Society of Thoracic Surgeons/ World Society for Pediatric and Congenital Heart Surgery/ European Congenital Heart Surgeons Association 2026 Clinical Practice Guidelines on Indications and Timing of Pulmonary Valve Replacement in Repaired Tetralogy of Fallot

Running/Short Title: Clinical Practice Guidelines for PVR

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Keywords: Tetralogy of Fallot; Pulmonary valve replacement; Congenital Heart Disease

Abstract

The Society of Thoracic Surgeons/ World Society for Pediatric and Congenital Heart Surgery/ European Congenital Heart Surgeons Association 2026 Clinical Practice Guidelines on Indications and Timing of Pulmonary Valve Replacement in Repaired Tetralogy of Fallot incorporate the most recent evidence for pulmonary valve replacement in this growing population. New evidence related to advanced imaging, electrophysiology testing, exercise testing, and transcatheter pulmonary valve replacement has emerged in the last 5 years. Compared to existing guidelines, the current update places emphasis on symptoms (children and adults), ventricular volumes (adults), need for invasive electrophysiology study prior to replacing the pulmonary valve (adults). This document is Part 1 and is accompanied by two other multi society documents: Part 2: Society of Thoracic Surgeons / World Society for Pediatric and Congenital Heart Surgery / European Congenital Heart Surgeons Association 2026 Expert Consensus Document on Timing, Indications, and Options for Pulmonary Valve Replacement in Children with Repaired Tetralogy of Fallot, and Part 3: Society of Thoracic Surgeons / World Society for Pediatric and Congenital Heart Surgery / European Congenital Heart Surgeons Association 2026 Expert Opinion on The Role of Exercise Testing in Determining Optimal Timing of Pulmonary Valve Replacement in Tetralogy of Fallot. With this three-part series, multidisciplinary team assessment, treatment planning, and long-term surveillance are also reinforced.

Abbreviations:

CMR = Cardiac Magnetic Resonance

ECHSA = European Congenital Heart Surgeons Association

EF = ejection fraction

EP = electrophysiology

LV = Left ventricular

LVEDP = left ventricular end diastolic pressure

LVEDV = left ventricular end diastolic volume

NSVT = non-sustained ventricular tachycardia

NYHA = New York Heart Association

PI = Pulmonary insufficiency

PICO = population, intervention, comparison, and outcomes

RV = Right ventricular

RVOT = right ventricular outflow tract

SCD = sudden cardiac death

STAT = Society of Thoracic Surgeons–European Association for Cardio-Thoracic Surgery

STS = Society of Thoracic Surgeons

TOF = tetralogy of Fallot

VA = ventricular arrhythmia

VO2 max = maximum volume of oxygen used during exercise

VT = ventricular tachycardia

WSPCHS = World Society for Pediatric and Congenital Heart Surgery

Background and Scope

It is estimated that approximately 50% of patients with repaired tetralogy of Fallot (TOF) will require pulmonary valve replacement (PVR) within 30 years after corrective surgery.[1] Across the age spectrum, PVR is associated with a 30-day mortality rate of <2% and can be performed surgically or percutaneously.[1,2] Determining the specific indications and best timing to perform PVR is essential to prevent irreversible ventricular dysfunction, while also minimizing the need for multiple re-operations.

In 2013 Geva outlined frequently referenced criteria for PVR.[3] However, these recommendations acknowledged the lack of outcomes data following PVR, and were based primarily on adult data. Despite the significant number of patients undergoing PVR, current guidelines are outdated and fail to provide specific, outcomes-based recommendations for adults and children with repaired TOF.

Both the American and European societies agree that PVR is recommended for adults with repaired TOF that develop symptoms from pulmonary valve insufficiency (PI). Also, PVR has been recommended for select asymptomatic patients that have evidence of moderate or greater right ventricular (RV) dysfunction.[1,4] These recommendations are part of guidelines focusing broadly on congenital heart disease, providing a concise discussion on PVR and only partially addressing the complexity of the issue. Furthermore, both guidelines are intended for adults and lack specific consideration for children.

A detailed contemporary review and synthesis of existing data is necessary to guide management of both the pediatric and adult populations while identifying differences in management, if present. New data on imaging measures and clinical outcomes provides an opportunity to refine previous recommendations. These clinical practice guidelines (CPG) were written to address knowledge gaps by updating and expanding guidelines to incorporate advancements in surgical management and new technologies.

Methodology

In 2022, the members of the guidelines writing committee were assembled with a primary goal to include a diverse group of recognized experts in the field. Representation from centers and professional societies from across the globe was also prioritized and included: the Society of Thoracic Surgeons (STS) for North American representation and association with the largest clinical database for congenital heart surgery in North America, and international societies representing global voices for congenital heart surgery, specifically the World Society

for Pediatric and Congenital Heart Surgery (WSPCHS) and the European Congenital Heart Surgeons Association (ECHSA). The purpose of this writing group was to develop a CPG on indications and timing of pulmonary valve replacement in repaired tetralogy of Fallot.

The writing committee reviewed the existing published literature related to pulmonary valve replacement in repaired TOF and identified five clinically impactful questions framed in a PICO (population, intervention, comparison, and outcomes) format.[5] A qualified medical librarian developed a comprehensive search strategy for PubMed and Embase databases with input from the writing group. The search strategy is detailed in supplementary Appendix A. Formal search results were limited to articles published on human subjects in the English language with no restriction on publication year (i.e., from inception to October 2023). The literature search focused on study designs such as randomized control trials (RCT), reviews such as meta-analysis, systematic reviews, observational studies, and case studies. A hand search of bibliographies of the selected articles and Google Scholar supplemented the literature search for all PICO topics, and articles published between October 2023 and March 2025 were also included by expert selection for the PICO questions related to arrhythmia risk assessment. The manuscript was structured based on the Preferred Reporting items for Systematic Review and Meta-analysis (PRISMA) framework.[6]

Initially, a total of 3,247 potentially relevant articles were retrieved via a comprehensive database search (n=3,231) and additional citation search (n=16). During the title and abstract screening, articles were categorized according to the clinical (PICO) question they addressed. The strength of the available evidence surrounding the PICO topics of PVR timing and approach in children and exercise testing was not sufficient to support the creation of clinical practice guidelines, and these topics are addressed as an Expert Consensus Document (PVR in Children, Part 2) and an Expert Opinion/White Paper (Exercise Testing in Repaired TOF, Part 3), respectively.

The focus of this CPG document was thus narrowed to the following five specific questions:

1. Does PVR reduce symptoms for children and adults with repaired TOF and PI?
2. How can arrhythmia risk assessment in adults best inform the need for invasive electrophysiology (EP) procedures?
3. Does PVR reduce the risk of sudden cardiac death in adults?
4. Are RV volume measures so closely associated with ventricular function and outcomes in adults that RV volume remains an appropriate guide for timing of PVR?

5. How does RV and LV function change over time in repaired TOF with and without PVR, and how do those changes in function predict long-term outcomes for adults, including exercise capacity (by exercise testing), mortality, arrhythmia, and other signs of heart failure?

After excluding duplicates, and narrowing the scope to specific questions of interest, a total of 112 articles were identified as relevant for developing the recommendations for this manuscript, as detailed in the PRISMA flow chart (Figure 1). Writing group members screened the studies based on inclusion and exclusion criteria for relevance using a two-step screening process using Covidence, a web-based collaboration software platform.[7] The first step involved screening of titles and abstractions, and the second step involved full text review for inclusion. Two reviewers were assigned to each PICO for screening and conflicts were resolved via discussion.

Two independent reviewers assessed the studies for risk of bias. The Newcastle-Ottawa scale was used for observational studies, Cochrane Risk of Bias Version 1 was used for RCTs, and A MeaSurement Tool to Assess Systematic Reviews (AMSTAR) was used to assess systematic reviews and meta-analysis (Supplemental Appendix B).

Recommendations were formulated and reviewed by all members of the writing committee following the IOM standards. The recommendations were graded according to the American College of Cardiology/ American Heart Association Recommendation System.[8,9] A modified Delphi consensus method was used to achieve consensus with responses required by all the members of the writing committee, and 75% agreement on recommendation statements, class and level of evidence.[10] 5-point Likert scale (where 1=strongly agree, 2=disagree,3=neutral, 4=agree, and 5=strongly disagree) was used to rate the recommendations. A single voting round was needed to achieve consensus. Statements are presented in Table 1, voting results are detailed in Supplemental Appendix C, and supporting evidence by topic is outlined in Supplemental Appendix D.

The manuscript was reviewed and approved by the STS Workforce on Evidence-Based Surgery, the Council on Quality, Research & Patient Safety, and the STS Executive Committee, along with a 2-week public comment period. Additionally, the manuscript and recommendations were reviewed and endorsed by the WSPCHS and the ECHSA.

Literature Review and Recommendations

1. Symptomatic patients (children and adults)

> Pulmonary valve replacement is recommended to improve symptoms attributable to moderate or greater pulmonary insufficiency that may include decreasing objective exercise tolerance (i.e., lower than expected exercise tolerance on exercise testing), exertional dyspnea, decreased arterial oxygen saturation (<90%), and fatigue. (Class I: LOE B-NR)

PICO question: Does pulmonary valve replacement (PVR) reduce symptoms for patients with repaired tetralogy of Fallot (TOF) and pulmonary insufficiency (PI)?

Background and context

Symptoms have previously been a class I indication for pulmonary valve replacement (PVR) in repaired tetralogy of Fallot in the presence of at least moderate pulmonary valve insufficiency (PI).[3,11]Symptoms related to chronic PI in repaired TOF have been traditionally difficult to define, as they vary in type and severity, and are frequently subtle, or may not be perceived by patients as abnormal after living with PI for the majority of their lifetime. A contemporary review of the literature was undertaken to synthesize the available evidence surrounding symptomatology of chronic PI, and to elucidate if PVR alleviates symptoms for patients who have undergone repair of TOF with resultant moderate or greater PI.

Data synthesis

Data from 51 relevant papers were analyzed to examine the impact of PVR on various symptoms in patients with repaired TOF (refer to the Appendix D: Topics and supporting literature). These studies encompass a range of age groups, though most included a mixed cohort of pediatric and adult populations. The New York Heart Association (NYHA) functional class was assessed in 34 studies with 32 (94%) reporting significant symptomatic benefit, indicating strong evidence to support improvement in functional class in patients undergoing PVR. Additional symptoms including arrhythmias, syncope, and quality of life were collected suggesting a potential benefit of PVR, however the heterogeneity in methods across studies limited the interpretability of these findings.[12-14] To address this limitation and clarify arrhythmia burden before and after PVR, a separate, comprehensive review of contemporary evidence related to arrhythmia risk and role of electrophysiology study (EPS) prior to PVR is outlined in the next section of this document. A large body of work also investigated exercise capacity with mixed findings, and an in-depth analysis of the impact of PVR on exercise capacity can be found in the Expert Opinion Part 3 of this series.

2. Ventricular Arrhythmia Risk Assessment and Risk of Sudden Death (adults)

- Ventricular arrhythmia risk assessment prior to PVR is reasonable in selecting those patients who would most benefit from an invasive EP study with electroanatomic mapping. (Class IIa; LOE B, NR)
- EP study with right ventricular electroanatomic mapping, prior to transcatheter or surgical PVR, may be reasonable to identify patients with a potentially modifiable substrate for ventricular arrhythmias. (Class IIb; LOE B, NR)
- Ventricular arrhythmias are reduced, but not eliminated, after PVR. Continued serial assessment of ventricular arrhythmia risk in repaired TOF is recommended. (Class I; LOE B, NR)

PICO Question: How can arrhythmia risk assessment best inform the need for invasive EP procedures?

Background and context

Historically, early after initial complete repair of TOF, it was recognized that there was an association with sudden cardiac death (SCD) and ventricular arrhythmias (VA).[15,16] The use of intracardiac EPS for assessment of SCD in patients with repaired TOF was first described in 1977 and suggested VA may be the cause of sudden death in these patients.[17]

In the current era, the most common VA in repaired TOF is macroreentrant monomorphic ventricular tachycardia (VT) supported by anatomic isthmuses created as a result of the surgical repair.[18] Invasive EP assessment of VA risk has evolved to include programmed ventricular stimulation for assessment of inducibility of monomorphic VT, inducibility of polymorphic VT, and electroanatomic assessment of slowly conducting anatomic isthmuses (SCAI).[18,19]

Since the early 2000's, numerous studies in patients with repaired TOF have attempted to find an association between inducible VA and SCD. However, there has been inconsistency in the field regarding utilization of clinical data, both noninvasive markers and invasive EP study, for risk assessment of clinical VA and SCD. Pulmonary valve replacement (PVR) has emerged as one of the most frequent interventions in patients with repaired TOF aimed at relieving RV outflow tract obstruction and the deleterious effects of chronic pulmonary insufficiency on the right ventricle. With advances in transcatheter PVR, in conjunction with contemporary advances in electrophysiology three-dimensional mapping technologies and therapies, opportunities for arrhythmia intervention around the time of PVR is now often discussed.

Data synthesis

The studies evaluated were primarily adult studies and for that reason, this document will focus on data and recommendations for arrhythmia evaluation in adults with repaired TOF. There is not sufficient data regarding arrhythmia assessment in children to include them in this review.

In the last two decades, multiple studies have demonstrated numerous clinical risk factors to be associated with VA in repaired TOF. Invasive testing including elevated left ventricular end diastolic pressure (LVEDP) and inducibility of VT/VF at EP study have been demonstrated to be associated with VA in repaired TOF.[20] Noninvasive clinical risk factors which have been demonstrated to be associated with VA in repaired TOF include: older age at complete repair,[21] decade of repair, non-sustained VT (NSVT) prior to PVR, RVEF prior to PVR, older age at PVR,[22] history of VT, atrial arrhythmias,[23] QRS duration,[22,24] QRS fragmentation,[25,26] history of Blalock-Taussig-Thomas shunt,[19,27] older age,[28,29] history of atrial arrhythmia, lower RVEF, lower LVEF, and higher LVEDV, amount and location of ventricular scar,[29] frequent ventricular ectopy, cardiac index,[30] RV systolic dysfunction,[21] RVOT dysfunction,[31] reduced peak oxygen uptake (VO_2) and elevated B-type natriuretic peptide.[32]

EP studies for assessment of VA in repaired TOF are often performed around the time of surgical or transcatheter PVR, as this presents an opportunity for VA intervention if needed. Several studies have demonstrated that EP procedures prior to PVR in adults with repaired TOF alter clinical management or identify patients who would go on to have VA or ICD therapies.[33-35] EP studies as a screening tool are invasive and costly and realistically cannot be performed as frequent as noninvasive testing. Therefore, identifying patients with the highest pretest probability of positive findings on invasive EPS should be the goal to minimize unnecessary costly and invasive testing in those whose clinical data suggests a low risk of VA and SCD. Various clinical risk factors have been associated with inducibility of VA or slowly conducting anatomical isthmuses during EPS, including atrial tachyarrhythmias, native outflow tract PV annulus diameter >26mm,[36] age,[37] QRS vector,[38] CT detected wall thickness or calcification or high-intensity gadolinium signals during MRI.[37,39] Figure 2 presents a ventricular arrhythmia risk stratification tool to guide decision-making related to EPS and intervention. The categorization of low vs. intermediate vs. high risk for SCD is based on numerous clinical risk factors identified to be associated with VA in repaired TOF.[32,40-42] There is not a current era risk score that incorporates all clinical risk factors to define low vs. intermediate vs. high risk of VA, and published risk scores utilize different clinical risk factors. The variability in clinical approach to VA risk assessment in repaired TOF across centers currently limits the validity of firm

definitions for categorical risk. It is reasonable to classify risk of VA utilizing a published risk score with the understanding this is not a standardized definition.

In considering the use of clinical risk factors for assessment of VA risk to determine which patients would most benefit from EPS, it is important to note the following:

- There are numerous VA risk factors which have been identified and mixed results from various studies, suggesting that a broad assessment of demographic, noninvasive and invasive risk factors is necessary.
- Some studies do not identify any clinical risk factors which predict VA events after PVR,[43] so it is unclear if a thorough risk assessment is adequate for VT and SCD screening.
- Negative EPS prior to PVR does not indicate there is zero risk of VA after PVR.[34,44]
- In some cases, placement of a transcatheter PVR may limit the accessibility of a critical VT isthmus for a subsequent endocardial ablation.

PICO question: Does PVR reduce the risk of sudden death?

Assessment of ventricular arrhythmic events in TOF varies amongst studies. Most studies combine primary outcomes to include all ventricular arrhythmic events, including SCD, sustained VT, and ICD therapies.

In assessing risk of VA after PVR, studies have shown mixed results. Some studies have shown VA decreased after PVR,[45-48] some have not shown a reduction in mortality or VA events,[23,49-51] some have shown VT events occurred more often in those patients with PVR versus those without PVR.[52] A large study of 1744 patients with repaired TOF demonstrated a decrease in VA in patients without VA prior to PVR but no decrease in VA in patients with known VA prior to PVR.[28]

Multiple studies have evaluated clinical risk factors associated with VA events after PVR and demonstrated older age at PVR, atrial tachyarrhythmias after PVR, lower RVEF after PVR, decreased LV systolic function, history of VT prior to PVR, history of ICD implant prior to PVR, and longer QRS duration to be associated with VA events (death, SCD, sustained VT).[53-55]

PVR may reduce VA events, but it does not eliminate them. Furthermore, the substrate found most responsible for monomorphic VT in EPS prior to PVR is the anatomic isthmus between the PV and VSD, an area that may be rendered inaccessible after transcatheter PVR. New VT has been demonstrated post PVR in several studies.[23,37,56]

3. Ventricular Size and Function (adults)

- In an asymptomatic adult with repaired TOF and moderate or greater pulmonary insufficiency (regurgitant fraction ≥25%), pulmonary valve replacement is reasonable with 2 or more of the following:
  - RVEDV ≥160 ml/m^2
  - RVESV ≥80 ml/m^2
  - RVEF <47%
  - LVEF <55% (Class IIa; LOE B-NR)
- In an asymptomatic adult with repaired TOF and moderate or greater pulmonary insufficiency (regurgitant fraction ≥25%), pulmonary valve replacement may be considered with RVESV ≥ 80 ml/m^2 alone. (Class IIb; LOE B-NR)

PICO question: Are RV volume measures so closely associated with ventricular function and outcomes that RV volume remains an appropriate guide for timing of PVR?

Background and context

Cardiovascular magnetic resonance (CMR) is the gold standard for measurement of ventricular size and systolic function. While symptoms have been a class I indication for PVR,[11,57] CMR has played an important role particularly in asymptomatic patients with TOF post repair, in determining need for intervention. The physiological changes of severe PI (right ventricular [RV] dilatation, systolic and diastolic dysfunction in both right and left ventricles) can be accurately and reliably measured with freedom from limitations of acoustic windows. Current guidelines include consideration of pulmonary valve replacement with RV dilatation and/or dysfunction. Typical cutoffs for dilatation include RV end-diastolic volume ≥150-160 ml/m^2 (or RV:LV diastolic volume ratio of 2:1), or RVESV ≥80 ml/m^2. This largely relates to association with normalization of RV volumes after pulmonary valve replacement.[58-61] However, RV size still improves beyond these cutoffs (although may not normalize), and the correlation of normalization of RV size and function with clinical outcomes or mortality is unclear. In the absence of randomized controlled data, improvement in mortality has been difficult to demonstrate.

Data Synthesis

Across numerous studies, RVEDV and RVESV improve after PVR, with stable to small (but statistically significant) increases in RVEF and LVEF.[12,50,51,53,56,58,60-91] Although larger RVEDV and lower RVEF are less likely to normalize, there are not clear cutoffs beyond which normalization cannot occur.[59,78] Normalization of RV size and function may be better predicted by preoperative RVESV than RVEDV.[92-93] However, normalization of RV size and function has not been related to differences in outcome.[53,59]

In repaired TOF patients who have not undergone PVR, ventricular function is more consistently related to clinical outcome than RV dilatation (Table 2). In patients undergoing PVR, ventricular function remains a consistent correlate of outcome (Table 3). Of RV volume measurements, RVESV is more consistently related to outcome than RVEDV.[47,55,79,93,94]
Key recent additions to literature include two large cohort studies. In a Scottish national cohort of 341 adults,[51] preoperative RVEDV was related to the primary outcome of death or ventricular tachycardia. However, PVR was based on an RVEDV cutoff (150 ml/m^2), and PVR during the cohort period was related to an increase in the primary outcome (HR 2.82, 95% CI 1.36-5.86, p=0.005). PVR referral based on RVEDV alone is thus concerning. A large international cohort (INDICATOR TOF registry)[47] was designed to compare outcomes in patients who did or did not undergo PVR. Both in the entire cohort of 1,143 patients and in the propensity-matched cohort of 524 patients, PVR was associated with decreased rate of the primary outcome of death or sustained ventricular tachycardia (adjusted HR 0.41, 95% CI 0.21-0.81, p=0.010 in the propensity-matched cohort). The effect of PVR was dependent on RV dilatation and dysfunction – PVR was associated with improved outcome in patients with RVESV ≥80 ml/m^2 (HR 0.32, 95% CI 0.16-0.62, p<0.001), but there was no association with outcome in those with lower RVESV (HR 0.86, 95% CI 0.38-1.92, p=0.70). This manuscript was notable, as it demonstrated the survival benefit of PVR. This manuscript was further compelling due to its size, the matched nature of the cohorts of those who did and did not have PVR, and the identification of a preoperative CMR cutoff (RVESV ≥80 ml/m^2) associated with the survival benefit.

Potential future directions include myocardial characterization. Native T1 mapping and increased extracellular volume, which measure myocardial fibrosis, have been related to outcome in this population.[29,95-98] However, there are not yet data describing changes after pulmonary valve replacement, or potential cutoffs which may identify patients who would benefit with improved clinical outcome after intervention. In addition, the majority of existing data were generated from adults across a spectrum of operative approaches and eras. Increasing data from children and in a more modern cohort, including changes in risk/benefit ratio with transcatheter (vs surgical) approaches will be essential for contemporary decision-making.

While RVEDV decreases after PVR, there are inadequate data to demonstrate a benefit in clinical outcomes. Elevated RVEDV alone, in an asymptomatic adult, should not merit referral for PVR, but should be considered with other CMR or clinical factors. Prophylactic PVR prior to these cutoffs has not demonstrated an improvement in outcomes.

When evaluating ejection fraction for the purpose of determining intervention, standard calculation of RVEF [(RVEDV – RVESV) / RVEDV] is recommended rather than the "corrected" RVEF (net forward pulmonary flow / RVEDV), as the majority of data relating RVEF to clinical outcome utilizes the standard calculation. Increase in RVEF and LVEF post PVR, although statistically significant in several studies, is typically small. Due to relation of adverse outcomes with depressed systolic function, optimal timing may involve referral of patients prior to more significant decline in function. In patients with moderate or worse RV or LV systolic dysfunction, perioperative risk should be considered, and consultation with adult congenital heart disease or heart failure specialists should be considered, as PVR may not be tolerated or sufficient. While elevated RVEDV alone is an inadequate indication for PVR, RVESV better predicts normalization of RV size and function, with multiple studies relating RVESV to clinical outcome. The interaction of RVESV with survival benefit of PVR most strongly supports this as an indication for intervention. This also fits the physiologic importance of RVESV as a marker of both dilatation and dysfunction, and its relation to end-systolic wall stress. Utilizing a cutoff of RVESV ≥80 ml/m^2 effectively maintains the approach of 2 CMR cutoffs, as either the RVEDV would be ≥160 ml/m^2, or the RVEF would be <50% at lower RVEDV.

Other indications and clinical factors may be considered beyond the scope of typically measured CMR variables, as described elsewhere in these guidelines.

In summary, while RV volume decreases post PVR in patients with repaired TOF and pulmonary insufficiency, decrease or normalization of volume alone is not related to outcomes after PVR. RVESV, RVEF and LVEF, rather than RVEDV, are more closely related to clinical outcomes. However, further data are necessary in a contemporary surgical cohort, particularly evaluating efficacy of PVR in children and adolescents.

Acknowledgements: The authors would like to thank the staff of the STS, in particular Jane Han, Bansari Upadhyay, and Louie Neri, for their support, the members of the STS Workforce on Evidence-Based Surgery, and the anonymous reviewers for their diligent evaluation of these documents, their expertise, and edits, which have been critically important to creating the final version of this document.

Funding Sources: The authors have no funding sources to disclose.

Figure 1. PRISMA Flow Diagram

Figure 2. Ventricular arrhythmia risk stratification tool to guide clinical decision-making related to electrophysiology testing and intervention for patients with repaired tetralogy of Fallot.

Tables

Table 1: The Society of Thoracic Surgeons/World Society for Pediatric and Congenital Heart Surgery/European Society 2026 Clinical Practice Guidelines for the Timing and Indications for Pulmonary Valve Replacement in Tetralogy of Fallot

Recommendations	COR	LOE
Recommendation for symptomatic patients		
Pulmonary valve replacement is recommended to improve symptoms attributable to moderate or greater pulmonary insufficiency that may include decreasing objective exercise tolerance (i.e., lower than expected exercise tolerance on exercise testing), exertional dyspnea, decreased arterial oxygen saturation (<90%), and fatigue.	I	B-NR
Recommendations for electrophysiology studies		
Ventricular arrhythmia risk assessment prior to PVR is reasonable in selecting those patients who would most benefit from an invasive EP study with electroanatomic mapping.	IIa	B-NR

Recommendations	COR	LOE
EP study with right ventricular electroanatomic mapping, prior to transcatheter or surgical PVR may be reasonable to identify patients with a potentially modifiable substrate for ventricular arrhythmias.	IIb	B-NR
Ventricular arrhythmias are reduced, but not eliminated, after PVR. Continued serial assessment of ventricular arrhythmia risk in repaired TOF is recommended.	I	B-NR
Recommendations for CMR-based metrics		
In an asymptomatic adult with repaired TOF and moderate or greater pulmonary insufficiency (regurgitant fraction ≥25%), pulmonary valve replacement is reasonable with 2 or more of the following: • RVEDV ≥160 ml/m^2 • RVESV ≥80 ml/m^2 • RVEF <47% • LVEF <55%	IIa	B-NR
In an asymptomatic adult with repaired TOF and moderate or greater pulmonary insufficiency (regurgitant fraction ≥25%), pulmonary valve replacement may be considered with RVESV ≥ 80 ml/m^2 alone.	IIb	B-NR

COR: Class of Recommendation, LOE: Level of Recommendations, TOF: Tetralogy of Fallot, RVEDV: Right Ventricular End-Diastolic Volume, RVESV: Right Ventricular End-Systolic Volume, RVEF: Right Ventricular Ejection Fraction, LVEF: Left Ventricular Ejection Fraction, EP: Electrophysiology, PVR: Pulmonary Valve Replacement, B-NR: Level B Nonrandomized

Table 2: Relation of CMR measures to clinical outcomes in repaired TOF patients without PVR

Study	n	Study type	Age (yrs)	Outcome	Comments
Right ventricular ejection fraction (RVEF)					
Giardini 2006[99]	61	Retrospective observational	23.1 ± 12.1	Exercise capacity	RVEF <40% associated with lower peak VO_2, VO_2 slope, VCO_2 slope
Meadows 2007[100]	37	Retrospective observational	26.8 ± 11.9	Exercise capacity	RVEF correlated with peak VO_2 and % predicted oxygen pulse; RVEF cutoff ≤47%
Lu 2010[101]	67	Retrospective observational	28.5 (14-69)	Functional health status	RVEF <45% had decreased Short Form 36 scores
Left ventricular ejection fraction (LVEF)					
Geva 2004[102]	100	Retrospective observational	24 (10-57)	NYHA class ≥3	LVEF significant on multivariate analysis; limiting to RV variables, RVEF and RV mass:volume were significant
Knauth 2008[103]	88	Case control	24.0 (10.0-57.6)	Death, sustained VT, NYHA class	RVEDV Z-score ≥7 and LVEF <55% associated with primary outcome on multivariate analysis
Yang 2015[104]	158	Prospective observational	29.5 ± 12.2	Exercise capacity	LVEF correlated with peak VO_2 and oxygen update efficiency plateau
Right ventricular end systolic volume (RVESV)					
Lu 2016[94]	28	Retrospective observational	33.5 (26-42)	Subsequent change in functional health status	Baseline RVESV correlated with subsequent decrease in Short Form 36 scores
Right ventricular end diastolic volume (RVEDV)					
Knauth 2008[103]	88	Case control	24.0 (10.0-57.6)	Death, sustained VT, NYHA class	RVEDV Z-score ≥7 and LVEF <55% associated with primary outcome on multivariate analysis
Gnanappa 2019[65]	126	Retrospective observational	17.3 ±7.6	VO_2 max	VO_2 max did NOT differ in those with RVEDV ≥ 170 ml/m_2 vs less

NYHA, New York Heart Association class; RVEDV, right ventricular end-diastolic volume; RVESV, right ventricular end-systolic volume; RVEF, right ventricular ejection fraction; LVESV, left ventricular end-systolic volume; LVEF, left ventricular ejection fraction; SCD, sudden cardiac death; VT, ventricular tachycardia; PVR, pulmonary valve replacement.

Table 3: Relation of CMR measures to clinical outcomes in patients post PVR

Study	n	Study type	Age at PVR (yrs)	Outcome	Comments
Right ventricular ejection fraction (RVEF)					
Heng 2017[89]	57	Retrospective observational	35.8 ± 10.1	Death, VT	Pre-PVR RVEF (but not RVEDV) related to primary outcome after PVR
Bokma 2018[23]	977	cohort	Not reported	Death, sustained VT	RVEF related to primary outcome in multivariate analysis of entire cohort (PVR or no) and in matched cohort; higher event rates if CMR criteria for PVR not met
Geva 2018[21]	1418	Retrospective observational	25.8 (18.6-37.6)	Death, aborted SCD, sustained VT	Pre-PVR RVEF <40%, RV mass:volume ≥0.45 associated with primary outcome (on multivariate analysis) after PVR
Pastor 2020[53]	189	Cross-sectional Study	23.5 ± 11.7	Death, sustained VT, aborted SCD, NYHA class	Post-PVR RVEF related to outcomes; neither pre-op nor post-op RVEDV related
Left ventricular ejection fraction (LVEF)					
Lee 2020[55]	190	Retrospective observational	19.0 (14.1-22.6)	Death, OHT, arrhythmia	Preop RVESV, LVESV and lower LVEF related to adverse events
Right ventricular end systolic volume (RVESV)					
Bokma 2016[93]	65	Retrospective observational	29 ± 8.3	Death, VT, heart failure	RVESV >95 ml/m^2 related to outcome
He 2019[79]	81	Case-control	21.0 ± 17.0	Death, OHT, arrhythmia, PVR, CHF	RVESV associated with adverse events in those without PVR; RVESV <120 ml/m^2 associated with normalization of RV size after PVR
Lee 2020[55]	190	Retrospective observational	19.0 (14.1-22.6)	Death, OHT, arrhythmia	Preop RVESV, LVESV and lower LVEF related to adverse events
Bokma 2023[47]	1143	Cohort	Not reported	Death, sustained VT	PVR beneficial if RVESV≥80 ml/m^2, PVR not beneficial if CMR criteria not met

Right Ventricular End Diastolic Volume (RVEDV)					
Ferraz Cavalcanti 2013[63]	3118	Meta-analysis	No aggregate data	NYHA status	Greater RVEDV had *less* improvement in NYHA
Dobson 2021[51]	341	Retrospective observational	Not reported	Death, arrhythmia	RVEDV related to primary outcome (many without PVR); PVR also associated with increased risk of primary outcome
Left Ventricular End Systolic Volume (LVESV)					
He 2022[56]	42	Retrospective observational	21.6 (15.4-24.8)	Death, arrhythmia, valve failure or repeat PVR	On multivariate analysis, only LVESV related to outcomes (RVEDV, RVESV RVEF, RVEF and LVEF only on univariate)

NYHA, New York Heart Association class; RVEDV, right ventricular end-diastolic volume; RVESV, right ventricular end-systolic volume; RVEF, right ventricular ejection fraction; LVESV, left ventricular end-systolic volume; LVEF, left ventricular ejection fraction; SCD, sudden cardiac death; VT, ventricular tachycardia; PVR, pulmonary valve replacement.

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Figure 1

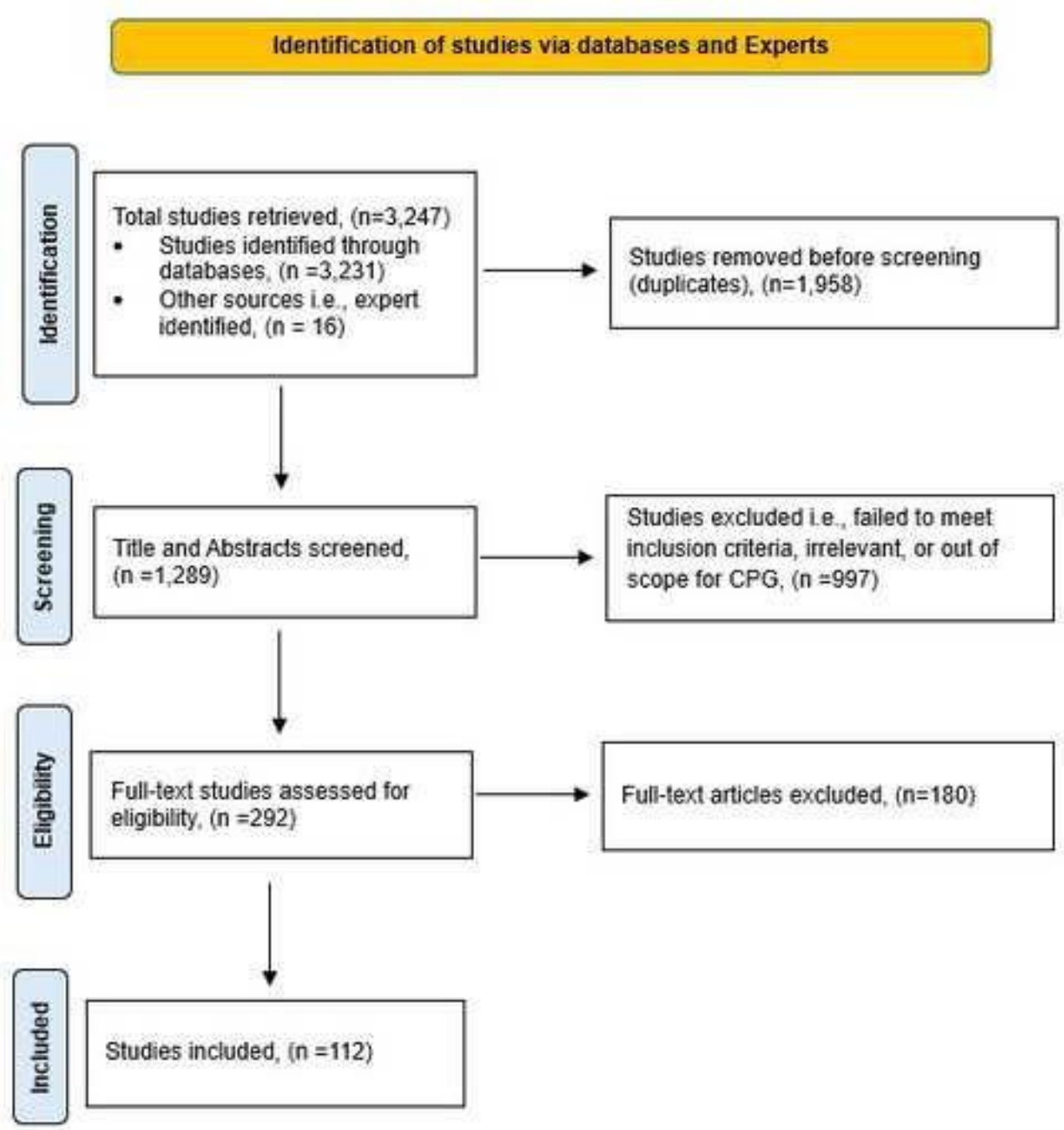

Figure 2