Because of major advances in congenital heart surgery and intensive care during the past 60 years, the number of adults living with congenital heart disease (CHD) has risen sharply. Currently, in the United States, there are more adults living with CHD than children.\textsuperscript{1} The estimated prevalence of adult congenital heart disease (ACHD) is \( \approx \) 3000 per million adults,\textsuperscript{2} and this number is expected to increase by 5\% per year.\textsuperscript{3} The late sequelae associated with surgical and nonsurgical palliation include valvular degeneration and regurgitation, ventricular systolic and diastolic dysfunction, and arrhythmias. For the vast majority of patients with moderate-to-complex congenital heart disease, serial cardiovascular imaging is required to monitor structural changes to determine need and timing of repeat intervention.

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In many patients with CHD, the right heart is most affected. Despite life-saving palliation, the right ventricle (RV) may experience persistent pressure or volume overload, and there may be intrinsic RV myopathy or processes that result in the RV functioning as the systemic or subaortic ventricle (Table 1). It has been demonstrated that patients with RV enlargement, dysfunction, and fibrosis have greater exercise limitation and are at risk for RV failure and lethal ventricular arrhythmias.\textsuperscript{4} There is a trend to recommend earlier reintervention to preserve RV function and decrease risk of chronic right heart failure.\textsuperscript{5} Careful quantification of RV size and function is critical in determining optimal timing of catheter-based intervention or surgical correction.

During the past decade, there have been many advances in noninvasive RV imaging. Echocardiography, cardiac computed tomographic angiography (CTA), and cardiac magnetic resonance (CMR) are the modalities of choice for evaluating the RV, each with unique advantages and short falls. CMR is currently the gold standard for quantifying RV function in patients with CHD because of its high level of spatial resolution, with precise definition of complex anatomy and excellent reproducibility. However, CMR is costly, requires patients to lie still for an extended period of time, expertise necessary for performing and interpreting CMR in this population is not widely available, and patients with devices such as pacemakers or implantable defibrillators may not be eligible. Echocardiography has traditionally provided mostly qualitative assessment of RV size and function but is far more widely available at significantly lower cost, and therefore, the most
practical method by which to perform serial assessments. We propose that recent advances in echocardiographic evaluation of RV function and standardization of RV measurements can rival CMR in quantitative RV assessment.

Echocardiographic Methods to Assess Subpulmonary Right Ventricular Function

Echocardiographic assessment of RV systolic function has largely been qualitative, relying on visual assessment of the longitudinal motion of the tricuspid annulus in the 4-chamber view and inward motion of the RV free wall and apex. This qualitative assessment or eyeball method is clearly limited when compared with quantitative assessment of systolic function with CMR.6 As a result, CMR has been considered the best available method for assessing RV function.7,8 In 2010, guidelines were developed for quantitative echocardiographic assessment of the right heart in adults.9 These guidelines provide standards for linear 2-dimensional (2D) volumetric and nonvolumetric assessment of RV function (Table 2). These guidelines have revolutionized the echocardiographer’s ability to provide standardized quantitative RV measures to monitor change with time. We review some of these parameters in detail including literature comparing these methods with CMR in the CHD population where available.

Right Ventricular Size

The RV geometry is more complex than that of the left ventricle (LV). It wraps around the LV, forming a crescent shape, and is defined by 3 distinct sections: an inlet portion, a trabecular portion involving the body and apex, and an outflow or infundibular portion generally free of trabeculation.4 The geometric complexity has limited the development of volumetric measurements of the RV.

Although widely used, 2D linear dimensions alone have not been shown to correlate well with CMR.10 Recent work by Greutmann et al11 evaluated a simple mathematical model using 2D echocardiographic parameters (Figure 1) to estimate CMR-derived RV ejection fraction (RVEF) and demonstrated
that quantitative assessment of RV end-diastolic volume index (RVEDVi) by echo is feasible and strongly correlates with RV EDDi measured by CMR in adults with tetralogy of Fallot (TOF) ($r=0.87; \ P<0.001$). Using the Bland–Altman method, no systematic error between CMR RVEDVi and the calculated RVEDVi derived by the 2D model was found (difference between RVEDVi means, 2.8 mL/m²). The 95% limits of agreement were −46 and +52 mL/m². In this study of 67 randomly selected patients, 8% were noted to have poor acoustic windows; however, no studies were excluded because of inadequate acoustic windows. Importantly, among patients in whom 2D echocardiographic estimated either underestimated or overestimated CMR RV EDDi at baseline, follow-up 2D imaging demonstrated similar degree of underestimation or overestimation, suggesting that echocardiography can be used for serial imaging to determine interval change and allow a greater interval between routine CMR studies among patients with TOF being followed for severe pulmonic regurgitation. The greatest promise for echocardiography is in 3D applications, which lead to more accurate estimation of RV volumes, less underestimation of RV end-diastolic and end-systolic volumes, and improved test–test variation when compared with 2D echocardiography.9 Right ventricular volume assessment by real-time 3D echocardiography has been validated in vitro against displacement of water with outstanding correlation ($r=0.99; \ P<0.001$).12 According to most recent guidelines, the lower reference limit for 3D-derived RVEF is 44% and the upper limit for RV EDDi is 89 mL/m².9 3D echocardiography is feasible with high rates of complete data acquisition in consecutive patients with ACHD ranging from 89% to 97% in recent studies,13,14 and assessment of the RV has been studied in both children and adults with CHD with significant correlation with CMR.15,16 One study evaluated 29 pediatric patients with single ventricle anatomy and excellent correlation of 3D echocardiography with CMR end-diastolic volume ($r=0.96; \ P<0.001$).15 Grewal et al16 evaluated the accuracy of 3D echocardiography compared with CMR in adult patients with repaired TOF and severe pulmonic regurgitation. Among 25 patients, there were strong correlation of both end-diastolic volume as well as end-systolic volume on 3D echocardiography and CMR ($r=0.88$ and $r=0.89$, respectively). Van der Zwaan et al18 published similar findings of excellent correlation among patients with ACHD with 3D echocardiography versus CMR end-diastolic volume correlation ($r=0.93$).

Limitations to 3D echocardiography include the following: techniques for volume quantification may be time consuming, suboptimal image quality may lead to incomplete data sets and decrease feasibility in some patients, and methods are operator dependent and require specialized software and training. Despite these limitations, 3D techniques are promising, and greater automation should lead to more widespread clinical adoption.

### Fractional Area Change and 3D RVEF

Fractional area change of the RV, defined as (end-diastolic area−(end-systolic area)/(end-diastolic area))×100; Figure 2), is a simple measure of RV systolic function that includes only the body and apex of the RV. Nevertheless, fractional area change has been shown to correlate with RVEF by CMR ($r=0.8$) in the general population but correlates less well in the CHD population.10,11,12,13 Given the complex geometry of the RV, 3D assessment provides much more accurate assessment of RVEF, and a recent study demonstrates excellent correlation of 3D RVEF with CMR RVEF in a cohort of 30 patients with TOF ($r=0.93; \ P<0.001$).14

### Tricuspid Annular Plane Systolic Excursion

The contractile pattern of the RV differs from that of the LV in that the RV mainly relies on longitudinal shortening rather than circumferential contraction. The tricuspid annular plane systolic excursion (TAPSE) is a method to measure the distance of systolic excursion of the RV annular segment along its longitudinal plane, from a standard apical 4-chamber view, representing longitudinal function of the RV. Importantly, when several markers of RV systolic function were studied in a general population, TAPSE was the only RV function parameter predicting survival.21 The advantages of TAPSE include its simplicity, lack of dependence on optimal image quality, and reproducibility. It can be performed with basic 2D echocardiography and does not require sophisticated equipment or software. Current guidelines recommend routine use of this simple method to assess RV function, with values <16 mm considered impaired.9 One recent study evaluating TAPSE in patients with CHD with either pulmonary hypertension or RV volume overload (TOF) demonstrated negative correlation with CMR RV EDDi ($r=−0.67$ and 0.42, respectively; $P<0.001$) and positive correlation with CMR RVEF (0.81 and 0.65, respectively; $P<0.001$), demonstrating that TAPSE is useful in both pressure- and volume-loaded CHD.14

Figure 2. Right ventricular (RV) fractional area change: endocardial border is traced in the 4-chamber view in patients with normal RV function (left), mildly reduced (middle), and severely reduced RV function (right). Reprinted from Rudski et al9 with permission from Copyright © 2010, American Society of Echocardiography. Authorization for this adaptation has been obtained both from the owner of the copyright in the original work and from the owner of copyright in the translation or adaptation.
Doppler-Based Measures

Tissue Doppler Imaging

Tissue Doppler imaging (TDI) provides insight into global and regional myocardial systolic and diastolic function. Velocities within the myocardium can be measured during cardiac cycle, and measuring these tissue Doppler velocities in various regions within the myocardium allows assessment of regional myocardial function. Color tissue Doppler was introduced as an alternative technique for measuring tissue velocities, with the advantage that tissue velocities can be documented simultaneously in different myocardial segments during the cardiac cycle. This has been demonstrated among healthy subjects that TDI is feasible for the assessment of RV function by using color TDI to measure regional velocities at the tricuspid valve annular, basal, mid, and apical regions of the RV free wall. Pulsed-wave TDI at the lateral tricuspid annulus allows the recording of RV peak myocardial contraction velocity (S'). In one study, RVEF <50% by CMR was best detected by S' <11 cm/s by TDI, and in another study S' <8.8 cm/s predicted an RVEF <45%. TDI confers an advantage in CHD in that these techniques can be applied to any chamber morphology and do not rely on assumptions of chamber geometry. Importantly, TDI may detect RV dysfunction in patients with apparently normal function as assessed by conventional echocardiographic parameters. Among 40 adult patients with repaired TOF, reduction in RV early diastolic velocity seems to be an early abnormality and is associated with occurrence of arrhythmic events, and may be useful in risk stratification.

Limitations of TDI include limited reliability of measured velocities under different loading conditions, angle dependence, and the assumption that function of a single segment represents the function of the entire RV.

Myocardial Performance Index

The myocardial performance index (also known as MPI, RIMP, or Tei index) is another method to measure global RV systolic and diastolic function. It is based on the relationship between ejection and nonejection work of the heart. It measures the ratio of the isovolumic time intervals to ventricular ejection time (IVRT+IVCT/ET; Figure 3). This ratio increases with worsening ventricular function as the isovolumic phase lengthens and reflects ventricular-vascular interaction. In a study of >200 patients with RV dysfunction, MPI >0.50 correlated with RVEF ≤30% by CMR (sensitivity 95%, specificity 85%). This measure has proved to be a useful indicator of RV function among patients with CHD. TDI-Tei index (TDI-MPI) differs from the conventional MPI in that it can be measured in a single cardiac cycle, which improves accuracy. This measure has been used in postoperative patients with TOF with negative linear correlation between the MPI and the RVEF by CMR (r=0.73; P<0.001). The advantage of the MPI is that it is independent of ventricular geometry and has low interobserver and intraobserver variabilities. Other advantages include its feasibility in most patients and good reproducibility. Disadvantages include the load-dependence of this measure, lack of reliability in patients with atrial fibrillation and elevated right atrial pressure.
Strain
Strain is defined as percentage change in myocardial deformation; whereas strain rate represents the rate of deformation of the myocardium with time. Strain rate has been closely correlated with myocardial contractility in in vitro and in vivo experimental settings. Decrease in longitudinal peak systolic strain may occur before detectable changes in RVEF, suggesting measurements of longitudinal peak systolic strain may be used as an early marker of dysfunction. Echocardiographic speckle-tracking strain–analysis may be a sensitive method to detect early deterioration of RV performance and strain at the lateral free wall as obtained by speckle-tracking echocardiography correlated with RVEF as assessed by CMR. Advantages of use of strain include relative independence on loading conditions and its applicability in a wide range of pathologies. Disadvantages include requirement for additional software, lack of normative data, and high degree of variability because the measures are complex. Angle dependence is an issue with tissue Doppler–based methods but not with speckle tracking.

RV Diastolic Function
Right ventricular compliance may be reduced in many patients with CHD. Conditions that lead to reduced compliance are usually, but not always, associated with RV pressure overload either because of RV outflow obstruction or pulmonary vascular disease. However, relative hypoplasia of the RV may occur in patients with palliated pulmonary atresia and intact ventricular septum, palliated tricuspid stenosis, and unbalanced atroventricular canal defects. In Ebstein’s anomaly, the effective RV size may be significantly reduced, and therefore overall chamber compliance is reduced. Current guidelines suggest evaluating RV diastolic function via transtricuspid E/A ratio, E/E’ ratio, and right atrial size, and these methods have been validated in acquired heart disease. Another Doppler indicator of poor RV compliance is the presence of antegrade forward flow into the pulmonary artery during atrial contraction (Figure 4). MPI encompassed both systolic and diastolic assessments of RV function as detailed above.

Exercise RV Function
It is clear that exercise capacity correlates with outcomes among patients with CHD. There is little data to assess echocardiographic or CMR measures for RV function at peak exercise in the ACHD population, and this is an area for potential fruitful investigation.

Assessing Systemic RV Function
Current echocardiographic guidelines for RV quantification do not apply to the systemic RV, and accurate measurement of ventricular function remains a challenge. In the systemic RV, the circumferential wall shortening is more predominant than longitudinal shortening, resulting in a contraction pattern of the systemic RV that is more similar to the contraction pattern of an anatomic LV rather than an anatomic RV, and echocardiographic global and longitudinal function variables do not correlate with CMR RVEF.

Among patients with systemic RVs, delineating the endocardial boundaries of the heavily trabeculated and hypertrophied RV may be challenging, and variability in tracing the endocardial borders can significantly affect the volumes measured even with CMR. However, echocardiography and CMR were directly compared in assessing function of systemic RV in 37 adults with D-transposition of the great arteries and previous atrial switch operation. Fractional area change <33% and RV pressure dP/dt <1000 mm Hg/s could identify RVEF < 50% with a sensitivity of 77% and 69% and a specificity of 58% and 87%, respectively.

Several novel echocardiographic techniques have been shown to be useful in assessing systemic RV function. Strain indices of the systemic RV have been an evolving area of interest recently. In patients with D-transposition of the great arteries after atrial switch surgery, measurement of global longitudinal strain rate based on speckle-tracking imaging significantly correlates with CMR-derived RVEF (r=0.62; P<0.001) and may be a potential useful index for serial echocardiographic assessment of the systemic RV function. In another study, reduced longitudinal global strain of the systemic RV was associated with increased risk for clinical events.

Echocardiography as a Mainstay in Monitoring RV Function in ACHD
Adult patients with CHD require serial monitoring to determine optimal timing for intervention, and echocardiography is the most widely applied noninvasive imaging technique for the routine diagnosis and follow-up. Imaging limitations because of suboptimal acoustic windows may decrease sensitivity in assessing the RV. Moreover, traditional qualitative assessment of RV function is clearly inadequate in serial monitoring of RV function in the ACHD population. Fortunately, echocardiographic quantification of RV size and function has rapidly evolved during the past 2 decades. Novel echocardiographic imaging modalities have emerged, which significantly improve accuracy of RV functional assessment and correlate with CMR-derived RVEF as discussed above. These may become valuable indicators of ventricular performance, compliance, and disease progression, which may rival routine use of CMR for RV assessment and monitoring.

Although CMR clearly provides exceptional spatial resolution and excellent reproducibility in assessing complex congenital heart anatomy and ventricular function, echocardiography has the advantage of significantly lower cost and much greater availability compared with CMR.

It is clear that during the past 15 years cardiac imaging has become one of the highest cost items in the medical budget of a health plan and one of the fastest growing items. There is wide variability in imaging cost; however, average cost of CMR in the United States is $74040 and may be more for the complex series of images and analyses required in patients with ACHD. When comparing cost of various cardiac imaging modalities, echocardiography is the most cost-effective resting cardiac...
imaging. If the average cost (not charge) of an echocardiogram is equal to 1 (as a cost comparator), the cost of a CTA is 3.1X, and CMR 5.51X, and right and left heart catheterization is 19.96X. This comparison assumes the cost of a standard 2D echocardiogram, and congenital echocardiography with 3D image acquisition is more costly at most centers. At one of our institutions (Dr Yeh) charges for a congenital CMR focused to evaluate RV volumes for TOF are 3X that of a congenital echocardiogram with 3D image acquisition for the same indication. Although the clear benefits of superior spatial resolution are evident for definition of complex congenital anatomy and precise assessment of ventricular systolic function, this must be carefully weighted with greater expense.

Once complex congenital anatomy has been clearly defined, echocardiography may be used for serial monitoring. One study evaluated the tailored use of CMR among 94 repaired patients with TOF who required serial RV monitoring. These authors found that indexed end-diastolic RV diameter and area on echocardiography were significantly associated with RV dilation on CMR (P<0.001). Arrhythmia history and increased MPI were significantly associated with low RVEF (P<0.001 and P=0.017, respectively). It has been proposed that close echo monitoring could reduce the use of serial CMR and result in significant cost savings.

Other limitations to routine use of CMR include much lower availability nation-wide and lack of portability; ≈2% of patients who undergo magnetic resonance imaging may experience significant claustrophobia, and younger patients may require sedation with anesthesia. An increasing proportion of ACHD patients may have contraindication to CMR because of certain types of pacers and implantable defibrillators, or other implantable devices such as stents, coils, stent-mounted valves, septal defect occluders, and surgical clips, which may significantly compromise image quality, and arrhythmia may also limit image acquisition. Additionally, scan times are longer ranging from 45 minutes to 2 hours or longer for more complex CHD cases. Finally, gadolinium carries some, although small, risk for nephrogenic systemic fibrosis, and caution must be used among patients with significant renal disease.

Cardiac CTA imaging is a rapidly growing modality and has been studied in the ACHD population; however, ionizing radiation exposure with cardiac CTA imaging is a major limitation for serial use with time. Although some centers are able to scan CHD patients and evaluate systolic ventricular function with significantly lower levels of radiation than previously reported, this is not currently widespread practice but does provide a promising alternative for assessment of RV function in time.

Echocardiography remains the noninvasive modality of choice for assessing and monitoring CHD. It is recommended to be the initial imaging modality for evaluation of patients with suspected CHD and should be performed before other modalities are used. We propose that among patients with RV pathology or risk for dysfunction, CMR and echocardiography could be initially performed to determine correlation and define anatomy, and among patients who require serial follow-up for assessment of RV parameters, echocardiography should be subsequently performed, and when significant changes are suspected, repeat CMR may be considered. Appropriate use of quantification and novel echocardiographic techniques correlate well with RVEF by CMR and should minimize high cost associated with serial CMR imaging.

Conclusions

The RV has long been been the forgotten ventricle and in 2006 the National Heart, Lung, and Blood Institute identified RV physiology as a priority in cardiovascular research. Accurate quantification of RV function is critical to appropriate determination of surgical timing and postoperative monitoring in patients with ACHD. CMR offers advantages compared with 2D echocardiography in terms of excellent spatial resolution, volumetric quantification, and delineation of complex anatomy; however, echocardiography remains a primary modality for assessment of RV systolic and diastolic functions with rapidly evolving novel techniques such as 3D imaging, TDI, and strain imaging that are quickly challenging the leading role of CMR in RV functional assessment, and come with increased availability and significantly lower cost.

Disclosures

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Response to Defaria Yeh and Foster

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I agree with Drs DeFaria Yeh and Foster’s statements that in patients with congenital heart disease, “Careful quantification of RV size and function is critical in determining optimal timing of catheter-based interventions or surgical correction” and that “CMR is currently the gold standard for quantifying RV function in patients with congenital heart disease because of its high level of spatial resolution, with precise definition of complex anatomy and excellent reproducibility.” Thus, the crux of the debate is not whether cardiac magnetic resonance (CMR) is the preferred method for evaluation of right ventricular size and function in patients with congenital heart disease (we all agree on that); instead, the challenge clinicians face is how to use CMR and echocardiography in a complementary fashion that leverages the strengths of each modality while striving to reduce resource use. This topic is addressed in both DeFaria Yeh and Foster’s article and in mine. It is important to note that both echocardiography and CMR continue to evolve. Recent advances in myocardial deformation imaging by both modalities, improved CMR techniques for assessment of myocardial characteristics (ie, scar tissue and diffuse fibrosis), and novel methods for evaluation of mechanical dyssynchrony are just few examples that highlight the dynamic nature of our field. Therefore, optimal application of a multimodality approach in patients with congenital heart disease will require continued critical evaluation of current and future imaging techniques.
Is MRI the Preferred Method for Evaluating Right Ventricular Size and Function in Patients With Congenital Heart Disease: MRI Is Not the Preferred Method for Evaluating Right Ventricular Size and Function in Patients With Congenital Heart Disease

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