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Timing of pulmonary valve replacement: can we use the same volumetric thresholds in repaired tetralogy of Fallot and pulmonary stenosis?

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Abstract

The timing of pulmonary valve replacement in patients with pulmonary regurgitation following treatment of pulmonary stenosis is undefined. Although cardiac magnetic resonance-based right ventricular volumes in tetralogy of Fallot patients have been used as a guide in pulmonary stenosis patients, anatomic differences between tetralogy of Fallot and pulmonary stenosis patients complicate their application to pulmonary stenosis patients and could result in late referral for pulmonary valve replacement. We sought to determine if pulmonary stenosis patients referred for pulmonary valve replacement were at greater risk for morbidity or need for tricuspid valve intervention at the time of pulmonary valve replacement. A retrospective cohort study was performed on all adult patients with a diagnosis of pulmonary stenosis or tetralogy of Fallot followed at our centre. Clinical and imaging-based exposures were collected. Pre-specified endpoints included need for concomitant tricuspid valve repair or replacement and pre- and post-pulmonary valve replacement cardiac magnetic resonance-based volumetric measurements. Between 1/1999 and 1/2020, 235 patients underwent pulmonary valve replacement for pulmonary regurgitation (52 with pulmonary stenosis, 183 with tetralogy of Fallot). Pulmonary stenosis patients were more likely to have at least moderate tricuspid regurgitation ($p = 0.010$), undergo concomitant tricuspid valve intervention ($p = 0.003$), and require tricuspid valve repair or replacement secondary to annular dilation ($p = 0.027$) compared to tetralogy of Fallot patients. There was no difference in pre-pulmonary valve replacement right ventricular size between pulmonary stenosis and tetralogy of Fallot patients. These findings suggest that referral for pulmonary valve replacement may be occurring later in the disease course for pulmonary stenosis patients.

Pulmonary valve stenosis represents 7–12% of all adult congenital cardiac defects[1,2] and often requires early palliation with surgical valvotomy or balloon valvuloplasty. As a result, many patients are left with chronic pulmonary regurgitation that can lead to right ventricular dilation and dysfunction. While pulmonary valve replacement can ameliorate pulmonary regurgitation and arrest aberrant ventricular remodelling, the longevity of bioprosthetic valves complicates decisions regarding surgical timing.

Cardiac MRI-based volumetric thresholds are utilised to facilitate timing of pulmonary valve replacement in tetralogy of Fallot patients.[3–11] Because of limited data regarding the optimal volumetric thresholds for pulmonary valve replacement in patients with pulmonary regurgitation following intervention for pulmonary stenosis, tetralogy of Fallot-based values are frequently extrapolated to the pulmonary stenosis population. However, the presence of a right ventricular outflow tract patch in tetralogy of Fallot patients who have undergone ventriculotomy or infundibulotomy as part of their repair can amplify right ventricular volumes relative to isolated pulmonary stenosis where an outflow tract patch is absent. Consequently, use of tetralogy of Fallot-derived cardiac MRI values in pulmonary stenosis patients may inappropriately delay pulmonary valve replacement, leading to tricuspid annular dilation and clinically significant tricuspid regurgitation. In addition, wide dissemination of guideline-based tetralogy of Fallot volumetric thresholds for pulmonary valve replacement has resulted in smaller right ventricular volumes at the time of pulmonary valve replacement referral in the tetralogy of Fallot population as compared to before there were guidelines.[12] Conversely, in the pulmonary stenosis population, the lack of established cardiac MRI criteria and consequent uncertainty regarding appropriate timing of pulmonary valve replacement referral may lead to greater right ventricular size at the time of intervention.

To aid in defining criteria for pulmonary valve replacement in pulmonary stenosis patients, we compared the clinical and volumetric characteristics of pulmonary stenosis and tetralogy of Fallot patients who underwent pulmonary valve replacement at our centre to determine if pulmonary stenosis patients were at greater risk for morbidity, including need for concomitant

tricuspid valve surgery at the time of pulmonary valve replacement; and whether the degree of right ventricular dilation at the time of referral for pulmonary valve replacement has changed over time for patients with tetralogy of Fallot and pulmonary stenosis.

Materials and method

Approval from the Columbia University Medical Center Institutional Review Board was obtained before data acquisition. We performed a retrospective cohort study of patients >18 years with pulmonary stenosis or tetralogy of Fallot followed at the Schneeweiss Adult Congenital Heart Center at Columbia University Medical Center from January, 1999 to January, 2020. Patients with additional right-sided volume loading lesions, unrepaired tetralogy of Fallot, a pulmonary homograft or right ventricle to pulmonary artery conduit, pulmonary valve replacement for predominant pulmonary stenosis, and all Ross patients were excluded. Patients defined as having tetralogy of Fallot included those with pulmonary atresia variants. To minimise bias, individuals who underwent primary transcatheter pulmonary valve replacement were excluded.

Clinical variables were defined prior to data collection and were extracted from the electronic medical record. Variables of interest included diagnosis, pulmonary valve replacement status, gender, age at initial surgical or balloon intervention, and age at pulmonary valve replacement or last follow-up. The presence of sustained or symptomatic supraventricular or ventricular arrhythmia was noted when applicable. Dyspnoea was considered present if documented by the patient's primary congenital cardiologist at the pre-pulmonary valve replacement clinical appointment. Indication for tricuspid valve intervention was determined through review of operative reports, when available.

For patients who underwent cardiac MRI, pre- and post-pulmonary valve replacement right ventricular end-diastolic volume, end-systolic volume, and ejection fraction were collected. To assess the change in right ventricular size over time for each cohort, mean indexed right ventricular end diastolic volume was collated and compared over two 10-year periods: 1/1/2000–1/1/2010 and 1/2/2010–1/1/2020.

Cardiac MRI studies were performed with breath holding and electrocardiogram gating at 1.5 T (Signa, General Electric, Milwaukee, WI) using an 8-channel phased array coil for signal reception. Gadolinium was not routinely administered unless specifically indicated. Short-axis cine images were acquired using a steady-state free precession pulse sequence (FIESTA) with the following parameters: TR/TE/flip 3.6/1.5/45 degrees, 24 views per segment, FOV = 35 cm, acquisition matrix 192 × 160, slice thickness 8 mm with no gap, and receiver bandwidth 125 kHz. Images were reviewed and analysed using ReportCARD software (GE Healthcare) until 2017 and Circle software thereafter. Cine loops were used to select images at end-diastole and end-systole. End-diastole was defined as the phase with the largest volume, and end-systole as the phase with the smallest volume for the right and left ventricles, independently. Manual tracing was performed on each end-diastole and end-systole short-axis view and summed to calculate right and left ventricular volumes. By convention, right ventricle trabeculations were considered part of the right ventricular cavity in both systole and diastole. Ejection fraction was calculated using the end diastolic volume and end systolic volume values [(end diastolic volume-end systolic volume/end diastolic volume) × 100%].

Endpoints included the need for tricuspid valve repair or replacement at the time of pulmonary valve replacement and the degree of tricuspid regurgitation at the time of pulmonary valve replacement. At our institution, concomitant tricuspid valve intervention at the time of pulmonary valve replacement was recommended when pre-operative transthoracic echocardiograms demonstrated moderate or more tricuspid regurgitation and the operating surgeon confirmed the presence of significant annular dilation or a primary tricuspid valve abnormality. The degree of tricuspid regurgitation was dichotomised based on the presence of moderate or greater regurgitation on transthoracic echocardiograms obtained immediately prior to pulmonary valve replacement and read by board-certified congenital cardiologists.

Data were expressed as n (%), mean, or median where appropriate. Univariable analyses comparing discrete clinical variables were performed with Chi-square tests. Continuous variables of interest were analysed by standard t-tests. Logistic regression was used to assess univariable and multivariable predictors for simultaneous tricuspid valve intervention among those patients who underwent pulmonary valve replacement. Multivariable logistic regression models were constructed to include criteria from the 2008 and 2018 American College of Cardiology/American Heart Association Guidelines for the Management of Adults with Congenital Heart Disease.[10,11] Statistical analysis was performed with Stata statistical software (version 14.1; Stata Corp, College Station, Texas).

Results

In total, 607 patients with a diagnosis of pulmonary stenosis or tetralogy of Fallot were identified from the medical records of the Schneeweiss Adult Congenital Heart Center. We excluded 4 patients with unrepaired tetralogy of Fallot, 13 patients who underwent pulmonary valve replacement for severe pulmonary stenosis, 18 patients with a pulmonary homograft as part of the initial repair, 29 patients with right ventricle to pulmonary artery conduits, 5 patients with additional right-sided volume loading lesions, and 10 patients with complex associated disease. Of the remaining 528 patients, 167 (32%) had pulmonary stenosis and 361 (68%) had tetralogy of Fallot. Totally, 235 (44%) of these patients with tetralogy of Fallot or pulmonary stenosis underwent pulmonary valve replacement secondary to post-intervention pulmonary regurgitation. Clinical features and pre-pulmonary valve replacement cardiac MRI measurements of right ventricular volume and function are presented in Table 1. There were no differences in pre- or post-pulmonary valve replacement cardiac MRI measurements of indexed right ventricular end diastolic volume between the two groups. There were no differences in the number of patients with dyspnoea, supraventricular tachycardia, or ventricular tachycardia between the two groups.

Of the 235 patients who underwent pulmonary valve replacement, 52 (22%) had pulmonary stenosis and 183 (78%) had tetralogy of Fallot. Patients with pulmonary stenosis who underwent pulmonary valve replacement were significantly more likely to have at least moderate tricuspid regurgitation ($p = 0.010$) and undergo concomitant tricuspid valve intervention at the time of pulmonary valve replacement ($p = 0.003$). After excluding one patient with pulmonary stenosis and one patient with tetralogy of Fallot with tricuspid valve injury secondary to implantable cardioverter defibrillator leads, operative reports were available on 36 (81%) patients who underwent tricuspid valve intervention at the time of pulmonary valve replacement. Notably, patients with

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Table 1. Demographics, clinical characteristics, and pre-PVR echocardiographic and cardiac MRI measurements of ventricular volume and function for patients who underwent PVR.

Variable	PS (n = 52)	TOF (n = 183)	p
Female (%)	24 (46%)	98 (54%)	0.35
Body mass index (kg/m^2)	26	26	0.54
Mean age at PVR (years)	35	33	0.37
Mean age at first intervention/surgery (years)	4	5	0.35
Dyspnea	21 (45%)	78 (51%)	0.63
NYHA class (≥ II)			
Supraventricular tachycardia (atrial flutter, atrial fibrillation)	12 (27%)	27 (18%)	0.18
Ventricular arrhythmias	1 (2%)	25 (16%)	0.015
Prior palliative shunt	2 (5%)	49 (35%)	<0.0001
Prior transannular patch	1 (3%)	85 (61%)	<0.0001
Prior RV to PA conduit	0 (0)	3 (2%)	0.38
Number of prior cardiac surgeries	1.1	1.3	0.037
Abnormal LF Function by echo	0 (0)	12 (9%)	0.0841
Severe RV dilation by echo	19 (56%)	88 (64%)	0.37
Abnormal RV systolic function by echo	8 (24%)	41 (30%)	0.74
Mitral regurgitation (≥ moderate) by echo	0	0	1
Baseline tricuspid regurgitation (≥ moderate) by echo	16 (32%)	27 (15%)	0.010
Baseline pulmonary regurgitation (≥ moderate) by echo	52 (100%)	183 (100%)	1
Pre-PVR LVEDV (mean, ml)	142	152	0.24
Pre-PVR LVESV (mean, ml)	63	68	0.41
Pre-PVR LVEF (mean)	57	55	0.53
Pre-PVR RVEDV (mean, ml)	296	316	0.22
Pre-PVR RVEDVi (mean, ml/m^2)	164	174	0.23
Pre-PVR RVESV (mean, ml)	164	181	0.19
Pre-PVR RVESVi (mean, ml/m^2)	89	98	0.16
Pre-PVR RVEF (mean)	47	44	0.09
Pre-PVR pulmonary regurgitation fraction	0.42	0.43	0.84
Medical therapy – diuretic	5 (15%)	9 (6%)	0.11
Medical therapy – aspirin	7 (21%)	13 (9%)	0.066
Medical therapy – anticoagulation	7 (21%)	9 (6%)	0.011
Medical therapy – other	14 (43%)	60 (45%)	0.98
Pre-PVR VO2 (mean, ml/kg/m^2)	28	25	0.10
Pre-PVR ICD and/or PPM	3 (7%)	21 (13%)	0.27

pulmonary stenosis were significantly more likely to undergo tricuspid valve intervention secondary to annular dilation when compared to patients with tetralogy of Fallot (100% versus 70%, p = 0.027).

Univariable risk factors and multivariable models for the need for tricuspid valve intervention at the time of pulmonary valve replacement are listed in Table 2. By univariate regression, a diagnosis of pulmonary stenosis and age at time of pulmonary valve replacement were associated with increased odds for concomitant tricuspid valve repair or replacement at the time of surgery. Importantly, a diagnosis of pulmonary stenosis remained a significant predictor for simultaneous tricuspid valve intervention after including patient age.

Pre-pulmonary valve replacement cardiac MRI measurements were available on 231 patients, 149 (64%) with tetralogy of Fallot and 82 (35%) with pulmonary stenosis. Figure 1 displays the mean indexed right ventricular end diastolic volume for patients with tetralogy of Fallot and pulmonary stenosis referred for pulmonary valve replacement across two decades: 1/1/2000–1/1/2010 and 1/2/2010–1/1/2020. Patients with tetralogy of Fallot referred in the second decade had a significantly smaller mean indexed right ventricular end diastolic volume than those referred in the first

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Table 2. Univariate and multivariable analyses of exposures of interest for concomitant TVR in all patients undergoing PVR.

	Univariable		Multivariable	
Variable	OR (95% CI)	p	OR (95% CI)	p
PS vs. TOF	2.86 (1.58–5.88)	0.004	2.72 (1.33–5.55)	0.006
RVEDVI	1.00 (0.98–1.01)	NS		
Age at PVR	1.02 (1.00–1.05)	0.04	1.02 (0.98–1.05)	0.064

PS = pulmonary stenosis; PVR = pulmonary valve replacement; RVEDVI = right ventricular end-diastolic volume indexed to body surface area; TOF = tetralogy of Fallot

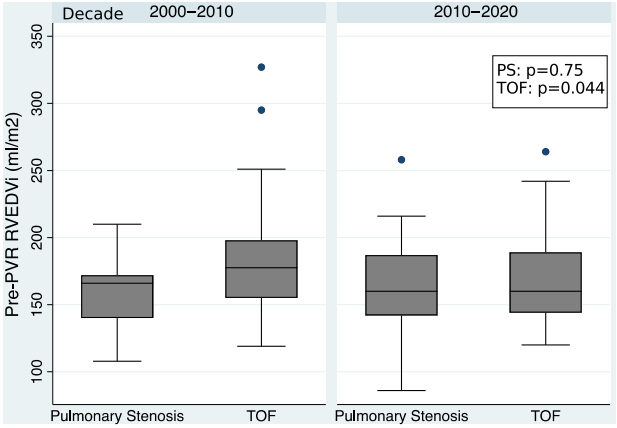

Figure 1. Indexed right ventricular end-diastolic volume at time of PVR by diagnosis and decade of referral. PVR: pulmonary valve replacement. RVEDVi: right ventricular end-diastolic volume indexed to body surface area; TOF: tetralogy of Fallot.

decade (168 ml/m^2 versus 184 ml/m^2, p = 0.044); there was no significant difference in mean indexed right ventricular end diastolic volume between the two periods, however, for patients with pulmonary stenosis (164 ml/m^2 versus 159 ml/m^2, p = 0.75). There was no significant difference in mean pre-pulmonary valve replacement indexed right ventricular end diastolic volumes (tetralogy of Fallot: 174 ml/m^2 versus pulmonary stenosis: 163 ml/m^2, p = 0.18), or mean pre-pulmonary valve replacement indexed right ventricular end-systolic volumes (tetralogy of Fallot: 97 ml/m^2 versus pulmonary stenosis: 89 ml/m^2, p = 0.19) between patients with pulmonary stenosis and tetralogy of Fallot. Patients with pulmonary stenosis did have a significantly higher right ventricular ejection fraction (tetralogy of Fallot: 44% versus pulmonary stenosis: 48%, p = 0.047). There was also no significant difference in the percent changes of indexed right ventricular end diastolic volume, indexed right ventricular end systolic volume, or right ventricular ejection fraction between pulmonary stenosis and tetralogy of Fallot patients following pulmonary valve replacement.

Discussion

Although pulmonary stenosis is one of the most common forms of CHD, criteria for pulmonary valve replacement in patients following repair of pulmonary stenosis with subsequent chronic pulmonary regurgitation are undefined. In our cohort, patients with pulmonary stenosis were more likely to have moderate or greater tricuspid regurgitation and require tricuspid valve repair or replacement at the time of pulmonary valve replacement compared to those patients with tetralogy of Fallot, despite having similar right ventricular volumes. Importantly, they were also more likely to have evidence of significant annular dilation at the time of surgery. These findings may suggest that pulmonary valve replacement referral in pulmonary stenosis patients is performed later in the disease course relative to patients with tetralogy of Fallot.

While similarities between pulmonary stenosis and tetralogy of Fallot exist, extrapolation of cardiac MRI-based volumetric thresholds for pulmonary valve replacement in tetralogy of Fallot to pulmonary stenosis patients is complicated by the presence of a right ventricular outflow tract patch in the majority of tetralogy of Fallot patients. Because the right ventricular outflow tract patch directly augments right ventricular volumes, the tetralogy of Fallot cardiac MRI-based thresholds incorporate an inherent enlargement of the right ventricle that is absent in most pulmonary stenosis patients. Consequently, application of tetralogy of Fallot-derived thresholds to the pulmonary stenosis population may yield functionally greater degrees of right ventricular dilation in the pulmonary stenosis cohort. In our study, despite the absence of an outflow tract patch, pulmonary stenosis patients had a similar indexed right ventricular end diastolic volume to those with tetralogy of Fallot at the time of pulmonary valve replacement, suggesting a greater degree of functional right ventricular dilation. It is important to note that this is may also be true for the subset of tetralogy of Fallot patients who had not undergone ventriculotomy or infundibulotomy as part of their initial repair and who also lacked an outflow tract patch.

Pulmonary stenosis patients had a significantly more tricuspid regurgitation and a greater need for simultaneous tricuspid valve intervention at the time of pulmonary valve replacement. Because annular dilation is more common in patients with significant right ventricular dilation and dysfunction, the need for tricuspid valve intervention and the degree of pre-pulmonary valve replacement tricuspid regurgitation is a valid proxy for excessive right ventricular remodelling from protracted pulmonary regurgitation. These findings suggest that referral for pulmonary valve replacement may be occurring at right ventricular volumes that exceed the optimal threshold for pulmonary stenosis patients.[13] Alternative explanations for this finding, including a higher relative rate of tricuspid valve abnormalities in the pulmonary stenosis population, are possible; primary tricuspid valve abnormalities, however, were significantly more common in the tetralogy of Fallot group. This finding is consistent with the fact that initial repair of tetralogy of Fallot is more likely to damage the tricuspid valve apparatus secondary to placement of the ventricular septal defect patch, as compared to pulmonary stenosis patients who generally undergo valvotomy alone. In fact, in our cohort, every patient with pulmonary stenosis who underwent tricuspid valve intervention at the time of pulmonary valve replacement had evidence of annular dilation. Thus, despite a higher rate of primary valve abnormalities in the tetralogy of Fallot group, patients with pulmonary stenosis required significantly more tricuspid valve interventions at the time of pulmonary valve replacement, suggesting a greater degree of maladaptive right ventricular remodelling.

While there are data to suggest that concomitant tricuspid valve annuloplasty improves tricuspid regurgitation in the short term, tricuspid intervention at the time of pulmonary valve replacement may not improve tricuspid regurgitation longitudinally.[14,15] As an additional right ventricular volume load, haemodynamically significant tricuspid regurgitation can contribute to progressive right ventricular enlargement and is associated with increased mortality, heart failure, and arrhythmias in patients with CHD.[16,17] Given the potential clinical consequences and the questionable durability of

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tricuspid valve annuloplasty in the adult CHD population, intervening on severe pulmonary regurgitation should be considered prior to the development of significant functional tricuspid regurgitation in pulmonary stenosis patients.

The optimal timing for pulmonary valve replacement following pulmonary stenosis repair remains undefined. While the 2008 American College of Cardiology/American Heart Association guidelines highlight dyspnoea, symptomatic or sustained supraventricular tachycardia, and moderate or more tricuspid regurgitation as indications for pulmonary valve replacement in the tetralogy of Fallot population, the 2018 guidelines emphasise cardiac MRI-derived metrics. In contrast, the 2018 guidelines do not provide specific cardiac MRI-based criteria as indications for pulmonary valve replacement in the pulmonary stenosis population. It is important to recognise that most of the studies that define optimal timing of pulmonary valve replacement in tetralogy of Fallot patients are based on the results of cardiac MRI-derived reduction in right ventricular volumes following pulmonary valve replacement. In our study, tetralogy of Fallot patients referred in the last 10 years had significantly less right ventricular dilation when compared to tetralogy of Fallot patients referred in the prior decade. This finding was not seen in the pulmonary stenosis population, suggesting a potential impact from the number of studies delineating volumetric thresholds for patients with tetralogy of Fallot on pulmonary valve replacement referral. While similar reductions in right ventricular volumes were achieved with pulmonary valve replacement, our cohort of pulmonary stenosis patients undergoing pulmonary valve replacement required a greater number of concomitant tricuspid valve interventions compared to their tetralogy of Fallot counterparts. These findings suggest that such extrapolation of tetralogy of Fallot-based volumetric thresholds may not be appropriate and highlight the need for additional studies to define lesion-specific criteria for intervention. This study was not powered to detect thresholds of poor recovery in the pulmonary stenosis population as our cohort underwent pulmonary valve replacement at a relatively consistent set of right ventricular volumes. Future prospective studies with a larger sample size are necessary to determine cut-offs at which post-operative recovery for this cohort of patients is lost.

As a retrospective single centre study spanning over 20 years, it is difficult to precisely define referral criteria for pulmonary valve replacement in the pulmonary stenosis population over the entirety of the study period. At present, our group will refer symptomatic pulmonary stenosis patients with moderate or more pulmonary regurgitation for pulmonary valve replacement. As a result of our findings, the recommendations for asymptomatic patients have evolved. We utilise smaller cardiac MRI-derived right ventricular volumes, in the range between 150 and 160 ml/m^2, in our decision making along with additional criteria including decreased exercise capacity, atrial or ventricular arrhythmias, and development of significant tricuspid regurgitation. Importantly, we will not wait until the development of significant tricuspid regurgitation to proceed with pulmonary valve replacement; however, we cannot exclude that possibility in our referral base. As such, it is possible that some pulmonary stenosis patients presented with significant tricuspid regurgitation at the time of pulmonary valve replacement as a consequence of this practice. However, the presence of such decision making would only underscore the need for the development of formal criteria, inclusive of volumetric thresholds, specific to the pulmonary stenosis population.

Other limitations are important to highlight. As a cross-sectional retrospective study, we were limited to data made available through chart review and did not have access to pre-pulmonary valve replacement data for the whole cohort. The evolution of provider practice over time for both the pulmonary stenosis and tetralogy of Fallot populations may also complicate our findings. Additional masked confounders that impact study results, including differences in provider attitudes regarding the clinical importance of tricuspid regurgitation in the pulmonary stenosis and tetralogy of Fallot populations, may also be present. Nonetheless, our study is the first to assess practice for referral for pulmonary valve replacement in patients with palliated pulmonary stenosis and chronic pulmonary regurgitation and highlights the need for future multi-centre studies to establish lesion-specific criteria for intervention in this patient population.

In our cohort, patients with pulmonary stenosis were more likely to have moderate or greater tricuspid regurgitation and more frequently required tricuspid valve intervention at the time of pulmonary valve replacement compared to patients with tetralogy of Fallot. This occurred in the absence of significantly different post-pulmonary valve replacement changes in right ventricular size or function. This suggests that referral for pulmonary valve replacement may be occurring later in the disease course for some patients with pulmonary stenosis.

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References

1. Marelli AJ, Ionescu-Ittu R, Mackie AS, et al. Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. Circulation 2014; 130: 749–756.
2. Cheatham JP, Coe JY, Kugler JD, Fletcher SE, Tower AJ. Successful transcatheter perforation of the atretic pulmonary valve membrane in a newborn using the new Coe radiofrequency end hole catheter. Cathet Cardiovasc Diagn 1998; 45: 162–166.
3. Geva T. Repaired tetralogy of Fallot: the roles of cardiovascular magnetic resonance in evaluating pathophysiology and for pulmonary valve replacement decision support. J Cardiovasc Magn Reson 2011; 13: 9.
4. Weinberg CR, McElhinney DB. Clinician update: pulmonary valve replacement in tetralogy of Fallot. Circulation 2014; 130: 795–798.
5. Therrien J, Provost Y, Merchant N, et al. Optimal timing for pulmonary valve replacement in adults after tetralogy of Fallot repair. Am J Cardiol 2005; 95: 779–782.
6. Oosterhof T, Van Straten A, Vliegen HW. Preoperative thresholds for pulmonary valve replacement in patients with corrected tetralogy of Fallot using cardiovascular magnetic resonance. Circulation 2007; 116: 545–551.
7. Beuchel ER, Dave HH, Kellenberger CJ, et al. Remodeling of the right ventricle after early pulmonary valve replacement in children with repaired tetralogy of Fallot: assessment by cardiovascular magnetic resonance. Eur Heart J 2005; 26: 2721–2727.
8. Lee CL, Kim YM, Lee CH, et al. Outcomes of pulmonary valve replacement in 170 patients with chronic pulmonary regurgitation after relief of right ventricular outflow tract obstruction: implications for optimal timing of pulmonary valve replacement. J Am Coll Cardiol 2012; 60: 1005–1014.
9. Earing MG, Connolly HM, Dearani JA, et al. Long-term follow-up of patients after surgical treatment for isolated pulmonary valve stenosis. Mayo Clin Proc 2005; 80: 871–876.
10. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease. J Am Coll Cardiol 2019; 73: e81–e192. DOI 10.1016/j.jacc.2018.08.1029.
11. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA guidelines for the management of adults with congenital heart disease: executive summary. Circulation 2008; 118: 2395–2451.

https://doi.org/10.1017/S1047951122000361 Published online by Cambridge University Press

12. Lewis MJ, O'Connor DS, Rozenshtien A, et al. Usefulness of magnetic resonance imaging to guide referral for pulmonary valve replacement in repaired tetralogy of Fallot. Am J Cardiol 2014; 114: 1406–1411.
13. Bokma JP, Winter MM, Oosterhof T, et al. Severe tricuspid regurgitation is predictive of adverse events in tetralogy of Fallot. Heart 2015; 101: 794–799.
14. Deshaies C, Trottier H, Khairy P, et al. Tricuspid valve intervention following pulmonary valve replacement in adults with congenital heart disease. J Am Coll Cardiol 2020; 75: 1033–1043.
15. Kogon B, Mori M, Alsoufi B, Kanter K, Oster M. Leaving moderate tricuspid valve regurgitation alone at the time of pulmonary valve replacement: a worthwhile approach. Ann Thorac Surg 2015; 99: 2117–2123.
16. Gatzoulis MA, Balaji S, Webber SA, et al. Risk factors for arrhythmia and sudden cardiac death after late repair of tetralogy of Fallot: a multicentre study. Lancet 2000; 356: 975–981.
17. Lewis MJ, Ginns JN, Ye S, et al. Postoperative tricuspid regurgitation after adult congenital heart surgery is associated with adverse clinical outcomes. J Thorac Cardiovasc Surg 2016; 151: 46–465.

https://doi.org/10.1017/S1047951122000361 Published online by Cambridge University Press