Serial Follow-Up of Two Surgical Strategies for the Repair of Tetralogy of Fallot

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Adverse events following tetralogy of Fallot repair include arrhythmia, reoperation, and death. While limiting the right ventriculotomy mitigates these events over the short and intermediate term, the impact over the long term is unknown. Children requiring tetralogy of Fallot repair were divided into 2 groups: extended right ventriculotomy vs limited right ventriculotomy with transatrial ventricular septal defect closure. Follow-up office ECGs and Holter monitoring, exercise stress tests, and echocardiograms were obtained in the first, second, and third postoperative decades. The primary outcome was cumulative events defined as reoperation, arrhythmia, or death. The extended and limited right ventriculotomy techniques were used in 21 and 17 children, respectively. Cumulative survival was 93.6% at 30 years and was not different between groups. While 10-year (42.9% vs 5.9%, $P = 0.02$) event rates favored the limited ventriculotomy technique, there were no significant differences at 20 or 25 years. Cox proportional hazard analysis demonstrated that the limited ventriculotomy technique was independently associated with lower events at 10 years (hazard ratio 0.03, 95% confidence interval 0.0016, 0.5697, $P = 0.01$). The limited ventriculotomy group had greater exercise capacity in the second decade but not the third. Right ventricular end-diastolic diameter Z score was not different at 10 or 20 years, but was significantly smaller in the limited ventriculotomy group at 30 years (5.55 ± 1.69 vs 4.14 ± 0.63, $P = 0.03$). Limiting the right ventriculotomy during tetralogy of Fallot repair limits 10-year events, improves exercise capacity at 20 years, and attenuates late right ventricular dilation.

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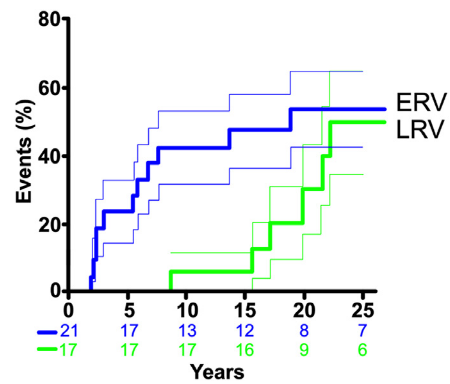
Keywords: Tetralogy of Fallot, Ventriculotomy, Congenital heart disease



Cumulative events in the extended and limited right ventriculotomy groups.

Central Message

Limiting the right ventriculotomy during tetralogy of Fallot repair limits events up to 10 years, improves exercise capacity at 20 years, and attenuates late right ventricular dilation.

Perspective Statement

The short- and intermediate-term results after a limited right ventriculotomy during tetralogy of Fallot repair have been promising. However, the long-term results are less understood. The current study reports the 10-, 20-, and 25-year outcomes following a limited ventriculotomy during tetralogy of Fallot repair.

INTRODUCTION

Avoidance of an extensive right ventriculotomy (ERV) has become a tenet of contemporary tetralogy of Fallot (TOF) repair[1,2] to prevent right ventricular (RV) dilation,[3] progressive RV dysfunction,[4] arrhythmias,[5] reoperation,[6] and sudden death.[5] Many surgeons currently prefer either a transatrial-transpulmonary approach[7] or transatrial ventricular septal defect (VSD)

Abbreviations: TOF, tetralogy of Fallot; ERV, extensive right ventriculotomy; LRV, limited right ventriculotomy; VSD, ventricular septal defect; RV, right ventricle; LV, left ventricle; RBBB, right bundle branch block; VO_2 max, maximal oxygen consumption; TTE, transthoracic echocardiogram; CMR, cardiac magnetic resonance imaging; RVEDD, right ventricular end-diastolic diameter; LVEDD, left ventricular end-diastolic diameter; LVEF, left ventricular ejection fraction

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closure with a limited right ventriculotomy (LRV) as dictated by the anatomy.[2] Although the short- and intermediate-term outcomes following the use of an LRV have demonstrated promising results,[6] the majority of patients will eventually require reoperation. Whether long-term outcomes beyond 30 years continue to favor a limited ventriculotomy approach is unknown.

In 1982 at SUNY Upstate Medical University, there was a change in the strategy for TOF repair, moving to an LRV approach for relief of RV outflow tract (RVOT) obstruction and transatrial VSD closure. Follow-up on the LRV approach has been reported twice, once in the short term[8] and again nearly a decade later in the intermediate term.[9] We sought to determine the long-term results following TOF repair to ascertain whether the LRV approach limited events such as reoperation, arrhythmia, and death into the third postoperative decade.

MATERIALS AND METHODS

Following institutional review board approval, data from children included in the original cohort who underwent TOF repair between January 1, 1976 and December 31, 1985 at SUNY Upstate Medical University were reviewed and divided into 2 groups based upon the type of repair: ERV vs LRV. Children who underwent pulmonary valve replacement at their initial repair and those lost to follow-up after less than 5 years were excluded. Serial follow-up was obtained and categorized into the first, second, and third postoperative decades. This was accomplished by recording results that were as close to 10, 20, and 30 years from the date of the initial TOF repair as possible.

Surgical Methods

Briefly, in the ERV group repaired between 1976 and 1981, an ERV was made from the infundibulum to the RV mid-body for VSD closure and generous infundibular muscle resection.[8,9] Outflow tract patching was performed as needed based on intraoperative assessment of RVOT obstruction. For the LRV group repaired 1982–1985, the VSD was closed through the tricuspid valve and a LRV (<2 cm) was made at the point of maximal obstruction as identified on preoperative catheterization. Fibrous tissue was excised and limited muscle excision was performed only if necessary to relieve obstruction. The ventriculotomy was closed with a patch to expand the RVOT anteriorly in all cases.[8,9] In both groups, a transannular patch was placed when the predicted peak postoperative RV:left ventricular pressure ratio based on preoperative catheterization or intraoperative Hegar sizing exceeded 0.75.[10]

Electrocardiography

Twelve-lead ECGs were performed at each instance of outpatient follow-up. Machine-calculated mean QRS duration was recorded where available; physician-calculated QRS duration was used otherwise. Presence of machine- or physician-detected complete right bundle branch block (RBBB) was defined as a QRS duration >120 ms and presence of an rsR' in the anterior precordium (V1–V3).[11] Twenty-four hour Holter recordings were taken according to clinical suspicion and monitoring of arrhythmias. Arrhythmias included supraventricular tachycardia (SVT) and ventricular ectopy defined as Lown grades 4A (couplets), 4B (salvos), and 5 (R on T phenomenon). High-grade ventricular ectopy and SVT were chosen owing to their frequent requirement of medical and/or procedural intervention and their association with sudden cardiac death.[12]

Exercise Stress Testing

Exercise stress testing was performed on a treadmill using a modified Bruce protocol, exercising patients to exhaustion.[13] Electrocardiographic monitoring was in place from before exercise through recovery. Maximal oxygen consumption (VO_2 max) was measured in standard fashion. Measurement of oxygen consumption during exercise testing was not available before the second postoperative decade.

Echocardiography

Transthoracic echocardiograms were performed and RV and LV end-diastolic diameters (RVEDD, LVEDD) were measured in M mode in the parasternal long axis view at the level of the atrioventricular valve leaflets. Ventricular end-diastolic diameter Z scores were calculated by normalizing values to body surface area.[14] Percent change in RVEDD and corresponding Z score were calculated to illustrate change over time, using the 10-year time point for comparison. Color Doppler tracings were used to determine the tricuspid insufficiency gradient.

CMR Studies

CMR studies were acquired in a 1.5T scanner (GE Healthcare, Milwaukee, Wisconsin) with a dedicated phased array cardiac coil and ECG gating. Ventricular volumes and ejection fractions were determined using steady state free precession to acquire short axis images from the base of the heart to the apex. Pulmonary valve regurgitant fraction was measured using phase-contrast flow mapping of the main pulmonary artery and ascending aorta. Images were analyzed offline using Segment software version 1.9 (Medviso AB, Lund, Sweden) with Z values calculated by normalizing measurements to patient age and body surface area[15] as previously described.[16]

Outcomes

The primary outcome was the cumulative event rate defined as reoperation, arrhythmia, or death. Secondary outcomes included exercise endurance time, VO_2 max during exercise stress tests, and RV dilation measured by transthoracic echocardiogram.

Statistical Analysis

All data are presented as mean ± standard deviation or median with interquartile range for continuous variables or as a frequency and percentage for noncontinuous variables. After ensuring the equality of variables using a Shapiro-Wilks test, univariate analysis was performed using a two-tailed *t* test or Mann-Whitney test where appropriate. One-way ANOVA was used to compare the whole cohort over time. Bonferroni correction for comparison of multiple time points was used where appropriate. Fisher's exact test or Chi-squared analysis was used for comparison of qualitative variables. Kaplan-Meier curves were generated to determine survival, freedom from reoperation, and incidence of cumulative events, and the log-ranks were compared. Despite longer follow-up, Kaplan-Meier curves were truncated at 25 years due to a limited number of subjects at risk at 30 years. A Cox proportional hazard model was used to determine whether the cumulative event differences at 10 years between the ERV and LRV groups were independent of preoperative patient characteristics. The age at operation, surgical era (1970s vs 1980s), gender, use of a preoperative aortopulmonary shunt, or use of a transannular patch were selected owing to their availability for comparison in nearly all patients. All statistics were completed using GraphPad Prism version 5.0b (GraphPad Software, San Diego, CA) or SAS 9.4 (SAS Institute Inc., Cary, NC) where a *P* value of less than 0.05 was considered statistically significant.

RESULTS

Demographics

From 42 children requiring TOF repair, 1 patient in the ERV group and 3 patients in the LRV group were excluded (Fig. 1). The ERV and LRV techniques were used in 21 and 17 children, respectively. Baseline demographics between the 2 groups were similar; however, children repaired using the LRV technique were younger at the time of their initial operation (Table 1). There were no significant differences in gender, rate of preoperative aortopulmonary shunting, or use of a transannular patch between groups (Table 1). With 100% follow-up of the cohort, cumulative follow-up obtained was 1071.2 patient-years at a median of 30.9 (19.5—35.8) years.

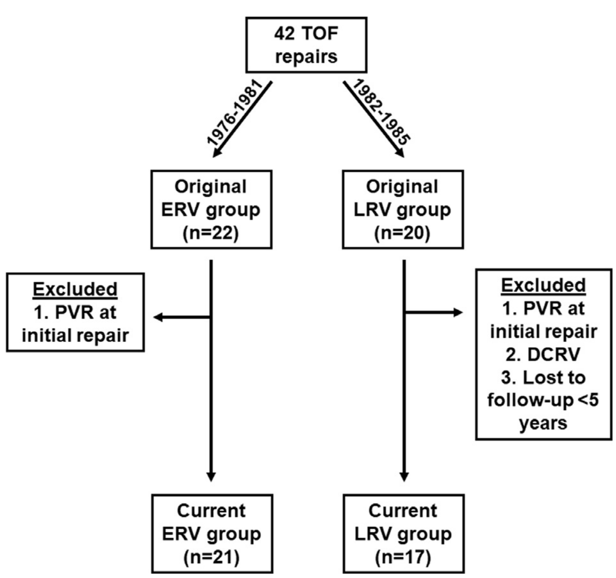


Figure 1. Schematic illustrating patient selection for the study. DCRV, dual chamber right ventricle; ERV, extended right ventriculotomy; LRV, limited right ventriculotomy; PVR, pulmonary valve replacement; TOF, tetralogy of Fallot.

Table 1. Perioperative Demographics

	ERV (*n* = 21)	LRV (*n* = 17)	*P* Value
Age at repair (y)	3.8 ± 1.0	2.7 ± 0.9	0.001*
Male gender	57.1% (12/21)	64.7% (11/17)	0.74
Previous aortopulmonary shunt	28.6% (6/21)	29.4% (5/17)	1.0
Transannular patch	85.0% (17/20)	58.8% (10/17)	0.13

ERV, extended right ventriculotomy; LRV, limited right ventriculotomy.
*denotes statistical significance

Survival and Reoperation

There were a total of 2 deaths in follow-up: 1 operative death in an ERV patient who was undergoing reoperation for RVOT patch revision 2.9 years after initial repair, and 1 LRV patient who died 25.9 years postoperatively following an unspecified illness. There were no operative deaths in the LRV group. Cumulative survival was 93.6% at 30 years and was not significantly different with the use of either the ERV or LRV technique. There were a total of 26 reoperations, and cumulative freedom from reoperation was 50.4% at 25 years. Twelve patients in the ERV group required 19 reoperations. These included 1 residual VSD closure, 3 RVOT expansions, 3 RVOT aneurysmorrhaphies, and 17 pulmonary valve replacements, 5 of which were in combination with the aforementioned RVOT reconstructions. Pulmonary valve replacements were performed between 2 and 31 years postoperatively, with 10/17 performed in the first postoperative decade. By comparison, 5 patients in the LRV group required 7 reoperations (1 RVOT expansion, 6 pulmonary valve replacements). Pulmonary valve replacements in LRV patients were performed between 16 and 27 years postoperatively.

While freedom from reoperation was significantly higher in the LRV group at 10 years (57.1% ERV vs 94.1% LRV, $P = 0.02$), there was no significant difference between groups at 20 years ($P = 0.45$) or 25 years ($P = 0.48$; Fig. 2). There was

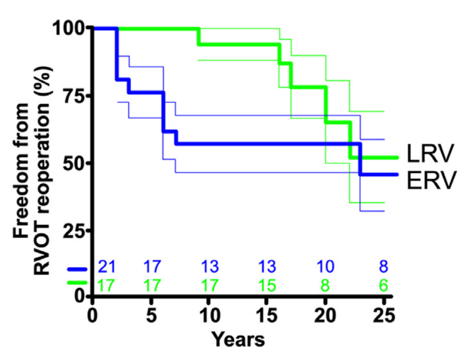


Figure 2. Kaplan-Meier curve with 95% confidence interval comparing freedom from reoperation following tetralogy of Fallot repair in the extended right ventriculotomy (ERV) and limited right ventriculotomy (LRV) groups. Freedom from reoperation is significantly higher up to 10 years but not afterward following repair with an LRV.

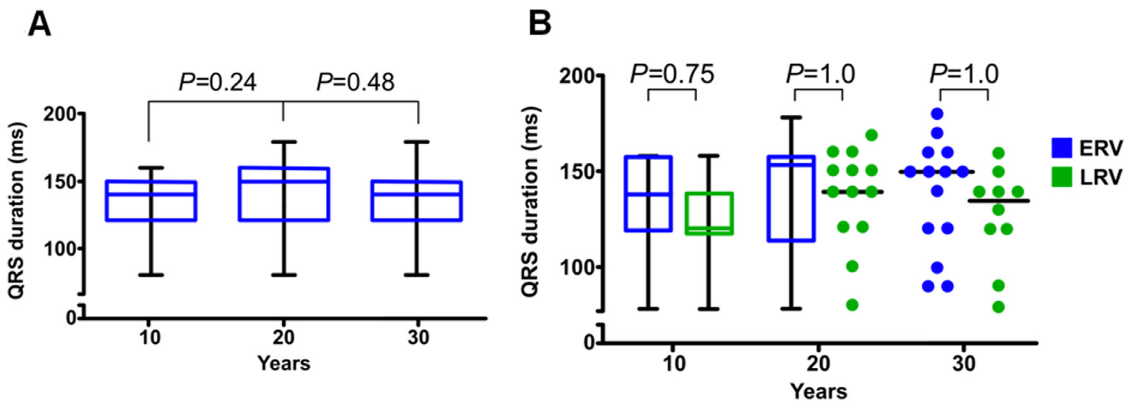


Figure 3. (A) Box and whisker plot of QRS duration for the entire cohort following tetralogy of Fallot repair. (B) Box and whisker plot of QRS duration following tetralogy of Fallot repair in the extended right ventriculotomy (ERV) and limited right ventriculotomy (LRV) groups. Tetralogy of Fallot repair results in QRS prolongation regardless of repair strategy. The median is the center colored bar while the upper and lower colored bars represent the upper and lower quartiles. The black bars represent the range, minimum to maximum. Where there were less than 15 data points, a scatter plot with a line for the median was used.

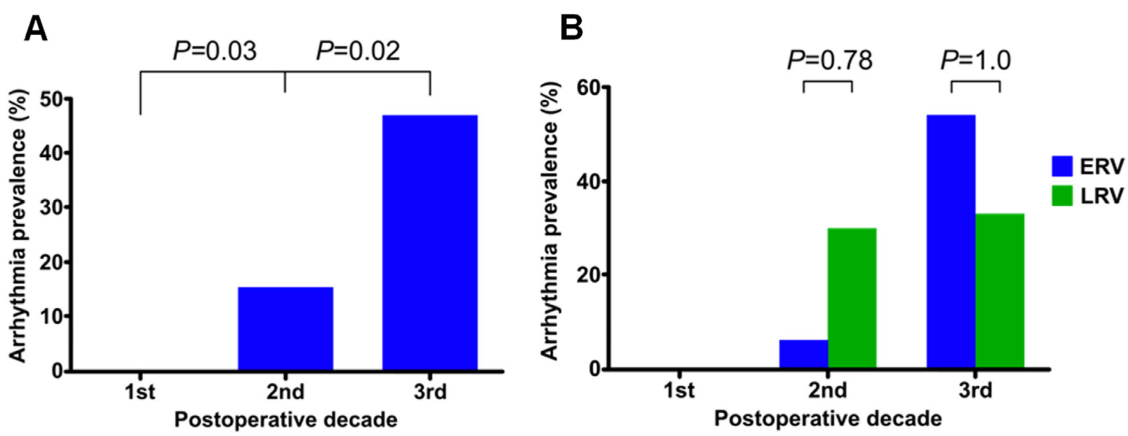


Figure 4. (A) Prevalence of ventricular arrhythmias or supraventricular tachycardia for the entire cohort following tetralogy of Fallot repair. (B) Prevalence of ventricular arrhythmias or supraventricular tachycardia following tetralogy of Fallot repair in the extended right ventriculotomy (ERV) and limited right ventriculotomy (LRV) groups. Tetralogy of Fallot repair results in a progressive increase in arrhythmias over time that is not related to the length of the ventriculotomy.

1 catheter-based reintervention for branch pulmonary artery stenosis in the LRV group and none in the ERV group, with no significant difference between groups ($P = 0.44$).

Electrocardiography and Arrhythmias

Twelve-lead ECGs were reviewed at 10.2 ± 0.9 years (ERV $n = 17$, LRV $n = 16$), 19.7 ± 2.1 years (ERV $n = 20$, LRV $n = 14$), and 29.8 ± 3.4 years (ERV $n = 15$, LRV $n = 11$) postoperatively with no difference in time elapsed since surgery between groups. QRS duration did not significantly change for the entire cohort over the follow-up period (Fig. 3A). Similarly, there were no significant differences in QRS duration between groups at any time point (Fig. 3B). RBBB was present in the majority of the cohort at 10 years (78.8% [26/33]) and did not increase significantly over time. From the entire cohort, there were 70 Holter monitor recordings available for review, 44 (2.15 ± 1.09 per patient) in the ERV group and 26 (1.53 ± 0.94 per patient) in the LRV group. These were reviewed at 4.4 ± 2.3 years (ERV $n = 17$, LRV $n = 10$), 13.4 ± 2.9 years (ERV $n = 16$, LRV $n = 10$), and 25.5 ± 4.99 years (ERV $n = 11$, LRV $n = 6$) postoperatively with no difference in time elapsed since surgery between groups. Unlike QRS duration, the prevalence of recorded arrhythmias in the entire cohort increased exponentially on serial follow-up (Fig. 4A). However, similar to QRS duration, there were no significant differences in the prevalence of arrhythmias between groups (Fig. 4B).

Cumulative Events

For the entire cohort, there were 38 events or 0.035 events per patient-year. There were 27 events in the ERV group and 11 events in the LRV group resulting in event rates of 0.040 vs 0.025 events per patient-year, respectively ($P = 0.17$). The majority of events were reoperations (26/38), and as a result, the cumulative event rate is similar to the reoperation rate (49.6% at 25 years) for the entire cohort. The 10-year (42.9% ERV vs 5.9% LRV, $P = 0.02$) cumulative event rate favored the

Table 2. Cox Proportional Hazards Model for 10-Year Cumulative Events

	Hazard Ratio	95% Confidence Interval	*P* Value
LRV approach	0.04	0.002, 0.947	0.04
Age (y)	0.594	0.265, 1.33	0.21
Male gender	0.262	0.005, 1.39	0.12
Previous aortopulmonary shunt	8.812	1.400, 55.474	0.02
Transannular patch	2.153	0.217, 21.361	0.51
1980s era (vs 1970s)	0.438	0.096, 1.991	0.29

LRV, limited right ventriculotomy.

LRV group. Comparison of the 10-year cumulative event rate in age-matched ERV and LRV cohorts ($n = 15$ each, Supplementary Table 11) also showed that events were significantly lower in the LRV group ($P = 0.02$; Supplementary Fig. 1). Additionally, a Cox proportional hazard analysis accounting for age at surgery, preoperative systemic to pulmonary artery shunt, gender, and use of a transannular patch demonstrated that only the LRV approach was independently associated with a lower event rate compared to the ERV group (Table 2). The multivariate analysis also demonstrated that preoperative aortopulmonary shunting was a significant predictor of 10-year events (Table 2). Rates of preoperative shunting were not different between groups (Table 1) or between surgical eras (3/12 1970s vs 8/26 1980s, $P = 1.0$). The 20-year LRV event rate (54.2% ERV vs 21.8% LRV, $P = 0.06$) was numerically but not significantly less than the ERV approach, with further convergence in event rates by 25 years (Fig. 5).

Cardiac Fitness and Imaging

Exercise stress tests were reviewed at 5.5 ± 1.9 years (ERV $n = 10$, LRV $n = 8$), 13.2 ± 2.5 years (ERV $n = 15$, LRV $n = 14$), and 20.7 ± 3.5 years (ERV $n = 9$, LRV $n = 5$) postoperatively

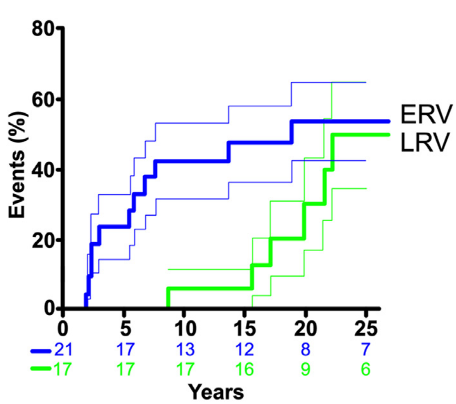


Figure 5. Kaplan-Meier curve with 95% confidence interval depicting 25-year cumulative events following tetralogy of Fallot repair in the extended right ventriculotomy (ERV) and limited right ventriculotomy (LRV) groups. The LRV approach limited cumulative events up to 10 years with convergence in event rates afterward.

with no difference in time elapsed since surgery between groups. For the entire cohort, exercise endurance time remained nearly static over the follow-up period (Fig. 6A). Exercise endurance time was not significantly different between groups in the first postoperative decade (Fig. 6B), after which measurement of maximal oxygen consumption became available to assess cardiorespiratory fitness. For the entire cohort, VO_2 max was significantly higher in the second postoperative decade than the third (Fig. 6C). In addition, VO_2 max was greater in the LRV group in the second postoperative decade (Fig. 6D), but this difference did not persist in the third postoperative decade.

Echocardiographic data including times elapsed since surgery and numbers of echocardiograms available for review at each time point are shown in Table 3. From the entire cohort, there were no significant differences in left ventricular chamber size or systolic function at any time. While there were no significant differences in the RV end-diastolic diameter and corresponding Z score between groups at 10 and 20 years, the LRV group had a significantly smaller RV end-diastolic diameter Z score at 30 years (Table 3). CMR data were available for only 4 patients in each group, performed between 24 and 40 years postoperatively. There were no significant differences in RV end-diastolic volume (254.4 ± 84.9 mL ERV vs 193.13 ± 21.2 mL LRV, $P = 0.21$) or corresponding index (131.0 ± 51.0 mL/m^2 ERV vs 107.0 ± 16.1 mL/m^2 LRV, $P = 0.42$). Pre- and postpulmonary valve replacement comparisons of echocardiographic and exercise parameters were also made in an effort to determine whether there is any difference in benefit from pulmonary valve replacement between groups (Supplementary Tables 2—5).

DISCUSSION

In a cohort of 38 patients with serial follow-up after TOF repair, limiting the right ventriculotomy was associated with improved outcomes in each decade following repair. The LRV approach was associated with decreased cumulative events in the first decade and improved cardiorespiratory fitness in the second decade. During the third decade after repair, the LRV approach limited RV dilation.

The LRV group had significantly fewer cumulative events at 10 years followed by a convergence at 25 years driven by an increasing number of pulmonary valve replacements in both groups. While earlier repair[17] has been previously associated with lower event rates, when accounting for age of operation in our series, only the LRV approach was protective from cumulative events out to 10 years. Though preoperative aortopulmonary shunting was a significant predictor of events in the current study, the rate of shunting was not significantly different between groups or surgical eras. Therefore, it is less likely to explain the difference in 10-year event rates for ERV vs LRV patients. The overall increase in events in our population over time is similar to other long-term reports on repaired TOF.[1,5] Although most patients repaired using an LRV will likely require reoperation, longer ventricular incisions appear

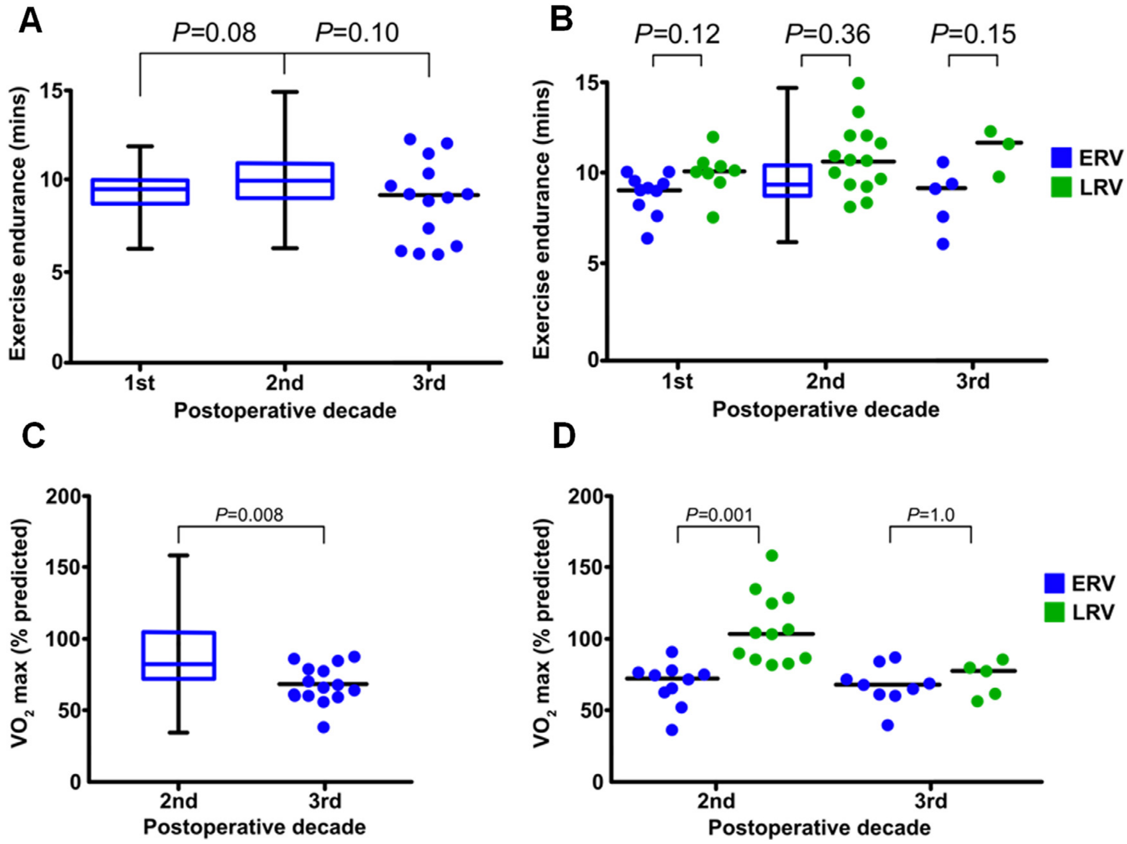


Figure 6. (A) Box and whisker plot of exercise endurance times for the entire cohort following tetralogy of Fallot repair. (B) Box and whisker plot of exercise endurance times following tetralogy of Fallot repair in the extended right ventriculotomy (ERV) and limited right ventriculotomy (LRV) groups. (C) Box and whisker plot of maximal oxygen consumption (VO_2 max) for the entire cohort following tetralogy of Fallot repair. (D) Box and whisker plot of VO_2 max following tetralogy of Fallot repair in the ERV and LRV groups. Limiting the right ventriculotomy in tetralogy of Fallot repair improves exercise capacity into the second postoperative decade but not afterward. The median is the center colored bar while the upper and lower colored bars represent the upper and lower quartiles. The black bars represent the range, minimum to maximum. Where there were less than 15 data points, a scatter plot with a line for the median was used.

to significantly shorten the reoperation-free interval. This informs our current practice in TOF repair, which is to avoid ventriculotomy whenever possible and make a <1 cm ventriculotomy only when adequate relief of RVOT obstruction (RVOT and pulmonary valve Z scores > 0) cannot be achieved through the tricuspid valve.[2] Like many other groups,[17] we prefer to repair TOF in infancy.

The use of an ERV has been associated with RV dysfunction,[7] akinesis,[7] and RV aneurysms.[18] Accordingly, 3 of the early reoperations in our ERV cohort included RV aneurysmorrhaphy, which partially accounts for the early divergence in freedom from reoperation. Despite the early advantage in freedom from reoperation in the LRV group, there was no difference at 20 or 25 years. This may be related to the frequent need for reoperation following transannular patching during TOF repair. Recent literature suggests that the use of a transannular patch is associated with a higher rate of late reoperation, without a significant decrease in survival.[19] In our cohort, nearly three-quarters of patients received a transannular patch, which may explain the high rate of reoperation compared to other studies.[3,5]

Arrhythmias following TOF repair have been shown to predict mortality.[5] The overall prevalence of arrhythmias in the current study was similar to other reports at 47.1% in the third postoperative decade.[5,20] However, only 12.5% of the arrhythmias were SVT compared to reported rates of 72.5%[5] and 67.1%[20] at similar time points. Historically, there has been a close association of arrhythmias with repaired TOF.[5] The lack of difference in QRS duration, RBBB, and arrhythmia prevalence between groups in this study was unexpected. The development of RBBB, progressive QRS prolongation, and arrhythmias after TOF repair is multifactorial. As the majority of patients in the study underwent transannular patch placement, a known risk factor for late arrhythmias,[5] we speculate that this explains the lack of difference in arrhythmia prevalence between groups.

The national trend in TOF repair has been toward earlier reoperation for pulmonary valve replacement.[21,22] It is interesting that the early difference in RV size between groups demonstrated in a previous review of this cohort[8] was no longer apparent at 10 or 20 years after many patients in the ERV group had undergone their first pulmonary valve

Table 3. Echocardiographic Follow-Up Data

TTE 10 Years	ERV ($n = 16$)	LRV ($n = 14$)	P Value
Time elapsed since operation (y)	10.9 ± 1.8	10.5 ± 1.8	1.0
10 y LVEDD (cm)	4.31 ± 0.36	4.11 ± 0.50	0.72
10 y LVEDD Z	−0.40 ± 0.75	−0.39 ± 1.19	1.0
10 y LVEF (%)	60.3 ± 15.3	64.7 ± 5.0	1.0
10 y RVEDD (cm)	3.17 ± 0.56	2.66 ± 0.67	0.12
10 y RVEDD Z	6.25 ± 2.46	5.19 ± 2.69	0.93
10 y TI >mild	7.1% (1/14)	0	1.0
10 y TI jet pressure (mm Hg)	34.6 ± 11.8	28.6 ± 9.2	0.72
10 y RVOT gradient (mm Hg)	25.7 ± 13.7	30.8 ± 17.6	1.0
10 y > moderate PI	40.0% (6/15)	46.2% (6/13)	1.0
TTE 20 Years	**ERV ($n = 17$)**	**LRV ($n = 11$)**	**P Value**
Time elapsed since operation (y)	20.8 ± 2.4	19.4 ± 2.7	0.66
20 y LVEDD (cm)	4.57 ± 0.54	4.77 ± 0.56	1.0
20 y LVEDD Z	−0.36 ± 1.34	−0.03 ± 1.40	1.0
20 y LVEF (%)	62.7 ± 7.8	64.6 ± 6.9	1.0
20 y RVEDD (cm)	3.35 ± 0.53	3.06 ± 0.61	1.0
Change from 10 year (%)	+5.7	+15.0	
20 y RVEDD Z	5.82 ± 1.93	4.75 ± 2.30	1.0
Change from 10 year (%)	−15.5	−8.5	
20 y TI > mild	6.7% (1/15)	10.0% (1/10)	1.0
20 y TI jet pressure (mm Hg)	39.6 ± 11.5	27.7 ± 8.9	0.06
20 y RVOT gradient (mm Hg)	30.9 ± 14.9	25.1 ± 10.5	1.0
20 y >moderate PI	41.2% (7/17)	45.5% (5/11)	1.0
TTE 30 Years	**ERV ($n = 14$)**	**LRV ($n = 8$)**	**P Value**
Time elapsed since operation (y)	28.6 ± 2.8	28.3 ± 3.5	1.0
30 y LVEDD (cm)	4.93 ± 0.60	5.09 ± 0.60	1.0
30 y LVEDD Z	0.48 ± 1.80	0.60 ± 1.58	1.0
30 y LVEF (%)	66.1 ± 9.9	64.9 ± 9.5	1.0
30 y RVEDD (cm)	3.25 ± 0.42	2.93 ± 0.24	0.12
Change from 10 year (%)	+2.5	+11.3	
30 y RVEDD Z	5.55 ± 1.69	4.14 ± 0.63	0.03*
Change from 10 year (%)	−11.2	−20.2	
30 y TI >mild	14.3% (2/14)	0	1.0
30 y TI jet pressure (mm Hg)	36.3 ± 17.5	24.0 ± 10.2	0.33
30 y RVOT gradient (mm Hg)	25.0 ± 14.7	17.5 ± 12.0	0.72
30 y >moderate PI	28.6% (4/14)	25.0% (2/8)	1.0

ERV, extended right ventriculotomy; LRV, limited right ventriculotomy; LVEDD, left ventricular end-diastolic diameter; LVEF, left ventricular ejection fraction; PI, pulmonary insufficiency; RVEDD, right ventricular end-diastolic diameter; RVOT, right ventricular outflow tract; TI, tricuspid insufficiency; TTE, transthoracic echocardiogram.
Incomplete echo data at 30 years for 16 patients are attributable to the following: mortality,[2] lost to follow-up at 21.8 ± 8.6 years postoperatively,[11] no echo performed at most recent follow-up[3]
*denotes statistical significance

replacement. However, after several LRV patients required pulmonary valve replacement in the second decade, RV size was once again significantly smaller in the LRV group at 30 years. This adds to existing evidence that an ERV is a risk factor for late RV dilation,[3] which has also been demonstrated by CMR data in long-term follow-up.[23] Ongoing investigation into the long-term effects of right ventriculotomy will be important to advancing our understanding of repaired TOF, particularly as CMR studies become available in 30-year follow-up.

LIMITATIONS

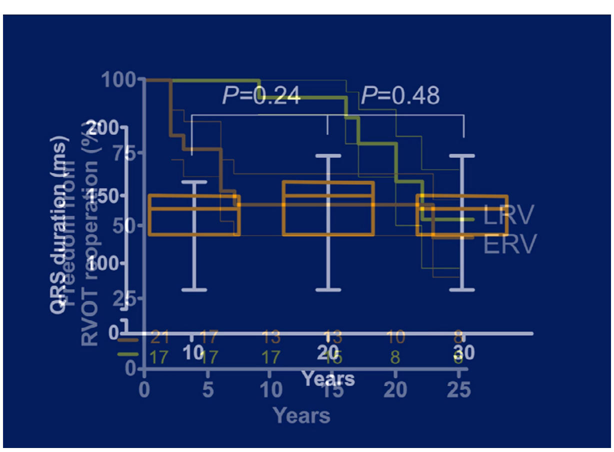
This study has all of the limitations expected of a single-center retrospective analysis. Restriction of the groups to the originally studied cohort made for a relatively small sample size. In addition, all repairs were performed using the technology and clinical practice of 2 close but noncontemporaneous periods, which may have influenced the results. Limitations of echocardiography in the studied era and performance of relatively few preoperative catheterizations precluded the use of preoperative anatomic factors like branch pulmonary artery size or pulmonary annular dimensions in the multivariate analysis. Similarly, quantification of pulmonary insufficiency was available in too few patients before their first reoperation to make a meaningful estimate of freedom from pulmonary insufficiency. Additionally, echocardiographic data were not completely obtainable in all patients at the most recent follow-up, and the available data did not allow us to determine the missing data structure. CMR was available in only 8 patients and would offer greater accuracy in analyses of RV size and function over time. While similar time points for Holter monitoring and exercise stress tests were compared between groups, they were performed less regularly and are not the same time points as were compared for 12-lead ECGs and echocardiograms. Given the advances in medical care and diagnostics over the study period (particularly echocardiography and CMR imaging) and subsequent variability in the timing of pulmonary valve replacement, cumulative events were used as the primary outcome in an effort to capture the earliest point of TOF repair failure. Even so, Holter and ECG monitoring are not 100% sensitive in diagnosing arrhythmias, and the cumulative event rate may still be underestimated as a result.

CONCLUSIONS

In a cohort of 38 patients with serial follow-up after TOF repair, limiting the right ventriculotomy was associated with improved early and late outcomes. Use of an LRV conferred a 10-year advantage in freedom from reoperation and limited cumulative events up to 10 years postoperatively. The LRV approach was also associated with better cardiorespiratory fitness in the second postoperative decade. Limiting the right ventriculotomy did not change the prevalence of arrhythmias in late follow-up, but did minimize RV dilation at 30 years. When a ventriculotomy is necessary during TOF repair, it should be as limited as possible to minimize events, improve cardiorespiratory fitness, and attenuate late RV dilation.

SUPPLEMENTARY MATERIAL

The following is the supplementary data to this article:



Video 1. In the associated video, the authors explain the importance of the current study and summarize the results.

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