Open Access | Opinion Research
Journal of Heart Valve Disease, 29(1), 2024 | PP: 30– 36
https://doi.org/10.36923/jhvd.v29i1.212

The Dilemma of Pulmonary Valve Replacement In Patients With Repaired Tetralogy of Fallot

Amir-Reza Hosseinpour[1], Matthias Kirsch[2], Maria-Helena Perez[3], Stefano Di Bernardo[4]

Abstract: Patients with repaired tetralogy of Fallot often have pulmonary valvar regurgitation with variable degrees of right ventricular dilatation, which may lead to dysfunction of both ventricles. Therefore, some patients may need pulmonary valve replacement. This calls for adequate patient selection, and there are ventricular volumetric guidelines to aid this. However, pulmonary valve replacement has caused a dilemma because: 1- It often does not reverse right ventricular dilatation back to normal nor prevent arrhythmias. 2- It is argued that right ventricular dilatation occurs early after repair of tetralogy of Fallot but remains stable thereafter. 3- The patient' prognosis is said to be affected mainly by the function of the left ventricle rather than that of the right ventricle. 4- Prosthetic valves on the right side of the heart are more prone to infection as compared to the left side since bacteria can reach them easily. All these concerns are examined in this Opinion Article. Overall, this dilemma is justified but usually not sufficiently elaborated and often gives the incorrect message that this procedure is of no benefit. The reality is, however, that pulmonary valvar regurgitation is a harmful condition and deserves to be remedied on hemodynamic grounds. The dilemma arises because hemodynamic improvement after valve implantation may be partial, and many think that this is not enough to justify an intervention, especially with the added risk of endocarditis. However, if valve replacement is to be offered, this should be done in a timely fashion; waiting too long is associated with a worse outcome.

Keywords: Pulmonary Valve Insufficiency, Pulmonary Valve Regurgitation, Pulmonary Valve Replacement, Tetralogy of Fallot, Adults with Congenital Heart disease

Article History:

Received: 07-09-2024
Accepted: 01-08-2024
Publication: 08-08-2024

Cite this article as:

Hosseinpour, A.-R., Kirsch, M., Perez, M.-H., & Di Bernardo, S. (2024). The dilemma of pulmonary valve replacement in patients with repaired tetralogy of Fallot. *Journal of Heart Valve Disease,* 29(1). 30-36. doi.org/10.36923/jhvd.v29i1.212

Corresponding Author(s):

Amir-Reza Hosseinpour
Service of Cardiac Surgery, Lausanne University Hospital, Switzerland. Email: reza@hosseinpour.net

1. Introduction

Patients with repaired tetralogy of Fallot (ToF) often have pulmonary valvar regurgitation [1]. This causes variable degrees of right ventricular (RV) dilatation, which may eventually lead to dysfunction of both the right and left ventricles (LV) and symptoms of heart failure. Therefore, some patients may need pulmonary valve replacement (PVR). This may be done surgically or percutaneously, depending on anatomic characteristics. Either way, PVR calls for adequate patient selection so that this treatment is offered before their RV becomes so dilated and dysfunctional that full recovery is unlikely. That point of no return is elusive, but current guidelines recommend PVR when the indexed right ventricular end-diastolic volume (RVEDV) reaches 160 ml/m², although other factors are also considered, including an end-systolic volume of 80 ml/m² and the rate of ventricular dilatation, even though the rate that should cause alarm is not known [2].

Actually, these volumetric criteria offer a rather limited view of the cardiac status of these patients, ignoring that no two such patients are alike. In fact, these patients vary considerably depending on whether they have had an initial shunt, infundibulotomy, transannular patch, patch repairs of branch pulmonary arteries and residual stenoses, the degree of residual pulmonary valvar regurgitation and/or stenosis, and their age at the time of repair. Ideally, all these factors should be included in the decision process for PVR. In practice, however, this would be too complicated, and the decision process has been simplified to the above-mentioned volumetric criteria.

Nevertheless, it is important to remind ourselves, as it is often forgotten, that these volumetric criteria have been proposed to help us decide when to offer PVR to patients who are asymptomatic or only mildly symptomatic. Such a guide is useful because symptoms of heart failure are not always present, which may give us a false sense of security. Indeed, symptoms often arise quite late when the benefits of valve replacement may be diminished due to excessive ventricular dilatation and dysfunction, i.e., waiting for symptoms is not a safe strategy. However, symptoms may arise early in some patients, in which case these volumetric criteria become irrelevant and PVR

[1]Service of Cardiac Surgery, Lausanne University Hospital, Switzerland. Email: reza@hosseinpour.net
[2]Service of Cardiac Surgery, Lausanne University Hospital, Switzerland. Email: matthias.kirsch@chuv.ch
[3]Service of Cardiac Surgery, Lausanne University Hospital, Switzerland. Email: Marie-Helene.Perez@chuv.ch
[4]Service of Cardiac Surgery, Lausanne University Hospital, Switzerland United Kingdom. Email: stefano.di-bernardo@chuv.ch

must be offered; it would be clinically unsound to deny PVR to symptomatic patients simply because their RV is not "sufficiently dilated" yet. Such a weak correlation between symptoms and RV dimensions may possibly reflect the fact that many factors other than RV dimensions are ignored in the decision-making process as mentioned above.

Independently of the factors used for the indication, however, PVR comes at a significant cost and is therefore not a simple decision. This is a clinical dilemma that is worth examining.

2. The root of this dilemma

This dilemma has emerged because PVR in patients with repaired ToF did not produce the results that many clinicians were expecting [3-10]. Indeed, expectations were high and, with the benefit of hindsight, unrealistic. Many were expecting a cure, thinking that PVR would 'normalize' these hearts, which it does not; the consequent disappointment undermined the enthusiasm for this treatment. This is a fundamental misunderstanding of what we can achieve in patients with ToF. We cannot cure ToF at present; we can only palliate it [11]. This statement may cause confusion because the term palliation in our specialty conventionally refers to the management of univentricular hearts, which is not what is meant here; we employ this term literally in its generic sense, i.e., treatments that we offer to ameliorate a condition that we cannot cure. This is the frame of mind in which we must assess our treatment options at present, including PVR; otherwise, these will all be disappointing, i.e., we must appreciate the lesser (palliative) benefits of our current treatment modalities.

This inability to cure is reflected in the following sobering findings in the long-term follow-up of patients with repaired ToF:

1. Progressive RV dilatation/dysfunction with wide QRS complexes and a high incidence of ventricular arrhythmias, even with a competent pulmonary valve, but more so with pulmonary valvar incompetence [1, 10, 12].
2. Adults with repaired ToF constitute the largest subgroup of patients with congenital heart disease who have received an implantable cardioverter-defibrillator [13, 14].
3. The LV also becomes dysfunctional, and this undermines prognosis [15-29].
4. Many of these patients die young. Indeed, 30-year mortality is 5% in the least complex cases, and 22% in the most complex ones [30].

Despite these findings, however, we must not undervalue the treatments that are currently available for ToF, which have enabled us to overcome the dismal natural history of this malformation. Clearly, patients now live much better and longer because of our treatments than they would otherwise. These treatments follow the same basic pattern seen in all areas of medicine—when cure is not possible, the next best objective is to reduce the deleterious effects of the illness, which is one way to define palliation. Indeed, severe pulmonary valvar regurgitation has significant deleterious effects in patients with repaired ToF [1, 12]; this should be rectified as a matter of adequate palliation. Nevertheless, the dilemma continues with four arguments, three of which can be argued against, but one that is difficult to deny.

3. The 3 arguments that can be argued against

One or more of the following three arguments are presented in various publications:

1. PVR does not achieve its intended objective, which is to reverse RV dilatation/dysfunction and prevent arrhythmias.
2. RV dilatation occurs early after the repair of ToF but remains stable thereafter, implying that PVR may not be needed.
3. The patient's prognosis is affected mainly by the function of the LV rather than the RV, implying that we need not worry so much about the RV.

These three statements should be examined more closely:

3.1. PVR does not achieve its intended objective

It is true that PVR often does not reverse RV dilatation/dysfunction or the widening of QRS complexes back to normal, and the propensity to ventricular arrhythmias continues [3-10]. Nevertheless, there is some RV remodeling with a decrease in volumes after PVR, even if not completely back to normal (although some do recover completely), i.e., the ventricle is suffering less, as shown in several studies [3, 5, 7, 31-39]. None of these studies could identify a threshold of RV volume above which the volume did not decrease at all after PVR, although normalization was unlikely with larger preoperative volumes. There is also a small but statistically significant decrease in QRS duration [35].

Interestingly, one of these studies shows that these improvements are not maintained; the RV dilates again in 7-10 years [36]. Again, this is disappointing, but in the context of palliation, an improved condition for 7-10 years is valuable.

3.2. RV dilatation occurs early after the repair of ToF but then remains stable

This statement emerged from short-term studies of patients who were either asymptomatic (the majority) or only mildly symptomatic, with only mild, or at the most moderate, RV dilatation [34, 40-43]. In other words, these studies focused on patients that nobody would consider for PVR in the first place. Patients who were accepted for PVR were excluded from all these studies, which are briefly summarized here.

Rutz et al. assessed patients who typically had RVEDVs of 110-120 ml/m² (i.e., only slightly dilated) and volumetric comparisons were done over a period of only about 3 years, which was the period between their first and last magnetic resonance imaging (MRI) [40]. Not surprisingly, these remained stable during this short study period. In addition, the authors provide no information as to the time lapse between the operation (ToF repair) and their first MRI.

Quail et al. assessed patients with an average RVEDV of only about 130 ml/m² and compared those who were offered PVR to those who were not [34]. The authors do not specify how much time had lapsed since ToF repair in these patients, but state that this was similar between the two groups. Again, RV volumes remained stable in those without PVR, but comparisons were done over a short period (<2 years). However, RV volumes were normalized in most patients who were offered PVR.

Luijnenburg et al. also assessed patients with RVEDVs of about 130 ml/m², but these were followed up for 5 years, although the authors do not specify how much time had lapsed since their corrective operation [41]. In these patients, RV volumes did increase, as did QRS duration, although slowly. In other words, by prolonging the follow-up period, volume instability becomes visible despite relatively low initial volumes.

A multicentric study between North America and Europe, led by Wald et al., assessed patients with RVEDVs of 140-145 ml/m², i.e., moderate dilatation without reaching the conventional threshold for surgical indication [42]. These patients were assessed over only 2 years. Despite such a short period, RV volumes increased in 15% of patients.

Finally, Hoelscher et al. conducted a study between Switzerland and Italy assessing patients with slightly larger average RVEDVs of about 150 ml/m² over a 3-year period [43]. Even though ventricular volumes of these patients were still below the level of surgical indication, and despite a relatively short study period, RV volumes increased rapidly in 25% of patients.

Clearly, these data do not tell us what would happen if PVR is not offered to patients who do meet the current criteria for this treatment, i.e., symptoms of heart failure and/or RVEDVs >160 ml/m². However, they do show that even among asymptomatic patients with smaller ventricles, RV volumes remain stable in the short term only in some, and this is unpredictable and less likely with larger ventricles.

3.3. Patient's prognosis is affected mainly by the function of the LV rather than the RV

This statement has emerged from several studies [15-29]. LV dysfunction in these patients is not surprising because there is no doubt that RV dysfunction will secondarily also cause LV dysfunction, although this may also be caused by chronic hypoxemia in patients who were operated on late, or due to inadequate myocardial protection in cases of protracted surgical correction [20, 21]. However, it has also been shown that LV function may improve if RV function is improved, which may be achieved with PVR [33-35, 37, 44].

4. The argument that is more difficult to deny

An ongoing concern is that prosthetic valves on the right side of the heart are more prone to infection compared to those on the left side, as bacteria can reach them more easily [48, 49]. Left-sided prosthetic valves are relatively protected because, in the absence of septal defects, bacteria would have to pass through the lungs to reach these prostheses, and the lungs are quite effective bacterial filters, both mechanically and immunologically. Although infective endocarditis is always a threat in many forms of congenital heart disease, the highest risk group appears to be patients with prostheses in the pulmonary position. This is associated with significant morbidity and mortality. This is undoubtedly the strongest argument against the implantation of prosthetic valves in the pulmonary position.

5. Discussion

The dilemma regarding PVR in patients with pulmonary valvar regurgitation following repair of ToF is justified but usually insufficiently elaborated, often giving an incorrect message. Indeed, the message often insinuated is that this procedure is of no benefit to the heart and may even make it worse. This is very misleading and makes no pathophysiologic sense. The reality is that the heart is better off without pulmonary valvar regurgitation than with it, as with any valve. This is not where the dilemma lies. The dilemma exists because PVR usually results in only partial cardiac improvement or, in the worst cases, it may possibly only serve to prevent or slow down further deterioration, and many think that this does not justify an intervention, especially with the added risk of endocarditis. Certainly, endocarditis is a more significant risk to patients' lives acutely than the chronic effects of pulmonary valvar regurgitation, hence the reluctance to offer valve implantation. However, to further fuel the dilemma,

if valve replacement is to be offered, this should be done in a timely fashion; waiting too long is associated with worse outcomes [10, 31, 32, 39, 45-47].

Acknowledgement Statement: The authors would like to thank the reviewers for providing comments in helping this manuscript to completion.

Conflicts of Interest: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

CRediT Author contribution statements: All authors contributed to the intellectual property of this manuscript and have been involved in the design, structure, data collation, writing, and reviewing of this manuscript.

Funding: This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Data Availability Statement: Data is available at request. Please contact the corresponding author for any additional information on data access or usage.

Disclaimer: The views and opinions expressed in this article are those of the author(s) and contributor(s) and do not necessarily reflect JHVD's or editors' official policy or position. All liability for harm done to individuals or property as a result of any ideas, methods, instructions, or products mentioned in the content is expressly disclaimed.

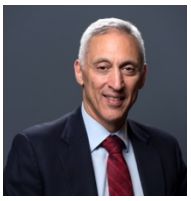
References

1. Gatzoulis MA, Till JA, Somerville J, Redington AN. Mechanoelectrical interaction in tetralogy of Fallot: QRS prolongation relates to right ventricular size and predicts malignant ventricular arrhythmias and sudden death. *Circulation*. 1995;92(2):231-237. https://doi.org/10.1161/01.CIR.92.2.231 PMid:7600655
2. Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, et al. 2018 AHA/ACC guidelines for the management of adults with congenital heart disease: A report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2019;73(12):1494-1563. https://doi.org/10.1016/j.jacc.2018.08.1028 PMid:30121240
3. Wald RM, Haber I, Wald R, Valente AM, Powell AJ, Geva T. Effects of regional dysfunction and late gadolinium enhancement on global right ventricular function and exercise capacity in patients with repaired tetralogy of Fallot. *Circulation*. 2009;119(10):1370-1377. https://doi.org/10.1161/CIRCULATIONAHA.108.816546 PMid:19255342 PMCid
4. Harrild DM, Berul CI, Cecchin F, Geva T, Gauvreau K, Pigula F, et al. Pulmonary valve replacement in tetralogy of Fallot: impact on survival and ventricular tachycardia. *Circulation*. 2009;119(3):445-451. https://doi.org/10.1161/CIRCULATIONAHA.108.775221 PMid:19139389 PMCid
5. Geva T, Gauvreau K, Powell AJ, Cecchin F, Rhodes J, Geva J, et al. Randomized trial of pulmonary valve replacement with and without right ventricular remodeling surgery. *Circulation*. 2010;122(11 Suppl). https://doi.org/10.1161/CIRCULATIONAHA.110.951178 PMid:20837914 PMCid
6. Scherptong RW, Hazekamp MG, Mulder BJ, Wijers O, Swenne CA, van der Wall, et al. Follow-up after pulmonary valve replacement in adults with tetralogy of Fallot: association between QRS duration and outcome. *J Am Coll Cardiol*. 2010;56(18):1486-1492. https://doi.org/10.1016/j.jacc.2020.04.058 PMid:32527397
7. Plymen CM, Finlay M, Tsang V, O'Leary J, Picaut N, Cullen S, et al. Haemodynamic consequences of targeted single- and dual-site right ventricular pacing in adults with congenital heart disease undergoing surgical pulmonary valve replacement. *Europace*. 2015;17(2):274-280. https://doi.org/10.1093/europace/euu281 PMid:25371427 PMCid
8. Bokma JP, Geva T, Sleeper LA, Babu Narayan SV, Wald R, Hickey K, et al. A propensity score-adjusted analysis of clinical outcomes after pulmonary valve replacement in tetralogy of Fallot. *Heart*. 2018;104(9):738-744. https://doi.org/10.1136/heartjnl-2017-312048 PMid:29092913
9. Cohen MI, Khairy P, Zeppenfeld K, Van Hare GF, Lakkireddy DR, Triedman JK. Preventing arrhythmic death in patients with tetralogy of Fallot: JACC review topic of the week. *J Am Coll Cardiol*. 2021;77(6):761-771.. https://doi.org/10.1016/j.jacc.2020.12.021 PMid:33573746
10. Van den Eynde J, Pompeu M, Vervoort D, Roever L, Meyns B, Budts W, et al. Pulmonary Valve Replacement in Tetralogy of Fallot: An Updated Meta-Analysis. *Ann Thorac Surg*. 2022;113(3):1036-46.. https://doi.org/10.1016/j.athoracsur.2020.11.040 PMid:33378694
11. Hosseinpour AR, González-Calle A, Adsuar-Gómez A, Ho SY. The predicament of surgical correction of tetralogy of Fallot. *Pediatr Cardiol*. 2021;42(6):1252-1257. https://doi.org/10.1007/s00246-021-02662-6 PMid:34175987
12. Khairy P, Aboulhosn J, Gurvitz MZ, Opotowsky AR, Mongeon FP, Valente AM, et al. Arrhythmia burden in adults with surgically repaired tetralogy of Fallot: a multi-institutional study. *Circulation*. 2010;122(9):868-875. https://doi.org/10.1161/CIRCULATIONAHA.109.928481 PMid:20713900
13. Khairy P, Harris L, Landzberg MJ, Viswanathan S, Barlow A, Gatzoulis MA, et al. Implantable cardioverter-defibrillators in tetralogy of Fallot. *Circulation*. 2008;117(3):363-370. https://doi.org/10.1161/CIRCULATIONAHA.107.726372 PMid:18172030

14. Koyak Z, Harris L, de Groot JR, Silversides CK, Oechslin EN, Bouma BJ, et al. Sudden cardiac death in adult congenital heart disease. *Circulation*. 2012;126(16):1944-1954. https://doi.org/10.1161/CIRCULATIONAHA.112.104786 PMid:22991410
15. Gonzalez de Alba C, Khan A, Woods P, Broberg CS. Left ventricular strain and fibrosis in adults with repaired tetralogy of Fallot: A case-control study. *Int J Cardiol*. 2021;323:34-39. https://doi.org/10.1016/j.ijcard.2020.08.092 PMid:32882293
16. Diller G-P, Kempny A, Liodakis E, Alonso-Gonzalez R, Inuzuka R, Uebing A, et al. Left Ventricular Longitudinal Function Predicts Life-Threatening Ventricular Arrhythmia and Death in Adults With Repaired Tetralogy of Fallot. *Circulation*. 2012;125(20):2440-2446. https://doi.org/10.1161/CIRCULATIONAHA.111.086983 PMid:22496160
17. Ait Ali L, Trocchio G, Crepaz R, Stuefer J, Stagnaro N, Siciliano V, et al. Left ventricular dysfunction in repaired tetralogy of Fallot: incidence and impact on atrial arrhythmias at long term follow up. *Int J Cardiovasc Imaging*. 2016;32(9):1441-1449. https://doi.org/10.1007/s10554-016-0928-7 PMid:27318953
18. Miyake M, Abe R, Matsutani H, Sakamoto J, Kondo H, Iwakura A, et al. Relationship between right and left ventricular diastolic dysfunction assessed by 2-dimensional speckle-tracking echocardiography in adults with repaired tetralogy of Fallot. *Int J Cardiovasc Imaging*. 2021;37(2):569-576. https://doi.org/10.1007/s10554-020-02045-7 PMid:33006716 PMCid
19. Andrade AC, Jerosch-Herold M, Wegner P, Gabbert DD, Voges I, Pham M, et al. Determinants of Left Ventricular Dysfunction and Remodeling in Patients With Corrected Tetralogy of Fallot. *J Am Heart Assoc*. 2019;8(17). https://doi.org/10.1161/JAHA.118.009618 PMid:31474177 PMCid
20. Egbe AC, Adigun R, Anand V, West CP, Montori VM, Murad HM, et al. Left Ventricular Systolic Dysfunction and Cardiovascular Outcomes in Tetralogy of Fallot: Systematic Review and Meta-analysis. *Can J Cardiol*. 2019;35(12):1784-1790. https://doi.org/10.1016/j.cjca.2019.07.634 PMid:31732195
21. Wald R, Oechslin EN. Looking to the left to get it right: left ventricular systolic dysfunction and risk stratification late after tetralogy of Fallot repair. *Can J Cardiol*. 2019;35(12):1623-1625. https://doi.org/10.1016/j.cjca.2019.10.003 PMid:31813498
22. Hickey EJ, Veldtman G, Bradley TJ, Gengsakul A, Manlhiot C, Williams WG, et al. Late risk of outcomes for adults with repaired tetralogy of Fallot from an inception cohort spanning four decades. *Eur J Cardiothorac Surg*. 2009;35(1):156-164. https://doi.org/10.1016/j.ejcts.2008.06.050 PMid:18848456
23. 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(1):9. https://doi.org/10.1186/1532-429X-13-9 PMid:21251297 PMCid
24. Tobler D, Crean AM, Redington AN, Van Arsdell GS, Caldarone CA, Nanthakumar K, et al. The left heart after pulmonary valve replacement in adults after tetralogy of Fallot repair. *Int J Cardiol*. 2012;160(3):165-170. https://doi.org/10.1016/j.ijcard.2011.04.007 PMid:21570135
25. Valente AM, Gauvreau K, Assenza GE, Babu-Narayan SV, Schreier J, Gatzoulis MA, et al. Contemporary predictors of death and sustained ventricular tachycardia in patients with repaired tetralogy of Fallot enrolled in The Indicator cohort. *Heart*. 2014;100(3):247-253. https://doi.org/10.1136/heartjnl-2013-304958 PMid:24179163 PMCid
26. Dragulescu A, Friedberg MK, Groose-Wortmann L, Redington A, Mertens L. Effect of chronic right ventricular volume overload on ventricular interaction in patients after tetralogy of Fallot repair. *J Am Soc Echocardiogr*. 2014;27(8):896-902. https://doi.org/10.1016/j.echo.2014.04.012 PMid:24846007
27. Orwat S, Diller G-P, Kempny A, Radke R, Peters B, Külne T, et al. Myocardial deformation parameters predict outcomes in patients with repaired tetralogy of Fallot. *Heart*. 2016;102(3):209-215. https://doi.org/10.1136/heartjnl-2015-308569 PMid:26715570
28. Pastor TA, Geva T, Lu M, et al. Relation of right ventricular dilation after pulmonary valve replacement to outcomes in patients with repaired tetralogy of Fallot. *Am J Cardiol*. 2020;125(6):977-981. https://doi.org/10.1016/j.amjcard.2019.12.017 PMid:31959431
29. Possner P, Tseng SY, Alahdab F, Bokma JP, Lubert AM, Khairy P, et al. Risk factors for mortality and ventricular tachycardia in patients with repaired tetralogy of Fallot: A systematic review and meta-analysis. *Can J Cardiol*. 2020;36(11):1815-1825. https://doi.org/10.1016/j.cjca.2020.01.023 PMid:32416063
30. Blais S, Marelli A, Vanasse A, et al. The 30-year outcomes of tetralogy of Fallot according to native anatomy and genetic conditions. *Can J Cardiol*. 2021;37(6):877-886. https://doi.org/10.1016/j.cjca.2020.10.002 PMid:33059007
31. Buechel ERV, Dave HH, Kellenberger CJ, Dodge-Khatami A, Pretre R, Berger F, Bauersfeld U. Remodelling 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(24):2721-2727. https://doi.org/10.1093/eurheartj/ehi581 PMid:16214832
32. Therrien J, Provost Y, Merchant N, Williams W, Colman J, Webb G. Optimal timing for pulmonary valve replacement in adults after tetralogy of Fallot repair. *Am J Cardiol*. 2005;95(6):779-782. https://doi.org/10.1016/j.amjcard.2004.11.037 PMid:15757612
33. Oosterhof T, van Straten A, Vliegen HW, Meijboom FJ, van Dijk APJ, Spijkerboer AM, et al. Preoperative thresholds for pulmonary valve replacement in patients with corrected tetralogy of Fallot using cardiovascular

magnetic resonance. *Circulation*. 2007;116(5):545-551. https://doi.org/10.1161/CIRCULATIONAHA.106.659664 PMid:17620511
34. Quail MA, Frigiola A, Giardini A, Muthurangu V, Hughes M, Lurz P, et al. Impact of pulmonary valve replacement in tetralogy of Fallot with pulmonary regurgitation: a comparison of intervention and nonintervention. *Ann Thorac Surg*. 2012;94(5):1619-1626. https://doi.org/10.1016/j.athoracsur.2012.06.062 PMid:22959579
35. Ferraz Cavalcanti PE, Barros Oliveira Sá MP, Andrade Santos C, Melo Esmeraldo I, Renda de Escobar R, Motta de Menezes A, et al. Pulmonary valve replacement after operative repair of tetralogy of Fallot: meta-analysis and meta-regression of 3,118 patients from 48 studies. *J Am Coll Cardiol*. 2013;62(23):2227-2243. https://doi.org/10.1016/j.jacc.2013.04.107 PMid:24080109
36. Hallbergson A, Gauvreau K, Powell AJ, Geva T. Right ventricular remodeling after pulmonary valve replacement: early gains, late losses. *Ann Thorac Surg*. 2015;99(2):660-666. https://doi.org/10.1016/j.athoracsur.2014.09.015 PMid:25483002
37. Heng EL, Gatzoulis MA, Uebing A, Sethia B, Uemuera H, Smith GC, et al. Immediate and midterm cardiac remodelling after surgical pulmonary valve replacement in adults with repaired tetralogy of Fallot: a prospective cardiovascular magnetic resonance and clinical study. *Circulation*. 2017;136(18):1703-1713. https://doi.org/10.1161/CIRCULATIONAHA.117.027402 PMid:29084778 PMCid
38. Mongeon FP, Ben Ali W, Khairy P, Bouhout I, Therrien J, Wald RM, et al. Pulmonary valve replacement for pulmonary regurgitation in adults with tetralogy of Fallot: a meta-analysis - A report for the writing committee of the 2019 update of the Canadian Cardiovascular Society Guidelines for the management of adults with congenital heart disease. *Can J Cardiol*. 2019;35(12):1772-1783. https://doi.org/10.1016/j.cjca.2019.08.031 PMid:31813508
39. Lee C, Choi ES, Lee CH. Long-term outcomes of pulmonary valve replacement in patients with repaired tetralogy of Fallot. *Eur J Cardiothorac Surg*. 2020;58(2):246-252. https://doi.org/10.1093/ejcts/ezaa030 PMid:32047919
40. Rutz T, Ghandour F, Meierhofer C, Naumann S, Martinoff S, Lange R, et al. Evolution of right ventricular size over time after tetralogy of Fallot repair: a longitudinal cardiac magnetic resonance study. *Eur Heart J Cardiovasc Imaging*. 2017;18(3):364-370. https://doi.org/10.1093/ehjci/jew273 PMid:28363200
41. Luijnenburg SE, Helbing WA, Moelker A, Kroft LJM, Groenink M, Roos-Hesselink JW, et al. 5-year serial follow-up of clinical condition and ventricular function in patients after repair of tetralogy of Fallot. *Int J Cardiol*. 2013;169(6):439-444. https://doi.org/10.1016/j.ijcard.2013.10.013 PMid:24182670
42. Wald RM, Valente AM, Gauvreau K, Babu-Narayan SV, Assenza GE, Schreier J, et al. Cardiac magnetic resonance markers of progressive RV dilation and dysfunction after tetralogy of Fallot repair. *Heart*. 2015;101(21):1724-1730. https://doi.org/10.1136/heartjnl-2015-308014 PMid:26276804
43. Hoelscher M, Bonassin F, Oxenius A, Seifert B, Leonardi B, Kellenberger CJ, et al. Right ventricular dilatation in patients with pulmonary regurgitation after repair of tetralogy of Fallot: How fast does it progress? *Ann Pediatr Cardiol*. 2020;13(4):294-300. https://doi.org/10.4103/apc.APC_140_19 PMid:33311917 PMCid
44. Balasubramanian S, Harrild DM, Kerur B, Marcus E, Del Nido P, Geva T, et al. Impact of surgical pulmonary valve replacement on ventricular strain and synchrony in patients with repaired tetralogy of Fallot: a cardiovascular magnetic resonance feature tracking study. *J Cardiovasc Magn Reson*. 2018;20(1):37. https://doi.org/10.1186/s12968-018-0460-0 PMid:29909772 PMCid
45. Geva T, Mulder B, Gauvreau K, Babu-Narayan SV, Wald RM, Hickey K, et al. Preoperative predictors of death and sustained ventricular tachycardia after pulmonary valve replacement in patients with repaired tetralogy of Fallot enrolled in the INDICATOR cohort. *Circulation*. 2018;138(19):2106-2115. https://doi.org/10.1161/CIRCULATIONAHA.118.034740 PMid:30030416
46. Dorobantu DM, Sharabiani MTA, Taliotis D, Parry AJ, Tulloh RMR, Bentham JR, et al. Age over 35 years is associated with increased mortality after pulmonary valve replacement in repaired tetralogy of Fallot: results from the UK National Congenital Heart Disease Audit database. *Eur J Cardiothorac Surg*. 2020;58(4):825-831. https://doi.org/10.1093/ejcts/ezaa069 PMid:32187367 PMCid
47. Rozzi G, Lo Muzio FP, Fassina L, Rossi S, Statello R, Sandrini C, et al. Right ventricular functional recovery depends on the timing of pulmonary valve replacement in tetralogy of Fallot: a video kinematic study. *Eur J Cardiothorac Surg*. 2021;59(6):1329-1336. https://doi.org/10.1093/ejcts/ezab026 PMid:33547473
48. Dardari M, Cinteza E, Vasile CM, Padovani P, Vatasescu R. Infective endocarditis among pediatric patients with prosthetic valves and cardiac devices: A review and update of recent emerging diagnostic and management strategies. *J Clin Med*. 2023;12(15):4941. https://doi.org/10.3390/jcm12154941 PMid:37568344 PMCid
49. Demir F, Varan C, Erdem S, Atmis A, Akbas T, Subasi B, et al. Infective endocarditis in childhood: a single-centre experience of 26 years. *Cardiol Young*. 2023;33(10):1950-1955. https://doi.org/10.1017/S1047951122003419 PMid:36419327

About the Author (s)



Professor Amir-Reza Hosseinpour is a British (of Iranian roots) congenital cardiac surgeon. He did all his studies and speciality training in the United Kingdom before moving to Seville in southern Spain in September 2006 to take up the position of Chief of Congenital Cardiac Surgery at the Virgen del Rocio University Hospital. He moved to Lausanne, Switzerland, in August 2022 to take up the same position at the Lausanne University Hospital. He is also an Associate Professor of Cardiac Surgery at the University of Lausanne.

Professor Matthias Kirsch is a German cardiac surgeon. He did all his studies and speciality training in France (Paris) except for a fellowship that he did in Brussels. After having worked several years in Paris, he moved to Lausanne in September 2015 where he became the Chief of the Cardiac Surgery Service at the Lausanne University Hospital. He is also the Professor of Cardiac Surgery at the University of Lausanne.

Doctor Maria-Helena Perez is a Swiss (of Spanish roots) pediatric intensive care specialist. She did her studies and specialization in Lausanne and became a member of the pediatric ICU staff of the Lausanne University Hospital after her training. In 2018, she became the chief of that unit.

Doctor Stefano Di Bernardo is a Swiss (of Italian roots) pediatric interventional cardiologist. He did his studies in Lausanne and his specialization in both Lausanne and Zurich. He joined the pediatric cardiology unit of the Lausanne University Hospital after his training. He is the deputy chief of that unit and is the coordinator of the interventional cardiology section.