

National Imaging Associates, Inc.*	
Clinical guideline	Original Date: October 2009
TRANSTHORACIC (TTE) ECHO	
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93308, +93320, +93321, +93325, +93356	
Guideline Number: NIA_CG_067	Implementation Date: January 20212

#### **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)

(Indications for pediatric patients follows 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:
  - o Mmedications, dehydration, or infection
- ECG evidence of prior MI (pathologic Q waves) defined below:
  - > 40 ms (1 mm) wide
  - o > 2 mm deep
  - > 25% of depth of QRS complex

#### **Murmur or Click**

- Initial evaluation when there is a reasonable suspicion for valvular or structural heart disease such as:
  - High grade ≥ 3/6

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<sup>1—</sup> Transthoracic (TTE) Echo

- o Holosystolic
- o Continuous
- Diastolic

## **Arrhythmias**

- Frequent premature ventricular contractions (PVCs, greater than 30 per hour on remote monitoring)
- 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

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

## **Perioperative Evaluation**

(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), 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 therapy (Saric, 2016)
- Screening test for pulmonary hypertension in patients with scleroderma

#### **Evaluation of Valvular Function**

(Doherty, 2017, 20198; Nishimura, 2014; Otto, 20210)

• 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 month
- Re-evaluation after starting medication in patients with low flow/low gradient severe aortic stenosis

## **Native Valvular Regurgitation with TTE**

(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 <u>valvular</u> aortic regurgitation
- Re-evaluation of asymptomatic patient every 6 12 months with severe mitral regurgitation

#### Prosthetic Valves with TTE

- 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 years after surgery and then annually
- Routine surveillance of surgical mitral valve repair: 1 year post-op and then every 2-3
  years
- Routine surveillance (≥ 3 yrs. after valve implantation) of prosthetic valve or native valve repair
- 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
- Annual evaluation of bioprosthetic heart valves older than 10 years

#### **Transcatheter Heart Interventions**

Transcatheter Aortic Valve Replacement (TAVR) (Doherty, 2017; Otto, 2017, Otto-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 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

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

- Pre-procedure evaluation
- Reassessment for degree of MR and left ventricular function (1, 6 months, and annually 12 months post procedure, and then annually to 5 yrs.)

## Closure of PFO or ASD (Doherty, 2019)

Pre-procedure evaluation

- Routine follow-up post procedure for device position and integrity (see Background section)
- Evaluation for clinical concern for infection, malposition, embolization, or persistent shunt
- Routine surveillance of an asymptomatic patient with a PFO is **not** indicated (Sachdeva, 2020)

# Left Atrial Appendage (LAA) Occlusion (Doherty, 2019)

Pre-procedure evaluation

#### **Pericardial Disease**

(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 Mass**

(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)

(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 ECKG
- 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 (Habib, 2010)

## **Thoracic Aortic Disease**

(Bhave, 2018; Erbel, 2014; Hiratzka, 2010, 2016; Hiratzka, 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 2<sup>nd</sup> degree relatives is reasonable
- Six-month follow-up after initial finding of a dilated thoracic aorta
- Annual <u>follow-follow-</u>up of enlarged thoracic aorta that is above top normal for age, gender, and body surface area
- Biannual (twice/year) follow-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
- Patients with Turner's syndrome should undergo imaging to assess for 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

- Screening of first-degree relatives of patients with a 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
- 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 <u>4-one</u> of the following:
  - Aortic diameter ≥ 4.5 cm
  - Rapid rate of change in aortic diameter when an annual growth rate of ≥ 0.5 cm is suspected.
  - o 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 annually (Svensson, 2013)
- Evaluation of sinus of valsalva aneurysms and associated shunting secondary to rupture (Terdjman, 1984). Echo imaging every 4-12 weeks is recommended during pregnancy and 6 months post-partum in patients with ascending aortic dilation (Regitz-Zagrosek, 2018)

### **Hypertension**

(Doherty, 20198)

- Initial evaluation of suspected hypertensive heart disease including but not limited to the following:
  - Left ventricular hypertrophy on E<u>C</u>KG
  - Cardiomegaly
  - Evidence of clinical heart failure

#### **Heart Failure**

(Doherty, 20198; 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:
  - 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

(Doherty, 201<u>9</u>8; <u>Maddox, 2021<sub>7</sub>; Ommen, 2020<sub>7</sub>; Gersh, 2011;</u> Patel, 2013; Regitz-Zagrosek, 2018; Yancy, 2013)

- Initial evaluation of suspected inherited or acquired cardiomyopathy, including but not limited to:
  - Restrictive
  - Infiltrative
  - 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 with recovered left ventricular ejection fraction defined as (must meet all 3 criteria):
  - 1) Documentation of a decreased LVEF <40% at baseline</li>
  - 2) ≥10% absolute improvement in LVEF
  - 3) A second measurement of LVEF >40% (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) (Dorbala, 2019)

## Hypertrophic Cardiomyopathy (HCM)

(Ommen, 2020Gersh, 2011)

Initial evaluation of suspected HCM

- Re-evaluation of patients with HCM with <u>a change in clinical status or a new clinical</u> <u>event new symptoms including but not limited to the following:</u>
- Chest pain
- Shortness of breath
- Palpitations
- Syncope
- 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 years for symptomatically stable patients 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
- Re evaluation of clinically unaffected patients with a first-degree relative with HCM every 5 yearsScreening 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

## **Imaging Surveillance for Cardiotoxic Chemotherapy**

(<del>Maleszewski, 2018; Plana, 2014; Zamorano, 2016)</del>

• TTE is the method of choice for the evaluation of patients prior to cardiotoxic chemotherapy, and subsequently for monitoring and follow-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 planned (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

(Doherty, 20198; 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
  - Infection
  - Murmur suggestive of aortic insufficiency
  - Worsening heart failure
- Monitoring <u>annually</u> for rejection in a cardiac transplant recipient

## **Cardiovascular Disease in Pregnancy**

(Davis, 2020; Regitz-Zagrosek, 2018)

- Valvular stenosis÷
  - Mild- can <u>be</u> evaluate<u>d</u> 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 <u>be</u> evaluate<u>d</u> 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 for for months postpartum

## **Adult Congenital Heart Disease**

(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:
  - o Chest Pain
  - Shortness of breath
  - New or increased murmur on physical exam
- Evaluation prior to surgical or transcatheter procedure
- For follow-tollow-up of specific lesions, see Overview

## **Coronary Anomalies**

(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)

(Campbell, 2014)

- Hypertension
- Renal failure
- Palpitations, if one:
  - Family history at age < 50 of either:</li>
    - 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
  - o Abnormal ECG
  - o Family history with unexplained sudden death or cardiomyopathy
- Syncope, if any of the followingone of:
  - Abnormal ECG
  - Exertional syncope
  - Family history at age < 50 of either one:</li>
    - 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
  - o 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 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 electrocardiogram (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
  - Thromboembolic events or stroke (Saric, 2016)
- 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:
  - o Cardiotoxic chemotherapy, before and following exposure
  - Radiation therapy to chest, before and long long term follow follow (Lancellotti, 2013)
- Inflammatory & Autoimmune, including any one of the following:
  - Suspected Rheumatic Fever
  - Systemic lupus erythematosus
  - Takayasu Arteritis
  - Kawasaki Disease (<u>McCrindle, 2017</u>Newburger, 2004)
- 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, Allagille syndrome, chromosomal abnormality associated with cardiovascular disease)
  - o 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 <u>first-first-</u>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

#### 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 (Hagain, 2017)) The guidelines for adult congenital heart disease (Stout, 2018) are not intended to be used for patients under 18 years of age.

- Congenital Heart Disease (CHD) with a change in clinical status or to guide therapy
- For fellow-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 d

  isease (McCrindle, 2017);
  - o \_\_Uupon diagnosis, 1-2 two-weeks later, and 4 to 6 weeks after diagnosis-
  - For patients with important and evolving If any coronary artery abnormalities during the acute illnessare present, 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. as clinically indicated (Newburger, 2004)
- Periodic screening of children of patients with hypertrophic cardiomyopathy every 12-18 months starting by age 12 or earlier if a growth spurt or signs of puberty are evident and/or when there are plans for engaging in intensive competitive sports or there is a family history of sudden cardiac death (Gersh, 2011)

#### **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:**

## Adult and Pediatric Congenital Heart Disease Follow-Up (Sachdeva, 2020)

- 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

Unrepaired Lesion	1-3 months	3- 6 months	6-12 months	1-2 years	3-5 years
Aortic stenosis (AS) and/or aortic regurgitation (AR) in a child	-		Moderate or more AS/AR and increasing aortic size	Stable aortic size (2-3 years)	
Bicuspid aortic valve with ≤ mild AS/AR and no aortic dilation in a child	-				Х
Atrial septal defect				Moderate size (6-12 mm)	Small size (3-6 mm)
Mitral regurgitation (MR): asymptomatic	Infant with ≥ moderate MR		Infant with mild MR, Child with ≥ moderate MR		th mild MR years)
Unrepaired Lesion	1-3 months	3-6 months	6-12 months	1-2 years	<del>3-5 years</del>

Mitral Stenosis (MS)	Infant with any MS	Child with ≥ moderate MS		Child with mild MS	
Tricuspid regurgitation (TR): asymptomatic		Infant with ≥ moderate TR	Child & Adult with ≥ moderate TR	Infant or child with mild TR	Adult with mild TR
Patent Ductus Arteriosus		Infant			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	Child with small muscular VSD; Adult
				location	with any VSD
Postprocedure: Surgical or Catheter-Based	1-3 months	3- 6 months	6-12 months	1-2 years	•
-	1-3 months  Infant with ≥ moderate AS or AR or LV dysfunction		_		VSD
Postprocedural treatment of AS or AR with repair or	Infant with ≥ moderate AS or AR or LV	months  Infant -with ≤ mild AS or AR and no LV	months  Child with  ≥  moderate	1-2 years  Child with ≤ mild AS	VSD
or Catheter-Based  Postprocedural treatment of AS or AR with repair or replacement  ASD device closure:	Infant with ≥ moderate AS or AR or LV dysfunction	months Infant -with ≤ mild AS or AR and no LV dysfunction	months  Child with  ≥  moderate  AS or AR	1-2 years  Child with ≤ mild AS	VSD 3-5 years

Unrepaired Lesion	1-3 months	3-6 months	6-12 months	1-2 years	<del>3-5 years</del>
MS or MR	Infant with ≥moderate MS or MR	Infant with mild MS or MR	Child with ≥moderate MS or MR	Child with mild MS or MR	-
Tricuspid valve surgery or catheter-based procedure: asymptomatic				Х	
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				Х	
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			X		
VSD: small residual shunt			Х		2-3 years
VSD: significant residual shunt, valvular or ventricular dysfunction,		3-12 months			

arrhythmias, or pulmonary hypertension			

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**

<u>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.</u>

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

CABG Coronary artery bypass grafting surgery

CAD Coronary artery disease

CMR Cardiovascular magnetic resonance
CRT Cardiac resynchronization therapy

CT Computed tomography ECG Electrocardiogram

HCM Hypertrophic cardiomyopathy

HF Heart failure

ICD Implantable cardioverter-defibrillator

LV Left ventricular

LVEF Left ventricular ejection fraction

MI Myocardial infarction
MR Mitral regurgitation
MS Mitral stenosis

PDA Patent ductus arteriosus
PFO Patent foramen ovale
PS Pulmonary stenosis

TAVR Transcatheter aortic valve replacement

TEE Transesophageal echocardiogram

TIA Transient ischemic attack

ToF Tetralogy of Fallot
TR Tricuspid regurgitation

TTE Transthoracic echocardiogram

PVC Premature ventricular contraction

VSD Ventricular septal defect VT Ventricular tachycardia

## **POLICY HISTORY**:

I OLICI IIISTORI	
<u>Date</u>	Summary
<b>March 2021</b>	<ul> <li>Added the following to prosthetic heart valves:</li> </ul>
	<ul> <li>Routine surveillance of surgical bioprosthetic valve: 5</li> </ul>
	and 10 years after surgery and then annually
	<ul> <li>Routine surveillance of surgical mitral valve repair: 1</li> </ul>
	year and then every 2-3 years
	<ul> <li>Added section on heart failure with recovered EF under</li> </ul>
	cardiomyopathy
	<ul> <li>Heart failure with recovered left ventricular ejection</li> </ul>
	fraction defined as:
	1) Documentation of a decreased LVEF <40% at
	baseline; AND
	<ul> <li>2) ≥10% absolute improvement in LVEF; AND</li> </ul>
	<ul><li>3) second measurement of LVEF &gt;40% (Wilcox,</li></ul>
	2020)
	Repeat echocardiogram every 6 months
	until 12-18 months of heart failure with

a recovered EF, then then annually for 2 vears, 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: 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: > 40 ms (1 mm) wide

	<del>,</del>
	> 2 mm deep[
	<ul><li>&gt; 25% of depth of QRS complex</li></ul>
	<ul> <li>Added 12 months to the following statement: Follow up of</li> </ul>
	aortic disease when there has been no surgical intervention:
	<ul> <li>Acute dissection: 1 month, 6 months, 12 months, then</li> </ul>
	<u>annually</u>
	<ul><li>Chronic dissection: annually</li></ul>
	<ul> <li>For heart failure removed the requirement for a clear</li> </ul>
	precipitating change in medication or diet.
	<ul> <li>Further defined signs/symptoms suggestive of VAD-related</li> </ul>
	complications as including but not limited to:
	O TIA or stroke
	O Infection
	<ul> <li>Murmur suggestive of aortic insufficiency</li> </ul>
	<ul> <li>Worsening heart failure</li> </ul>
	<ul> <li>Known adult congenital heart disease with a change in clinical</li> </ul>
	status or cardiac exam including but not limited to:
	<ul> <li>Chest Pain</li> </ul>
	<ul> <li>Shortness of breath</li> </ul>
	<ul> <li>New or increased murmur on physical exam</li> </ul>
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- 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 1<sup>st</sup> and 2<sup>nd</sup> 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)

	<ul> <li>Evaluation after any surgical or catheter-based repair</li> </ul>
	(3-12 months) for a patient with heart failure
	<u>symptoms</u>
	<ul> <li>Complete chart added to include timing of TTE follow-</li> </ul>
	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
	<ul> <li>Added separate section on follow-up of patients with</li> </ul>
	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:
	<ul> <li>Routine surveillance (2-5 years) in an asymptomatic</li> </ul>
	patient with anomalous right coronary artery from the
	left aortic sinus
	<ul> <li>Routine surveillance (2-5 years) in an asymptomatic</li> </ul>
	patient with small 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
	opuated and added new references
November 2019	Added CPT code +93356
November 2013	Added CF1 Code +955550
July 2010	a Added indication for hymotopsion of suspected conding
<u>July 2019</u>	Added indication for hypotension of suspected cardiac     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
	<ul> <li>Added indication for exercise induced syncope</li> </ul>

- 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)

# Review date: July 2019 Review Summary:

- 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)
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- 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)

## November 2019

Added CPT code +93356

#### Review Date: March 1, 2020

#### **Review Summary:**

- 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 reevaluation 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 1<sup>st</sup> and 2<sup>nd</sup> 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
- 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

## **Review Date:** August 2020

## **Review Summary:**

- 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:
  - > 40 ms (1 mm) wide
  - → 2 mm deep[
  - > 25% of depth of QRS complex
- Added 12 months to the following statement: 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
- For heart failure removed the requirement for a clear precipitating change in medication or diet.
- Further defined signs/symptoms suggestive of VAD related complications as including but not limited to:
  - TIA or stroke
  - → Infection
  - Murmur suggestive of aortic insufficiency
  - Worsening heart failure
- Known adult congenital heart disease with a change in clinical status or cardiac examincluding but not limited to:
  - Chest Pain
  - Shortness of breath
  - New or increased murmur on physical exam

## 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:
    - 1) Documentation of a decreased LVEF <40% at baseline; AND</p>
    - 2) ≥10% absolute improvement in LVEF; AND
    - 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
- <u>Created a separate section on hypertrophic cardiomyopathy (HCM) with the following additions:</u>
  - 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:
    - <u>Children from genotype-positive families and families with early onset</u> <u>disease every 1-2 years</u>
    - 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.

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