Initial evaluation for CRT device optimization after implantation
- Re-evaluation for CRT device optimization in a patient with worsening heart failure
- Known implanted pacing device with symptoms possibly due to device complication or suboptimal pacing device settings

Ventricular Assist Devices (VADs) and Cardiac Transplantation[10, 42]

- To determine candidacy for VAD
- Optimization of VAD settings and assessment of response post device
- Re-evaluation for signs/symptoms suggestive of VAD-related complications, including but not limited to:
    - TIA or stroke
    - Infection
    - Murmur suggestive of aortic insufficiency
    - Worsening heart failure

Post Heart Transplant Surveillance Imaging

- Monitoring every 6 months (or at the discretion of the transplant center) for rejection in a cardiac transplant recipient. May be approved for more frequent monitoring in the first-year post-transplant[43].

Cardiovascular Disease in Pregnancy[32, 44]

- Valvular stenosis
  - Mild can be evaluated each trimester and prior to delivery
  - Moderate-severe can be evaluated monthly
- Valvular regurgitation
  - Mild-moderate regurgitation can be evaluated each trimester and prior to delivery
  - Severe regurgitation can be evaluated monthly
- Pre-pregnancy evaluation with mechanical or bioprosthetic heart valves (if not done within the previous year)
- Prior Postpartum Cardiomyopathy: can be repeated at the end of the 1st and 2nd trimesters, 1 month prior to delivery, after delivery prior to hospital discharge, 1 month postpartum, and serially including up to 6 months after normalization of ejection fraction
- Aortopathic syndromes (i.e., Marfan's, Ehlers-Danlos, Loeys-Dietz, or Turner's) or known dilated aortic root or ascending aorta: may be approved for pre-pregnancy planning and for monitoring each trimester during pregnancy and again several weeks post-partum. More frequent imaging may be approved depending on aortic diameter, aortic growth rate and comorbidities predisposing to dissection (i.e., presence of an aortopathic syndrome, HTN)[27].

Adult Congenital Heart Disease[16, 45, 46]

- Initial evaluation of suspected adult congenital heart disease
- Known adult congenital heart disease with a change in clinical status or cardiac exam, including but not limited to:
  - Chest Pain
  - Shortness of breath
  - New or increased murmur on physical exam
- Evaluation prior to surgical or transcatheter procedure
- For follow-up of specific lesions, see Table 1 **and** Table 2**: Adult and Pediatric Congenital Heart Disease Follow-up**

Inflammatory & Autoimmune

- Including any one of the following:
  - Suspected rheumatic fever[47]
  - Systemic lupus erythematosus[48]

- Takayasu arteritis[49]
- Multisystem Inflammatory Syndrome (MIS): at baseline and for surveillance when there is documented concern for coronary involvement or other late sequelae[50]
- Kawasaki disease[51]
    - Upon diagnosis, 1-2 weeks later, and 4 to 6 weeks after diagnosis
    - For patients with important and evolving coronary artery abnormalities during the acute illness, echocardiograms may need to be more frequent. In the setting of increasing size of coronary aneurysms, echocardiogram can be performed up to twice per week until dimensions have stopped progressing, then at least once per week in the first 45 days of illness, and then monthly until the third month after onset.
    - For persistent coronary aneurysm after the acute illness, echocardiogram surveillance intervals are based on the size of the aneurysm:
        - Small: at 6 months. and then yearly
        - Medium: at 3, 6 and 12 months and then every 6-12 months
        - Large/Giant: at 3, 6, 9 and 12 months and then every 3-6 months

COVID-19[52]

- Acute infection
    - Cardiopulmonary signs or symptoms (ECG abnormalities, elevated biomarkers, chest pain, dyspnea, syncope, palpitations)
- Post-Acute Sequelae (PASC) defined as new or returning cardiopulmonary symptoms 4 or more weeks and persisting more than 2 months following confirmed COVID infection, not explained by an alternative diagnosis (WHO definition).
- Post Vaccination
    - Symptoms or signs of myocarditis (ECG abnormalities, chest pain, elevated biomarkers)

Surveillance for Neuromuscular Disorders[53]

Asymptomatic surveillance intervals (genetically affected individuals with no signs or symptoms of cardiac involvement). Development of signs or symptoms of cardiac involvement necessitates more frequent assessment.

- Duchenne muscular dystrophy (DMD) and Becker muscular dystrophy (BMD)
    - age <10 years, TTE every 2 years
    - age 10 years or older, TTE annually
- Emery-Dreifuss muscular dystrophy (EDMD)
    - X-linked form: at least annual TTE
    - Autosomal form: TTE at initial diagnosis, surveillance TTE only if initial TTE abnormal

- Myofibrillar myopathy (MFM)
    - Annual TTE
- Barth (BTHS)-X linked recessive (only males develop disease)
    - Infant males TTE every 6 months
    - Age 1 year or older, annual TTE
- Limb-Girdle muscular dystrophy (LGMD)
    - TTE may be performed annually
- Friedrich's ataxia (FA)
    - TTE can be performed at least annually
- Myotonic dystrophy (DM)
    - TTE every 2-4 years

PEDIATRIC PATIENTS - INDICATIONS FOR TRANSTHORACIC ECHOCARDIOGRAPHY (TTE) (PATIENTS UNDER THE AGE OF 18)[54]

- Hypertension (see section: Hypertension (Pediatric))
- Renal failure
- Palpitations, if one:
    - Family history at age < 50 of either:
        - Sudden cardiac death/arrest **OR**
        - Pacemaker or ICD
    - History or family history of cardiomyopathy
- Chest pain, if one or more of the following:
    - Exertional chest pain
    - Abnormal ECG
    - Family history with unexplained sudden death or cardiomyopathy
- Syncope, if any of the following:
    - Abnormal ECG
    - Exertional syncope
    - Family history at age < 50 of either one:
        - Sudden cardiac death/arrest **OR**
        - Pacemaker or ICD
    - Family history of cardiomyopathy
- Signs and/or symptoms of heart failure, including, but not limited to:
    - Respiratory distress
    - Poor peripheral pulses
    - Feeding difficulty
    - Decreased urine output
    - Edema
    - Hepatomegaly
- Abnormal physical findings, including any one of the following:
    - Clicks, snaps, or gallops

    - Fixed and/or abnormally split S2
    - Decreased pulses
    - Central cyanosis
- Arrhythmia, if one of the following:
    - Supraventricular tachycardia
    - Ventricular tachycardia
- Murmur
    - Pathologic sounding or harsh murmur, diastolic murmur, holosystolic or continuous murmur, late systolic murmur, grade 3/6 systolic murmur or louder, or murmurs that are provoked and become louder with changes in position
    - Presumptively innocent murmur, but in the presence of signs, symptoms, or findings of cardiovascular disease
- Abnormal basic data, including any one of the following:
    - Abnormal ECG
    - Abnormal cardiac biomarkers
    - Desaturation on pulse oximetry
    - Abnormal chest x-ray
- Sickle cell
    - One time screening for risk stratification for pulmonary hypertension in children ≥ 8 years of age[55]
- Suspicion of Structural Disease, including any one of the following:
    - Premature birth where there is suspicion of a Patent Ductus Arteriosus
    - Vascular Ring, based upon either one:
        - Difficulty breathing with stridor and eating solid foods that might suggest a vascular ring
        - Abnormal barium swallow or bronchoscopy suggesting a vascular ring
- Genetic & Syndrome Related, including any one of the following:
    - Genotype positive for cardiomyopathy, family history of hypertrophic cardiomyopathy or heritable pulmonary arterial hypertension
    - Patient with a known syndrome associated with congenital or acquired heart disease (Down's syndrome, Noonan's syndrome, DiGeorge syndrome, William's syndrome, Trisomy Thirteen, Trisomy Eighteen, Alagille syndrome, chromosomal abnormality associated with cardiovascular disease)
    - Abnormalities of visceral or cardiac situs
    - Known or suspected connective tissue diseases that are associated with congenital or acquired heart disease. (e.g., Marfan's, Loeys-Dietz)
    - Patients with a first-degree relative with a genetic abnormality, such as cardiomyopathies (hypertrophic, dilated, arrhythmogenic right ventricular dysplasia, restrictive, left ventricular noncompaction).
- Maternal-Fetal related, including any one of the following:
    - Maternal infection during pregnancy or delivery with potential fetal/neonatal cardiac sequelae

- Maternal phenylketonuria
- Suspected cardiovascular abnormality on fetal echocardiogram

ADULT AND PEDIATRIC CONGENITAL HEART DISEASE FOLLOW-UP[16] ‡*
[‡All surgical or catheter-based repairs allow evaluation PRIOR to the procedure and POSTPROCEDURAL evaluation (within 30 days)]

- For all lesions, TTE is indicated for change in clinical status and/or development of new signs or symptoms
- Infant with any degree of unrepaired valvular AS/AR may have surveillance TTE every 1 – 4 weeks as needed
- Surveillance interval for patients with subvalvular stenosis **plus** aortic regurgitation will be dictated by the magnitude of the more significant abnormality (e.g., mild stenosis with moderate regurgitation would have surveillance interval as though stenosis were also moderate).
- Infant with any degree of unrepaired MS may have surveillance TTE every 1 – 4 weeks as needed
- After any surgical or catheter-based repair, evaluation (3-12 months) for a patient with heart failure symptoms
- Annual surveillance in a child with normal prosthetic mitral valve function and no LV dysfunction
- Surveillance (3-12 months) in a child with prosthetic mitral valve and ventricular dysfunction and/or arrhythmia
- Annual surveillance for incomplete or palliative repair (including but not limited to Glenn shunt, Fontan procedure and RV-PA conduit)
- TTE may be unnecessary in a year when cardiac MRI is performed unless clinical indication warrants otherwise

[*Note: See tables below for specific surveillance intervals.]

Infancy is defined as between birth and 1 year of age; childhood from 1-11 years of age; and adolescence from 11 to 21 years of age[56]

Table 1: Unrepaired Lesion Follow-Up‡
‡Blue shading indicates lifetime surveillance interval

Unrepaired Lesion	Surveillance Intervals				
	1-3 months	3-6 months	6-12 months	1-2 years	3-5 years
Aortic Stenosis (AS) and/or aortic			Child Asymptomatic ≥ moderate AS/AR	Child Asymptomatic mild AS/AR	

regurgitation (AR) (See section above for surveillance intervals for infants)					
Bicuspid aortic valve with ≤ mild AS/AR and no aortic dilation in a child					**3 Years**
Atrial septal defect				**Moderate size (6-12mm)**	**Small size (3-6mm)**
Double outlet right ventricular (DORV): with balanced systemic and pulmonary circulation	Infant	Child			
Mitral regurgitation (MR)	Infant with ≥ moderate MR		Infant with mild MR. Child with ≥ moderate MR.		Child with mild MR (2-5 years)
Mitral Stenosis (MS) (See section above for surveillance intervals for infants)		Child with ≥ moderate MS		Child with mild MS	
Congenitally corrected transposition of the Great Arteries (ccTGA)		Infant	**Moderate or greater A-V valve regurgitation**	**< Moderate A-V valve regurgitation**	
Tricuspid regurgitation (TR)		Infant with ≥ moderate TR	Child with ≥ moderate TR	Child with mild TR	
Unrepaired Lesion	**Surveillance Intervals**				
	1-3 months	**3-6 months**	**6-12 months**	**1-2 years**	**3-5 years**
Patent Ductus Arteriosus		Infant		Child	**Adult**
		Infant		Child	

Pulmonary stenosis (PS)				**Adult**	
Coarctation		Infant		Child **Adult**	
Ventricular septal defect (VSD)	Infant with ≥ moderate VSD			Child with non-muscular VSD	Child with small muscular VSD **Adult with any VSD**
Anomalous coronary arteries				**Moderate to large coronary fistula**	**Small coronary fistula or RCA arising from left coronary sinus (2-5 years)**
Subvalvular AS See section above for information on surveillance intervals for stenosis **plus** regurgitation	Infant with any degree of stenosis		Child with ≥ moderate stenosis **Adult with ≥ moderate stenosis**	Child with mild stenosis **Adult with mild stenosis**	
Supravalvular AS		Infant with any degree of stenosis	Child with ≥ moderate stenosis **Adult with ≥ moderate stenosis**	Child with mild stenosis **Adult with mild stenosis**	**2-5 years** **Adult with ≥ moderate stenosis**
Total anomalous pulmonary venous connection (TAPVC)	Prior to planned repair or for change in clinical status and/or development of new signs and symptoms				

Note: Despite surgical or catheter-based procedures, most patients with congenital heart disease are left with disorders or **sequelae** that are known consequences of the reparative intervention. These disorders can include arrhythmias, valvular and myocardial dysfunction, and vascular and non-cardiovascular abnormalities. These sequelae can be categorized as mild, moderate, or severe. Use clinical judgement to assess the nature of the sequelae when adjudicating cases based on the follow-up criteria below.

Table 2: Postprocedural Follow-up‡
‡Blue shading indicates lifetime surveillance interval

Post-procedure: Surgical or Catheter-Based	Surveillance Intervals				
	1-3 months	3-6 months	6-12 months	1-2 years	3-5 years
Post-procedural treatment of AS or AR with repair or replacement	Infant with ≥ moderate AS or AR or LV dysfunction	Infant with ≤ mild AS or AR and no LV dysfunction	Child with ≥ moderate AS or AR	Child with ≤ mild AS or AR	
ASD device closure: no or mild sequelae	Within 1st year	Within 1st year	At 1 year		**2-5 years**
ASD surgical repair: no or mild sequelae			Within 1st year		**2-5 years**
ASD: device closure or surgical repair with residual shunt, valvular or ventricular dysfunction, arrhythmias, or pulmonary hypertension		**3-12 months**			
DORV: no or mild sequelae			Within 1st year	**1-2 Years**	
DORV: valvular or ventricular dysfunction, outflow obstruction, arrythmias, branch pulmonary artery stenosis, presence of RV-PA conduit		**3-12 months**			

Post-procedure: Surgical or Catheter-Based	Surveillance Intervals				
	1-3 months	3-6 months	6-12 months	1-2 years	3-5 years
Tricuspid valve surgery or catheter-based procedure: no or mild sequelae				**1-2 years**	
Tricuspid valve surgery or catheter-based procedure: valvular or ventricular dysfunction or arrhythmias			Child	**Adult**	
Pulmonary Stenosis: no or mild sequelae			Child with moderate or severe sequelae	Child with no or mild sequelae	**Adult**
Coarctation: no or mild sequelae		Within 1st year		**After 1st year**	
PDA: no or mild sequelae				Annually within 1st two years	**Five years after 1st two years***
PDA: post-procedural left PA stenosis or aortic obstruction				**1-2 years**	
Tetralogy of Fallot (ToF): after transcatheter pulmonary valve replacement, with no or mild sequelae	1 month	6 months		**Annually**	

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Post-Procedure: Surgical or Catheter-Based	Surveillance Intervals				
	1-3 months	3-6 months	6-12 months	1-2 years	3-5 years
ToF: patient with conduit dysfunction valvular or ventricular dysfunction, pulmonary artery stenosis, or arrhythmias			6-12 months		
Congenitally corrected transposition on the Great Arteries (ccTGA): no or mild sequelae		Within 1st year		1-2 years	
ccTGA: valvular or ventricular dysfunction, outflow obstruction, ventricular - PA conduit		3-12 months			
d-TGA: no or mild sequelae	Infant with moderate sequelae	Within 1st year		1-2 years	
d-TGA: moderate or greater valvular or ventricular dysfunction, outflow obstruction, branch pulmonary artery stenosis or arrhythmias, presence of RV-PA conduit		3-12 months			

Post-Procedure: Surgical or Catheter-Based	Surveillance Intervals				
	1-3 months	3-6 months	6-12 months	1-2 years	3-5 years
d-TGA: dilated neoaortic root and increasing Z-Score or neoaortic regurgitation				1-2 years	
Truncus Arteriosus (TA): no or mild sequelae	Within 1st year		After 1st year		
TA: moderate or greater truncal stenosis / regurgitation		3-6 months			
TA: residual VSD, RV-PA conduit, branch pulmonary artery obstruction		3-12 months			
VSD: no or mild sequelae or small residual shunt			Within 1st year		2-3 years
VSD: significant residual shunt, valvular or ventricular dysfunction, arrhythmias, or pulmonary hypertension		3-12 months			

<table>
<tr><th rowspan="2">Post-procedure: Surgical or Catheter-Based</th><th colspan="5">Surveillance Intervals</th></tr>
<tr><th>1-3 months</th><th>3-6 months</th><th>6-12 months</th><th>1-2 years</th><th>3-5 years</th></tr>
<tr><td>Anomalous coronary arteries</td><td>Within 1st year</td><td>Infant with or without ventricular or valvular dysfunction

Child or adult with ventricular or valvular dysfunction</td><td></td><td>Annually</td><td></td></tr>
<tr><td>Subvalvular AS
See section above for information on surveillance intervals plus regurgitation</td><td>Infant with ≥ moderate stenosis</td><td>Infant with ≤ mild stenosis</td><td></td><td>Child with ≤ mild stenosis and/or AR
Adult with ≤ mild stenosis and/or AR</td><td></td></tr>
<tr><td>Subvalvular AS
continued</td><td></td><td colspan="2">3-12 months
Child ≥ moderate stenosis
3-12 months
Adult ≥ moderate stenosis</td><td></td><td></td></tr>
<tr><td>Supravalvular AS</td><td></td><td></td><td>Patient with ≥ moderate stenosis</td><td></td><td>2-5 years
Patient with ≤ mild stenosis</td></tr>
<tr><td>Total anomalous pulmonary venous connection</td><td></td><td>Infant with mild or no sequelae</td><td></td><td>Child with mild or no sequelae</td><td>Adult with mild or no sequelae</td></tr>
</table>

***PDA lifetime surveillance applies only to device closure; PDA lifetime surveillance is not indicated for surgical closure.**

BACKGROUND

Transthoracic echocardiography (TTE) uses ultrasound to image the structures of the heart in a real time format, providing 2-dimensional, cross-sectional images. The addition of Doppler ultrasound derives hemodynamic data from flow velocity versus time measurements, as well as from color-coded two-dimensional representations of flow velocities.

TTE's safety and versatility in examining cardiac structure, function, and hemodynamics lends to its utility for numerous indications in children and adults.

TEE (transesophageal echocardiography) widens the scope of utility for echocardiographic imaging, and its indications are covered in a separate guideline.

Abbreviations:

AS	Aortic stenosis
AR	Aortic regurgitation
ASD	Atrial septal defect
BNP	B-type natriuretic peptide or brain natriuretic peptide
CABG	Coronary artery bypass grafting surgery
CAD	Coronary artery disease
ccTGA	Congenitally corrected transposition of the Great Arteries
CMR	Cardiovascular magnetic resonance
CRT	Cardiac resynchronization therapy
CT	Computed tomography
CVA	Cerebrovascular accident
DORV	Double outlet right ventricle
d-TGA	D-Transposition of the Great Arteries
ECG	Electrocardiogram
EF	Ejection fraction
HCM	Hypertrophic cardiomyopathy
HTN	Hypertension
HF	Heart failure
ICD	Implantable cardioverter-defibrillator
LAA	Left atrial appendage
LV	Left ventricular/ventricle
LVEF	Left ventricular ejection fraction
LVH	Left ventricular hypertrophy
MI	Myocardial infarction
MR	Mitral regurgitation
MS	Mitral stenosis
PA	Pulmonary artery
PAC	Premature atrial complex
PDA	Patent ductus arteriosus
PFO	Patent foramen ovale
PS	Pulmonary stenosis
PVC	Premature ventricular contraction
RV	Right ventricular/ventricle
TA	Truncus arteriosus
TAVR	Transcatheter aortic valve replacement
TEE	Transesophageal echocardiogram
TIA	Transient ischemic attack
ToF	Tetralogy of Fallot
TR	Tricuspid regurgitation
TTE	Transthoracic echocardiogram
VAD	Ventricular assist device

VF	Ventricular fibrillation
VSD	Ventricular septal defect
VT	Ventricular tachycardia

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POLICY HISTORY

Date	Summary
April 2023	• Expanded and clarified indications based upon ECG abnormalities • Clarified arrhythmias (premature atrial complexes (PAC)) which do not meet criteria for approval. • Expanded and clarified surveillance imaging criteria for thoracic aortic aneurysm in Turner's syndrome • Added Takotsubo cardiomyopathy to section on surveillance for cardiomyopathy with recovered left ventricular ejection fraction • Expanded indication for screening in suspected cardiac sarcoidosis • Expanded section on post heart transplant surveillance • Added screening in children with sickle cell disease • Expanded section on aortopathic syndromes, cardiovascular disease in pregnancy • Clarified syncope indications • Pulmonary hypertension: added section for annual screening in certain diseases, added indication for repeat following pulmonary embolism evaluate for chronic thromboembolic pulmonary hypertension • Cardiomyopathy: added examples of infiltrative processes, added intervals for repeat testing in different forms of amyloidosis • Added indication for surveillance following radiation therapy • Hypertrophic cardiomyopathy: added statement on imaging related to Camzyos therapy • Clarified surveillance related to exposure to cardiotoxic medication • Added section on COVID • Added section on inflammatory and autoimmune diseases • Added section on neuromuscular disorders • Reorganized Pediatric section for clarity • Added sections on supravalvular and subvalvular AS and total anomalous pulmonary venous connection to congenital heart disease table • Added statement on clinical indications not addressed in this guideline
June 2022	• Within the Hypertrophic Cardiomyopathy section, added To guide therapy
February 2022	• Modified definition of pathological Q waves • Added indications for murmur evaluation • Clarified definition of frequent PVC • Added annual surveillance TTE following palliative procedures in congenital heart disease.

	• Added post op atrial switch for d-TGA surveillance intervals (table) • Screening for PH in sickle cell added • Revised surveillance indications post op prosthetic valve and native valve repair • Expanded guidelines for AS/AR, MS/MR, TR, PS, ASD, TOF, DORV, TGA, TA, and coronary anomalies • Reorganized pediatric indications for clarity • Added section for pediatric hypertension (both initial evaluation and follow-up)

Reviewed / Approved by Clinical Guideline Committee

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