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Analysis of Ventricular Function by Computed Tomography

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Abstract

The assessment of ventricular function, cardiac chamber dimensions and ventricular mass is fundamental for clinical diagnosis, risk assessment, therapeutic decisions, and prognosis in patients with cardiac disease. Although cardiac computed tomography (CT) is a noninvasive imaging technique often used for the assessment of coronary artery disease, it can also be utilized to obtain important data about left and right ventricular function and morphology. In this review, we will discuss the clinical indications for the use of cardiac CT for ventricular analysis, review the evidence on the assessment of ventricular function compared to existing imaging modalities such cardiac MRI and echocardiography, provide a typical cardiac CT protocol for image acquisition and post-processing for ventricular analysis, and provide step-by-step instructions to acquire multiplanar cardiac views for ventricular assessment from the standard axial, coronal, and sagittal planes. Furthermore, both qualitative and quantitative assessments of ventricular function as well as sample reporting are detailed.

Keywords

Ventricular function; computed tomography; ejection fraction; left ventricular mass; wall motion abnormality; arrhythmogenic right ventricular dysplasia

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INTRODUCTION

Although the mainstay for cardiac computed tomography (CT) is the assessment of coronary artery disease, the isotropic sub-mm spatial resolution, high temporal resolution and good contrast between ventricular lumen and myocardium make CT very well suited to obtain valuable information on ventricular function. ¹⁻³ Currently, several noninvasive imaging techniques are available for the assessment of ventricular function – each with their own limitations. Cardiac magnetic resonance imaging (CMR) is considered the gold standard.⁴ However, CMR is costly, time-consuming, and has limited availability to predominantly tertiary medical centers. Furthermore, some patients may not be able to undergo a CMR examination, for example due to metallic devices or clinical conditions such as claustrophobia and the inability to lay flat.² Transthoracic echocardiography (TTE) is the cheapest and most routinely used method for the measurement of ventricular function but may be limited by poor acoustic windows in patients with obesity, chronic obstructive pulmonary disease (COPD), narrow rib intercostal spaces, or prior cardiothoracic surgery. 5-7 Furthermore, assessment of the right ventricle can be difficult especially when assessing structural abnormalities such as arrhythmogenic right ventricular cardiomyopathy/ dysplasia (ARVC/D).8 Cardiac CT, with some modifications to the acquisition protocol, can be utilized to obtain accurate ventricular assessments comparable to CMR. Tube current modulation with improved noise reduction strategies have allowed for reduction of radiation dose with preserved image quality. 10-12 For patients that are difficult to image by echocardiography and are undergoing CT imaging for coronary artery disease (CAD) without previous ventricular assessment, especially with a history of acute coronary syndrome (ACS) or heart failure (HF), or for patients who need specific assessment of right ventricular (RV) function or morphology, evaluation of ventricular contractile function by cardiac CT should be considered.

MULTIMODALITY COMPARISON OF LV VOLUMES, FUNCTION, AND MASS

As temporal resolution has improved, cardiac CT can be utilized to obtain important information on ventricular function, regional wall motion, and LV mass and results are comparable to measurements obtained via CMR. ^{13–16} Unlike the evaluation of the coronary arteries, cardiac function can be accurately assessed without the need to alter heart rate via administration of beta-blockers. ¹⁷ In retrospective analysis of 40 patients with suspected CAD where cardiac CT (with 64-slice single source, 64-slice dual-source and 128-slice dual source CT) and 1.5 Tesla CMR were performed 7 days apart, excellent correlations were observed between CT and CMR for LV volumes, function, and mass when both modalities were measured with exclusion of the papillary muscles (LV end systolic volume [LVESV] r = 0.98; LV end diastolic volume [LVEDV] r = 0.96; LV ejection fraction (LV EF) r = 0.94; and LV mass r = 0.97). Additionally, applying high-strength noise reduction strategy on post-processed images of electrocardiographic (ECG) tube current modulated datasets improves correlation of EF to CMR and reduces the variability of function and mass measurements by increasing the contrast-to-noise ratio. ¹⁰ In a prospective study of 79 patients who underwent 64-slice single source cardiac CT for CAD evaluation, 1.5 Tesla CMR was performed within 1 week and very good agreement between the two imaging modalities was reported (LV EF: CT $52\pm14\%$ vs. CMR $52\pm14\%$; r = 0.73); LVEDV): CT

74±21 ml/m² vs. CMR 76±25 ml/m²; r = 0.59); LVESV CT 37±19 ml/m² vs. CMR 38±23 ml/m²; r = 0.76). 18

When comparing 16-slice CT in 88 patients with other imaging modalities (TTE and biplane cine-ventriculography) to 1.5 Tesla CMR within 48 hours, global and regional LV function was more accurate with CT than cine-venticulography, and CT was superior to TTE for global function. A meta-analysis of 27 studies using at least 64-slice CT (15 studies comparing CT and TTE and 12 studies comparing CT to MRI) showed no difference in LVEF between CT and CMR nor was a difference seen in LVEF between CT and TTE. In direct comparison with CMR, good correlation has been demonstrated for cardiac CT to estimate LV size, volume and function at a wide range of EF from severely reduced to hyperdynamic function (30–72%), including patients with significant valvular disease or orthotopic heart transplant. A patients

INDICATIONS FOR CARDIAC CT

Per the most recent guidelines, the appropriate use criteria for the assessment of left ventricular (LV) or RV function by cardiac CT include the following²²:

- Assessment of LV function following ACS or HF patients with inadequate images from other noninvasive methods.
- Quantitative assessment of RV function.
- Assessment of RV morphology in suspected ARVC/D.

Assessment of LV function in ACS

The assessment of LV function in the post-ACS setting has prognostic and treatment implications. ^{23, 24} Previous studies have demonstrated good correlation and reproducibility with TTE in patients with CAD for the evaluation of global EF.²⁵ The identification of regional wall motion abnormalities and perfusion defects have shown to have good sensitivity (94%) and specificity (97%) in patients with ACS with excellent interobserver reliability for EF quantification (r = 0.83). Furthermore, the concomitant assessment of ventricular function and wall motion abnormalities by cardiac CT provides considerable incremental value in low-intermediate risk patients with suspected ACS in addition to coronary artery anatomy and perfusion assessment.²⁷ Resting regional LV function has incremental value beyond coronary stenosis for determining ACS, especially in those presenting with acute chest pain. 28 Regional LV function assessment by CT showed an improvement of positive predictive value (PPV) from 35% to 89% in patients with significant stenosis for detecting ACS.^{28, 29} However, this incremental value of having functional datasets must be weighed with the risk-benefit ratio from the concerns of higher radiation exposure with retrospectively-gated scans, ³⁰ and practice pattern change within the cardiac CT community towards prospectively-triggered scans to achieve the lowest radiation dose scan possible in keeping with the "As Low As Reasonably Achievable" ALARA principle at the expense of loss in functional analysis.³¹

Assessment of ventricular function in heart failure (HF)

Assessment of left ventricular systolic function has well known prognostic implications as a predictor of mortality and congestive heart failure. 32, 33 As discussed in previous sections, cardiac CT has been shown to provide accurate and reproducible assessment of LV size, volume and function in patients with HF reduced EF (HFrEF), significant valvular disease, and orthotopic heart transplants. 19-21 For patients with technically difficult studies and/or conflicting assessments of function on multiple non-invasive modalities that have contraindications to CMR, cardiac CT has significant utility in providing accurate and definitive assessment of ventricular function. Some examples of patients include patients with difficult acoustic echocardiographic windows and patients with contraindication to CMR such as those with metal in the eye, intracardiac devices (pacemaker, defibrillators, cardiac resynchronization therapy [CRT]), claustrophobia, and inability to lay flat for a prolonged period of time. For patients with HFrEF with low probability for CAD (i.e. nonischemic cardiomyopathy such as viral etiology), cardiac CT can provide data on LV EF and more importantly be used to exclude the presence of obstructive CAD, making an invasive diagnostic cardiac catheterization unnecessary. 34 With respect to HFrEF patients under evaluation for cardiac resynchronization therapy (CRT) where device implantation is indicated for LV EF of <35% 35, 36 or <30% 37, 38 cardiac CT may in addition be useful for coronary venous assessment before biventricular lead placement, ³⁴ and if a retrospectively ECG-gated scan was performed, CT provided additional information on global and also on regional LV function. One area of research in this cohort is the utility of cardiac CT for assessing intraventricular dyssynchrony to predict the response to CRT and potentially for pre-procedural planning regarding left ventricular lead placement. 39, 40

Furthermore, cardiac CT may play an important role in the assessment of HFrEF patients with left ventricular assist devices (LVADs) and can detect LVAD dysfunction as well as device related complications. Al Although echocardiography is the primary imaging modality to monitor patients with LVADs, it has its own limitations as previously discussed. In addition to anatomic information, a retrospectively-gated scan can evaluate inflow and outflow cannula position and angulation, cannula thrombi, pericannula fluid collection, position of blood pump, pericardial effusion and driveline position. In a small study of 14 patients with LVAD and persistent HF symptoms, retrospectively-gated 64-slice CT was performed and found abnormalities in 8 patients, of which 6 patients had their medical management altered as a direct result of the CT findings and underwent surgical intervention (device exchange and heart transplant).

While the clinical indication for CT in patients with HFrEF is better defined, the utility of CT with respect to patients with HF preserved EF (HFpEF) is less clear. Scant data is available for cardiac CT and its use to assess for diastolic function. A feasibility study of 70 patients who underwent 64-slice CT and 2D TTE with tissue Doppler imaging reported good correlation between cardiac CT transmitral velocity (r = 0.73) and CT mitral septal tissue velocity (r = 0.87) as compared to TTE.⁴² However, larger studies are needed to validate these initial findings.

Assessment of right ventricular function

RV function plays an important role in the pathogenesis of many cardiovascular diseases and serves as an integral diagnostic and prognostic marker in many cardiopulmonary conditions that include CHF, pulmonary hypertension and pulmonary embolism. 43, 44 TTE is the first line imaging modality for the assessment of the RV but can be technically difficult for the same reasons as the LV. In addition, the complex geometry of the RV, its thin walls and heavy trabeculation may further complicate edge recognition. ^{18, 45, 46} CMR is considered the reference standard but has the same contraindications as stated previously. 18 For these patients, cardiac CT is an alternative option for RV function evaluation. Adequate contrast enhancement of the right ventricular lumen is required, but it can typically be achieved by extending the duration of contrast injection by 10 seconds as compared to standard contrast injection protocols for coronary artery imaging. ²² To minimize the overall contrast load, a second phase injection using a reduced flow rate or a diluted mixture of contrast at the same flow rate can be administered. ⁴⁷ Assessment of RV function by cardiac CT has been validated in multiple studies with good accuracy and reproducibility utilizing CMR as the reference standard. 19, 48 A cohort that may frequently benefit from the CTbased assessment of RV volumes and function are the complex adult congenital heart disease patients, such as those after Tetralogy of Fallot repair. These patients frequently have pacemakers or defibrillators, yet the decision for optimal timing of pulmonary valve replacement is heavily dependent on accurate assessment of RV size and function. Pulmonary valve replacement should be undertaken before the RV end-diastolic volume index (RVEDVI) reaches 170 mL/m² or the RV end-systolic volume index (RVESVI) reaches 85 mL/m^{2.49} In patients with chronic pulmonary regurgitation, timing of pulmonary valve replacement should be considered before RVEDVI exceeds 163 mL/m² or RVESVI exceeds 80 mL/m^{2.50}

Assessment of ARVC/D

Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia (ARVC/D) is a rare inherited cardiomyopathy predominantly involving the RV with progressive fibrofatty tissue and presence of ventricular arrhythmias.⁵¹ TTE is the first line imaging modality but is limited in its ability to detect RV dilatation and RWMA.8 Cardiac CT can assess RV dilatation, reduction of RV ejection fraction, severe segmental dilatation, and regional hypokinesis – all part of the major or minor criteria per the original task force criteria of the diagnosis of ARVC/D (Table 1).^{47, 52–54} The 2010 ACC/AHA appropriate use criteria designates cardiac CT as "Appropriate" for the evaluation of suspected right ventricular anomalies such as ARVC/D.²² One study with the use of major or minor criteria showed moderate to excellent sensitivity and positive predictive value. 55 Per the guidelines, RVEDVI 110 mL/m² for males and 100 mL/m² for females or RV EF 40% are part of the major criteria values, while RVEDVI 100 mL/m² for males and 90 mL/m² for females or RV EF 45% are considered part of the minor criteria (Table 1). 56, 57 Additionally, the presence of fatty tissue, bulging appearance, and dilatation of the RV are suggested in a small study of 77 patients to aid in the differentiation of ARVC/D from ventricular tachyarrhythmias.⁵⁵ Similar to the assessment of LV function, the role of cardiac CT for ARVC/D assessment should be reserved for patients with inadequate TTE images and contraindications for CMR.

DATA ACQUISITION PROTOCOLS

Cardiac CT imaging should be performed using at least 64-slice CT scanner technology according to the vendor specific protocol for the evaluation of ventricular function. General recommendations for cardiac CT typically include a minimal contrast flow rate of 5ml/sec, but for specific analysis of ventricular function, lower injection rates may be sufficient. Retrospectively ECG-gated image reconstruction is necessary to reconstruct images through all phases of the cardiac cycle. Tube current modulation techniques are often applied for coronary artery imaging. They limit tube output in systole, a time period that is typically not used for image reconstruction in coronary CT angiography. Even with tube modulation, reconstruction of data sets for functional assessment is usually possible. 30, 58 Functional analysis can also be performed by prospectively ECG-triggered protocols if sufficient "padding" is added to cover most of the cardiac cycle.⁵⁹ However, the benefit of using such protocols over the standard retrospectively ECG-gating protocols with tube current modulation is unclear. The timing of contrast administration is standardized for arterial phase imaging (typically, scan acquisition starts 4–6 seconds after time to peak aortic opacification). Contrast injection for left ventricular and coronary imaging is typically performed for a time period that is equal to the duration of data acquisition (but at least 10 seconds if data acquisition is less than 10 seconds). To obtain adequate RV opacification, an additional 10 seconds of contrast injection should be added but may be given at a slower flow rate (e.g., 2 ml/sec), if the contrast injector permits such injection protocols, to minimize the amount of contrast given to patients.

IMAGE PROCESSING AND EVALUATION

While there lacks data for how best to post-process functional datasets, in our experience the following pointers may be taken into consideration. For functional analysis, a multiphase reformatted dataset of maximally 1.5 mm thick axial images without overlap should be reconstructed at 10% increments (10 phases) for single source CT scanners or 5% increments (20 phases) for dual-source CT scanners throughout the cardiac cycle from the onset of the R-wave for the assessment of global and regional LV function in a cine mode. Thinner slices can be reconstructed with overlap to improve spatial resolution but such fine spatial resolution, while important for coronary artery assessment, is not necessary for ventricular function analysis and would result in unnecessary large Digital Imaging and Communications in Medicine (DICOM) files. The rationale for reconstructing at 10% increments (10 phases) for single source CT scanners is due to the temporal resolution of these scanners of ~165–175 milliseconds (ms). At a heart rate of 60 beats per minute (bpm), at 10 phases of the cardiac cycle, the difference between each phase is 100 ms. Given the temporal resolution of the CT scanner itself, no benefit can be expected from reconstructing more phases. With dual-source CT scanners, the temporal resolution may be as low as 65; therefore, at a heart rate of 60 bpm, reconstructing 10 phases would not make full use of temporal resolution and reconstructing at 5% increments (20 phases) may be recommended.

Global and Regional LV function using a 17-segment model

The 17-segment model (Figure 1) creates a distribution of 35%, 35%, and 30% for the basal, mid-cavity, and apical thirds of the heart respectively.⁶⁰ Global and regional LV function

can be assessed visually with the multiphase reformatted dataset in cine mode to correspond to those used in echocardiography, including LV horizontal long-axis (4-chamber view), vertical long-axis (2-chamber view), LV outflow tract long-axis (parasternal long axis view), and short-axis views. Global LV function is reported as hyperdynamic, normal or reduced (mild, moderate, or severe). Regional wall motion (RWM) of the myocardium is assessed to be normal or abnormal (hypokinesia, akinesia, dyskinesia, aneurysm) in short-axis segments of the LV corresponding to the AHA/ACC/ASE 17-segment model. Abnormalities have to be present in at least two contiguous myocardial segments or in one segment visualized in two different views. Figure 2 demonstrates an example of a significant segmental wall motion abnormality.

Quantification of the left and right ventricles

To quantify cardiac chamber size and function, measurement of the LV internal boundaries are performed at end-diastole and end-systole.⁶¹ External boundaries have to be added to obtain information on myocardial thickening and mass. Figure 3 details a step-wise approach to obtain the correct planes that allow for left ventricular functional measurement. End-diastole can be defined at the onset of the QRS, but is preferably defined as the frame following mitral valve closure or the frame in the cardiac cycle in which the cardiac dimension is largest which is after atrial contraction in patients in sinus rhythm.⁶¹ Endsystole is best defined as the frame preceding mitral valve opening or the time in the cardiac cycle in which the cardiac dimension is smallest. ⁶¹ Not surprisingly, there is prognostic value in CT-based LV volumes and function. In 7758 patients from the Coronary CT Angiography Evaluation for Clinical Outcomes: An International Multicenter (CONFIRM) registry, worsening LV EF, and larger LV volumes predicted mortality and were associated with CAD. 62 In this registry, LV EF was classified as normal (55%), mildly reduced (55%) but 45%), moderately reduced (45% but 35%), and severely reduced (<35%). Abnormal LV end-systolic volume (LVESV) was defined as 90 mL and abnormal LV end-diastolic volume (LVEDV) as 200 mL.⁶²

Scant data is available on normative values for RV volume and function with cardiac CT. A small study of 103 normotensive, nonobese adults undergoing 64-slice CT reported the lower 95% confidence interval (CI) of RV EF to be 42.2%. ⁶³ A larger Multi-Ethnic Study of Atherosclerosis (MESA) of 487 patients undergoing CMR reported sex-differences in RV EF with lower limits of the 95% CI to be 51% in men and 58% in women. ⁵⁷ For most cardiac software package, automated LV function is readily available and more robust while that of RV function is limited and more frequently than not requires manual correction, which can be extremely time intensive.

MANUAL VERSUS AUTOMATED QUANTIFICATION

It remains a trade-off with automated post-processing software versus manual contours for delineating the endocardial and epicardial boundaries with respect to time required for quantification. The development of automated EF assessments has reduced the time needed for post-processing and has shown good agreement for the estimation of EF compared to semiautomated or manual assessments even if significant differences in LVEDV were observed.⁶⁴ Automated post-processing software can automatically delineate the ventricular

chamber throughout systole and diastole (Figure 4), and can provide an accurate and fully automated assessment of global LV function with significantly reduced processing time compared with manual or semi-automated methods.

Depending on the software algorithm, papillary muscles may be included or excluded. The terminology of papillary muscle "inclusion" and "exclusion" is confusing. Most modern automatic software vendors use a blood pool extraction method for extracting volumes, whereby the papillary muscles, due to its different Hounsfield unit from contrast enhanced blood, are not included in determining the volumes and thus are "included" as part of the endocardial tracing, but are "excluded" from the volumetric measurements, yielding smaller volumes. Manual quantification of CT volumes and function, which values have traditionally been the ones compared to CMR, "excludes" the papillary muscles and are typically performed by drawing circular endocardial contours in radial short-axis stacks and applying modified Simpson's summation of discs to calculate volumes. The exclusion of papillary muscles from the endocardial contours yields larger volumes. ^{10, 13–16, 18} The inclusion or exclusion of papillary muscle can change the volumes measured, as well as sex differences for male and female for volumes, but not for EF. 65, 66 In a study that included 179 subjects, significant difference was observed in LV parameters between both papillary muscles included and excluded groups. The difference ranges were 5.6% to 30.1% for LV volumes, 5.8% to 9.4% for LV mass, and 4.3% to 6.0% for LV EF.65 These findings emphasize the importance of including the papillary muscles in the measurement of LV cavity, which is essentially the traditional "exclusion" of the papillary muscle method from the LV volumetric contour. 65, 67-69 There are now software vendors that have capabilities to "exclude" papillary muscles and delineate the endocardial contours similar to that which would be performed manually. These details may seem minutia, but if not aware of the differences may lead to inappropriate over- or under-estimation of reported volumes.

DATA ELEMENTS TO BE INCLUDED IN THE REPORT

Data elements suggested to be included in a report regarding ventricular function are as follows:

- Cardiac morphology of the left and right atria and ventricles.
- Qualitative global LV and RV function (if requested)
- Regional wall motion abnormalities
- Quantitative LV and RV (if requested) ejection fractions and volumes including indexing to body surface area

Table 2 depicts a sample report.

CARDIAC CT LIMITATIONS

Compared to other modalities, assessment of ventricular function by cardiac CT requires radiation exposure and administration of contrast dye. Patients with significant contrast dye allergies or renal insufficiency are not candidates for contrast-enhanced cardiac CT. Premature atrial and ventricular beats and atrial fibrillation can cause arrhythmia and

misregistration artifacts, resulting in poor visualization of the endocardial and epicardial contours as well as high radiation dose due to the scanners' built-in arrhythmia algorithms, which opens the temporal image acquisition window to full tube current when acquiring images in retrospectively-gated scan mode. Severe obesity can compromise image quality and thus reduce diagnostic accuracy or require higher doses of radiation.

CONCLUSION

In addition to the assessment of coronary artery disease, cardiac CT can be utilized to investigate LV function, RV function, and ventricular morphology It serves as alternative option for functional assessment particularly when other imaging modalities such as echocardiography yield inadequate images or in patients with contraindications to CMR.

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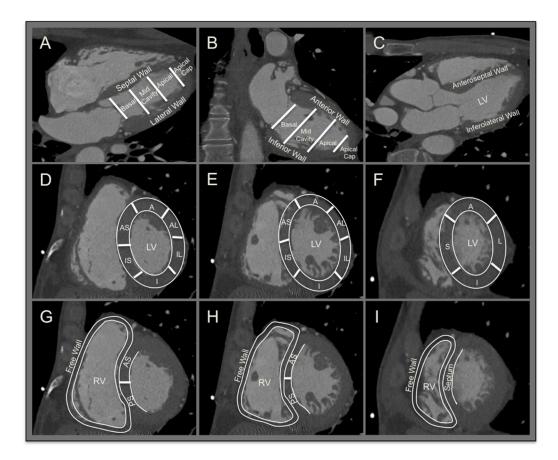


Figure 1. LV and RV Segmentation including the 17-segment Model

A. Four-chamber view showing basal, mid-cavity and apical segments and the apical cap. B. Two-chamber view. C. Vertical long-axis 3-chamber (parasternal long axis) view. D, E and F. Short-axis view of the basal, mid-cavity and apical segments showing LV segmentation; anterior (A), anteroseptal (AS), anterolateral (AL), inferior (I), inferoseptal (IS), inferolateral (IL) and septal (S) segments. G, H and I. Short-axis view of the basal, mid-cavity and apical segments showing RV segmentation; free wall, anteroseptal (AS) and posteroseptal (PS) segments.

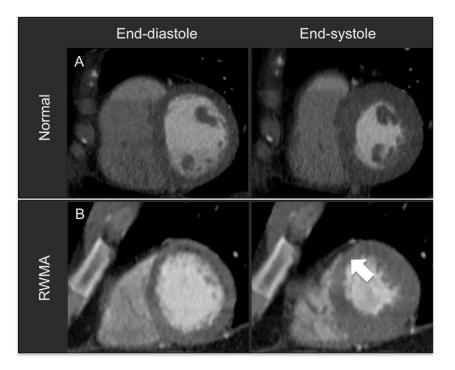


Figure 2. Example of Normal and Abnormal Regional Wall Motion **A.** Short-axis view in end-diastole and end-systole of a normal patient. **B.** Short-axis view in end-diastole and end-systole demonstrating regional wall motion abnormality in anteroseptal segment (white arrow).

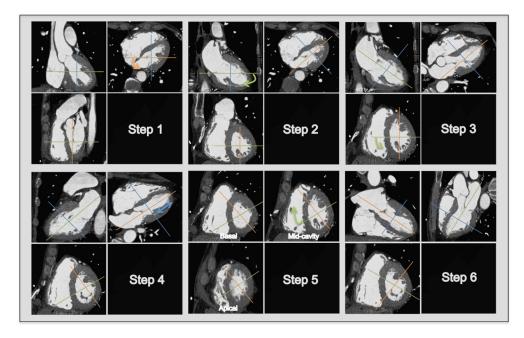


Figure 3. Stepwise Approach in Creating Planes for Ventricular Function Analysis
Step 1. In the axial image, scroll to a modified 4-chamber level and rotate the reference line
(orange) to be parallel to the LV cavity and positioned in the center of the mitral valve
coursing through the apical cap. Step 2. In the 2-chamber view, rotate the reference line
(green) to be parallel to the LV cavity and positioned in the center of the mitral valve
coursing through the apical cap. These first 2 steps will get the double-oblique true shortaxis. Step 3. In the short-axis view at the mid ventricular level, rotate the reference line
(green) towards the RV crux to remove the LV outflow tract (LVOT) from the image to get
the true 4-chamber view and 2-chamber view, which can be used for image interpretation.
Step 4. In the 4-chamber view, move the reference line (blue) up and down to get the basal,
mid-cavity and apical short-axis views for image interpretation. Step 5. In the mid-cavity
short-axis, rotate the reference line (green) towards the 10 o'clock position to bring out the
vertical long-axis 3-chamber (parasternal) view (Step 6).

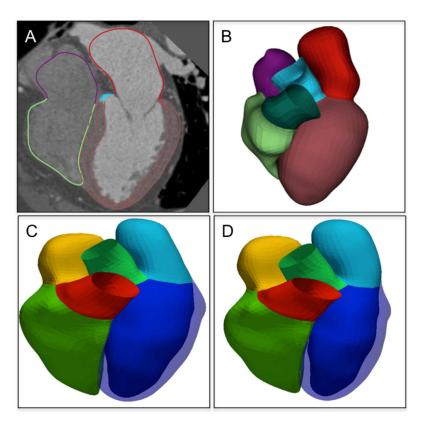


Figure 4. Automated Heart Chamber Segmentation **A.** Four-chamber segmentation with multiplanar reformatting. **B–D.** Three-dimensional (3D) reconstruction of reformatted 4-chamber segmentation (B) at end-diastolic phase (C) and end-systolic phase (D). Courtesy of Guanglei Xiang PhD.

Table 1

Revised Task Force Criteria for the Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/ Dysplasia (ARVC/D)⁵⁴

I. Global or regional dysfunction and structural alterations

Major criteria

By 2D echo:

- · Regional RV akinesia, dyskinesia, or aneurysm
- and 1 of the following (end diastole):
 - PLAX RVOT 32 mm (corrected for body size [PLAX/BSA] 19 mm/m²)
 - PSAX RVOT 36 mm (corrected for body size [PSAX/BSA] 21 mm/m²)
 - or fractional area change 33%

By MRI:

- Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
- and 1 of the following:
 - Ratio of RV end-diastolic volume to BSA 110 mL/m² (male) or 100 mL/m² (female)
 - or RV ejection fraction 40%

By RV angiography:

· Regional RV akinesia, dyskinesia, or aneurysm

Minor criteria

By 2D echo:

- Regional RV akinesia or dyskinesia
- and 1 of the following (end diastole):
 - PLAX RVOT 29 to <32 mm (corrected for body size [PLAX/BSA] 16 to <19 mm/m²)
 - PSAX RVOT 32 to <36 mm (corrected for body size [PSAX/BSA] 18 to <21 mm/m²)
 - or fractional area change >33% to 40%

By MRI:

- Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
- and 1 of the following:
 - $\qquad \text{Ratio of RV end-diastolic volume to BSA} \quad 100 \text{ to } < 110 \text{ mL/m}^2 \text{ (male) or} \quad 90 \text{ to } < 100 \text{ mL/m}^2 \text{ (female)}$
 - or RV ejection fraction >40% to 45%

II. Tissue characterization of wall

Major criteria

Residual myocytes <60% by morphometric analysis (or <50% if estimated), with fibrous replacement of the RV free wall
myocardium in 1 sample, with or without fatty replacement of tissue on endomyocardial biopsy

Minor criteria

Residual myocytes 60% to 75% by morphometric analysis (or 50% to 65% if estimated), with fibrous replacement of the RV free
wall myocardium in 1 sample, with or without fatty replacement of tissue on endomyocardial biopsy

III. Repolarization abnormalities

Major criteria

 Inverted T waves in right precordial leads (V₁, V₂, and V₃) or beyond in individuals >14 years of age (in the absence of complete right bundle-branch block QRS 120 ms)

Minor criteria

- Inverted T waves in leads V_1 and V_2 in individuals >14 years of age (in the absence of complete right bundle-branch block) or in V_4 , V_5 , or V_6
- Inverted T waves in leads V₁, V₂, V₃, and V₄ in individuals >14 years of age in the presence of complete right bundle-branch block

IV. Depolarization/conduction abnormalities

Major criteria

• Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads $(V_1 \text{ to } V_3)$

Minor criteria

- Late potentials by signal average ECG in 1 of 3 parameters in the absence of a QRS duration of 110 ms on the standard ECG
- Filtered QRS duration (fQRS) 114 ms
- Duration of terminal QRS <40 μV (low-amplitude signal duration) 38 ms
- Root-mean-square voltage of terminal 40 ms $20 \mu V$
- Terminal activation duration of QRS 55 ms measured from the nadir of the S wave to the end of the QRS, including R', in V₁, V₂, or V₃, in the absence of complete right bundle-branch block

V. Arrhythmias

Major criteria

• Nonsustained or sustained ventricular tachycardia of left bundle-branch morphology with superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL)

Minor criteria

- Nonsustained or sustained ventricular tachycardia of RV outflow configuration, left bundle-branch block morphology with inferior axis (positive QRS in leads II, III, and aVF and negative in lead aVL) or of unknown axis
- >500 ventricular extrasystoles per 24 hours (Holter)

VI. Family history

Major criteria

- · ARVC/D confirmed in a first-degree relative who meets current Task Force criteria
- ARVC/D confirmed pathologically at autopsy or surgery in a first-degree relative
- Identification of a pathogenic mutation categorized as associated or probably associated with ARVC/D in the patient under evaluation

Minor criteria

- History of ARVC/D in a first-degree relative in whom it is not possible or practical to determine whether the family member meets current Task Force criteria
- Premature sudden death (<35 years of age) due to suspected ARVC/D in a first-degree relative
- ARVC/D confirmed pathologically or by current Task Force Criteria in second-degree relative

PLAX: parasternal long-axis view; RVOT: RV outflow tract; BSA: body surface area; PSAX: parasternal short-axis view; aVF: augmented voltage unipolar left foot lead; and aVL: augmented voltage unipolar left arm lead.

Diagnostic terminology for original criteria: The diagnosis of ARVC/D is fulfilled by the presence of 2 major, or 1 major plus 2 minor criteria or 4 minor criteria from different groups. Diagnostic terminology for revised criteria: Definite diagnosis: 2 major or 1 major and 2 minor criteria or 4 minor from different categories; Borderline: 1 major and 1 minor or 3 minor criteria from different categories; Possible: 1 major or 2 minor criteria from different categories.

Table 2

Sample Cardiac CT Report of Ventricular Morphology and Function

CARDIAC MORPHOLOGY AND FUNCTION: The right and left atria and ventricles are morphologically normal. Normal qualitative global LV function. No regional wall motion abnormality.

Calculated BSA: m²

LVEF: %

LV end diastolic volume: cc

LV end diastolic volume index: cc/m²

LV end systolic volume: cc

LV end systolic volume index: cc/m²

RVEF: %

RV end diastolic volume: cc

RV end diastolic volume index: cc/m²

RV end systolic volume: cc

RV end systolic volume index: cc/m²