New 2024

American College of Radiology
ACR Appropriateness Criteria®

Evaluation of Coronary Artery Anomalies

Variant: 1 Adult. Suspected coronary artery anomaly. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRA coronary arteries without and with IV contrast	Usually Appropriate	O
MRA coronary arteries without IV contrast	Usually Appropriate	O
CTA coronary arteries with IV contrast	Usually Appropriate	☢☢☢
US echocardiography transthoracic resting	May Be Appropriate	O
Arteriography coronary	May Be Appropriate	☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
US echocardiography transthoracic stress	Usually Not Appropriate	O
MRA chest with IV contrast	Usually Not Appropriate	O
MRA chest without and with IV contrast	Usually Not Appropriate	O
MRA chest without IV contrast	Usually Not Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Not Appropriate	O
MRI heart function and morphology without IV contrast	Usually Not Appropriate	O
MRI heart function with stress without and with IV contrast	Usually Not Appropriate	O
MRI heart function with stress without IV contrast	Usually Not Appropriate	O
CT chest with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without IV contrast	Usually Not Appropriate	☢☢☢
CT coronary calcium	Usually Not Appropriate	☢☢☢
CTA chest with IV contrast	Usually Not Appropriate	☢☢☢
CTA chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT heart function and morphology with IV contrast	Usually Not Appropriate	☢☢☢☢
Rb-82 PET/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢

Variant: 2 Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging.

Procedure	Appropriateness Category	Relative Radiation Level
Arteriography coronary	Usually Appropriate	☢☢☢
CTA coronary arteries with IV contrast	Usually Appropriate	☢☢☢
US echocardiography transthoracic resting	May Be Appropriate (Disagreement)	O
US echocardiography transthoracic stress	May Be Appropriate	O
MRA coronary arteries without and with IV contrast	May Be Appropriate	O
MRA coronary arteries without IV contrast	May Be Appropriate	O
MRI heart function and morphology without and with IV contrast	May Be Appropriate	O
MRI heart function and morphology without IV contrast	May Be Appropriate	O
MRI heart function with stress without and with IV contrast	May Be Appropriate	O
MRI heart function with stress without IV contrast	May Be Appropriate	O

CTA chest with IV contrast	May Be Appropriate	☢☢☢
Rb-82 PET/CT MPI rest and stress	May Be Appropriate (Disagreement)	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	May Be Appropriate	☢☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
MRA chest with IV contrast	Usually Not Appropriate	O
MRA chest without and with IV contrast	Usually Not Appropriate	O
MRA chest without IV contrast	Usually Not Appropriate	O
CT chest with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without IV contrast	Usually Not Appropriate	☢☢☢
CT coronary calcium	Usually Not Appropriate	☢☢☢
CTA chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT heart function and morphology with IV contrast	Usually Not Appropriate	☢☢☢☢

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Summary of Literature Review

Introduction/Background

Anatomy of coronary arteries can vary significantly among individuals. The incidence of coronary artery anomalies (CAAs) is reported as approximately 1% in the general population, according to angiographic and autopsies studies [1]. Although patients can often be asymptomatic, anomalies can be the cause of symptoms in some patients. Accurate classification and diagnosis can guide treatment planning and management. The outcomes of patients with CAAs are variable, however, studies suggest that CAAs account for up to 61% sudden cardiac death in young military recruits [2,3]. Initial imaging should help characterize the anomaly in terms of the vessel from which the artery arises, location of ostium, vessel course, and terminus and its relationship to adjacent cardiovascular structures. Proper characterization of CAAs is necessary for preoperative planning and optimizing the approach for endovascular or surgical treatment.

The purpose of this document is to provide guidance on imaging for CAAs for both diagnosis and treatment planning. For guidance on imaging patients with acute or chronic chest pain please see the ACR Appropriateness Criteria® topics on "Chest Pain-Possible Acute Coronary Syndrome" [4], "Chronic Chest Pain-High Probability of Coronary Artery Disease" [5], and "Chronic Chest Pain-Noncardiac Etiology Unlikely: Low to Intermediate Probability of Coronary Artery Disease" [6]. For guidance on imaging congenital or acquired heart disease including anomalous coronary arteries (ACAs) in a child please see the ACR Appropriateness Criteria® topic on "Congenital or Acquired Heart Disease" [7].

Initial Imaging Definition

Initial imaging is defined as imaging at the beginning of the care episode for the medical condition defined by the variant. More than one procedure can be considered usually appropriate in the initial imaging evaluation when:

- There are procedures that are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care)

OR

- There are complementary procedures (ie, more than one procedure is ordered as a set or simultaneously where each procedure provides unique clinical information to effectively manage the patient's care).

Discussion of Procedures by Variant

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.

Initial imaging is used to identify and characterize a CAA in terms of origin of vessel, location of ostium, vessel course, and terminus and its relationship to adjacent cardiovascular structures.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
A. Arteriography coronary

Invasive catheter angiography can assess anatomy of the coronary arteries and delineate the origin, course, and termination of the coronary arteries. CAAs have a prevalence ranging up to 2.1% by catheter angiography (8). The largest angiographic series of 126,595 patients by Yamanaka and Hobbs [8] reported a 1.3% incidence of ACA. However, it may not be suitable as the first-line test for assessment of CAAs because there are other alternative imaging options.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
B. CT chest with IV contrast

There is no relevant literature to support the use of nongated chest CT with intravenous (IV) contrast for evaluation of CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
C. CT chest without and with IV contrast

There is no relevant literature to support the use of nongated chest CT without and with IV contrast for evaluation of CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
D. CT chest without IV contrast

There is no relevant literature to support the use of nongated chest CT without IV contrast for evaluation of CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
E. CT coronary calcium

There is no relevant literature to support the use of CT coronary calcium for evaluation of CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
F. CT heart function and morphology with IV contrast

There is no relevant literature to support the use of CT heart function and morphology with IV contrast for evaluation of CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
G. CTA chest with IV contrast

There is no relevant literature to support the use of CT angiography (CTA) chest with IV contrast for evaluation of CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
H. CTA chest without and with IV contrast

There is no relevant literature to support the use of nongated chest CTA chest without and with IV contrast for evaluation of CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
I. CTA coronary arteries with IV contrast

Coronary CTA (CCTA) can delineate variations in course, origin of coronary arteries, and their relationship to adjacent structures. The reported prevalence of ACAs is higher with CCTA than invasive catheter angiogram (7.9% versus 2.1%) [9]. CCTA allows assessment of anomalous origin and course of coronary arteries as well as delineation of the location and number of ostia [10,11]. The course of a coronary artery, through the aortic wall (transmural), through the myocardium (myocardial bridging), and intraseptal, prepulmonic, retroaortic, and/or interarterial have been reported on CCTA examinations [12].

Multiple national and international societies have issued recommendations regarding the role of CCTA in the context of ACA delineation [13-15].

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
J. MRA chest with IV contrast

There is no relevant literature to support the use of MR angiography (MRA) chest with IV contrast for evaluation of suspected CAAs. MRA chest with IV contrast encompasses assessment of the entire chest and larger vessel vasculature.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
K. MRA chest without and with IV contrast

There is no relevant literature to support the use of MRA chest without and with IV contrast for evaluation of suspected CAAs. MRA chest with IV contrast encompasses assessment of the entire chest and larger vessel vasculature.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
L. MRA chest without IV contrast

There is no relevant literature to support the use of MRA chest without IV contrast for evaluation of suspected CAAs. MRA chest with IV contrast encompasses assessment of the entire chest and larger vessel vasculature.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
M. MRA coronary arteries without and with IV contrast

MRA of the coronary arteries without and with IV contrast is specifically targeted for evaluating the coronaries with a small field of view and echocardiogram gating. This technique can assess origin and course of the proximal coronary arteries; the spatial resolution may limit the assessment of the distal arterial course. Like other cross-sectional modalities, the relationship of coronary arteries to adjacent cardiovascular structures can be assessed. Flow techniques can also define degree of shunting with coronary cameral fistulae. Several societies have issued recommendations for MRA of the coronary arteries as screening tools [13,15].

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
N. MRA coronary arteries without IV contrast

There are limited data on the use of MRA for assessment of CAAs in the adult population. A small study by Albrecht et al [16], primarily in a pediatric patient population, showed high accuracy and sensitivity (92%) for noncontrast MRA of the coronary arteries to assess coronary anomalies. White et al [17] found that coronary MRA is a useful adjunctive technique in the evaluation of the relationship of anomalous coronary arteries to the great arteries.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
O. MRI heart function and morphology without and with IV contrast

There is no relevant literature to support the use of MRI heart function and morphology without and with IV contrast for evaluation of suspected CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
P. MRI heart function and morphology without IV contrast

There is no relevant literature to support the use of MRI heart function and morphology without IV contrast for evaluation of suspected CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
Q. MRI heart function with stress without and with IV contrast

There is no relevant literature to support the use of MRI heart function with stress without and with IV contrast for evaluation of suspected CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
R. MRI heart function with stress without IV contrast

There is no relevant literature to support the use of MRI heart function with stress without IV contrast for evaluation of suspected CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
S. Rb-82 PET/CT MPI rest and stress

There is no relevant literature to support the use of Rb-82 PET/CT heart for evaluation of suspected CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
T. SPECT or SPECT/CT MPI rest and stress

There is no relevant literature to support the use of single-photon emission computed tomography (SPECT) or SPECT/CT myocardial perfusion imaging (MPI) rest and stress for evaluation of suspected CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
U. US echocardiography transesophageal

There is no relevant literature to support the use of US echocardiography transesophageal for evaluation of suspected CAAs.

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.

V. US echocardiography transthoracic resting

Transthoracic echocardiography (TTE) allows for structural assessment of the heart. Location of coronary artery ostia can be identified for some patients. TTE may be a tool for screening a diverse set of cardiac abnormalities, one study found 1 of 336 rugby players to have CAA [18]. One Chinese study suggests that diagnosis of anomalous origin of left coronary artery from pulmonary artery is as high as 68.8% with TTE [19]. Diagnostic accuracy of this study can depend on operator proficiency and patient factors with a reported diagnostic sensitivity and specificity of ostial identification of 100% [19].

Variant 1: Adult. Suspected coronary artery anomaly. Initial imaging.
W. US echocardiography transthoracic stress

There is no relevant literature to support the use of TTE stress for evaluation of suspected CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging.

Pretreatment planning for known CAA may include specific anatomic details the guide surgical versus interventional options for treatment as well as decision-making as to hemodynamic significance of an anomaly to guide if treatment is warranted. Knowledge of the exact ostial location, relation to the valve cusp commissures, intercoronary pillar, length and location of a variant course, sites of abnormal connection, size of vessels and branch vessel location and patency can guide endovascular and surgical planning. Hemodynamic assessment of shunt ratios as well as if the variant causes myocardial ischemia can guide treatment planning [20].

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging.
A. Arteriography coronary

Arteriography of the coronary system can be used to delineate anatomic origins and distal connections. There is no relevant literature to support the use of coronary artery angiography for pretreatment evaluation of CAAs. Dynamic flow information regarding the direction and rate of flow pattern may aid in the detection of ischemia and collateral flow to help guide interventional planning.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging.
B. CT chest with IV contrast

There is no relevant literature to support the use of chest CT with IV contrast for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging.
C. CT chest without and with IV contrast

There is no relevant literature to support the use of chest CT without and with IV contrast for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging.
D. CT chest without IV contrast

There is no relevant literature to support the use of chest CT without IV contrast for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging.
E. CT coronary calcium

There is no relevant literature to support the use of CT coronary calcium for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. F. CT heart function and morphology with IV contrast

There is no relevant literature to support the use of CT heart function and morphology with IV contrast for pretreatment evaluation of CAAs. CT heart function and morphology is targeted at assessing structural heart disease including function, myocardial disease, valvular disease, and characterization of masses.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. G. CTA chest with IV contrast

There is no relevant literature to support the use of chest CTA with IV contrast for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. H. CTA chest without and with IV contrast

There is no relevant literature to support the use of chest CTA without and with IV contrast for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. I. CTA coronary arteries with IV contrast

Coronary artery anatomical variants warranting intervention may be treated by endovascular, minimally invasive, or open sternotomy approaches and may involve correction via reimplantation of a coronary artery, alteration of the anomalous course without reimplantation, or bypass and occlusion. A study by Ashrafpoor et al [21] suggests that CCTA can be used to evaluate imaging and anatomical characteristics resulting in increased risk of major adverse cardiac events when compared with similar subjects without such features, therefore identifying patients who may require treatment.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. J. MRA chest with IV contrast

There is no relevant literature to support the use of MRA chest with IV contrast for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. K. MRA chest without and with IV contrast

There is no relevant literature to support the use of MRA chest without and with IV contrast for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. L. MRA chest without IV contrast

There is no relevant literature to support the use of MRA chest without IV contrast for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. M. MRA coronary arteries without and with IV contrast

There is no relevant literature to support the use of MRA coronary arteries without and with IV contrast for pretreatment evaluation of CAAs. MRA of the coronaries can assess for origin and termination points as well as flow dynamics.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. N. MRA coronary arteries without IV contrast

There is no relevant literature to support the use of MRA coronary arteries without IV contrast for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. O. MRI heart function and morphology without and with IV contrast

There is no relevant literature to support the use of MRI heart function and morphology without and with IV contrast for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. P. MRI heart function and morphology without IV contrast

There is no relevant literature to support the use of MRI heart function and morphology without IV contrast for pretreatment evaluation of CAAs. MRI heart function and morphology allows for myocardial tissue analysis.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. Q. MRI heart function with stress without and with IV contrast

A recent study by Stagnaro et al [22] showed that dobutamine stress cardiac MRI at risk for or with previously diagnosed coronary artery disease had a sensitivity of 100% (confidence interval [CI] 2.5%-100%), specificity 92% (CI 64%-100%) when compared to CTA; this study was performed in pediatric patients.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. R. MRI heart function with stress without IV contrast

There is no relevant literature to support the use of MRI heart function with stress without IV contrast for pretreatment evaluation of CAAs. MRI heart function with stress testing allows for ischemia evaluation and may guide surgical decision making.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. S. Rb-82 PET/CT MPI rest and stress

There is no relevant literature to support the use of Rb-82 PET/CT heart for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. T. SPECT or SPECT/CT MPI rest and stress

Limited studies evaluate the use of SPECT/CT MPI for the pretreatment imaging in patients with ACA. However, a small case series suggests that when coupled with CCTA, PET-MPI may provide added value for the management of patients with ACA [23]. Ischemia assessment may contribute to surgical planning decisions.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. U. US echocardiography transesophageal

There is no relevant literature to support the use of US echocardiography transesophageal for pretreatment evaluation of CAAs.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. V. US echocardiography transthoracic resting

There is no relevant literature to support the use of TTE resting for pretreatment evaluation of CAAs. TTE allows for assessment of cardiac function that may guide decision-making for planned cardiac interventions.

Variant 2: Adult. Pretreatment planning for known coronary artery anomaly. Initial Imaging. W. US echocardiography transthoracic stress

There is no relevant literature to support the use of TTE stress for pretreatment evaluation of CAAs. TTE allows for assessment of cardiac function that may guide decision-making for planned cardiac interventions.

Summary of Highlights

This is a summary of the key recommendations from the variant tables. Refer to the complete narrative document for more information.

Variant 1: Initial imaging for suspected CAAs in adults with contrast-enhanced CTA of the coronary arteries is recommended to delineate the origin, course, and terminus of the coronary arteries. It allows for visualization of all cardiac structures in conjunction with aortic root assessment. Alternatively, both noncontrast and contrast-enhanced MRA of the coronary arteries are also usually appropriate alternatives to assess the variant anatomy of the coronary arteries. TTE may be appropriate, particularly to establish a baseline and acquire functional imaging.

Variant 2: When imaging is used for presurgical planning in adult patients with established CAAs, CTA of the coronary arteries with IV contrast is the recommended test. Alternatively, invasive catheter angiography is also suitable to assess anatomy and guide the surgical strategy, especially if there is coexistent coronary atherosclerosis. TTE, during both stress and rest, may be appropriate to delineate potential myocardial ischemia due to a coronary anomaly. MRA of the coronary arteries, both with and without IV contrast, may also be appropriate and further assess myocardial viability, particularly in conjunction with stress imaging to delineate potential resulting myocardial ischemia. CTA of the chest with IV contrast may be appropriate to delineate the anatomy of the thoracic aorta, pulmonary arteries, as well as both pulmonary and systemic veins, which is often necessary for presurgical planning. Both Rb 82 PET/CT and SPECT during both rest and stress may be appropriate to assess for potential myocardial ischemia in the setting of planned coronary artery transposition, bypass, or repair.

Supporting Documents

The evidence table, literature search, and appendix for this topic are available at https://acsearch.acr.org/list. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents, please go to the ACR website at https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria.

Gender Equality and Inclusivity Clause

The ACR acknowledges the limitations in applying inclusive language when citing research studies that predates the use of the current understanding of language inclusive of diversity in sex, intersex, gender, and gender-diverse people. The data variables regarding sex and gender used in the cited literature will not be changed. However, this guideline will use the terminology and definitions as proposed by the National Institutes of Health.

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared with those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® Radiation Dose Assessment Introduction document [25].

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Disclaimer

of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.

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