

## Tetralogy of Fallot: in good shape?

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Tetralogy of Fallot (TOF) is the most common cause of cyanotic congenital heart disease and is associated with a high prevalence of pulmonary regurgitation following repair often requiring later pulmonary valve replacement (PVR) [1–13]. Adults with repaired TOF and significant chronic pulmonary regurgitation are at risk for progressive right ventricular (RV) dilatation and dysfunction [14–23]. Helbing et al. [1] showed that impaired relaxation and restriction to filling affected RV function in patients with repair of TOF and pulmonary regurgitation. Uebing et al. [2] demonstrated that RV end-systolic volume is a useful measure for estimating RV function after TOF repair depicting parameters of

systolic and diastolic RV function. Assessment of RV function is important in the management of these patients in particular when it comes to optimal timing of surgery for PVR. Timing of surgery must be carefully considered, weighing the up-front risks of surgery and possible repeat surgery against the risk of ongoing pulmonary regurgitation [24, 25]. Therefore, monitoring RV volume and function is useful for managing patients with TOF.

Detailed and precise evaluation of RV function by cardiac imaging has been a long-standing challenge in clinical medicine. Two-dimensional (2D) imaging has been used to this purpose, but the complex shape of the RV has precluded accurate estimation of RV volume by 2D echocardiography using geometric models. Accordingly, all 2D echocardiographic methods show a rather poor performance in comparison with cardiovascular magnetic resonance imaging (CMR) [26]. Consequently, three-dimensional (3D) imaging methods combined with analysis using the multiple slice technique are more accurate because reliance on geometric modeling is eliminated. Over the past years, a vast experience had been built with CMR to investigate LV and RV dynamics [27–41]. As such, CMR has become the gold standard for quantitative analysis of RV anatomy, function, and shape [42–47].

Kayser et al. [48] evaluated the effect of through-plane motion on tricuspid flow measurements performed with CMR velocity mapping in nine normal subjects and 15 patients with RV disease. Eight

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Editorial comment to the paper entitled “Correlation of right ventricular ejection fraction and tricuspid annular plane systolic excursion in tetralogy of fallot by magnetic resonance imaging” by Sheehan. Doi: [10.1007/s10554-008-9387-0](https://doi.org/10.1007/s10554-008-9387-0).

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parameters of RV diastolic function were derived from the tricuspid flow measurements, both before and after a correction for through-plane motion. Measurements of E-peak, A-peak, and time-to-peak filling rate changed significantly after correction for through-plane motion. Tricuspid flow as a marker of RV diastolic function should be corrected for the effect of through-plane motion to improve functional evaluation of the RV.

Greenberg et al. [49] recently identified E- and A-wave flow patterns across the tricuspid valve in TOF patients. Results from CMR phase contrast velocity-encoded flow quantification correlated well with measurements of right ventricular enlargement. The authors studied 33 children following TOF repair who had CMR examinations that included cine imaging to quantify ventricular size and function and flow analysis across the atria-ventricular valves to evaluate ventricle in-flow patterns. A reduction in the E:A wave ratio across the tricuspid valve was associated with RV diastolic dysfunction and correlated well with RV enlargement. As a result, reduction in the E:A wave ratio across the tricuspid valve was considered a new useful criterion for determining the timing of valved pulmonary conduit surgery in children following TOF repair.

Over the past years, evaluation of RV shape has been proposed using tricuspid annular plane systolic excursion (TAPSE). Previous 2D echocardiographic studies have shown that a TAPSE value of less than 18 mm was associated with increased RV systolic dysfunction [50], whereby a TAPSE of less than 15 mm was associated with an adverse prognosis [51]. TAPSE has been shown to correlate highly with RV ejection fraction in normal subjects and patients with ischemic heart disease [52]. However, correlations between TAPSE and RVEF using CMR have not previously been studied in patients with repaired TOF.

In the current issue of the *International Journal of Cardiovascular Imaging*, Morcos et al. [53] evaluated whether TAPSE measurements by CMR correlated with RVEF in surgically repaired TOF patients. TAPSE was measured from systolic displacement of the RV-free wall/tricuspid annular plane junction in the apical 4-chamber view in 7 normal subjects and 14 TOF patients. All TOF patients had signs of tricuspid regurgitation. Because the authors had previously observed discrepancy between TAPSE

and RV ejection fraction in the presence of regional dysfunction [54], they also analyzed RV wall motion in terms of regional stroke volume at 20 short axis slices from apex to tricuspid annulus. TAPSE assessed with CMR proved to be an unreliable measure of RV ejection fraction in patients with repaired TOF. These findings are in line with previous 3D echocardiographic studies which also demonstrated a weak correlation with RV ejection fraction and TAPSE [55]. The different observations made in other previous studies [42, 43] might be due to (1) the exclusion of the RV volume out of the plane in the 4-chamber view in case of 2D imaging, (2) the absence of tricuspid regurgitation, or (3) the presence of regional wall dysfunction.

In a recent study by the same group, Sheehan et al. [56] showed that the RV remodels in several directions rather than following a shape continuum. To that purpose, 15 patients with repaired TOF and 8 normal subjects by CMR in long- and short-axis views were evaluated. The RV was constructed in three dimensions using the piecewise smooth subdivision surface method. Shape was analyzed from cross-sectional contours generated by intersecting the RV with 20 planes evenly spaced from apex to tricuspid annulus. RV shape in patients with TOF differed from normal subjects in several ways. First, the RV had a larger normalized cross-sectional area in patients with TOF. Second, the cross-sectional shape was rounder in patients with TOF. In addition, the RV in patients with TOF exhibited bulging basal to the tricuspid valve which was amplified by tilting of the tricuspid annulus. Consequently, characterization of RV remodeling from 3D reconstructions provides novel insights.

In conclusion, quantitative RV shape analysis faces the same problems as volume analysis—the geometric models developed for the LV can not be applied to the RV. Neither radial nor rectangular coordinate system methods fit the RV in long or short axis views. 3D imaging methods using the multiple slice technique are more accurate because reliance on geometric modeling is eliminated. TAPSE provides useful information on RV function and shape but appears of limited use in conditions that exhibit abnormal regional contraction such as in patients with TOF. Further studies of TAPSE in conditions that alter RV anatomy, wall motion pattern, and/or tricuspid function are therefore warranted.

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