

National Imaging Associates, Inc.*	
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TRANSTHORACIC (TTE) ECHO	
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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. All prior relevant imaging results and the reason that alternative imaging cannot be performed must be included in the documentation submitted.

ADULT PATIENTS - INDICATIONS FOR TRANSTHORACIC ECHOCARDIOGRAPHY (TTE)1

(Indications for <u>pediatric patients</u> follow this section) (Douglas, 2011)

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
- 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
 - ECG evidence of prior MI (pathologic Q waves) defined below:
 - Q wave must be present in at least two contiguous leads
 - → > 40 ms (1 mm) wide
 - → 2 mm deep

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^{1—} Transthoracic (TTE) Echo

Murmur or Click

- Initial evaluation when there is a reasonable suspicion for valvular or structural heart disease such as:
 - •<u>o</u> HHigh 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)
- 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)
- New left bundle branch block (as documented in MD notes and on ECG)

Syncope^{2, 3}

(Doherty, 2017; Shen, 2017)

- 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:
 - Hypertrophic cardiomyopathy
 - o Heart failure
- Exercise-induced syncope

Perioperative Evaluation^{4, 5}

(Fleischer, 2014; Lentine, 2012)

 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 (can be done every 6-12 months) (Nazzareno, 2016), 6 such as:
 - New chest pain

- Worsening shortness of breath
- Syncope
- Increased murmur
- Worsening rales on lung examination
- Evaluation of patients with pulmonary embolism to risk stratify and initiate appropriate therapy⁷ (Sarie, 2016)
- Screening test for pulmonary hypertension in patients with scleroderma or sickle cell anemia.

•

Evaluation of Valvular Function^{2, 8-10}

(Doherty, 2017, 2019; Nishimura, 2014; Otto, 2021)

• 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, 11, 12}

(Bonow, 2020; Doherty, 2017; Lancellotti, 2013)

- 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: 5 and 10 every 3 years after surgery -
- Routine surveillance of surgical <u>bioprosthetic and</u> mechanical valve: -<u>at 10</u> years <u>postoperatively and annually thereafter</u>
- 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
 - O 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, 10, 13

(Doherty, 2017; Otto, 2017, 2021)

- 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, included including but not limited to:
 - o Chest pain
 - o Shortness of breath
 - New or increased murmur on heart examination
- Assessment of stroke post TAVR

Percutaneous Mitral Valve Repair^{2, 10, 11}

(Bonow, 2020; Doherty, 2017; Otto 2021)

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

Closure of PFO or ASD⁸

(Doherty, 2019)

- Pre-procedure evaluation
- Routine follow-up post procedure for device position and integrity (see <u>Background</u> sectionTable 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** indicated¹⁴ (Sachdeva, 2020)

Left Atrial Appendage (LAA) Occlusion⁸ (Doherty, 2019)

Pre-procedure evaluation

Pericardial Disease^{2, 7, 15, 16}

(Chiabrando, 2020; Doherty, 2017; Klein, 2013; Saric, 2016)

- 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 Mass²

(Doherty, 2017)

- 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

Infective Endocarditis (Native or Prosthetic Valves)^{2, 9, 17}

(Doherty, 2017; Habib, 2010; Nishimura, 2014)

- 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
 - o 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 year¹⁷ (Habib, 2010)

Thoracic Aortic Disease 18-23

(Bhave, 2018; Erbel, 2014; Hiratzka, 2010, 2016; Svensson, 2013; Terdjman, 1984)

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 a thoracic aortic aneurysm (defined as ≥ 50% above normal) or dissection, or if an associated high-risk mutation is present
- 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 \geq 4.5 or expanding \geq 0.5 cm/yr
- Patients with Turner's syndrome should undergo imaging to assess for assess bicuspid aortic valve, coarctation of the aorta or dilation of the ascending or thoracic aorta. If the initial imaging is normal and there are no additional risk factors for dissection, imaging can be done every 5 10 years. If an abnormality exists, annual imaging is recommended
- Re-evaluation of known ascending aortic dilation or history of aortic dissection with one
 of the following:
 - New chest pain
 - Shortness of breath
 - Syncope
 - o 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
- Re-evaluation (< 1 yr, generally twice a year) of the size and morphology of the aortic sinuses and ascending aorta in patients with a bicuspid aortic valve with one of the following:
 - o Aortic diameter ≥ 4.5 cm
 - Rapid rate of change in aortic diameter when an annual growth rate of ≥ 0.5 cm is suspected
 - Family history (first-degree relative) of aortic dissection
- Follow-up of aortic disease when there has been no surgical intervention:
 - o Acute dissection: 1 month, 6 months, 12 months, then annually
 - Chronic dissection: annually
- Follow-up post either: Root repair or AVR plus ascending aortic root/arch repair: baseline post-op, then annually²¹ (Svensson, 2013)
- Evaluation of sinus of valsalva aneurysms and associated shunting secondary to rupture (Terdjman, 1984).²² Echo imaging every 4-12 weeks is recommended during pregnancy and 6 months post-partum in patients with ascending aortic dilation²⁴ (Regitz Zagrosek, 2018)

Hypertension (HTN) (Adult)8

(Doherty, 2019)

- 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)²⁵

- 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:
 - o 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^{8, 26-28}

(Doherty, 2019; Nagueh, 2016; Patel, 2013; Yancy, 2013)

- Initial evaluation of suspected heart failure (HF) (systolic or diastolic) based on symptoms, signs, or abnormal test result, including but not limited to:
 - o Dyspnea
 - o 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^{8, 24, 27-30}

(Doherty, 2019; Maddox, 2021; Ommen, 2020; Patel, 2013; Regitz Zagrosek, 2018; Yancy, 2013)

- Initial evaluation of suspected inherited or acquired cardiomyopathy, including but not limited to:
 - Restrictive
 - Infiltrative
 - Dilated
 - Hypertrophic
 - Infective
- 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 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%³¹ (Wilcox, 2020):
 - 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
- 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)³² (Dorbala, 2019)

Hypertrophic Cardiomyopathy (HCM)²⁹

(Ommen, 2020)

- 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 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

Imaging Surveillance for Cardiotoxic Chemotherapy^{33, 34}

(Plana, 2014; Zamorano, 2016)

 TTE is the method of choice for the evaluation of patients prior to cardiotoxic chemotherapy, and subsequently for monitoring and follow-up. The frequency of testing should be left to the discretion of the ordering physician, but generally no more often than at baseline and every 6 weeks thereafter while on active therapy.

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 planned³⁵ (Al-Khatib, 2017)

- 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^{8, 36}

(Doherty, 2019; Stainback, 2015)

- 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
 - o Infection
 - Murmur suggestive of aortic insufficiency
 - Worsening heart failure
- Monitoring annually for rejection in a cardiac transplant recipient

Cardiovascular Disease in Pregnancy^{24, 37}

(Davis, 2020; Regitz Zagrosek, 2018)

- 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
- Syndromes potentially involving the aorta (i.e., Marfan's, Ehlers-Danlos, Loeys-Dietz, or Turner syndrome): for mildly dilated aorta can repeat TTE every 12 weeks; for severely dilated aorta can repeat TTE monthly; can evaluate for 6 months postpartum

Adult Congenital Heart Disease^{14, 38, 39}

(Sachdeva, 2020; Stout, 2019; Warnes, 2008)

- 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 <u>Tables 1 and 2: Adult and Pediatric Congenital</u> <u>Heart Disease Follow-upOverview</u>

Coronary Anomalies¹⁴

(Sachdeva, 2020)

Routine surveillance (2-5 years) in an asymptomatic patient with anomalous right coronary artery from the left aortic sinus

Routine surveillance (2 5 years) in an asymptomatic patient with small coronary fistula and 1-2 years for moderate or large coronary fistula

PEDIATRIC PATIENTS - INDICATIONS FOR TRANSTHORACIC ECHOCARDIOGRAPHY (TTE) (PATIENTS UNDER THE AGE OF 18)⁴⁰

(Campbell, 2014)

- Hypertension (see section: Hypertension (Pediatric))
- Renal failure
- Palpitations, if one:
 - o 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
 - o 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
 - o 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-are become 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
- Suspected pulmonary hypertension
- Signs and symptoms of endocarditis
- Thromboembolic events
 - o Patients on anticoagulants, when required to evaluate for thrombus
 - o Thromboembolic events or stroke (Saric, 2016)⁷
- Systemic hematologic diseases that are associated with cardiac findings
 - Sickle cell disease and other hemoglobinopathies
 - HIV infection
- Chemotherapy or radiation therapy, any one of the following:
 - Cardiotoxic chemotherapy, before and following exposure
 - o Radiation therapy to chest, before and long-term follow-up¹² (Lancellotti, 2013)
- Inflammatory & Autoimmune, including any one of the following:
 - Suspected <u>rRheumatic fFever</u>
 - Systemic lupus erythematosus
 - Takayasu aArteritis
 - Kawasaki dDisease⁴¹
 - 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. (McCrindle, 2017)
- Suspicion of Structural Disease, including any one of the following:
 - o 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)
 - Known or suspected muscular dystrophies associated with congenital heart disease
 - Mitochondrial or metabolic storage disease (e.g., Fabry's disease)
 - 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 <u>sequelae</u>
 - Maternal phenylketonuria
 - Suspected cardiovascular abnormality on fetal echocardiogram

ADULT AND PEDIATRIC CONGENITAL HEART DISEASE FOLLOW-UP¹⁴ ‡* INDICATIONS FOR FOLLOW-UP ECHOCARDIOGRAPHY IN PEDIATRIC PATIENTS

Specific Indications for Follow-Up Echocardiograms in Pediatric Patients
(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 (Hagan, 2017))⁴¹

- Congenital Heart Disease (CHD) with a change in clinical status or to guide therapy
- For follow-up of specific lesions with CHD, see Overview
- 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 arrhythmias
- Kawasaki disease⁴⁰ (McCrindle, 2017)
 - 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.

Adult and Pediatric Congenital Heart Disease Follow-Up¹⁴

[*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(Sachdeva, 2020)
- •
- Infant with any degree of unrepaired AS/AR may have surveillance TTE every 1 4
 weeks as needed
- Infant with any degree of unrepaired MS may have surveillance TTE every 1 4 weeks as needed
- All surgical or catheter based repairs allow evaluation prior to the procedure and postprocedural evaluation (within 30 days)
- 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.]

<u>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 age⁴²</u>

Table 1: Unrepaired Lesion Follow-Up[±]

[‡]Blue shading indicates lifetime surveillance interval

Unrepaired Lesion	1-3 months	36 months	6-12 months	1-2 years	3-5 years
Aortic stenosis (AS) and/or aortic regurgitation (AR)-in-a child (See section above for surveillance intervals for infants)	-	-	Moderate or more AS/AR and increasing aortic size Child Asymptomatic ≥ moderate AS/AR	Stable aortic size (2-3 years)	
Bicuspid aortic valve with ≤ mild AS/AR and no aortic dilation in a child	-				<mark>Х</mark> З years
Atrial septal defect				Moderate size (6-12 mm)	Small size (3-6 mm)
Double outlet right ventricle (DORV): with balanced systemic and pulmonary circulation	<u>Infant</u>	<u>Child</u>			
Mitral regurgitation (MR): asymptomatic	Infant with ≥ moderate MR		Infant with mild MR, Child with ≥ moderate MR	Child with mild MR (2-5 years)	Child with mild MR (2-5 years)
Mitral Stenosis (MS) (See section above for surveillance intervals for infants)	Infant with any MS	Child with ≥ moderate MS		Child with mild MS	
Congenitally corrected transposition of the Great Arteries (ccTGA)		<u>Infant</u>	Moderate or greater A-V valve regurgitation	Moderate A-V valve regurgitation	
Transposition of the great arteries (d-TGA)					
Tricuspid regurgitation (TR): asymptomatic		Infant with ≥ moderate TR	Child & Adult-with ≥ moderate TR	Infant or child Child with mild TR	Adult with mild TR

Unrepaired Lesion	1-3 months	36 months	6-12 months	1-2 years	3-5 years
Patent Ductus Arteriosus		Infant		<u>Child</u>	Adult
Pulmonary stenosis (PS) : asymptomatic		Infant		Child & Adult +	
Coarctation		Infant		Child & Adult +	
Ventricular septal defect (VSD)	Infant with ≥ moderate VSD			Child with VSD in other locationnon -muscular VSD	Child with small mus- ceular VSD+ Adult with with any VSD
Anomalous coronary arteries				Moderate to large coronary fistula	Small coronary fistula or RCA arising from left coronary sinus (2-5 years)
Postprocedure: Surgical or Catheter Based	1-3 months	3-6 months	6-12 months	1-2 years	3-5 years
Postprocedural 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: asymptomatic	×	×	1 year	-	2-5 years
ASD surgical repair: asymptomatic	-	-	×	-	2-5 years

Unrepaired Lesion	1-3 months	36 months	6-12 months	1-2 years	3-5 years
ASD: device closure or surgical repair with residual shunt, valvular or ventricular dysfunction, arrhythmias, or pulmonary hypertension	-	3-12 months	-	-	-
Tricuspid valve surgery or catheter-based procedure: asymptomatic	-	-	-	×	-
Tricuspid valve surgery or catheter based procedure: valvular or ventricular dysfunction or arrhythmias	-	-	Child	Adult	-
Pulmonary stenosis: asymptomatic child	-	-	Moderate or severe sequelae	No or mild sequelae	-
Coarctation: asymptomatic	-	Within the 1st year	-	After the 1st year	-
PDA: asymptomatic	-	-	-	Annually within 2 years	5 years after first 2
PDA: postprocedural left PA stenosis or aortic obstruction	-	-	-	×	-
Tetralogy of Fallot (ToF): asymptomatic after transcatheter pulmonary valve replacement	1 month	6 months	-	Annually	-
ToF: patient with conduit dysfunction, valvular or ventricular dysfunction, pulmonary artery stenosis, or arrhythmias	-	-	×	-	-

Unrepaired Lesion	1-3 months	36 months	6-12 months	1-2 years	3-5 years
VSD: small residual shunt	-	-	*	-	2-3 years
VSD: significant residual shunt, valvular or ventricular dysfunction, arrhythmias, or pulmonary hypertension	-	3-12 months	-	-	-

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	1-3 months	36 months	6-12 months	1-2 years	3-5 years
Postprocedural treatment of AS or AR with repair or replacement	Infant with ≥ moderate AS or AR or LV dys-function	Infant with ≤ mild AS or AR and no LV dys-function	Child with ≥ moderate AS or AR	Child with ≤ mild AS or AR	
ASD device closure: no or mild sequelaeasympto matic	Within 1 st year ^X	Within 1 st year ^X	At 1 year		2-5 years
ASD surgical repair: no or mild sequelaeasympto matic			X <u>Within</u> 1st year		2-5 years
ASD: device closure or surgical repair with residual shunt, valvular or ventricular dysfunction, arrhythmias, or pulmonary hypertension		3-12 mo	nths		
DORV: post- operational			×		
DORV: asymptomatic with no or mild sequelae				×	×

Post-procedure: Surgical or Catheter-Based	1-3 months	36 months	6-12 months	1-2 years	3-5 years
DORV: no or mild			Within 1st	1 – 2 voors	
<u>sequelae</u>			<u>year</u>	<u>1 – 2 years</u>	
DORV: valvular or					
ventricular					
dysfunction,					
outflow					
obstruction,		¥ <u>3 − 12 m</u>			
arrhythmias,					
branch pulmonary					
artery stenosis,					
presence of RV-PA					
conduit Tricuspid valvo					
Tricuspid valve					
surgery or catheter-based					
procedure: <u>no or</u>				V1 2 voors	
mild					
sequelaeasympto					
matic					
Tricuspid valve					
surgery or					
catheter-based					
procedure:					
valvular or			Child	Adult	
ventricular					
dysfunction or					
arrhythmias					
Pulmonary			Child with		
stenosis: <u>no or</u>			mModerat	Child with	
mild			e or	<u>n</u> No or mild	<u>Adult</u>
<u>sequelae</u> asympto			severe	sequelae	
matic child			sequelae		
Coarctation: no or					
<u>mild</u>		Within the 1st		After the 1st	
sequelae asympto		year		year	
matic					
PDA: <u>no or mild</u>				Annually	5 years after first
<u>sequelae</u> asympto				within <u>1st</u>	<u>1st</u>
matic				2 years	2 <u>years*</u>

Post-procedure: Surgical or	1-3 months	36 months	6-12 months	1-2 years	3-5 years
Catheter-Based					
PDA:					
postprocedural				<u> </u>	
left PA stenosis or					
aortic obstruction					
Tetralogy of Fallot					
(ToF):					
asymptomatic					
after					
transcatheter	1 month	6 months		Annually	
pulmonary valve					
replacement <u>, with</u>					
no or mild					
<u>sequelae</u>					
ToF: patient with					
conduit					
dysfunction,					
valvular or			¥ <u>6-12</u>		
ventricular			months		
dysfunction,			<u>IIIOIILIIS</u>		
pulmonary artery					
stenosis, or					
arrhythmias					
Congenitally					
<u>corrected</u>					
transposition of		Within 1st		1 – 2 years	
the Great Arteries		<u>year</u>		1 - 2 years	
(ccTGA): no or					
mild sequelae					
ccTGA: valvular or					
<u>ventricular</u>					
dysfunction,					
<u>outflow</u>		<u>3 – 12 ma</u>			
obstruction,					
ventricular-PA					
conduit					

Post-procedure:					
Surgical or	1-3 months	36 months	6-12	1-2 years	3-5 years
Catheter-Based			months	,	,
Transposition of					
the great arteries					
(d TGA):					
surveillance after				V	
atrial switch				×	
(Mustard or					
Senning					
procedure)					
d-TGA: no or mild	<i>Infant</i> with	Within 1st			
	<u>moderate</u>			<u>1 – 2 years</u>	
<u>sequelae</u>	<u>sequelae</u>	<u>year</u>			
d-TGA: moderate					
or greater					
valvular or					
<u>ventricular</u>					
dysfunction,					
<u>outflow</u>					
obstruction,		<u>3 – 12 ma</u>			
<u>branch</u>					
pulmonary artery					
stenosis or					
arrhythmias,					
presence of RV-					
PA conduit					
d-TGA: dilated					
neoaortic root					
and increasing Z-				<u>1 – 2 years</u>	
score or neoaortic					
<u>regurgitation</u>					
<u>Truncus</u>					
Arteriosus (TA):	Within 1st		After 1st		
no or mild	<u>year</u>		<u>year</u>		
<u>sequelae</u>					
TA: moderate or					
greater truncal		3 – 6 months			
stenosis/		<u>3 0 months</u>			
regurgitation					

Post-procedure: Surgical or Catheter-Based	1-3 months	36 months	6-12 months	1-2 years	3-5 years
TA: residual VSD, RV-PA conduit, branch pulmonary artery obstruction		<u>3 – 12 m</u>	onths		
VSD: no or mild sequelae or small residual shunt			Within 1st year		<u>2 – 3 years</u>
VSD: small residual shunt	-	-	×	-	2-3 years
VSD: significant residual shunt, valvular or ventricular dysfunction, arrhythmias, or pulmonary hypertension		3-12 3 – 12	months		
Anomalous coronary arteries	Within 1 st year	Infant with or without ventricular or valvular dysfunction Child or adult with ventricular or valvular dysfunction		<u>Annually</u>	

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

Double Outlet Right Ventricle, Transposition of the Great Arteries, and Truncus Arteriosus Unrepaired:

- Routine surveillance (1-3 months) in an asymptomatic infant
- Routine surveillance (3-6 months) in an asymptomatic child

Post-procedure: Surgical or Catheter-based

- Routine surveillance if asymptomatic with mild sequelae at 6 months, 1-2 years, and 3-5
 years
- Routine surveillance if valvular or ventricular dysfunction, outflow tract obstruction, branch pulmonary artery stenosis, or arrhythmias at 3-12 months and 1-2 years

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.

OVERVIEW

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
СТ	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 <u>/ventricle</u>
LVEF	Left ventricular ejection fraction
LVH	<u>-Left ventricular hypertrophy</u>
MI	Myocardial infarction
MR	Mitral regurgitation
MS	Mitral stenosis
PA	Pulmonary artery
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 PVC
VSD	Ventricular septal defect
VT	Ventricular tachycardia

Premature ventricular contraction

POLICY HISTORY

Date	Summary
<u>June 2022</u>	 Within the Hypertrophic Cardiomyopathy section, added To guide therapy
February 20223	 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)
March 2021	 Added the following to prosthetic heart valves: Routine surveillance of surgical bioprosthetic valve: 5 and 10 years after surgery and then annually Routine surveillance of surgical mitral valve repair: 1 year and then every 2-3 years Added section on heart failure with recovered EF under cardiomyopathy Heart failure with recovered left ventricular ejection fraction defined as:
	 3) second measurement of LVEF >40% (Wilcox, 2020) Repeat echocardiogram every 6 months until 12-18 months of heart failure with a

recovered EF, then 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 Created a separate section on hypertrophic cardiomyopathy (HCM) with the following additions: o 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 (genotype-positive and phenotype-negative): Children 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 from genotype-positive families and families with early onset disease every 1-2 years All other children every 2-3 years Adults every 3-5 years Additional information added under follow-up with Kawasaki's disease 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. August 2020 For prosthetic valve with TTE specified routine surveillance as ≥ 3 yrs. (after valve implantation) of prosthetic valve or native valve repair Valvular dysfunction defined as including but not limited to: Chest pain Shortness of breath New or increased murmur on heart examination Clarified syncope can be either known or suspected Further definition of ECG evidence of prior MI (pathologic Q waves) defined as below: o > 40 ms (1 mm) wide

	 > 2 mm deep[
March 2020	 Added general information section as Introduction which outlines requirements for documentation of pertinent office notes by a licensed clinician, and inclusion of laboratory testing and relevant imaging results for case review. Added clarification of abnormal EKG to include evidence of prior myocardial infarction, including pathologic Q waves Added clarification of indication for frequent PVCs to include greater than 30 per hour on remote monitoring Added clarification that annual evaluation of bioprosthetic heart valves older than 10 years, to replace prosthetic heart valves Added statement about routine surveillance of PFO not indicated Separated sections on pericardial disease and cardiac source of emboli/ cardiac mass Added clarification cardiac source of emboli to include the following: Embolic source in patients with recent transient ischemic attack (TIA), stroke, or peripheral vascular emboli Added clarification of cardiac mass to include the following: evaluation of mass and re-evaluation when findings would alter therapy

- Added clarification of hypertensive heart disease to include asymptomatic left ventricular hypertrophy, cardiomegaly, or evidence of clinical heart failure
- Added indication for suspected cardiac amyloid to monitor disease progression and/or response to therapy, and to guide initiation and management of anticoagulation (TEE may be preferred)
- Added clarification of imaging for surveillance for cardiotoxic chemotherapy to include the following: TTE is the method of choice for the evaluation of patients prior to cardiotoxic chemotherapy, and subsequently for monitoring and follow up. The frequency of testing should be left to the discretion of the ordering physician, but generally no more often than at baseline and every 6 weeks thereafter.
- Added separate section on indications for TTE during pregnancy to include the following:
 - Valvular stenosis: mild-can evaluate each trimester and prior to deliver; moderate to severe can evaluate monthly
 - Valvular regurgitation: mild-moderate can evaluate each trimester and prior to delivery; severe regurgitation can evaluate monthly
 - Pre-pregnancy evaluation with mechanical or bioprosthetic heart valves if not done within the previous year
 - Prior postpartum cardiomyopathy: can repeat 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
 - Syndromes potentially involving the aortia (I,e, Marfan's, Ehlers-Danlos, Loeys-Dietz, or Turner syndrome): for mildly dilated aorta can repeat TTE every 12 weeks; for severely dilated aorta can repeat TTE monthly.
 Continued evaluation allowable for 6 months postpartum
- Extensive update to adult and pediatric congenital heart disease sections to include the following:
 - Evaluation prior to surgical or catheter-based procedure and postprocedural evaluation (within 30 days)
 - Evaluation after any surgical or catheter-based repair (3-12 months) for a patient with heart failure symptoms

	 Complete chart added to include timing of TTE follow-up in infants, children, and adults based on the lesion present and whether the lesion was unrepaired or surgical or catheter-based repair had been performed Added separate section on follow-up of patients with double outlet right ventricle, transposition of the great arteries, and truncus arteriosus Removed chart and background information regarding physiologic stages of adult CHD Added separate section for coronary anomalies to include the following: Routine surveillance (2-5 years) in an asymptomatic patient with anomalous right coronary artery from the left aortic sinus Routine surveillance (2-5 years) in an asymptomatic patient with small coronary fistula and 1-2 years for moderate or larger coronary fistula and 1-2 years for moderate or larger coronary fistula Updates to TTE in pediatric patients include the following: Clarification of congenital heart disease with a change in clinical status with the addition of "or to guide therapy" Added annual surveillance in a child with normal prosthetic mitral valve function and no LV dysfunction Added surveillance (3-12 months) in a child with prosthetic mitral valve and ventricular dysfunction and/or arrhythmias Updated and added new references
November 2019	Added CPT code +93356
July 2019	 Added indication for hypotension of suspected cardiac etiology Removed indication for respiratory failure or hypoxemia of uncertain etiology Clarification of murmur indication with "when there is a reasonable suspicion of valvular heart disease such as high grade, holosystolic, continuous, or diastolic murmur" Clarified frequent PVCs as greater than 30 per hour Added indication for unevaluated left bundle branch block Added indication for exercise induced syncope For perioperative evaluation for solid organ transplantation, added annual study prior to transplantation Removed indication for re-evaluation (<1 yr) in patients with moderate or severe aortic stenosis, who will be subjected to

- increased hemodynamic demands (e.g. noncardiac surgery, pregnancy)
- Removed tertiary syphilis or Takayasu's Arteritis indication
- Pulmonary hypertension:
 - Clarified re-evaluation for a change in clinical status or cardiac exam, or to guide therapy (every 6 - 12 months, or more frequently to guide therapy). Annual indication removed.
 - Screening for scleroderma added
- Removed indications for history of rheumatic heart disease and exposure to medications that could result in valvular heart disease
- Added mild valvular regurgitation as an indication for testing every 3 years
- Added indication for annual evaluation of prosthetic heart valves older than 10 years
- In depth indications for HOCM
- LVAD and transplant indications added
- Removed chart on specific chemotherapeutic agents
- Added detailed indications for adult congenital heart disease and serial follow up
- Removed indications for presyncope for pediatric patients
- Revised murmur indication in pediatric patients with more criteria for pathologic murmur
- Added definitions of age groups for pediatric patients (infancy, childhood, and adolescence)

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Reviewed / Approved by NIA Clinical Guideline Committee

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ADDITIONAL RESOURCES

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Reviewed / Approved by NIA Clinical Guideline Committee

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