Surgical Repair of Tetralogy of Fallot With Absent Pulmonary Valve: Favorable Long-Term Results

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Tetralogy of Fallot with absent pulmonary valve syndrome (ToF-APVS) is a rare variant of tetralogy, associated with severe pulmonary valve regurgitation and aneurysmal dilation of the pulmonary arteries (PAs). Reported outcomes after surgical corrections are limited to single center, older series and might not reflect the current outcome. We aim to use data from a national registry to evaluate short- and long-term outcomes after surgical repair of ToF-APVS, to serve for counselling and planning. All children undergoing ToF-APVS repair in the UK between 2002 and 2013 were included. Survival and freedom from reintervention were estimated using the Kaplan-Meier method, and univariable analysis was done using the Weibull regression model. A total of 98 children, 45% male, 10% with DiGeorge syndrome, median age of 213 days (1 day to 13 years) were included. Mortality at 30 days was 3.3%, higher for neonates (6.7% vs 2.7%, $P = 0.4$) and those on preoperative mechanical ventilatory support (16.7% vs 1.3%, $P = 0.04$). Survival was 92.1% and freedom from pulmonary valve or conduit replacement (PVR) 73.2% at 10 years. Neonates had worse survival (hazard ratio [HR] 6.2, $P = 0.02$), freedom from PVR (HR 4.5, $P = 0.01$), freedom from PAs arterioplasty (HR 6.6, $P = 0.001$), and overall freedom from any reintervention (HR 5.3, $P < 0.001$). Low weight at repair was associated with worse freedom from PVR ($P = 0.02$) and from PAs arterioplasty ($P = 0.009$), preoperative ventilatory support with increased mortality ($P = 0.009$), the presence of DiGeorge syndrome was associated with worse freedom from PVR (HR 4, $P = 0.02$). Surgical repair of ToF-APVS can be performed with low early and late mortality, with improving results even in those with preoperative mechanical ventilatory support. The need for right ventricular outflow tract reintervention, including on the PAs, is an expected issue in the long term for the majority of patients.

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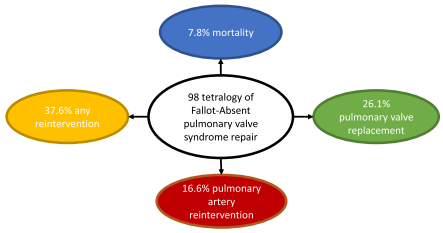
Keywords: Tetralogy of Fallot absent valve, Pulmonary valve replacement, Pulmonary arterioplasty, 22q11.2 deletion



Outcomes at 10 years after tetralogy of Fallot absent pulmonary valve syndrome repair.

Central Message

Mortality after correction of tetralogy of Fallot-Absent pulmonary valve syndrome is low. The need for right ventricular outflow tract and pulmonary artery reinterventions remains a long-term issue.

Perspective Statement

Early and late mortality after complete repair of tetralogy of Fallot with absent pulmonary valve is the single digits even at 10 years, having improved from earlier series. The need for reintervention is frequent and should be taken into consideration during follow-up, and especially when transitioning to adult care. Patient counseling and planning can benefit from results from this multicenter, national audit.

Abbreviations: BT, Blalock-Taussig; HR, hazard ratio; IQR, interquartile range; MAPCA, major aortopulmonary collateral; NCHDA, UK National Congenital Heart Disease Audit; NICOR, National Institute for Cardiovascular Outcomes Research; PAR, pulmonary arteries reinterventions; PAs, pulmonary arteries; PDA, persistent ductus arteriosus; PVR, pulmonary valve or conduit replacement; RVOT, right ventricular outflow tract; ToF-APVS, tetralogy of Fallot with absent pulmonary valve syndrome

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INTRODUCTION

Tetralogy of Fallot with absent pulmonary valve syndrome (ToF-APVS) is a rare variant, associated with severe pulmonary valve regurgitation and aneurysmal dilation of the pulmonary arteries (PAs).

Currently reported outcomes show significant long-term mortality, 8–22% at 10 years, but are limited to single-center or older series and might not reflect contemporary results.[1–4] We aim to use data from the UK National Congenital Heart Disease Audit (NCHDA) to evaluate outcomes after surgical repair of ToF-APVS to assist with counselling and planning for intervention.

METHODS

The National Institute for Cardiovascular Outcomes Research (NICOR) collects key validated data on cardiac procedures from all UK units through the NCHDA, detailed previously.[5] All patients under 16 years with mostly isolated ToF-APVS and repair performed between 2000 and 2013 were identified. Statistical analysis of long-term outcomes included Kaplan-Meier estimates and univariable Cox regression (more details on patients' selection and statistical methodology in Supplemental Material).

RESULTS

A total of 98 consecutive children undergoing ToF-APVS repair were included. Table 1 shows demographic, clinical, and procedural characteristics.

Mortality at 30 days was 3.3%, higher for neonates (6.7% vs 2.7%, $P = 0.4$) and those on preoperative mechanical ventilatory support (16.7% vs 1.3%, $P = 0.04$).

Table 1. Patient Demographic, Clinical, and Procedural Characteristics of 98 Children With ToF-APVS Undergoing Surgical Repair

Patient	
Age (d, median IQR)	213 (67–475)
Age group (*n*, %)	
Neonate	15 (15)
Infant	54 (55)
Children	29 (30)
Weight (kg, median IQR)	6.2 (3.8–8.9)
Male (*n*, %)	44 (45)
22q11.2 deletion (*n*, %)	10 (10)
Procedure	
Previous intervention	
BT shunt	3 (3)
MAPCA occlusion	3 (3)
PDA closure	2 (2)
Preoperative mechanical ventilator support	13 (13.3)
Bypass time	123 (97–159)
Cross-clamp time	72 (54–99)
30-day mortality	3 (3.4)

BT, Blalock-Taussig; IQR, interquartile range; MAPCA, major aortopulmonary collateral; PDA, persistent ductus arteriosus; ToF-APVS, tetralogy of Fallot-absent pulmonary valve syndrome.

Figure 1A shows survival, freedom from pulmonary valve or conduit replacement (PVR), PAs surgical/transcatheter reintervention (PAR) and overall PVR/PAR at 10 years. The following were predictors of late mortality: neonatal repair (hazard ratio [HR] = 6.2, $P = 0.02$), preoperative mechanical ventilatory support (HR = 11, $P = 0.009$). Specifically, mortality at 10 years was 21.2% in neonates and 23.8% for those on mechanical ventilatory support. Late PVR was associated with: neonatal repair (HR = 4.5, $P = 0.01$), low weight (HR 0.7/kg, $P = 0.02$),

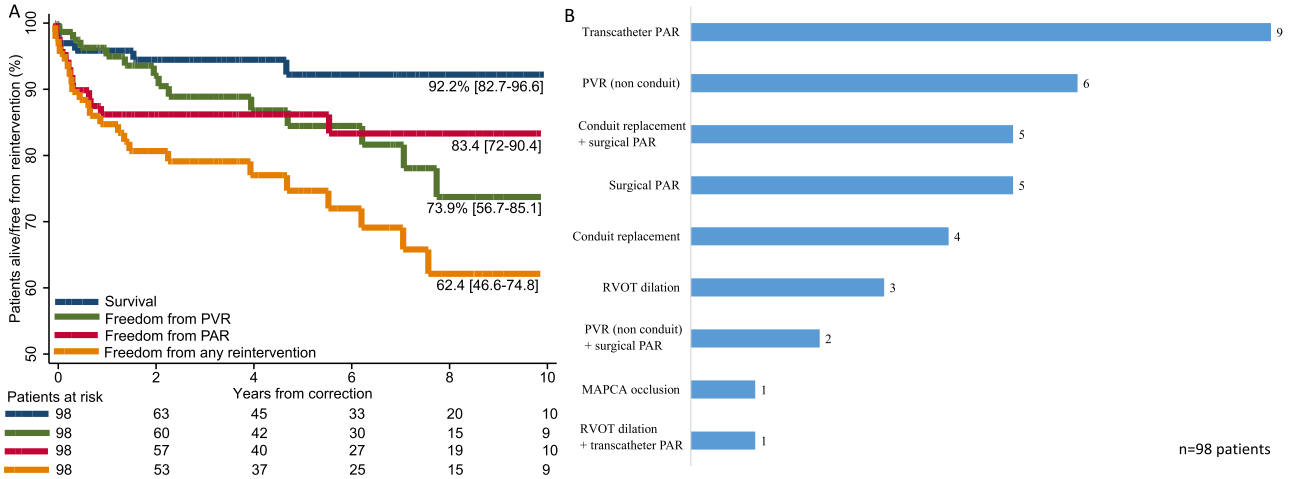

Figure 1. (A) Outcomes after initial repair in 98 children with tetralogy of Fallot absent valve syndrome. The curves represent unadjusted Kaplan-Meier survival functions for each of the 4 main outcomes. Values below the curves represent survival/freedom from reintervention at 10 years and 95% confidence intervals in brackets. (B) Types or reintervention performed and their prevalence. Numbers to the right of the bars represent total procedures of the respective type observed during follow-up. Nine percent of patients underwent more than 1 reintervention. PAR, pulmonary arteries reintervention; PVR, pulmonary valve replacement; RVOT, right ventricular outflow tract; MAPCA, major aortopulmonary collateral arteries.

and 22q11.2 deletion (HR = 4, $P = 0.02$). Late PAR was associated with: neonatal repair (HR = 6.6, $P = 0.001$), mechanical ventilatory support before repair (HR = 4.2, $P = 0.02$), low weight (HR = 0.74/kg, $P = 0.009$), and low age (HR = 0.18/y, $P = 0.03$). Mortality was higher in those undergoing a reintervention, but the differences were not statistically significant (HR = 2.8, $P = 0.2$).

Figure 1B shows the types of reinterventions performed, PA dilation or surgical repair, PVR/conduit replacement or a combination of the 2 being most prevalent. Out of the 9 PAs dilations, 3 included a stent placement. Of the 8 nonconduit PVRs, 5 were with a homograft prosthesis.

DISCUSSIONS

This is the largest contemporary and multicenter cohort of ToF-APVS patients undergoing surgical repair published to date, reporting outcomes of 98 consecutive patients. The late mortality was low at 8%, but the reintervention rate by the age of 10 was high at 38% (Central Figure). The early and late mortality were higher in neonates and those in critical respiratory distress.

Previously reported early mortality is dependent on operation era. Yong et al showed a drastic reduction from 18% before 2000 to 0% afterward.[1] Nørgaard et al report a figure of 5% before 2001,[2] Hew et al 15% before 1998,[4] while a cohort of 72 children operated after 2005 had a 5.5% early mortality.[3] The UK early mortality of 3% compares favorably to previous reports.

We observed a significantly lower long-term mortality, 8% at 10 years, compared to about 20% in older studies,[1,2,4] but similar to the more recent cohort from India,[3] possibly due to better operative techniques, intensive care, and possibly patient selection during the last decade. Of note are the improving results in those with preoperative ventilatory mechanical ventilation, which in our study have an early mortality of 16.7% and a late mortality of 21.2%, compared to previously reported 25−50% and 44−45%, respectively.[2,4]

On the other hand, in our group, there was a high reintervention rate, of nearly 40% at 10 years, higher than in other reported series (11−45%).[1−4] This may reflect some differences in follow-up and possibly lower thresholds for reintervention.

Compared to our previous report of simple ToF repair in infants from the same NCHDA database, we describe higher late mortality (8% vs 4.6%), PVR rate (27% vs 10%), PAR rate (17% vs 8%), and overall reintervention rate (40% vs 23%) in the current cohort.[5] This shows that even though outcomes are improving for the subgroup of ToF-APVS, they are still worse than in patients with simple ToF.

LIMITATIONS

This is a retrospective study based on a cardiac audit. We did not have data on the clinical presentation, imaging, degree of bronchial obstruction, or thoracic procedures. Additionally, there were no data on the type and size of conduits and valves used.

CONCLUSIONS

Surgical repair of ToF-APVS can be performed with low early and late mortality, with improving results even in those with preoperative mechanical ventilatory support. The need for right ventricular outflow tract reintervention, including on the PAs, is an expected issue in the long term for the majority of patients.

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