

Computed tomography of cardiomyopathies

Kevin Kalisz¹, Prabhakar Rajiah²

¹University Hospitals Cleveland Medical Center, Cleveland, Ohio, USA; ²Cardiothoracic Imaging, Radiology Department, UT Southwestern Medical Center, Dallas, Texas, USA

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Correspondence to: Prabhakar Rajiah, MD. Associate Professor, Department of Radiology, Cardiothoracic Imaging, UT Southwestern Medical Center, E6.120 B, Mail code 9316, 5323 Harry Hines Boulevard, Dallas, TX, 75390-8896, USA. Email: radprabhakar@gmail.com.

Abstract: Cardiac computed tomography (CT) is increasingly used in the evaluation of cardiomyopathies, particularly in patients who are not able to undergo other non-invasive imaging tests such as magnetic resonance imaging (MRI) due to the presence of MRI-incompatible pacemakers/defibrillators or other contraindications or due to extensive artifacts from indwelling metallic devices. Advances in scanner technology enable acquisition of CT images with high spatial resolution, good temporal resolution, wide field of view and multi-planar reconstruction capabilities. CT is useful in cardiomyopathies in several ways, particularly in the evaluation of coronary arteries, characterization of cardiomyopathy phenotype, quantification of cardiac volumes and function, treatment-planning, and post-treatment evaluation. In this article, we review the imaging techniques and specific applications of CT in the evaluation of cardiomyopathies.

Keywords: Computed tomography (CT); cardiomyopathies; heart; cardiac

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Introduction

Cardiomyopathies are a heterogeneous group of diseases of the myocardium associated with mechanical and/or electrical dysfunction typically exhibiting inappropriate ventricular hypertrophy or dilatation in the absence of underlying ischemia, hypertension, valvular or congenital heart disease (1). Heart failure and cardiomyopathy are common entities affecting approximately 6.5 million people in the United States and becoming increasingly prevalent in recent years (2). Ischemic heart disease is by far the most common etiology of heart failure, with dilated cardiomyopathy being the most common primary cardiomyopathy (3). Several non-invasive imaging modalities are used in cardiomyopathies to effectively and efficiently diagnose, categorize, and guide subsequent therapies, which are of critical importance. Computed tomography (CT) is playing an increasingly important role in the evaluation of

cardiomyopathies. In this article, we review the role of CT in the evaluation of cardiomyopathies.

Role of imaging

Imaging plays a crucial role in the screening, detection, and characterization of cardiomyopathies. There are several available imaging modalities, each with its own strengths and limitations, particularly echocardiography, nuclear medicine, magnetic resonance imaging (MRI) and invasive coronary angiography (ICA).

Echocardiography

Echocardiography is the initial imaging modality used in the evaluation of cardiac failure and cardiomyopathy as it is a widely available, portable technique that allows for real-

time evaluation of chamber size and morphology, systolic and diastolic function, and valvular abnormalities as well as blood flow, all without the use of ionizing radiation (4). Limitations of echocardiography include inadequate soft tissue characterization and suboptimal field-of-view in the setting of poor acoustic windows as in obesity, obstructive lung disease, or chest wall deformities. While transesophageal echocardiography (TEE) can overcome some limitations of poor surface windows, it is invasive with more limited availability.

Nuclear medicine

Nuclear medicine is also valuable in evaluation of cardiomyopathy (5,6), especially allowing for evaluation of myocardial perfusion with single-photon emission CT (SPECT). SPECT has high accuracy in evaluation of coronary artery disease (CAD) and also provides prognostic information (7,8). It also helps in distinguishing ischemic from non-ischemic etiologies (9). Nuclear imaging techniques have also shown useful in other areas such as left ventricle (LV) analysis (10), myocardial viability utilizing positron emission tomography (PET) (11), and diagnosis of some cardiomyopathy subtypes (5). For example, in the evaluation of cardiac amyloidosis, technetium-pyrophosphate has shown potential in distinguishing light chain from transthyretin amyloidosis subtypes, while ^{123}I -metaiodobenzylguanidine (MIBG) can be useful in functional evaluation of amyloid cardiomyopathy (12). ^{123}I -MIBG and carbon-11 labeled hydroxyephedrine are utilized for sympathetic innervation evaluation as well as potential future applications for monitoring molecular interventions (13). Disadvantages of nuclear imaging include the use of ionizing radiation, limited spatial resolution, poor tissue characterization, and relatively long imaging times.

ICA

ICA is considered the gold standard in the evaluation of CAD with fractional flow reserve (FFR) proven to be superior to luminal assessment in guiding treatment and improving outcomes (14). It also allows for intervention in patients with significant coronary artery stenosis. However, the main disadvantage of angiography is radiation exposure and invasiveness of the procedure with the potential for uncommon, yet serious, complications (15).

MRI

MRI provides excellent information in the evaluation of cardiomyopathy without the need for radiation or contrast. It has good spatial and temporal resolution and provides accurate and reproducible measurements of cardiac volumes and function, along with good soft-tissue characterization (16,17), which is done with an array of sequences such as T1, T2, T2*, steady state free precession (SSFP), short tau inversion recovery (STIR), perfusion, delayed enhancement, and T1 mapping (18-20) enabling differentiation of several cardiomyopathy phenotypes (21,22). However, cardiac MRI is not widely available, typically requires higher levels of skills, and has several contraindications including some pacemakers/implantable cardioverter defibrillators (ICD), claustrophobia and severe renal dysfunction.

CT

CT is not typically considered a first-line imaging modality in the evaluation of cardiomyopathies. However, it is increasingly gaining importance in this population, particularly in patients who have contraindications for other imaging tests or have suboptimal results, for example due to poor acoustic window in echocardiogram or contraindications for MRI. CT is widely available and has a rapid turnaround, with most scans performed in a matter of seconds. CT has excellent spatial resolution for evaluation of even small structures such as coronary arteries and modern scanners have high temporal resolution up to 66 milliseconds. CT angiography (CTA) is excellent in the evaluation of coronary arteries, which is an important component of cardiomyopathy evaluation as discussed below. CT can also provide functional evaluation as well as some tissue characterization and can be used before and after treatment (23). The principle disadvantages of CT include the use of ionizing radiation and iodinated contrast material. Iodinated contrast should be used with caution in patients with renal dysfunction and contrast allergy.

CT techniques

Recent progress in CT technology has allowed for the effective and efficient imaging of the heart for numerous applications. Key approaches to selecting among different cardiac techniques are summarized in *Table 1*. Specific scan parameters for several techniques are listed in *Table 2*. Advances in hardware have enabled acquisition of images

Table 1 Summary of cardiac CT technique factors

Technique factors	Considerations
Spatial resolution	Up to 0.35 mm Optimize for detailed evaluation of small structures (i.e., coronaries)
Temporal resolution	Up to 66 milliseconds in a third-generation dual source scanner Optimize to reduce cardiac motion artifact
ECG gating	Prospective: evaluation of coronary arteries; requires slow and regular heart rate (<60 beats per minute); lower radiation Retrospective: functional information, wall motion assessment, LVAD evaluation; allows for greater heart rates; higher radiation
Medications	Heart rate control: beta blockers (oral, intravenous) Coronary vasodilation: nitrates (sublingual)
Contrast protocol	Biphasic protocol: opacify left heart and coronaries without contrast on RV or SVC Triphasic protocol: opacify both right and left heart chambers without SVC streak artifact
Dose reduction	Prospective ECG triggering (regular/high pitch) High pitch helical mode ECG-based tube current modulation (retrospective) Body part-dependent tube current modulation Body habitus-based low kVp or mAs Thicker slices Iterative reconstruction algorithms

CT, computed tomography; ECG, electrocardiogram; LVAD, left ventricular assist device; RV, right ventricle; SVC, superior vena cava; kVp, kilovoltage peak; mAs, milliamperage-seconds.

Table 2 Scan parameters for selected applications

Technique	Gating technique	Contrast technique	Coverage	Slice thickness	Medications
Coronary artery assessment	Prospective	Biphasic	Carina to diaphragm	0.9 mm × 0.5 mm	Nitroglycerin 0.4 mg sublingual Metoprolol (50–100 mg PO; 5–25 mg IV)
Functional evaluation	Retrospective	Triphasic	Carina to diaphragm	2 mm × 1 mm	None
Perfusion (stress)	Prospective	Biphasic	Carina to diaphragm	0.9 mm × 0.5 mm	Stress agent (regadenoson 0.4 mg over 10 s; adenosine 140 µg/kg/min for at least 2 min)
Perfusion (rest)	Prospective	Biphasic	Carina to diaphragm	0.9 mm × 0.5 mm	Nitroglycerin 0.4 mg sublingual Metoprolol (50–100 mg PO; 5–25 mg IV)
Delayed enhancement	Prospective	No additional contrast than the early phase	Carina to diaphragm	0.9 mm × 0.5 mm	No additional medication

PO, per os (orally); IV, intravenous.

at high spatial and temporal resolutions, with smaller detectors and improved detector electronics and faster gantry rotation times. Most centers perform cardiac CT with 16 to 64 slice scanners with 64 slice multi-detector CT (MDCT) systems able to produce isotropic spatial resolution as high as 0.35 mm and a temporal resolution as good as 165 ms (24,25). Multi-detector scanners with up to 320 slices allows increased z-coverage of up to 16 cm, which allows single heart beat scanning. There are also dual source scanners, which have improved temporal resolution (up to 66 milliseconds) and have the option of high-pitch helical mode in which data can be obtained rapidly without data gaps due to the presence of a second tube (26). There are also several types of dual-energy scanners, such as dual-source, rapid kilovoltage peak (kVp) switching, dual spin and dual-layer technologies, which allow material characterization beyond what is possible with single energy CT (27).

The CT protocol for the evaluation of the cardiomyopathies can be varied depending on the specific clinical question to be answered. Cardiac CT is performed with electrocardiogram (ECG) gating to avoid motion artifacts. For the evaluation of coronary arteries, prospective ECG-triggering is the default mode to minimize radiation dose, although this is possible only if the heart rate is slow and regular (less than 60–65 beats per minute, depending on the scanner). Retrospective ECG-gating is used in evaluation of coronary arteries if the heart rate is high or irregular. This mode is also used in specific scenarios such as in the evaluation of cardiac function and volumes, wall motion or valvular abnormalities, and LV assist device (LVAD) position and complications. ECG-based tube current modulation is used with retrospective mode in order to minimize radiation dose. Non-ECG gated/helical high-pitch techniques can be used for the evaluation of pulmonary veins (28). For evaluation of coronary arteries, beta-blockers are administered either orally or intravenously if the heart rate is high or irregular (29). Sublingual nitroglycerine is also administered to dilate the coronary arteries. However, these medications are not required for non-coronary arterial indications, such as evaluation of cardiac function or LVAD evaluation. Intravenous contrast protocols are also optimized depending on the clinical indication. Recommended contrast injection protocols according to the Society of Cardiovascular CT are as follows: for evaluation of coronary arteries, a biphasic protocol is used where injection of contrast (typically 50–120 mL at 5–7 mL/s) is followed by a saline bolus (typically 40–50 mL at 5–7 mL/s) to opacify the left heart and coronary arteries, with no contrast in the right ventricle

(RV) or superior vena cava (SVC) (30,31). For evaluation of cardiac volumes and function, a triphasic protocol is used, where injection of contrast at 5–7 mL/s is followed by either a slower rate of contrast injection at 2 mL/s or a contrast-saline admixture at same rate (50:50 of contrast:saline at 5–7 mL/s), followed by a smaller volume saline bolus to opacify all the cardiac chambers without streak artifact in the SVC (30).

Several cardiac CT techniques allow for additional cardiac evaluation beyond anatomic and basic function assessment. Perfusion imaging relies on the distribution of contrast material during the first pass through the myocardium which is dependent on the myocardial arterial blood supply (32). CT myocardial perfusion exams have been described with both single and dual energy techniques. In the single energy technique, images are acquired in the early arterial phase (i.e., 8–16 s) during first pass through the myocardium with perfusion abnormalities presenting as areas of decreased myocardial attenuation secondary to reduced blood flow (*Figure 1*). Images are obtained at rest and after pharmacological stress, with variable order in which these are done (33). Using dual energy techniques, iodine perfusion maps displayed as color maps can evaluate myocardial perfusion differences with the value of quantified iodine serving as a marker of myocardial perfusion (34). Delayed myocardial enhancement (Delayed iodine enhancement) imaging is an indicator of myocardial fibrosis/scar analogous to late gadolinium enhanced MRI. This is done 5–15 min after intravenous administration of iodinated contrast and can be done either in the single or dual-energy mode. Single-energy technique is limited by poor signal-to-noise ratio (SNR) and contrast-to-noise ratio (CNR) as well as beam hardening artifact. Improved SNR and CNR can be seen in virtual monoenergetic images and iodine maps of dual-energy scanners. This technique can be used for the characterization of cardiomyopathies based on the pattern of enhancement, particularly in patients who have contraindications for MRI. This can also be used for the evaluation of myocardial viability in patients with infarction (35). Multiple studies have shown sensitivities of 88–92% and specificities of 72–77% in detection of chronic myocardial infarction (MI) (35,36). There is also potential for estimating the extracellular volume fraction (ECV), which is a biomarker for myocardial fibrosis. This technique is not widely used and requires data from larger patient cohorts (37).

Strain imaging is used for the evaluation of regional cardiac function in longitudinal, radial, circumferential and

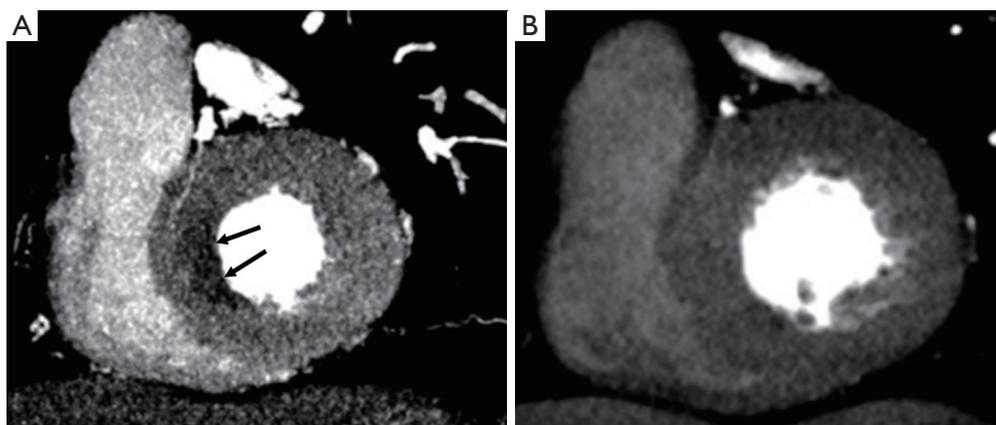


Figure 1 CT perfusion. (A) Short axis stress perfusion CT image obtained at 45% of R-R interval in a patient with chest pain shows a sub-endocardial perfusion defect in the basal septum (arrows); (B) rest CT perfusion image in the same patient at the same level does not show the defect. These findings are consistent with reversible myocardial ischemia. CT, computed tomography.

torsional directions by evaluating velocity gradients between two objects in space. This is usually performed using echocardiography or MRI; however, CT can also be used for evaluating myocardial strain. A recent study showed that quantitative LV strain is feasible using CT feature tracking, with comparable accuracy as that of echocardiography and has potential for evaluation of regional function in patients with poor echo windows or contraindications for MRI (38). This technique has potential in detecting regional functional abnormalities prior to the onset of overt cardiac failure. CT metrics such as changes in wall thickness, wall motion and volume over time can also be used to quantify LV dyssynchrony, which is useful for predicting response to cardiac resynchronization therapy (39).

Although radiation dose is a concern with the use of cardiac CT (40), it can be minimized using the principle of As Low As Reasonably Achievable (ALARA) by using a variety of technique such as prospective ECG-triggering including helical mode (41), ECG-based tube current modulation in retrospective acquisitions (42,43), automatic-tube current modulation based on body-part size (42), body-habitus based low kVp or milliamperage-seconds (mAs) (43,44), higher-pitch (45,46), thicker slices, and iterative reconstruction techniques (40,47).

Role of CT in cardiomyopathy assessment

There are several established roles for CT in the evaluation of cardiomyopathies. Broadly, these include: evaluation of coronary arteries, characterization cardiomyopathy type,

evaluation of cardiac volumes and function, treatment planning, and post-treatment evaluation, which are summarized on *Table 3*.

Evaluation of coronary arteries

Cardiac failure and cardiomyopathy is often suggested through history, physical exam, and echocardiography. However, accurate characterization is necessary for optimal patient management. The initial step in characterization of heart failure is to diagnose or exclude ischemic cardiomyopathy (48), since LV functional recovery can be achieved in these patients by coronary revascularization procedures (11). Although traditionally the work-up for ischemic cardiomyopathy was done by ICA (49), CTA is now considered the first-line non-invasive imaging test for exclusion of significant coronary artery stenosis, especially in the low to intermediate risk population (*Figure 2*). A particular advantage of coronary CTA (CCTA) in assessment of CAD is its high specificity (95–98%) and negative predictive value (95–100%) (50–52). In patients without previously known coronary disease presenting with and LV ejection fraction (LVEF) of less than 35%, CCTA demonstrated a sensitivity of 98% and specificity of 97% for the diagnosis of CAD (53). As a result of this and other studies, CCTA has been given a high appropriateness rating for the evaluation of ischemic etiology in patients presenting with heart failure and ischemic symptoms and may be even appropriate in imaging heart failure patients without ischemic symptoms (54). With latest technologies such

Table 3 Role of CT in cardiomyopathy assessment

Role	Description
Evaluation of coronary arteries	Evaluation of coronary artery stenosis Distinguish between ischemic and non-ischemic cardiomyopathy
Characterization of cardiomyopathies	Evaluate morphologic type of cardiomyopathy Perfusion (differentiate myocardial scar and ischemia) and delayed iodine assessment (fibrosis pattern)
Volume and function evaluation	Ventricular and atrial volume and function quantification Qualitative evaluation of wall motion abnormalities
Treatment planning	Myocardial viability assessment using delayed iodine assessment Cardiac resynchronization therapy planning (anatomy, scar assessment) Atrial fibrillation ablation planning (pulmonary vein and left atrial anatomy, left atrial appendage thrombus)
Post-treatment evaluation	Follow-up of cardiac function after cardiomyopathy treatment Evaluate post-procedural complications (left ventricular assist device, ablation) Evaluate heart transplant complications

CT, computed tomography.

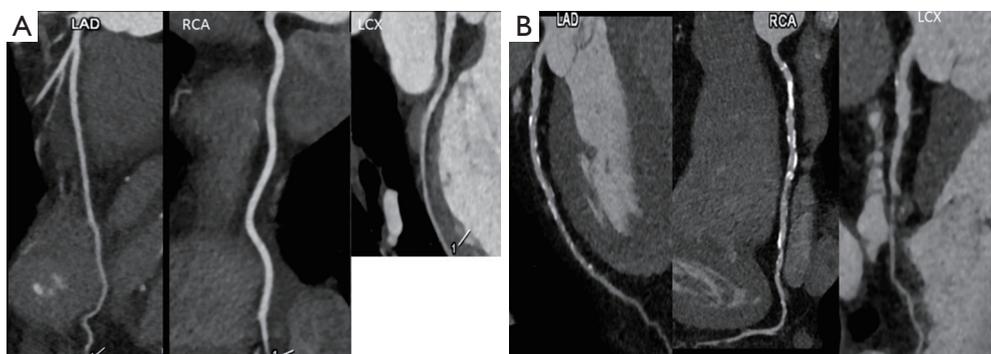


Figure 2 Coronary artery evaluation. (A) Cardiac CT angiography performed in a patient with new onset heart failure shows normal coronary arteries without any atherosclerotic plaque or luminal stenosis; (B) cardiac CTA in another patient with heart failure shows multiple calcific and non-calcific atherosclerotic plaques in all the coronary arteries. CT, computed tomography; CTA, CT angiography. LAD, left anterior descending; RCA, right coronary artery; LCx, left circumflex coronary artery.

as CT perfusion, CT-FFR or transcatheter attenuation gradient, the hemodynamic significance of coronary artery stenosis can also be characterized, which helps in triaging patients for ICA and selecting appropriate patients for revascularization, thus improving outcome (32,34,55-58). CT can also evaluate the plaque burden and vulnerability which also carry prognostic significance (59).

Characterization of cardiomyopathies

Characterization of the different phenotypes of

cardiomyopathies is important as the treatment varies depending on the etiology. For example, sarcoidosis benefits from corticosteroids, whereas iron-overload cardiomyopathy requires chelation therapy. Although MRI is the imaging modality that is typically used for characterizing cardiomyopathies, CT can also be effective in patients who cannot have MRI. The conventional CT images obtained using biphasic or triphasic injection protocol can provide morphological information, which can be used for diagnosis of several cardiomyopathies. With retrospective ECG-gated techniques, the qualitative and

Table 4 CT findings in cardiomyopathies

Cardiomyopathies	CT findings
Ischemic cardiomyopathy	Focal wall motion abnormalities in coronary distribution Subendocardial or transmural DIE in coronary distribution Decreased enhancement on myocardial perfusion Assessment of coronary arteries
Dilated non-ischemic cardiomyopathy	Global systolic dysfunction Dilated ventricle Linear mid-myocardial DIE Absence of significant coronary disease
Hypertrophic cardiomyopathy	Asymmetric hypertrophy of basal interventricular septum (classic form) Systolic anterior motion of mitral valve Patchy or diffuse DIE
Myocarditis	Global/regional hypokinesis Myocardial/sub-epicardial DIE
Sarcoidosis	Global/regional hypokinesis Focal wall thickening (acute) or thinning (chronic) Myocardial/sub-epicardial DIE
Amyloidosis	Ventricular thickening with biatrial enlargement Diffuse sub-endocardial or transmural DIE Expansion of ECV
Left ventricular non-compaction	Ratio of trabeculated to non-trabeculated myocardium of >2.3 in end-diastole
Stress-induced cardiomyopathy (Takotsubo)	Hyperkinesis of the basal segments with hypokinesis/akinesia of the apical segments Absence of DIE
Arrhythmogenic right ventricular cardiomyopathy	Presence of fat within RV Major focal RV wall motion abnormalities with either RV dilation or systolic dysfunction

CT, computed tomography; DIE, delayed iodine enhancement; ECV, extracellular volume; RV, right ventricle.

quantitative analysis of the cardiac volumes and function can be performed, which is also useful in the diagnosis. Both of these can also be used to exclude other diseases such as congenital and valvular heart diseases. CT myocardial perfusion can be used for evaluation of myocardial ischemia and infarct, with the former showing perfusion defects only with stress and the latter showing defect in both stress and rest images. Delayed iodine enhancement (DIE) images can be obtained 5–15 min after administration of intravenous contrast (60), either in the single or dual-energy mode to evaluate for myocardial fibrosis and scar analogous to MRI, but with much lower CNR and associated radiation dose.

Based on the pattern and distribution of enhancement, different types of cardiomyopathies can be distinguished (61,62). More recently, similar to cardiac MRI, cardiac CT has been shown to have the potential to quantify the myocardial ECV, which is a surrogate for myocardial fibrosis (63). Cardiac CT has shown good correlation in ECV calculation in several heart failure cohorts when compared with the cardiac MRI (64,65). *Table 4* summarizes CT findings for various cardiomyopathies described in greater detail here.

In ischemic cardiomyopathy, focal regional wall motion abnormalities in a vascular distribution can

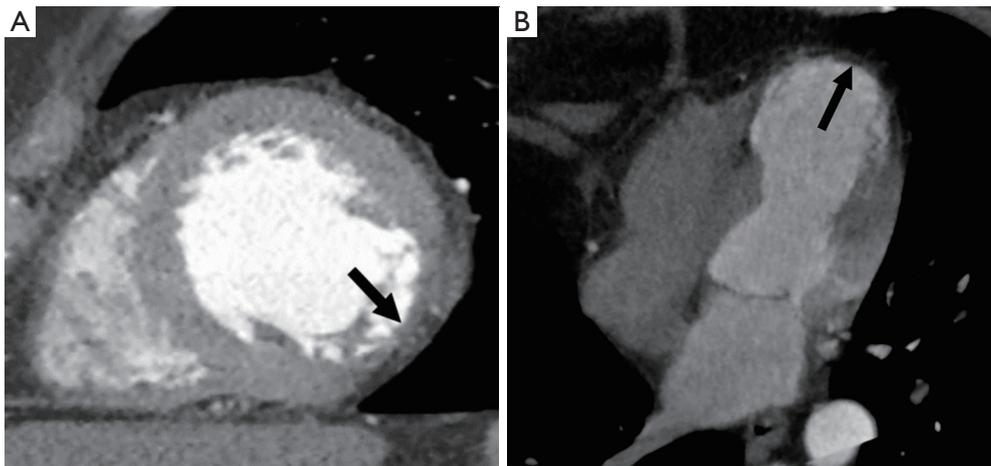


Figure 3 Ischemic cardiomyopathy. (A) Short axis contrast enhanced CT scan shows thinning of the inferolateral wall (arrow), with regional hypokinesis (not shown here), in a patient with infarct of the left circumflex artery; (B) 4-chamber post contrast CT in another patient shows aneurysm of the LV apex with fatty metaplasia of the thinned apical segments (arrow). Images courtesy: Dr Harold Goerne, UT Southwestern Medical Center, Dallas. CT, computed tomography; LV, left ventricle.

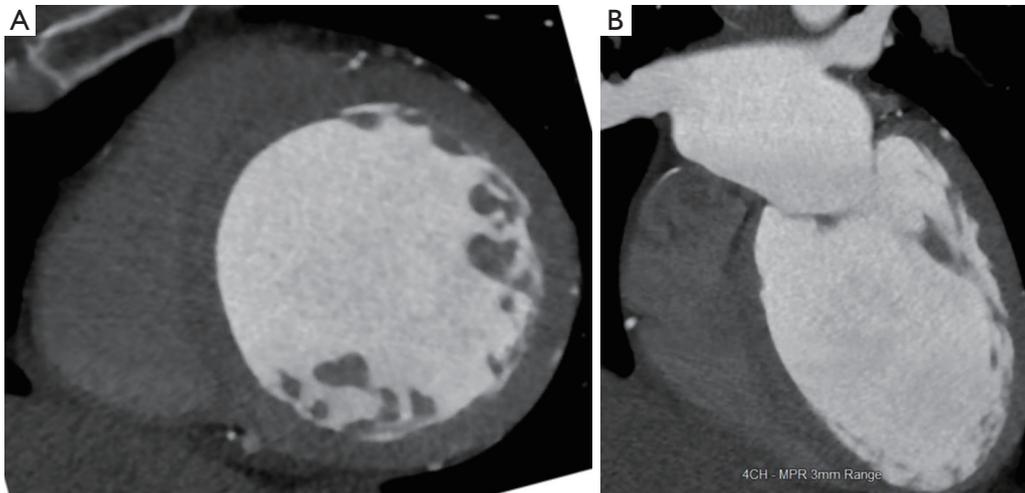


Figure 4 Dilated cardiomyopathy. (A) Short-axis CT image shows severe dilation of the left ventricle with end-diastolic diameter of 7.1 cm; (B) 4-chamber CT image also shows severe dilation of the left ventricle. The ejection fraction in this patient was 15% and the constellation of findings is consistent with dilated cardiomyopathy. CT, computed tomography.

be evaluated using cine images. Wall thinning or fatty metaplasia may be seen in chronic infarcts (*Figure 3A*). On DIE images, areas of enhancement are seen in a sub-endocardial or transmural location corresponding to a vascular distribution. Myocardial perfusion techniques will demonstrate areas of decreased enhancement on first-pass images, both at stress and rest and may delineate a greater territory of ischemic myocardium (32). The percentage of

stenosis of the supplying coronary arteries as well as plaque characterization can also be readily assessed. Complications of MI including aneurysm (*Figure 3B*), pseudoaneurysm, rupture and thrombus can also be detected in CT.

Dilated cardiomyopathy is characterized by a dilated LV and global systolic dysfunction without regional wall motion abnormalities (*Figure 4*). A linear mid-myocardial pattern of DIE may also be seen in idiopathic cases. CT



Figure 5 Hypertrophic cardiomyopathy. (A) Short axis reconstruction at the level of mid-ventricle in diastole shows asymmetrical hypertrophy of the ventricular septum (arrow) with normal thickness of the lateral wall; (B) short-axis reconstruction at the level of mid-ventricle in diastole shows measurements of all the myocardial segments; (C) 3-chamber reconstruction in systole shows severe narrowing of the LVOT (arrow). There is also additional thickening of the anterior leaflet of the mitral valve (arrowhead). LVOT, left ventricular outflow tract.

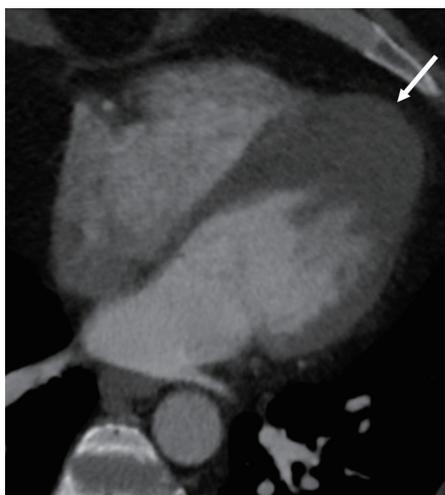


Figure 6 Apical hypertrophic cardiomyopathy. Four-chamber CT image shows hypertrophy of the apical segments (arrow) in this patient with apical hypertrophic cardiomyopathy. CT, computed tomography.

has been shown to effectively distinguish patients with ischemic and non-ischemic dilated cardiomyopathies by the exclusion of significant coronary disease as well as absence of typical subendocardial or transmural DIE enhancement patterns (66). However, CT and other imaging modalities are often unable to differentiate between the various causative etiologies of dilated cardiomyopathy.

Hypertrophic cardiomyopathy is characterized most commonly by asymmetrical hypertrophy of the basal septum, with narrowing of the LV outflow tract and presence

of systolic anterior motion of the mitral valve (Figure 5). Atypical forms include apical (Figure 6), mid ventricular, and concentric hypertrophy. Myocardial fibrosis has been shown in DIE images with either focal, typically located near the RV insertion points, or diffuse (61,62). The amount of fibrosis present in these patients has been shown to correlate with the presence of life-threatening arrhythmias and risk of sudden death (67).

In myocarditis, cine images demonstrate global or regional wall motion abnormalities. On DIE images, sub-epicardial/mid-myocardial enhancement may be seen (68). Contrary to MRI, myocardial edema is poorly assessed at cardiac CT. Differentiating myocarditis from ischemic heart disease may be challenging given similar presentation of chest pain and elevated cardiac biomarkers but is crucial as myocarditis is often self-limiting with anti-inflammatory therapy and supportive care.

Infiltrative cardiomyopathies are characterized by concentric LV thickening and CT has been shown to correlate well with MRI in the characterization of these cardiomyopathies, including sarcoidosis and amyloidosis (69,70). Cardiac involvement in sarcoidosis has been shown to be present in up to 5% of patients with systemic sarcoidosis (71). CT may demonstrate focal wall thickening or thinning in the acute and chronic stages, respectively, with regional motion abnormalities. On DIE images, mid-myocardial or sub-epicardial DIE indicative of fibrosis may be seen similar to that seen in cases of myocarditis (Figure 7) (69). The presence of fibrosis has shown to predict adverse cardiac outcomes of these patients (72). In

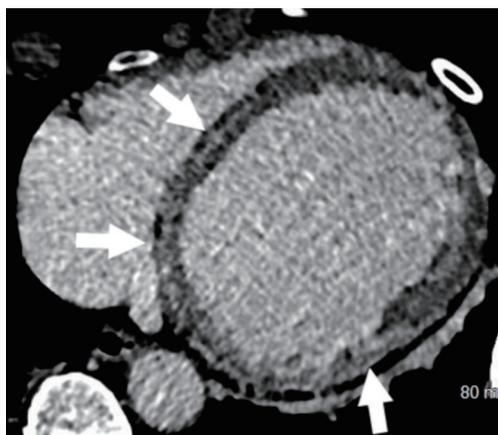


Figure 7 Sarcoidosis. Axial delayed enhancement CT obtained 10 min after contrast in a patient with sarcoidosis shows mid-myocardial delayed iodine enhancement in the septum and the lateral wall (arrows). CT, computed tomography.

the acute phase, the myocardium is thickened and there may be focal areas of granulomas. In the chronic phase, the myocardium is thinned.

Amyloidosis is characterized by abnormal deposition of fibrillar amyloid proteins within the myocardial extracellular space with various clinical subtypes including light-chain (AL), familial transthyretin-related (ATTR), secondary, and senile systemic forms. Structurally, amyloidosis typically demonstrates ventricular thickening with biatrial enlargement. On DIE images, there is a distinctive diffuse sub-endocardial to transmural enhancement pattern (73). CT ECV techniques have also demonstrated expansion of ECV (74).

LV non-compaction is characterized by prominent and excessive LV trabeculations due to an arrest of myocardial compaction, which may be either familial or sporadic. At imaging, the diagnosis can be made by a ratio of trabeculated to non-trabeculated myocardium of greater than 2.3 in end-diastole (75) (Figure 8). Areas of trabecular and subendocardial delayed contrast enhancement can be seen (76). Thrombosis, LV dysfunction and arrhythmias are complications associated with LV noncompaction (LVNC).

Stress induced cardiomyopathy (Takotsubo) is a reversible LV systolic dysfunction in the absence of coronary disease often seen after periods of severe emotional stress. Evaluation of CT cine images is key there is a characteristic pattern hyperkinesis of the basal segments with hypokinesis/akinesias of the apical segments, resulting in apical ballooning. An atypical form has also been reported with hyperkinesis of the apical segments and akinesis of basal segments (77,78). CT

is particularly useful in excluding coronary disease in these patients. DIE images will often be normal.

Arrhythmogenic RV cardiomyopathy (ARVD) is characterized by fibrofatty replacement of the RV with variable involvement of the LV and presents clinically as arrhythmias, typically at a young age. While CT may show fat in the RV myocardium, contributing diagnostic findings at imaging according to Task Force criteria include the presence of major wall motion abnormalities (akinesia, dyskinesia, or dyssynchrony) with either a dilated RV (end-diastolic volume greater than 110 mL/m^2 for male, greater than 100 mL/m^2 for female) or RV dysfunction (ejection fraction less than 40%) (Figure 9) (79,80).

Quantification of cardiac volumes and function

CT provides accurate measurements of the cardiac volumes and functions, and is utilized when quantification is not possible with other imaging modalities such as echocardiogram due to poor acoustic windows or MRI due to presence of MRI-incompatible pacemakers/ICDs or other contraindications. For quantification, retrospective ECG-gated acquisition throughout the cardiac cycle is required, along with a tri-phasic contrast injection to visualize both the ventricles. Radiation dose is minimized by using ECG-based tube current modulation and thicker slices. Beta blockers and nitroglycerine are not needed for this type of indication. The retrospective data is typically reconstructed from 0% to 100%, at 10% intervals, with the 0% image usually correlating with end-diastole and 30–40% image correlating with end-systole. The CT images are reconstructed in the short-axis plane, usually at 2-mm thickness, although may be variable. Cine images can be used to qualitatively evaluate for morphology and function.

Cardiac volumes and function can be calculated by various methods, including the area-length method, Simpsons method, and threshold based segmentation. In the area-length method, the volume is based on assumption of ellipsoid shape of ventricle and measuring the area and length of the ventricle in the long axis. In Simpson's method, volumes are determined by summing the areas of LV cavity at each short axis slice multiplied by the slice thickness (Figure 10A). Simpson's method has been shown superior to the area-length technique, which overestimates both end-diastolic and end-systolic volumes and stroke volume (81). Threshold-based segmentation utilizes the differences in attenuation between the blood pool and myocardium. Ventricular volume is calculated by summing

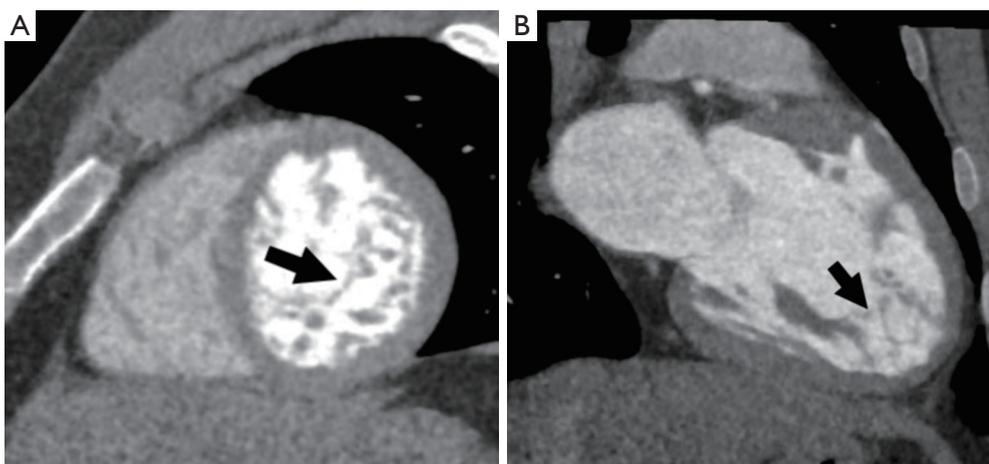


Figure 8 LV non-compaction. (A) Short axis CT reconstruction in end-diastole shows prominent trabeculations (arrow) in the LV apical region, consistent with non-compaction; (B) 2-chamber CT reconstruction also shows the prominent trabeculations (arrow), with end-diastolic ratio of non-compacted to compacted myocardium of 3.4, which is consistent with LV non-compaction. CT, computed tomography; LV, left ventricle.

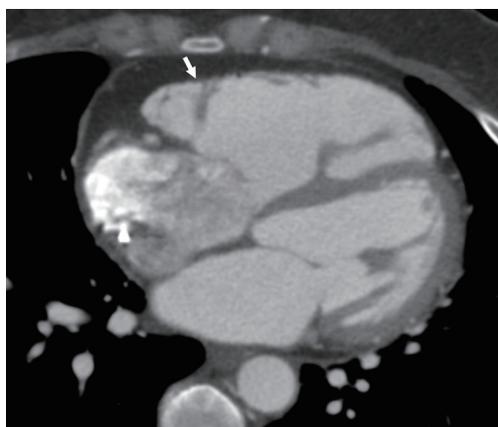


Figure 9 ARVD. Axial CT scan in a patient with ARVD shows dilated right ventricle and presence of fat in the RV free wall (arrow). ARVD, arrhythmogenic right ventricular cardiomyopathy; CT, computed tomography.

of all the contiguous voxels whose attenuation is above that of a predefined threshold. This technique has been shown to be accurate and reproducible with low interobserver variability with short processing time (82). Regional wall motion can also be evaluated including wall attenuation, systolic percentage wall thickening, wall motion/shortening and regional ejection fraction (*Figure 10B*). Lastly, while assessment of diastolic function is typically performed with echocardiography, it has also been shown to be feasible with

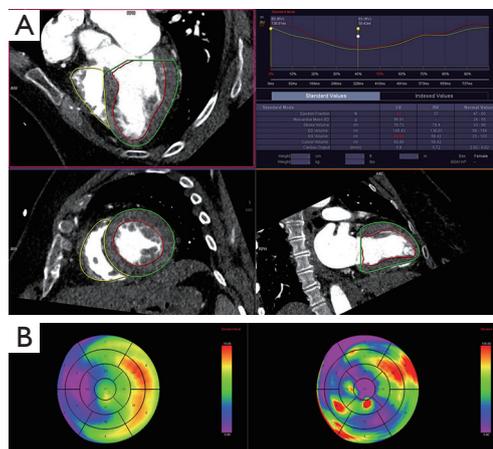


Figure 10 Functional evaluation. (A) CT measurement of ventricular volumes and function after drawing contours in the ventricles (red, LV endocardium; green, LV epicardium; yellow, RV endocardium); (B) regional functional evaluation with polar maps showing wall motion (in mm) and wall thickening (in percentage). CT, computed tomography; LV, left ventricular; RV, right ventricular.

CT. CT derived transmitral velocity, mitral septal tissue velocity, and estimation of LV filling pressures have shown good correlation with echocardiogram (83).

The LVEF and volumes obtained by CT have been shown to correlate with other imaging modalities such as echocardiography (81,84,85), SPECT (86) and MRI (87,88),

although there has been mild but consistent overestimation of LVEF (81,84). The temporal resolution of CT is not as good as echocardiography and hence the measurements may be limited in the setting of elevated heart rates (89). Compared to MRI, the acquisition is fast and the images can be reconstructed even at thinner slices if more accurate quantification is desired (90).

Cardiac CT is also valuable in assessing the volumes and function of other cardiac chambers, including the RV (91). CT has been shown to be accurate compared to MRI in the evaluation of RV volumes and functions (79,92,93), particularly in patients who cannot have MRI, since echocardiography provides limited acoustic windows in adults for the evaluation of RV. Atrial volumes and function can also be quantified using CT (94). Excellent correlation has been shown between CT and MRI in patients on sinus rhythm, but lower accuracy is seen in patients with atrial fibrillation (94,95).

Treatment planning

CT is also increasingly used in the treatment planning of various cardiomyopathies, particularly for viability assessment, cardiac synchronization therapy (CRT) and radiofrequency ablation of pulmonary veins for atrial fibrillation. Myocardial viability is important in patients being planned for coronary revascularization, since the success of this procedure is less if there are no viable segments present. This assessment is traditionally performed using cardiac MRI or PET. But, in patients who cannot undergo imaging at either of these modalities, CT is a good alternative imaging test. Infarcted myocardium is seen as a perfusion defect that persists in rest and stress myocardial perfusion images. Scar shows high iodine uptake in DIE images, either with single or dual energy technique (60). Studies have good accuracy of DIE in evaluating myocardial viability compared to MRI, both for single-energy (sensitivity, 92%, specificity, 100%, and accuracy, 94% when correlated with infarct size) (60,96) and dual energy CT (36).

CRT is typically used in treatment of cardiac dysfunction the setting of wide complex rhythms. There is high failure rate of CRT with high rates of response failure and suboptimal lead implantation (30% and 10%, respectively) (97,98). Key elements required for successful CRT include: knowledge of precise area of latest mechanical activation, presence of viable myocardium, and presence of accessible venous tributaries draining this area. Knowledge of the venous anatomy prior to the procedure aids in evaluating if the

procedure is feasible and to minimize the procedure time (99). Cardiac venous anatomy can be evaluated by delaying the acquisition of a coronary CTA protocol. Variations in the venous anatomy, such as absence of venous tributaries may result in a wasted procedure. A recent study showed a lower prevalence of coronary venous tributaries draining the lateral LV wall, i.e., posterolateral and left marginal veins in ischemic cardiomyopathy (68% and 48%), which potentially hinders access for optimal positioning of LV lead and causes suboptimal CT response (97). Presence of scar in the lateral LV wall in the region of LV lead placement has been associated with reduced effectiveness of CRT due to lesser response to electrical stimulation (100,101). Although this scar is typically assessed using cardiac MRI or PET, DIE can also show this and helps in selecting appropriate patients. CT could also potentially be used prospectively in CRT planning to map areas of LV scarring.

Ablation techniques for atrial fibrillation involve interrupting abnormal pathways of conduction between the pulmonary vein ostia and the left atrium. CT is used in the pre-procedural evaluation for assessment of pulmonary venous and left atrial anatomy and left atrial thrombus. Pulmonary vein mapping CT is usually acquired with prospective ECG triggering, but non-ECG gated techniques including high pitch helical mode can be used to minimize the radiation dose (102). CT provides information on the pulmonary vein ostial anatomy, particularly in noting the location, size, anatomic variants and adjacent structures that may be affected by the ablation. The ostial measurements are necessary for selecting the optimal catheter size and have been shown to be comparable to MRI and superior to TEE (103,104). Pre-procedural CT has been shown to reduce the procedure time, increase the success of sinus rhythm restoration, and reduce arrhythmia recurrence (105,106). Furthermore, CT can also be used to evaluate the complications of ablation including pulmonary vein stenosis (107). Left atrial volume and narrow left atrial ridges especially about the appendage and between venous ostia are also important factors, which if ignored may lead to incomplete ablation lines or damage of veins resulting in stenosis. Presence of narrow ridges on pre-procedural imaging may subsequently alter the site of ablation (108). CT-derived left atrial volume has been shown to be superior to that of echocardiography in predicting the success of ablation (109). CT is also used in the detection of left atrial appendage thrombus, which is an absolute contraindication for ablation procedure. CT has a high negative predictive value (up to 98%), but modest specificity

(85–88%) compared to TEE (108). It may be challenging to distinguish left atrial appendage thrombus from contrast admixture artifact. Thrombus and contrast admixture can be distinguished by measuring the attenuation (lower in thrombus), ratio of attenuation in LAA and aorta (lower in thrombus), iodine quantification with dual energy CT scanner (lower in thrombus), delayed phase acquisition (thrombus persists in delayed phase) or a split-bolus contrast injection protocol. Similarly, CT is also useful in evaluation of left atrial appendage anatomy and thrombus in patients with atrial fibrillation patients prior to undergoing atrial appendage occlusion device implantation (110).

Post-treatment evaluation

In patients with contraindications to MRI, CT is also a valuable imaging modality in the follow up of treatment. CT can provide accurate estimates of the ventricular volumes and function, which typically show improvement with several treatments. CT can also be used in the follow-up of procedures such as atrial ablation to evaluate for complications such as pulmonary venous stenosis, thrombosis and fistula with adjacent organs. CT is also useful in the evaluation of the position and complications of LVAD for patients with heart failure. Retrospective ECG gated CT can identify the position of the LV inflow and outflow cannulas. CT is also useful in the evaluation of other complications of LVAD including thrombus, hemorrhage, tamponade, aortic valve lesions, infection and fluid collections (111). CT has role in the evaluation of patients with heart transplants. Specifically, CT has been used in the setting of cardiac allograft vasculopathy (CAV), which is an accelerated form of CAD characterized by areas of fibrosis and intimal hyperplasia within transplant coronary vessels and is a major determinant in long-term survival in cardiac transplant patients (112). Traditional non-invasive imaging method such as echocardiography and nuclear medicine studies have been shown to be ineffective in evaluation of CAV (113). However, cardiac CT is a promising modality for the evaluation of CAV as it simultaneously evaluates coronary luminal irregularities as well as the coronary artery wall. CT has high sensitivity, specificity, and negative and positive predictive values in evaluation of CAV compared to both ICA and intravascular ultrasound (114,115).

Conclusions

Cardiac CT provides a comprehensive assessment of many

facets of cardiomyopathies including characterization of the phenotype, evaluation of coronary arteries, quantification of function, treatment planning and post-treatment evaluation. CT is mostly utilized in cardiomyopathies in specific situations where echocardiography or MRI is not possible (due to contraindications) or suboptimal (due to poor windows in echo or artifacts in MRI).

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Footnote

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