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Review article

Impact of Percutaneous Pulmonary Valve Implantation on the Timing of Reintervention for Right Ventricular Outflow Tract Dysfunction

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ABSTRACT

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect. Early surgical repair has dramatically improved the outcome of this condition. However, despite the success of contemporary approaches with early complete repair, these are far from being curative and late complications are frequent. The most common complication is right ventricle outflow tract (RVOT) dysfunction, affecting most patients in the form of pulmonary regurgitation, pulmonary stenosis, or both, and can lead to development of symptoms of exercise intolerance, arrhythmias, and sudden cardiac death. Optimal timing of restoration of RVOT functionality in asymptomatic patients with RVOT dysfunction after TOF repair is still a matter of debate. Percutaneous pulmonary valve implantation, introduced almost 2 decades ago, has become a major game-changer in the treatment of RVOT dysfunction. In this article we review the pathophysiology, the current indications, and treatment options for RVOT dysfunction in patients after TOF repair with a focus on the role of percutaneous pulmonary valve implantation in the therapeutic approach to these patients.

Impacto del implante percutáneo de válvula pulmonar en cuanto al momento de reintervenir por disfunción del tracto de salida del ventrículo derecho

Palabras clave:
Válvula pulmonar
Percutáneo
Tetralogía de Fallot
Cardiopatías congénitas

RESUMEN

La tetralogía de Fallot (TDF) es la cardiopatía congénita cianótica más frecuente. La reparación quirúrgica temprana ha mejorado radicalmente su pronóstico. Sin embargo, a pesar del éxito de los abordajes quirúrgicos contemporáneos con reparación completa a edades tempranas, estos distan de ser curativos y las complicaciones tardías son frecuentes. La disfunción del tracto de salida del ventrículo derecho (TSVD) es la complicación más frecuente, afecta a la mayoría de los pacientes en forma de insuficiencia pulmonar, estenosis pulmonar o ambas y puede llevar a la aparición de síntomas de intolerancia al ejercicio, arritmias o muerte súbita. El momento óptimo para restaurar la función del TSVD sigue siendo objeto de debate. El implante percutáneo de válvula pulmonar, introducido hace casi 2 décadas, ha supuesto un punto de inflexión en el tratamiento de la disfunción del TSVD. En este artículo se revisa la fisiopatología, las actuales indicaciones y opciones terapéuticas para la disfunción del TSVD en pacientes con TDF reparada, con especial énfasis en el papel del implante percutáneo de válvula pulmonar en el abordaje terapéutico de estos pacientes.

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Abbreviations

PPVI: percutaneous pulmonary valve implantation
PR: pulmonary regurgitation
PVR: pulmonary valve replacement
RV: right ventricle
RVEDVi: indexed right ventricular end-diastolic volume
RVESVi: indexed right ventricular end-systolic volume
RVOT: right ventricular outflow tract
TOF: tetralogy of Fallot

INTRODUCTION

The overall prevalence of congenital heart disease in adults is estimated to be of 3000 per million.[1] Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect, accounting for 10% of all congenital cardiac malformations.[2] Early surgical repair has dramatically improved the outcome of this condition, from a survival rate to adulthood < 25% without surgery[3] to a survival of approximately 90% at 30 years in patients undergoing complete repair surgery in infancy.[4] The therapeutic approaches have evolved from initial surgical palliation with Blalock-Taussig shunts[5] and the first described intracardiac repair,[6] an era of staged repair with shunt palliation prior to intracardiac repair, and finally an approach of direct complete repair early in infancy in the past 2 decades. Surgical techniques for complete repair evolved from a right, sometimes large ventriculotomy to close the ventricular septal defect and to resect the infundibular stenosis together with a transannular patch to relieve the right ventricular outflow tract (RVOT) obstruction to transatrial and transpulmonary approaches aiming to preserve the pulmonary valve annulus and, whenever possible, the pulmonary valve, and to minimize ventricular scarring.[7,8]

However, this contemporary approach with early complete repair is far from being curative and late complications after repaired TOF are frequent. In a very large cohort of patients with repaired TOF, half of the survivors had undergone a reoperation 30 years after repair.[4] RVOT dysfunction is the most common complication, affecting most patients in the form of pulmonary regurgitation (PR), especially patients with transannular patches.[9]

In some cases, the cardiac anatomy precludes complete surgical repair, such as in patients with pulmonary atresia, absent pulmonary valve, or in the presence of an anomalous coronary artery that crosses the RVOT. In these cases, a conduit from the right ventricle (RV) to the pulmonary artery is necessary to relieve the RVOT obstruction. These conduits are also used in other types of congenital heart surgery such us in the repair of a common arterial trunk or some forms of complex transposition of the great arteries (Rastelli procedure), as well as in procedures to relieve left heart obstructions such as the Ross or Ross-Konno procedures. Degeneration of these conduits can also lead to RVOT dysfunction.

In this context, restoration of RVOT functionality often becomes necessary. Percutaneous pulmonary valve implantation (PPVI), introduced almost 2 decades ago, has become a major game-changer in the treatment of RVOT dysfunction.

In this article, we review the pathophysiology, current indications, and treatment options for RVOT dysfunction with a focus on the role of PPVI in the therapeutic approach to these patients.

PATHOPHYSIOLOGY OF RVOT DYSFUNCTION

More than half of the patients after primary TOF repair develop RVOT dysfunction at some point in their lives. Similarly, patients with a RV to pulmonary artery conduit sooner or later experience a deterioration in conduit function leading to stenosis, regurgitation, or both.

It is well known that chronic PR causes RV volume overload, which is generally well tolerated over the years,[10] but if maintained over time may lead to RV dilation and dysfunction[11] (Figure 1), which are in turn associated with atrial[12] and ventricular arrhythmias, sudden cardiac death,[13–15] exercise intolerance, heart failure, and excess mortality.[16–19]

In addition, residual RVOT obstruction at all levels (infundibulum, pulmonary valve, main pulmonary artery or its branches) can also contribute to RV dysfunction. Pulmonary stenosis leads to RV pressure overload and in turn to RV dysfunction due to increased RV mass:volume ratio, which has been shown to be predictive of ventricular arrhythmias and death in a large retrospective study.[15]

Due to this common progression to RV dysfunction of both volume- and pressure- overloaded RVs after TOF repair and its association with clinical events, the restoration of the functionality of the RVOT by means of pulmonary valve replacement (PVR) is considered when these structural changes translate into clinical problems. It is accepted that symptomatic patients with RVOT dysfunction benefit from intervention in terms of relief of symptoms,[20–24] especially those patients with predominant pulmonary stenosis. However, a consistent improvement in objective functional capacity on cardiopulmonary exercise testing has not been demonstrated.[20,22,24,25] Similarly, PVR improves right ventricular hemodynamic parameters such us RV size[20]; however,

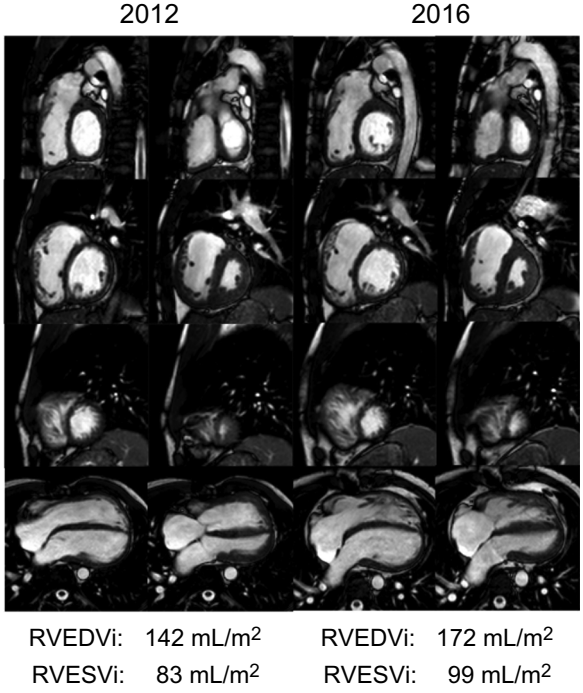

Figure 1. Progressive dilation of the right ventricle in a patient after repaired tetralogy of Fallot and pulmonary insufficiency demonstrated by cardiac magnetic resonance imaging. Short axis stack (basal, upper row; midventricular, second row; apical, third row) and 4-chamber view (lower row) of cine steady-state free precession end-diastolic (first and third columns) and end-systolic (second and fourth columns) images of the same patient in 2012 and 2016. Note the progression of the dilation of the right ventricle. RVEDVi, right ventricle end-diastolic volume index; RVESVi, right ventricle end-systolic volume index.

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