Right Heart-Pulmonary Circulation Unit in Congenital Heart Diseases

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KEYWORDS

• Congenital heart disease • Right ventricle • Tetralogy of Fallot • Ebstein anomaly

KEY POINTS

- A multimodality combination is currently a mandatory diagnostic approach for the evaluation of the right ventricle in congenital heart disease, particularly in the adult population.
- Echocardiography is a first-line technique; advanced echocardiographic modalities (speckle-tracking echocardiography and 3-D echocardiography) have added new physiopathologic and prognostic information.
- Cardiac MRI and cardiac CT are helpful in providing clinically relevant information for the follow-up and management of these patients.

INTRODUCTION

The main goal of cardiology is to assess ventricular function. Considerable knowledge has been achieved in the study of the left ventricular (LV) function; conversely, the assessment of right ventricular (RV) function is still a challenge. Recent developments in cardiac imaging and the more frequent multimodality approach, involving echocardiography, cardiac magnetic resonance (CMR), and CT, have deeply changed the understanding of RV anatomy and function. There are still numerous questions to be answered, however, regarding the RV function and its contributions to cardiovascular disease prognosis, particularly in the field of congenital heart disease. This article focuses on the RV mechanics in tetralogy of Fallot (ToF), Ebstein anomaly, and systemic RV by using a multimodality approach.

THE RIGHT VENTRICLE IN TETRALOGY OF FALLOT

Besides single ventricle lesions and complete transposition of the great arteries (TGA), ToF is the most common severe congenital heart defect, with a reported prevalence of 0.34 per 1000 live births worldwide.[1]

The main morphologic features comprise anterocephalad deviation of the muscular outlet septum and hypertrophy of the septoparietal trabeculations. Both together create a muscular subvalvar RV outflow tract (RVOT) obstruction. There is typically a large malalignment ventricular septal defect (VSD) with overriding of the aorta, which so has a biventricular origin. The pulmonary valve is often small and stenotic.[2,3]

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Surgical Management and Considerations

Surgical management strategies have emerged over time since Lillehei and colleagues[4] reported the first complete ToF repair in 1955. At the beginning, strategies were based on transventricular closure of the VSD, extensive resection of the RVOT obstruction, and generous use of a transannular patch.[3,4] These had the consequences of chronic pulmonary insufficiency, RVOT aneurysms, and akinetic RV regions,[5] altogether associated with RV dilatation and adverse function.[6] Studies in the past century using a transatrial-transpulmonary access led to surgical improvements by avoiding or minimizing RV incision.[7,8] Further changes are, above others, attributed to data showing that a restrictive enlargement of the pulmonary annulus does not lead to significant RV pressure load but can lower the transannular patch rate and minimize pulmonary regurgitation (PR).[3,9,10]

Modern surgical repair techniques, therefore, use a transatrial-transpulmonary approach and aim to have limited RVOT patching with preservation of the pulmonary valve function.[5,10,11] Early surgical repair has shown to have several advantages, including reduced RV hypertrophy and fibrosis as well as reduced risk of arrhythmias.[12,13] A recent histopathology study found marked RV and LV hypertrophy and fibrosis in late-repaired and unrepaired hearts of ToF patients after the first decade of life.[14] Other histologic studies found circumferential fibers in the RV midwall and an increased amount of circumferential fibers in the hypertrophied subpulmonary infundibulum in ToF, both demonstrating disorganization in the RV myocardial architecture.[15,16]

Currently, surgical repair is performed in young infants or even neonates with good early results,[17,18] although a recent meta-analysis showed that neonatal repair is associated with increased mortality, longer ICU stays, and longer total hospital length of stay.[19] Long-term survival of patients after ToF repair, however, is worse than for the general population, and physical compromise as well as residual anatomic and hemodynamic abnormalities is common.[20,21] Although it can be assumed that patients who undergo surgery now have better long-term results, PR with RV dilatation, pulmonary artery stenosis, RVOT aneurysms, and tricuspid regurgitation (TR) as well as residual septal defects are frequently seen and require careful lifelong follow-up.

Chronic Pulmonary Regurgitation and the Right Ventricle

Over the past 2 to 3 decades, surgical policy has turned towards repair of ToF in infancy with careful planning of RVOT reconstruction to preserve the pulmonary valve function. Chronic PR is still an important complication, however, not only seen in late survivors of ToF repair. Surgical repair in early infancy or neonates, in particular, coincides with increasing transannular patch rates and consequently chronic PR.[18,22] Although PR is usually well tolerated in childhood, it is no longer considered a benign lesion.[5] Note only does the increase in RV stroke volume lead to progressive RV dilatation but also chronic PR has been shown to have deleterious effects, such as RV dysfunction, reduced exercise capacity, ventricular arrhythmias, and sudden cardiac death (**Fig. 1**).[13,23–25]

Echocardiography is normally the first-line imaging modality, and markers suggestive of significant PR include diastolic flow reversal in the pulmonary arteries together with a PR jet width greater than or equal to 50% of the pulmonary annulus as well as a Doppler flow profile showing early termination of the PR.[26,27] Although 3-D echocardiography has improved over the past years, accurate assessment of RV size and systolic function in ToF as well as quantification of PR is still challenging and CMR imaging remains the gold standard.[28]

The effects of chronic volume load on RV mechanics have been a focus of research of the past decades. Studies in patients with atrial septal defects have shown that RV volume overload adversely affects RV and LV geometry and function.[29,30] In ToF, an increase of RV end-diastolic and end-systolic volumes as well as deterioration of RV function has been well demonstrated (see **Fig. 1**).[31] Kato and colleagues[32] hypothesized that cardiac enlargement and rotation into the left hemithorax lead to elevated left lung pulmonary vascular resistance and attenuates left pulmonary artery flow. They suggest that this in turn increase RV volume and pressure load with further RV dilatation and dysfunction. In the setting of chronic PR, a restrictive RV physiology was supposed to be beneficial,[33,34] but this remains controversial.[35,36]

Chronic volume load not only affects RV systolic function but also diastolic function.[37] Friedberg and colleagues[37] demonstrated RV diastolic dysfunction in children after ToF repair with significant PR using tissue Doppler and speckle-tracking echocardiography. That alterations in RV geometry and function can have an adverse impact on LV size and function has been demonstrated in several studies.[38,39] Explanations for this are shared myofibers and pericardial space as well as altered mechanical properties of the interventricular septum leading to a leftward septal shift.[40,41] LV function is an important risk factor