Cardiology in the Young

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Stenting of the right ventricular outflow tract as an initial intervention in Tetralogy of Fallot with pulmonary stenosis and major aortopulmonary collateral arteries

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Abstract

Objectives: To assess the role of right ventricular outflow tract stenting as the primary intervention in Tetralogy of Fallot with pulmonary stenosis and major aortopulmonary collateral arteries. *Background:* The management of a subset of infants with Tetralogy of Fallot with pulmonary stenosis and major aortopulmonary collateral arteries requires a staged approach including rehabilitation of diminutive native pulmonary arteries, conventionally using an aortopulmonary shunt. We share our experience of pulmonary artery rehabilitation with right ventricular outflow tract stenting. *Methods:* Retrospective review of all patients with Tetralogy of Fallot with pulmonary stenosis who underwent right ventricular outflow tract stenting as primary intervention over an 8-year period. *Results:* Ten patients underwent right ventricular outflow tract stent insertion at a median age of 61 days (interquartile range (IQR) 8.3–155 days). Median weight at stent deployment was 4.2 kg (IQR 3.2–5.7 kg). Oxygen saturations improved from a median of 79% (IQR 76–80%) to 92% (IQR 90–95%), $p = 0.012$. The median right and left pulmonary artery z score increased from −3.51 (IQR −4.59 to −2.80) and −2.07 (IQR −3.72 to 0.15) to a median of −1.17 (IQR −2.26 to 0.16) $p < 0.05$, and 0.24 (IQR −1.09 to 1.84) $p < 0.05$, respectively, at subsequent angiogram. Nine patients underwent further catheterisation. Four patients underwent complete anatomical repair. Only one patient required unifocalisation, as most patients had a native supply to all-important lung segments. *Conclusion:* Right ventricular outflow tract stenting is a useful procedure in the subset of patients with Tetralogy of Fallot with pulmonary stenosis and major aortopulmonary collateral arteries, where native pulmonary arterial growth is required to facilitate repair.

Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries is a rare and heterogeneous form of CHD.[1] The development and arborisation of the hypoplastic native confluent pulmonary arteries varies, as does the severity of pulmonary stenosis. Furthermore, the number and nature of lung segments supplied by major aortopulmonary collateral arteries vary extensively. These patients often present early with desaturation and require augmentation of pulmonary blood supply. In the presence of significant native pulmonary artery hypoplasia, early repair carries a significant risk of morbidity and mortality.[2,3]

Although Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries and pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries share many anatomic and physiological features, few studies focus on the management of this rare entity in isolation.[4] The best results in the management of pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries were demonstrated with a systematic approach of early unifocalisation of pulmonary blood supply in 78% of patients, and single-stage complete repair in 85% of these.[5,6] Only 22% of patients in this series with diminutive native pulmonary arteries to all lung segments and/or poor quality major aortopulmonary collateral arteries, therefore, were managed by an initial procedure to encourage growth of the native vessels. Other institutions have adopted an approach focussed on the rehabilitation of the native pulmonary arteries.[7] Lower rates of complete repair, and importantly, with resultant low right ventricular pressures have been shown with this approach. Importantly, and in contrast to cases with pulmonary atresia, the vast majority of patients with Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries will have a native pulmonary blood supply to all or virtually all segments of the lung. An approach of pulmonary artery rehabilitation may therefore be effective, and not require recruitment of major aortopulmonary collateral arteries in many patients. The critical determinant of outcomes with this approach, however, is maximising growth of the native pulmonary arteries, while avoiding distortion and growth restriction. Surgical shunts created to augment pulmonary

blood flow have a known incidence of pulmonary artery distortion,[8] and augmentation of the native right ventricular outflow tract blood flow has the potential for avoiding this.

Right ventricular outflow tract stenting is a widely accepted interventional method to increase antegrade pulmonary blood flow, and demonstrates superior pulmonary artery growth compared to arterial shunting in simple Tetralogy of Fallot.[9] It is at present unknown whether the same benefits could be achieved in patients with Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries, and whether this leads to improved outcomes. The purpose of this study was to evaluate our experience of right ventricular outflow tract stenting in patients with Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries as the primary staging procedure before complete repair.

Methods

We performed a retrospective review of consecutive Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries patients over an 8-year period (2011–2019). Follow-up was for a median of 3 years (IQR 1.1–7.6 years) after initial palliation. Ten patients underwent right ventricular outflow tract stenting as their primary intervention.

Patient demographics (including non-cardiac co-morbidities), procedure-related morbidity and complications, mortality, hospital stay, number and type of reintervention, and time until surgical intervention were collected. Angiographic measurements of the relevant anatomical and procedural information were collected. Vessel growth was assessed using both z scores and Nakata index at each subsequent angiogram.[10,11]

The decision for initial palliation was taken based on clinical presentation and feasibility of complete or staged surgical repair. Our institutional practice is for an early comprehensive definition of all pulmonary blood supply and pressure data through cardiac catheterisation prior to any surgical intervention. Low saturation at birth, indicating a prostaglandin-dependent pulmonary circulation or progressively decreasing saturations (<80%) were the primary indication for intervention. Infants felt to have inadequate forward flow across the pulmonary valve and insufficient major aortopulmonary collateral artery supply complementing native pulmonary blood supply, yet with adverse risk factors for complete repair were also palliated.

Within our institution, right ventricular outflow tract stenting is the first-choice palliation in this instance; those deemed unsuitable are palliated with either a right ventricle to pulmonary artery conduit or patent ductus arteriosus stent. Further support for right ventricular outflow tract stenting was provided by risk factors for surgical shunt palliation such as low weight, diminutive native pulmonary arteries, or severe co-morbidities (i.e., gastrointestinal, respiratory, neurological). The decision to occlude major aortopulmonary collateral arteries, either at the time of right ventricular outflow tract stenting or later, was based on clinical symptoms of increased pulmonary flow, dual supply, and competitive flow-limiting native pulmonary artery growth and feasibility of closure. The experience of our centre is that following post-right ventricular outflow tract stenting, patients require close clinical review and the duration of palliation can be extended with elective reintervention to increase the diameter of the right ventricular outflow tract stent in order to further improve saturations.

An outline of our centres right ventricular outflow tract stent procedure has been published previously.[12] In smaller children in whom a shorter term palliation was anticipated, coronary stents (Liberté Bare-Metal, Boston Scientific, Natick, Massachusetts, United States of America) were used. For those expected to require medium to long-term palliation, bare-metal Formula 418 stents (Cook Incorporated, Bloomington, Indiana, United States of America) were used.

Statistics

Continuous variables were analysed as medians and interquartile ranges (IQR). Categorical data is presented as counts and percentages. Comparison of pre-stent and post-stent parameters was performed using the Wilcoxon signed-rank test (IBM SPSS Statistics version 1.0.0–3906 SSPS Inc., Chicago, Ill, United States of America).

Ethics

This retrospective study was registered with our Institutional Research and Development Office, in accordance with the United Kingdom NHS National Research Ethics Service guidance. Individual informed consent nor formal Research Ethics Committee review was not required, because the study was undertaken by the direct clinical team using information previously collected in the course of routine care.

Results

Ten patients underwent right ventricular outflow tract stent insertion over a period of 8 years (January, 2011–July, 2019). Patient demographic information at the time of initial intervention is shown in Table 1. Seven patients were diagnosed antenatally. Two patients underwent major aortopulmonary collateral artery occlusion at the time of right ventricular outflow tract stenting and two patients underwent simultaneous ductus arteriosus stenting to the left pulmonary artery.

Table 2 demonstrates data obtained from angiography at palliation. Cook Formula 418 stents were used in three patients with Liberté Bare-Metal coronary stents used in all others. Median stent diameter was 5 mm (IQR 4.5–5.8 mm) and length of 20 mm (IQR 17–20 mm). Median saturations increased from 79% (IQR 76-80%) to 92% (IQR 90–95%) post-procedure, $p = 0.012$.

Pulmonary artery growth was assessed using both the z scores and Nakata index for the patient on subsequent angiograms over the course of their follow-up. Of the 10 patients that initially underwent right ventricular outflow tract stenting, 9 patients underwent repeat catheterisation. The median duration to the first subsequent angiogram was 245 days (IQR 178−335 days). The median right pulmonary artery z score increased from −3.51 (IQR −4.59 to −2.80) at the initial procedure to a median of −1.17 (IQR −2.26 to 0.16) ($p < 0.05$) at subsequent angiogram. The median left pulmonary artery z score increased from −2.07 (IQR −3.72 to 0.15) at stent placement to 0.24 (IQR −1.09 to 1.84) ($p < 0.05$) (Table 3, Fig 1). One patient had no morphologically identifiable left pulmonary artery. The median Nakata index increased from 45.75 (IQR 34.1−119) at stent insertion to 106.7 (IQR 77.6−178.8) ($p < 0.05$).

The median intensive care and hospital stays were 0 days (range 0–28 days) and 4 days (IQR 2–7 days), respectively. One patent with several co-morbidities (ex-prematurity, very low birth weight, and chromosomal 3 and 10 unbalanced translocations) had disconnected left pulmonary artery, warranting a ductal stent to the left pulmonary artery and right ventricular outflow tract stent to the right pulmonary artery. The patient required intensive care

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Table 1. Patient demographic data

Variable	
Patients	10
Male (%)	7 (70%)
Weight at palliation in kg (median, IQR)	4.2 (3.2–5.7)
Weight at palliation <5 kg (%)	6 (60%)
Age at palliation in days (median, IQR)	61 (8.3–155)
Prostaglandin infusion pre-palliation (%)	4 (40%)
O_2 saturations pre-palliation (median, IQR)	79% (76–80%)
Stent diameter in millimeters (median, IQR)	5 (4.5–5.8)
Number of co-morbidities (median, IQR)	1 (0–1)

Table 2. Angiographic data at initial palliation

Angiogram	
PV diameter (median, IQR)	4.6 mm (3.2–6.26 mm)
Functional atresia on angiogram	1
PV z score (median, IQR)	−4.24 (−4.79 to −3.2)
MPA diameter (median, IQR)	4.15 mm (3.2–6.05 mm)
MPA z score (median, IQR)	−4.04 (−5.12 to −2.11)
RPA diameter (median, IQR)	2.26 mm (1.7–2.97 mm)
RPA z score (median, IQR)	−3.51 (-4.59 to -2.8)
LPA diameter (median, IQR)	3.7 mm (2.1–5.08 mm)
LPA z score (median, IQR)	−2.07 (−3.72 to −0.15)
Nakata index (median, IQR)	45.7 (34.1–119)
Median number of right-sided MAPCAs	2 (1–2.8)
Median number of left-sided MAPCAs	1 (1–2)
Screening time (median, IQR)	39 min (25.5–47.8 min)
Radiation dose (cGy/cm^2)	200 (87.3–355)
Procedure time (median, IQR)	81 min (65.5–115 min)
Contrast volume per kg (median, IQR)	7.5 (5.87–10.4)
Complications	1

following stenting to manage pulmonary overcirculation. This patient had a cardiorespiratory arrest 28 days post-procedure and died. There were no other complications for the remaining patients. There was one subsequent death during the follow-up period on day 178 post-procedure following a respiratory arrest secondary to respiratory infection unrelated to the procedure.

Following right ventricular outflow tract stenting, nine patients underwent further interventions prior to surgical repair (Table 4). The median duration between reinterventions was 287 days (IQR 159–392 days) and the median number of interventions was 2 per patient. Interventions included right ventricular outflow tract stent balloon dilatation (nine interventions), balloon dilatation of branch pulmonary arteries (three interventions), occlusion of major aortopulmonary collateral arteries (five interventions), and right ventricular outflow tract restenting to a larger size diameter stent (three interventions).

To date, four patients have undergone complete repair as described in Table 4. One patient underwent repair with a transannular patch of the right ventricular outflow tract. Three patients underwent repair with a right ventricle-pulmonary artery valved conduit to the native pulmonary arteries. This included one patient undergoing patch augmentation of the pulmonary arteries in the same instance. The median time from right ventricular outflow tract stenting to complete repair was 536 days, and median patient weight at repair was 10.6 kg. One patient required further surgical intervention in the form of homograft patch augmentation extension and downsizing of the right ventricle to pulmonary artery conduit. Two patients underwent a flow-limiting right ventricle-pulmonary artery conduit with the ventricular septal defect left open. Of these, one patient had severely hypoplastic native pulmonary arteries with peripheral branch pulmonary artery stenosis, and a large major aortopulmonary collateral artery to the right lung. The second patient had relatively well-developed central pulmonary arteries, but severe peripheral branch pulmonary artery stenosis. Two patients remain well palliated and clinically stable, and are currently awaiting complete repair. The median age of surviving patients was 5.4 years (IQR, 3.4–8.4 years).

Table 3. Change in parameters post-intervention

Parameter	Pre-RVOT-stenting median (IQR)	At first angiogram post-RVOT-stenting, median (IQR)	p-value
Saturations	79% (76%–80%)	92% (90–95%)	0.012
RPA z score	−3.21 (−4.07 to −2.74)	−1.17 (−2.26 to −0.16)	0.038
LPA z score	−1.41 (−3.66 to 0.14)	0.24 (−1.09 to 1.84)	0.017
Nakata index	45.7 (34.1–119)	106.7 (77.6–178.8)	0.021

Discussion

Due to the rarity of this condition, Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries has been much less extensively studied than its more severe counterpart pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries.[4] Although there are certain similarities, with both groups having hypoplastic native pulmonary arteries and a ventricular septal defect, in Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries, there is antegrade flow through the stenotic pulmonary valve to native pulmonary arteries which are usually confluent. These pulmonary arteries typically have normal or near-normal arborisation and the regions of the lung supplied by major aortopulmonary collateral arteries tend to have a dual blood supply.[6]

There has been little focus on an interventional approach for patients with Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries and the subsequent outcomes. An early study where patients with forward flow were palliated with repeated pulmonary valvuloplasty and branch pulmonary artery balloon angioplasty demonstrated improved pulmonary artery growth.[4] Our study sought to assess the value of right ventricular outflow tract stenting in Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries by assessing the effects on pulmonary artery growth, improvement in saturations, surgical outcome, and survival. It is our opinion that right ventricular outflow tract stenting provides a significant improvement over balloon valvuloplasty, with a more

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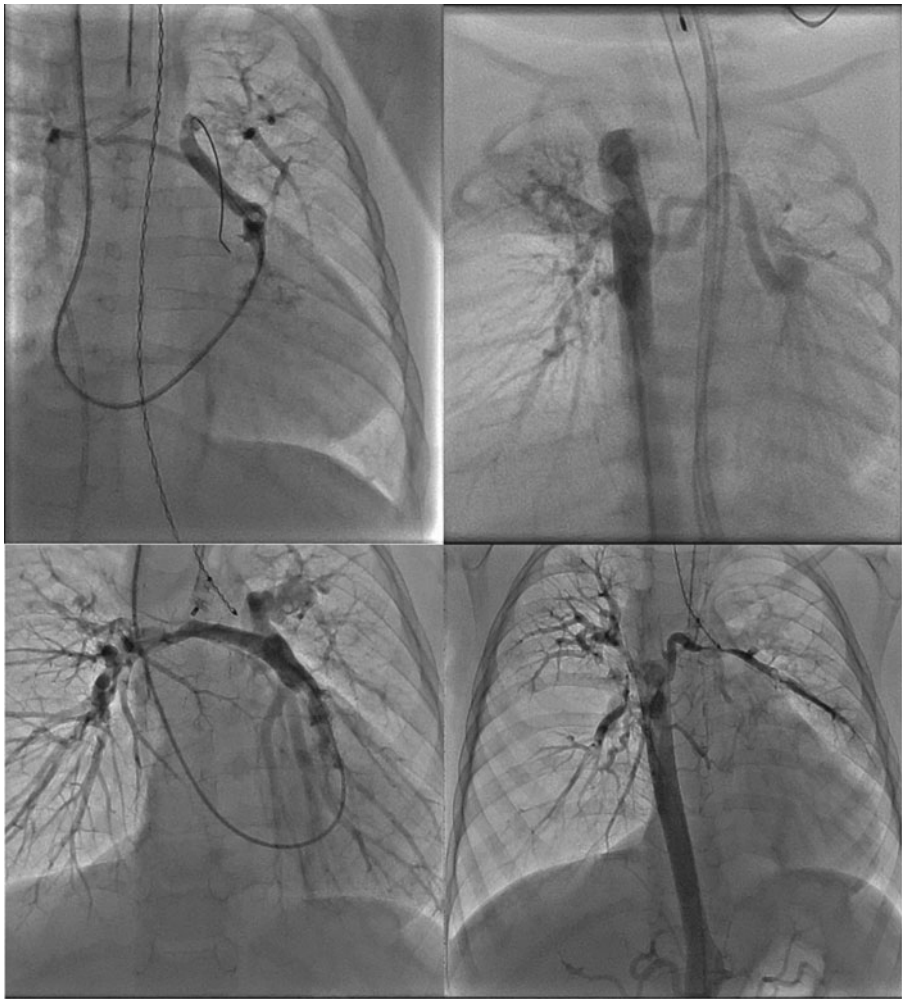

Figure 1. Pulmonary artery growth and MAPCA regression 22 months after RVOT stent. Top left panel – diminutive pulmonary arteries pre-intervention. Top right panel – MAPCA supply to lungs pre-intervention. Bottom right panel – pulmonary arteries post-intervention. Bottom panel – MAPCA stenosis and regression post-intervention.

durable improvement in saturations and a greater promotion of pulmonary artery growth.

We have demonstrated a significant improvement in pulmonary artery size in relation to body surface area over a median follow-up duration of 220 days following right ventricular outflow tract stent insertion. Furthermore, there was a significant improvement in oxygen saturations following the procedure, allowing for a safe delay of surgical correction in the majority of patients. Two patients died during the follow-up period. One patient with a disconnected left pulmonary artery with isolated supply from the arterial duct developed pulmonary overcirculation after right ventricular outflow tract stenting and concomitant patent ductus arteriosus stenting to the disconnected left pulmonary artery. The second patient died at 5 months of age following an unrelated respiratory infection.

Although our study has a limited number of patients, the rarity of the lesion results in there being few comparable reports in the literature. Our findings support the previous publication of Sandoval *et al,* which included 13 infants with Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries. This group also found that right ventricular outflow tract stenting demonstrates good pulmonary artery growth in this cohort.[13] In contrast to Sandoval *et al*, we found that intensive care duration and hospital stays were short in all but the two patients that subsequently died.[13] Although we have not been able to identify a suitable surgical comparator cohort, the overall short inpatient stay compares very favourably to that of early surgical repair in the published literature.

The decision whether to occlude major aortopulmonary collateral arteries at the time of right ventricular outflow tract stenting or to defer to a later stage was individualised. The natural history of these vessels suggests they will undergo spontaneous regression. However, if the vessels were large and supplied the same lung segment as the native pulmonary arteries, they were occluded in order to prevent pulmonary over circulation. The reintervention rate was not insignificant in our study; the requirement for reintervention was almost exclusively as an anticipated planned elective admission to upsize the diameter of the right ventricular outflow tract stent, either through restenting or balloon dilatation, or by major aortopulmonary collateral artery occlusion. We demonstrate similar rates of reintervention to Sandoval *et al.*[13] Complete repair of the lesion was achieved in four patients to date. Two further patients required an interim surgical staging procedure with a flow-limiting conduit, but without ventricular septal defect closure due to severe peripheral branch pulmonary artery stenosis. One of these, and two further patients that at present remain stented, are expected to undergo complete repair in the near future.

In pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries, more than 21% of patients will require early intervention to secure pulmonary blood flow to an area with duct-dependent supply, to increase flow to the native pulmonary arteries[3] in an attempt to "rehabilitate" these, or to

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Table 4. Description of individual patient and outcome data

Patient Number	Cardiac anatomy	Weight (kg)	Age at intervention (days)	Co-morbidity	Pre-intervention saturations (%)	Total number of MAPCAs	Lung segments with isolated MAPCA supply	Initial intervention	Stent dimensions (diameter/length)	Post-intervention Saturations (%)	Subsequent interventions *(days post-initial intervention)*	Repair type and subsequent intervention
1	TOF/PS/ MAPCAs	8.5	222	-	79	5	0	RVOT stent and MAPCA occlusion	7 mm/16 mm	94	1. MAPCA occlusion *(343 days)*	**Tetralogy of Fallot repair and PA augmentation**
2	DORV with malposed great arteries, hypoplastic MPA and Pulmonary valve with forward flow to LPA. Disconnected RPA supplied by MAPCA	3.6	81	1. Chromosome 3/10 unbal anced trans locations 2. Ex 34-week gestation. 3. Low birth weight (1.6 kg)	79	2	3	RVOT and MAPCA stent	RVOT: 6 mm/24 mm MAPCA: 4 mm/16 mm	77		None. Died day 28 post-intervention
3	TOF/PS/ MAPCAs	5.7	176	1. 22q11 deletion 2. Bilateral broncho-malacia 3. Right-sided emphysematous lung 4. Cleft palate 5. Left side hemiparesis due to multiple small infarcts	80	4	0	RVOT stent and MAPCA occlusion	5 mm/20 mm	90	1.RVOT balloon dilatation *(245 days)* 2.RVOT balloon dilatation *(686 days)*	**MAPCA unifocalisation to 12 mm clipped Hancock conduit with VSD left open**
4	TOF/PS/ MAPCAs	10.2	580	1. 22q11 deletion	75	2	0	RVOT stent	6 mm/20 mm	95	1. RVOT balloon dilatation and occlusion of right MAPCA *(413 days)*	-
5	TOF/PS/ MAPCAs	2.6	7	1. 22q11 deletion	70	4	0	RVOT stent	4.5 mm/16 mm	89	1. RVOT restenting (195 days) 2. Balloon dilation of RVOT and occlusion of one left MAPCAs *(578 days)* 3. Occlusion of MAPCAs (768 days)	**Tetralogy of Fallot repair with 19 mm aortic homograft**

6	TOF/PS/ MAPCAs	4.0	40	-	79	3	0	RVOT stent	5 mm/20 mm	92	1. Balloon dilata tion of RVOT/ LPA/RPA *(160 days)* 2. Balloon dilata- tion of RVOT stent *(545 days)* 3. Balloon dilata- tion of RVOT/ LPA/RPA *(1175 days)* 4. Balloon dilata- tion of RVOT/ LPA *(1665 days)*	**14 mm RV-PA Hancock conduit and percutaneous occlusion of MAPCAs** 1. Balloon dilata- tion of the PAs
7	TOF/PS/ MAPCAs	2.8	5	-	73	2	0	RVOT stent	4.5 mm/16 mm	99	1. RVOT restenting and MAPCA occlusion *(178 days)*	**Tetralogy of Fallot repair with RV-PA conduit**
8	TOF/PS/ MAPCAs	5.6	95	1. P2Y12 receptor deficiency platelet disorder	79	4	0	RVOT stent	5 mm/20 mm	92	1. Ballooning of RVOT stent *(335 days)*	**Tetralogy of Fallot repair with hilum-to-hilum homo- graft patch aug- mentation of the PAs and RV-PA conduit** 1. Take down of homograft and downsizing of RV-PA conduit 2. MAPCA occlusion 3. Balloon dilata- tion of RPA 4. LPA stent place- ment.
9	TOF/PS/ MAPCAs	4.3	5	-	87	4	3	RVOT and PDA stent	RVOT:4.5 mm/20 mm PDA: 4 mm/24 mm	90	1. Balloon dilation of arterial duct stent *(38 days)*	None. Died day 171 post- procedure
10	TOF/PS/ MAPCAs	3.1	12	-	90	3	1	RVOT stent	4.55 mm/20 mm	98	1. RVOT restenting *(307 days)*	-

DORV=Double outlet right ventricle; LPA=Left pulmonary artery; MAPCAs=Major aortopulmonary collaterals; PA=Pulmonary artery; PDA=Patent ductus arteriosus; RV=Right ventricle; RPA=Right pulmonary artery; RVOT=Right ventricular outflow; TOF = Tetralogy of Fallot; VSD=Ventricular septal defect

maximise growth of the native vessels prior to complete repair. Some institutions have favoured a rehabilitative approach in all or most patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries due to a low perceived long-term utility of the recruited major aortopulmonary collaterals to the pulmonary vascular bed. We have found that the management of this condition cannot rely on the native pulmonary arteries alone,[5] and the best outcomes have been demonstrated with an approach including early unifocalisation of pulmonary blood supply in the significant majority of patients.[3] Due to the inherently better native pulmonary artery arborisation in Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries, a strategy aimed towards maximising the growth of these vessels may be optimal.

Other factors may also influence the choice of early repair versus an initial procedure to increase flow to the native pulmonary arteries, including the patient's clinical condition, weight, pulmonary artery anatomy, and co-morbidities. In the past, a surgical systemic-to-pulmonary shunt has been the most frequent modality employed in this regard, but has several limitations. Early post-surgical haemodynamic lability and shunt thrombosis remain a concern,[14,15] and distortion of the native vessel may impede optimal growth.[8] Right ventricular outflow tract stenting as the primary intervention in Tetralogy of Fallot has become well established, being our primary palliation in cyanotic infants unsuitable or at increased risk from early repair.[9,12,13,16,17] We have found this approach to be superior to both ductal stenting and the modified Blalock-Taussig-Thomas shunt in this setting, conferring superior haemodynamic stability with maintained diastolic systemic blood pressure and reduced coronary steal. We have also previously shown that right ventricular outflow tract stenting aids superior branch pulmonary artery growth in comparison to modified Blalock-Taussig-Thomas.[9] Furthermore, unlike patent ductal arteriosus stenting, it allows for a longer duration of palliation due to a larger luminal diameter and avoids the distortion or jailing of branch pulmonary arteries.

This cohort of patients demonstrates that right ventricular outflow tract stenting with or without occlusion of major aortopulmonary collateral arteries provides an effective first stage intervention in those at increased risk from early surgical repair. It demonstrates significant growth of the native pulmonary arteries, allowing for complete repair in more than 1/3 of our patients. A larger study is required to robustly assess this condition against matched surgical cases, however; due to the rarity of this condition, we provide one of the largest bodies of evidence that this approach confers improved pulmonary artery growth.

Limitations

The retrospective nature and lack of a control group impose some limitations in the conclusions that can be drawn from our study. Tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries remains a rare lesion, limiting our cohort size, and comparative studies would require the involvement of multiple centres with the additional complexity of differing management paradigms. Four patients had not yet reached definitive endpoints in their management during the study period.

z scores were used as an indicator of pulmonary artery growth, but as they are derived from echocardiographic measurements, the application to angiographic measurements is limited.

The study does not allow the demonstration of the quality of the peripheral pulmonary vasculature in the effect on an outcome. This is evidenced in the two patients that underwent right ventricle-pulmonary artery conduit without ventricular septal defect closure, where the morphology of the peripheral pulmonary vasculature is extensively abnormal with multiple areas of stenosis.

Conclusion

Right ventricular outflow tract stenting is an effective way to improve pulmonary blood flow and pulmonary artery growth in Tetralogy of Fallot patients with antegrade pulmonary blood flow, diminutive pulmonary arteries, and major aortopulmonary collateral arteries. Though primarily applied to those with risk factors for complete repair or a surgical staged repair, the procedure may prove to be a less invasive method of palliation, with superior pulmonary artery growth, for all patients with this rare and challenging anomaly.

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Conflicts of interest. None.

Ethical standards. This work is the result of full participation among all listed authors and was approved by institutional research boards of each member.

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