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Tetralogy of Fallot and abnormal coronary artery: use of a prosthetic conduit is outdated†

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Key question

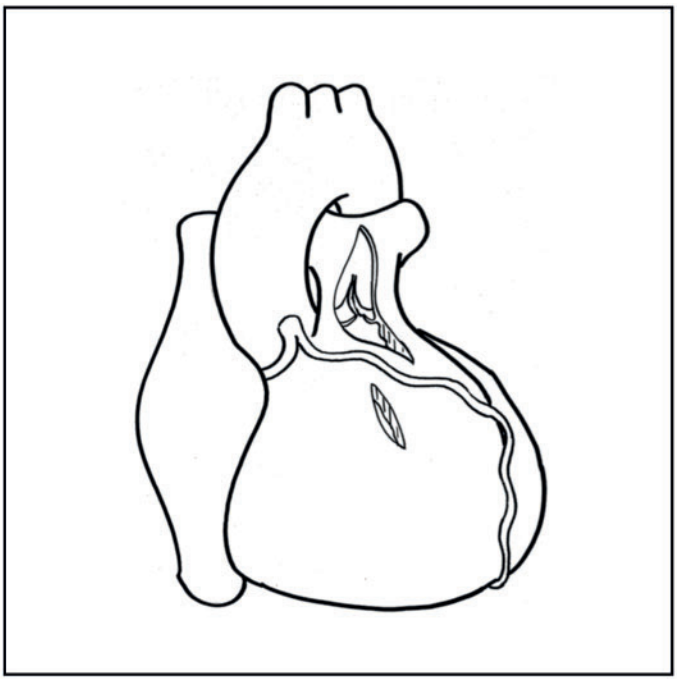
What is the outcome of tetralogy of Fallot with abnormal coronary artery repair without systematic use of a prosthetic conduit?

Key finding(s)

Annulus preservation in 53%; only 1 valved conduit. Right ventricle/left ventricle pressure ratio is a risk factor for right ventricular outflow tract reoperation.

Take-home message

Both transatrial-transpulmonary and transannular approaches offer excellent outcome. Insertion of right ventricle-to-pulmonary artery conduit is outdated.



Abstract

OBJECTIVES: Repair of tetralogy of Fallot (ToF) can be challenging in the presence of an abnormal coronary artery (CA) in 5–12% of cases. The aim of this study was to report our experience with ToF repair without the systematic use of a right ventricle-to-pulmonary artery (RV-PA) conduit.

METHODS: We conducted a monocentric retrospective study from 2000 to 2016, including 943 patients with ToF who underwent biventricular repair, of whom 8% ($n = 76$) presented with an abnormal CA. Mean follow-up time was 50 months (1 month–18 years).

RESULTS: The most frequent CA anomaly was the left descending artery arising from the right CA ($n = 47$, 61.8%). The median age at repair was 7.7 months (1.8 months–16 years). Thirteen patients (17%) required prior palliation, mostly systemic pulmonary shunts for anoxic spells in the neonatal period. Surgical repair allowed us to preserve the annulus in 40 patients (53%) by combining PA trunk plasty,

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commissurotomy and infundibulotomy under the abnormal CA. If the annulus had to be opened ($n = 35$, 46%), a transannular patch was inserted after a vertical incision of the PA trunk and extended obliquely on the RV over the anomalous crossing CA (with an infundibulotomy under the abnormal CA). Three patients (4%) required the insertion of an RV-PA conduit (1 valved tube and 2 RV-PA GORE-TEX tubes with annulus conservation). The early mortality rate was 4% ($n = 3$); none of the deaths was coronary related. Four patients (5%) required reoperation (2 early and 2 late reoperations) for residual pulmonary stenosis, 3 of whom had annulus preservation during the initial repair. The mean RV/left ventricle (LV) pressure ratio and an RV/LV pressure ratio >2/3 were identified as risk factors for right ventricular outflow tract (RVOT) reinterventions ($P = 0.0026$, $P = 0.0085$, respectively), RVOT reoperations ($P = 0.0002$ for both) and reoperation for RVOT residual stenosis ($P = 0.0002$, $P = 0.0014$, respectively). Two patients underwent pulmonary valve replacement. Freedom from late reoperation was 100% at 1 year, 97% at 5 years and 84% at 10 and 15 years.

CONCLUSIONS: Repair of ToF and abnormal CA can be performed without an RV-PA conduit, with an acceptable low reintervention rate. The high early mortality rate in this series remains a concern. If any doubt remains about the surgical relief of the RVOT obstruction, the RV/LV pressure ratio should always be measured in the operating room.

Keywords: Tetralogy of Fallot • Abnormal coronary artery • Surgical treatment • Surgery • Outcomes • Reoperation • Risk analysis • Transatrial approach • Right ventricle-to-pulmonary artery conduit • Pulmonary valve

INTRODUCTION

Tetralogy of Fallot (ToF) represents 5–7% of all congenital heart diseases and is the most common form of cyanotic congenital heart disease. Coronary artery (CA) anomalies occur in 5–12% of ToF cases [1–7]. Because this anomalous CA (ACA) most frequently crosses the infundibular region, it can lead to technical difficulties during the surgical repair and to increased morbidity and mortality rates. A CA lesion can occur if this abnormal course was undiagnosed before or during surgery, leading to severe myocardial ischaemia and inadequate or imperfect surgical correction of the right ventricular outflow tract (RVOT) obstruction [8].

Several surgical repair procedures have been described in the literature to relieve RVOT obstruction without damaging the ACA, the most common being insertion of a right ventricle (RV)-to-pulmonary artery (PA) prosthetic conduit over the ACA or infundibular resection through a right atriotomy and, when surgically possible, an infundibular incision either above or below the ACA, or both [1, 9–17]. An RV-to-PA conduit insertion implies future reinterventions, leading to significant surgical cumulative risk because the conduit is likely to require revision.

The twin aims of this retrospective study were to report the outcomes for ToF repair with associated ACA in our institution and to evaluate the results of different surgical techniques aimed at avoiding the use of RV-to-PA prosthetic conduits.

MATERIALS AND METHODS

A retrospective monocentric study was conducted in our institution from 2000 to 2016. We reviewed our paediatric cardiac surgery database to identify the patients with ToF who underwent biventricular repair. The Paris V University ethics committee granted approval for review of health records. The need for individual consent was waived due to the retrospective nature of the study.

From 2000 to 2016, 1132 patients with ToF underwent surgery in our institution. We excluded patients with ToF with pulmonary atresia and/or atrioventricular (AV) septal defect, patients who had prior surgical repair in another institution and patients who could not benefit from biventricular repair. Patients with ToF displaying a minor infundibular coronary branch were not included.

On the 943 patients with ToF included in our cohort, 76 (8%) had an ACA. An ACA was either diagnosed from preoperative transthoracic echocardiography or discovered at surgery. CA anatomy is described in Table 1. The main ACA were a left CA arising from the right CA ($n = 47$, 61.8%), a major infundibular CA ($n = 15$, 19.7%), a right CA arising from the left CA ($n = 5$, 6.5%), a single CA from 1 sinus ($n = 5$, 6.5%) or a right CA arising from the left sinus ($n = 4$, 5.2%). The mean follow-up period was 50 months (1 month–18 years).

Surgical management

Thirteen patients (17%) required prior palliation at a median age of 41 days (9 days–2 years): 11 systemic pulmonary shunts (85%), 1 RV-to-PA opening and 1 combined RV-to-PA opening and systemic pulmonary shunt. The main indication for a palliative procedure was anoxic spells mainly before 3 months of age (7/13) followed by deep cyanosis.

Surgical repair was performed via a median sternotomy, with standard aortic and bicaval venous cannulation and normothermic cardiopulmonary bypass (CPB) (37°C). Cardioplegia was obtained by infusion of anterograde normothermic hyperkalaemic blood in the aortic root that was subsequently repeated every 10 min or sooner when myocardial activity resumed. Care

Table 1: Anatomy of the abnormal coronary arteries in patients with tetralogy of Fallot

Abnormal anatomy of the coronary artery	Number of patients (%)
LAD from the RCA	
Main LAD from the RCA	32 (42.1)
Accessory LAD from the RCA	15 (19.7)
Major infundibular branch	
Infundibular branch from the RCA	12 (15.8)
Infundibular branch from the LAD	3 (3.9)
RCA from the LAD	5 (6.6)
Single CA	
From right sinus	4 (5.3)
From left sinus	1 (1.3)
RCA from the left coronary sinus	4 (5.3)

CA: coronary artery; LAD: left anterior descending artery; RCA: right coronary artery.

was taken after the pericardiotomy to observe the infundibulum and diagnose or confirm the presence of an ACA.

The repair was always conducted with the same chronological steps. First, the right atrium was opened to close the ventricular septal defect and to allow muscular resection of the proximal component of the RV obstruction. Secondly, the main PA was opened longitudinally for pulmonary valve inspection (and plasty if needed); then, an infundibulotomy was performed beneath the ACA. The muscular resection was performed mainly through the infundibulotomy but also through the pulmonary valve or the transannular incision if done.

Surgical repair allowed preservation of the annulus in 40 cases (53%). The association of both the transatrial and the transpulmonary/infundibular approach was used for infundibular resection in these cases, combined with PA trunk patch enlargement, pulmonary valve commissurotomy and patch enlargement of the infundibulotomy. Annulus preservation with dysplastic valve resection was performed in 5 patients. Two of the 40 patients with annulus preservation required the interposition of an extra-anatomical GORE-TEX tube: isosystemic RV pressures prevented the surgeon from weaning the patient from CPB; the annulus was preserved; and the tubes were inserted between the infundibulotomy and the main PA patches.

If the annulus was determined from preoperative echocardiography to be too narrow to allow preservation or if infundibular resection and pulmonary valve commissurotomy did not allow the annulus to accept the appropriate Hegar dilator, the annulus had to be opened ($n = 35$, 46%): a transannular patch was inserted after vertical incision of the PA trunk and extended obliquely on the RV (with an infundibulotomy under the abnormal CA) (Fig. 1).

One patient (6%) required an RV-PA valved conduit insertion because the RVOT remained too narrow despite annulus opening and muscular resection.

After repair and bypass weaning, RV pressures were always monitored, either via a transoesophageal echocardiogram or by a direct puncture in the RV and compared with the arterial systemic pressures.

Median age and weight at repair were 7.9 months (1.8 months–16 years) and 13.3 kg (3.7–26 kg). Mean CPB time and aortic clamping time were 115 ± 35 min (range 81–235) and 74 ± 20 min (range 45–154), respectively.

Statistical analysis

Continuous data are expressed as median and range or mean and standard deviation. Categorical data are expressed as number and percentage. Univariable analysis was performed using either the χ^2 or the Fisher's exact test for categorical data or the Student's *t*-test for continuous data. Overall survival and freedom from events were calculated using the Kaplan–Meier method with curves compared using the log-rank test. Data were analysed using GraphPad Prism version 6 software (GraphPad Software, La Jolla, CA, USA).

RESULTS

Patient demographics are displayed in Table 2. There were no statistical differences between patients who had annulus preservation and those in whom the annulus was divided, especially in terms of residual gradient ($P = 1.0$), the RV/left ventricle (LV) ratio ($P = 0.4005$), deaths ($P = 1.0$) or reinterventions ($P = 0.3616$).

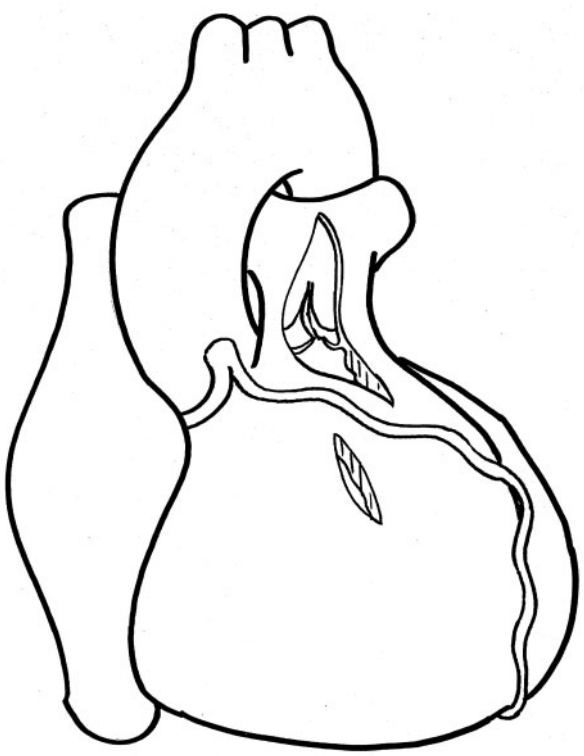

Figure 1: Transannular technique for correcting tetralogy of Fallot and abnormal coronary artery. The vertical incision in the pulmonary artery trunk extends obliquely on the right ventricle, parallel to the abnormal coronary artery. Infundibulotomy is realized under the abnormal coronary artery.

Early outcomes

Median stays in the intensive care unit and in the hospital were 3 days (1–15 days) and 1 week (5 days–1 month), respectively. Half of the patients were weaned from mechanical ventilation less than 24 h after surgery (maximum mechanical ventilation 9 days).

One patient required central extracorporeal life support after surgery: surgical repair included annulus preservation but with pulmonary valve leaflet resection, infundibular resection and interposition of 2 pericardial patches on the infundibulum and the main PA. Due to persistent muscular ventricular septal defects and systemic right ventricle pressures, a second GORE-TEX tube had been inserted between the 2 patches during the same intervention to avoid compression of the left anterior descending (LAD) artery (from the right CA). The patient was weaned from extracorporeal life support after 4 days, and the chest was closed 10 days after surgical repair. The patient is still alive as of the last follow-up, 7 years after surgery.

One patient required pacemaker implantation for complete AV block in the early postoperative period.

Survival

Overall survival was 96.1% [95% confidence interval (CI) 93.4–100] at 1, 5 and 15 years.

Table 2: Patient demographics

Patient characteristics	Population ($n = 76$)	Preserved annulus ($n = 40$)	Non-preserved annulus ($n = 36$)	P-value
Preoperative data				
Male sex	48 (63.2)	25 (62.5)	24 (66.7)	0.8116
Age (months)	7.9 (1.8–192)	18 (2.2–112.5)	7.1 (1.8–192)	0.8335
Weight (kg)	8 (3.7–26)	9.1 (3.7–26)	7 (4.2–25)	0.2936
Palliation	13 (17.1)	2 (5.0)	11 (30.6)	0.0048
Peri- and postoperative data				
CPB (min)	127 (81–235)	113 (81–235)	117 (129–188)	0.2655
Aortic clamping (min)	75 (45–154)	88 (45–154)	89 (60–110)	0.5164
Ventilation (h)	4 (2–216)	4 (2–216)	4 (2–24)	0.4073
ICU stay (days)	2 (1–20)	2 (1–20)	2 (1–9)	0.9020
Hospital stay (days)	7 (5–30)	7 (5–30)	7 (5–15)	0.9887
Mean gradient >20 mmHg	26	14	12	1.0
RV/LV ratio	0.55 (0.3–1)	0.6 (0.3–1)	0.5 (0.35–0.8)	0.4005
RV/LV ratio >2/3	16	10	6	0.4123
RV/LV ratio >0.5	38	21	17	0.8185
Mortality	4 (5.3)	2 (5.0)	2 (5.6)	1.0
Early	3 (3.9)	1 (2.5)	2 (5.6)	0.6006
Late	1 (1.3)	1 (2.5)	0	1.0
RVOT reinterventions	8 (10.5)	4 (10)	4 (11.1)	1.0
RVOT reoperations	5 (6.6)	4 (10)	1 (2.8)	0.3616
Early	2 (2.6)	2 (5.0)	0	0.4947
Late	3 (3.9)	2 (5.0)	1 (2.8)	1.0
For residual stenosis	4 (5.3)	3 (7.5)	1 (2.8)	0.6170

Data are shown as n (%) or median (range).

CPB: cardiopulmonary bypass; ICU: intensive care unit; RV/LV: right ventricle/left ventricle; RVOT: right ventricular outflow tract.

The early mortality rate was 3.9% ($n = 3$). There were no coronary-related deaths. Two patients with a major infundibular branch from the right CA, who had required a transannular patch, died 9 and 11 days postoperatively, respectively: the first patient died of an intravenous perfusion malfunction, with a massive gaseous embolism; the second patient died in her sleep at home, 3 days after discharge. Necropsy revealed a chylothorax. One patient with an LAD from the right CA and preserved annulus died 10 days after surgery from pulmonary oedema. The postoperative transpulmonary gradient was 20 mmHg with an RV/LV pressure ratio <0.5. The patient was 2 years old, extremely cyanotic before repair and part of a humanitarian programme, which explains why he was operated on so late. It was impossible to know if this was late pulmonary reperfusion oedema or an acute coronary malperfusion leading to left ventricular dysfunction.

There was 1 late death (1/73, 1.3%). The patient was 27 years old and died of ventricular dysfunction 10 years after homograft pulmonary valve replacement. He presented with a single CA from the right coronary sinus. Surgical repair had been performed at 10 years of age. The annulus had been preserved but the dysplastic valve had been resected. During follow-up, no electric signs of myocardial ischaemia had appeared on the echocardiogram, and the results of the last coronary angiography examination were reported as normal.

Reinterventions

Four patients (5%) required reoperation (2 early and 2 late reoperations) for residual pulmonary stenosis, of whom 3 had annulus preservation during the initial repair. Freedom from reoperation for RVOT residual stenosis was 97.4% (95% CI 95.4–100) at 1 year, 93.9% (95% CI 89.5–100) at 5 years, 87.2% (95% CI 78.4–100) at 10 and 15 years (Fig. 2).

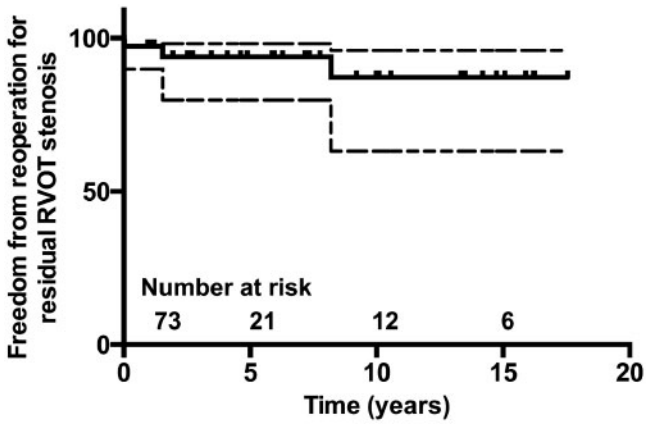

Figure 2: Freedom from reoperation for residual right ventricular outflow tract stenosis. Four patients required reoperation for early ($n = 2$) or late ($n = 2$) residual pulmonary stenosis. Freedom from reoperation for right ventricular outflow tract residual stenosis was 97.4% [95% confidence interval (CI) 95.4–100] at 1 year, 93.9% (95% CI 89.5–100) at 5 years, 87.2% (95% CI 78.4–100) at 10 and 15 years. RVOT: right ventricular outflow tract.

Early reinterventions. Two patients (2.6%) with annulus preservation required early reoperations for residual stenosis.

The first patient presented with a right CA from the LAD. He required an early reoperation on the same day because of isosystemic right ventricular pressure. Iterative infundibular muscular resection and infundibular patch enlargement with preservation of the annulus resulted in a decrease in the RV/LV pressure ratio to 0.6 and the residual gradient was 50 mmHg.

The second patient was 4 months old at the time of repair and had an LAD from the right CA. He required annulus transection and iterative infundibular resection 6 days after the surgery for an

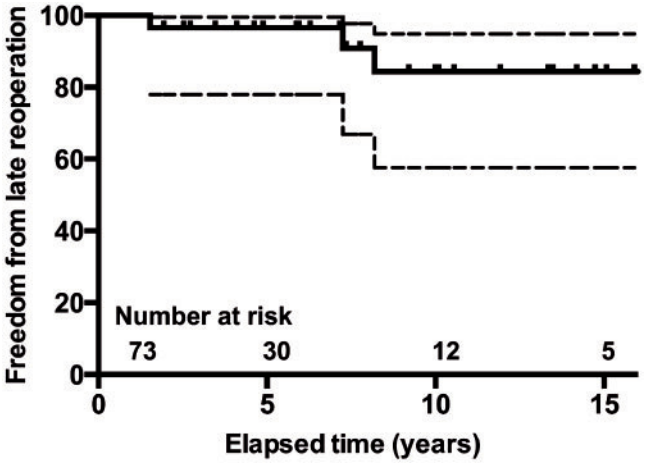

Figure 3: Freedom from late reoperation on the right ventricular outflow tract. Freedom from redo surgery for pulmonary stenosis or valve replacement was 100% at 1 year, 96.6% [95% confidence interval (CI) 93.6–100] at 5 years and 84.4% (95% CI 73.9–100) at 10 and 15 years.

RV/LV pressure ratio of 0.80. The postoperative gradient was 20 mmHg. Fifteen years after surgery, the patient is doing well and does not actually meet the criteria for pulmonary valve replacement.

Late reinterventions. Four patients (5.3%) required late reintervention on the RVOT: 2 for recurrence of RVOT stenosis and 2 for pulmonary valve replacement (1 surgical and 1 with a Melody^{TM} implant).

None of the 3 patients who required a tube insertion (1 repaired with an RV-to-PA valved conduit, 2 with annulus preservation and an additional RV-to-PA GORE-TEX tube).

Indication for late reoperation for residual pulmonary stenosis was considered when the RV pressure was isosystemic or when the stenosis was associated with RV dysfunction. Both patients who required late reoperation for RVOT obstruction had an LAD from the right CA: 1 had required a transannular patch whereas the other had a preserved annulus. Redo surgery comprised infundibular muscular resection and infundibular patch enlargement, respectively, 2 and 8 years after the initial repair. The postoperative course was uneventful.

Pulmonary valve replacement was performed 7 years after initial repair for the homograft implant (single CA, preserved annulus but resected valve) and at 13 years for the Melody^{TM} implant (LAD from the right CA, transannular repair).

Two additional patients required cardiac catheterization for stenting of pulmonary branches.

Freedom from late reoperation was 100% at 1 year, 96.6% (95% CI 93.6–100) at 5 years and 84.4% (95% CI 73.9–100) at 10 and 15 years (Fig. 3).

Risk factors

We failed to identify any risk factors for early and overall mortality.

Annulus preservation, conduit insertion and a mean postoperative gradient >20 mmHg were not identified as risk factors for reinterventions or reoperations. A mean RV/LV pressure ratio and an RV/LV pressure ratio >2/3 were identified as risk factors for RVOT reinterventions (*P*-values 0.0026 and 0.0085), RVOT reoperations (both *P*-values 0.0002) and reoperation for RVOT residual stenosis (*P*-values 0.0002 and 0.0014) (Table 3).

DISCUSSION

Tetralogy of Fallot and abnormal coronary artery

Survival rates for patients with ToF have increased over the years thanks to improvements in preoperative diagnosis, the development of various surgical techniques for repair and improvements in postoperative care. Nevertheless, it is widely assumed, like Saygi *et al.* [18] reported, that the presence of coronary anomalies is a risk factor for death.

To prevent coronary injury, the 2008 American College of Cardiology/American Hospital Association guidelines recommend that when ToF is associated with ACA, the anatomy of the CA should be determined before any intervention on the RVOT is undertaken [19]. We adhere completely to these guidelines. Following this preoperative diagnostic procedure allows one to adapt the surgical strategy to the patient and to have the most experienced members of the surgical team in the operating room. It might almost preclude the eventuality of major coronary injury and offer the option of repair without the use of a conduit. The preoperative diagnosis of ACA is usually made with echocardiography if it is performed by senior cardiologists. Computed tomography is a non-invasive imaging technique with high sensitivity and specificity for CA anatomical analysis, but cardiac catheterization remains the gold standard for accurate coronary diagnosis, although it is not mandatory before surgery [4, 5, 7, 20]. We do not recommend mandatory computed tomography imaging when ACA is suspected (because it would not change the indication for surgery) except for patients who had a palliative operation elsewhere without a specific description of the coronary anatomy at first surgery.

Surgical techniques for repair

Numerous surgical techniques have been described for patients with ToF and ACA to relieve RVOT obstruction and to avoid CA injury: infundibulotomy with CA ligation, RV-to-PA connection with a prosthetic conduit, the transatrial–transpulmonary approach, main PA translocation, mobilization of the ACA to slide the patch underneath or even ACA translocation [1, 9–17]. There is no consensus on the ideal surgical management of this subtype of ToF. The transatrial–transpulmonary approach has often been described to display the best long-term outcomes in ToF either associated with or without CA anomalies [1, 6, 13, 17]. Preservation of the pulmonary annulus was possible in more than half of our patients: surgery included extensive muscular infundibular resection via both a transatrial approach and through the infundibulotomy, pulmonary valve plasty with a commissurotomy and, if required, perioperative balloon dilatation and pericardial patch enlargement of both the PA trunk and the infundibulum. Five of the 40 patients had annulus preservation but pulmonary valve leaflet resection, and 2 had an additional tube inserted for residual stenosis. If the annulus needed to be opened, a vertical incision of the PA trunk, extending obliquely on the RV just above the ACA, frequently associated with an infundibulotomy under it, yielded satisfactory postoperative results. The 2 different approaches did not differ significantly in

Table 3: Risk factors for reintervention and reoperation

	Patient data	*P*-value
RVOT reintervention (*n* = 8)		
RV/LV pressure ratio	Mean ratio 0.73 ± 0.16 (vs 0.55 ± 0.13)	0.0026
RV/LV pressure ratio >2/3	5/8 patients (vs 11/68)	0.0085
RV/LV pressure ratio >0.5	7/8 patients (vs 31/68)	0.0561
RVOT reoperation (*n* = 5)		
RV/LV pressure ratio	Mean ratio 0.8 ± 0.12 vs 0.55 ± 0.13	0.0002
RV/LV pressure ratio >2/3	5/5 patients (vs 11/71)	0.0002
RV/LV pressure ratio >0.5	5/5 patients (vs 33/71)	0.0543
Reoperation due to residual RVOT stenosis (*n* = 4)		
RV/LV pressure ratio	Mean ratio 0.825 ± 0.125 vs 0.55 ± 0.125	0.0002
RV/LV pressure ratio >2/3	4/4 patients (vs 11/72)	0.0014
RV/LV pressure ratio >0.5	4/4 patients (vs 34/72)	0.1151

Data are presented as number or mean ± standard deviation.

RV/LV: right ventricle/left ventricle; RVOT: right ventricular outflow tract.

terms of postoperative residual gradients, RV/LV ratios, survival or freedom from reinterventions. To our knowledge, both techniques can be justified, depending on the surgeon's appreciation of the pulmonary valve and annulus regardless of the ACA anatomy, because they offer similar outcomes. In our experience, the key point and the success of the surgery depend on the aggressive muscular resection beneath the crossing CA. This goal can be achieved from the superior and inferior approaches (through the pulmonary annulus and via the infundibulotomy), with particular attention to the posterior wall of the CA. We use very fine ophthalmic scissors to proceed with this resection and always finish with an anterograde blood cardioplegia reinjection in the aortic root to exclude any injury. The final result has to be systematically evaluated by a transoesophageal echocardiogram and determination of the pressure gradient between the RV and the main PA.

Mortality and coronary artery damage

Some authors of case reports or of studies with larger cohorts have also reported their experience with the surgical management of ToF and associated ACA [1, 8, 15, 17]. Although the early mortality rate was reported to be 8–15% before the 1990s, it has clearly dropped below 10% during the last 20 years and now ranges between 0% and 6% [1, 8, 10–14, 17].

Although it occurs rarely nowadays, inadvertent transection of the ACA while opening the infundibulum is every surgeon's nightmare because it can result in serious myocardial infarction and operative death. In such cases, a CA bypass is required, and one should always use the internal mammary artery for long-term patency [21, 22].

Data from the European Association for Cardio-Thoracic Surgery congenital database indicate an overall hospital mortality rate of 2.58% after ToF repair (6654 patients, 1999–2011) [23]. Causes of death and coronary patterns were not specified. The early mortality rate in our cohort of patients with ToF and ACA was 3.9% with an overall survival rate of 96% at 15 years, close to the survival rates described in the literature [24]. None of the deaths were coronary related but in case of any abnormal postoperative course, a coronary angiogram should strongly be considered in an emergency. Comments in previous publications make it appear that anomalies of the anatomy of the CA should no longer be considered a risk for increased mortality in ToF repair [1, 14]. Nevertheless, it can be dangerous to assume that CA in patients with ToF is no longer an issue. In our large-volume centre, we still feel that patients with ToF with CA are more demanding than those without CA and need special attention and appropriately trained surgeons if one wants to avoid the systematic use of the RV-to-PA conduit.

Reoperation on the right ventricular outflow tract

Kalfa *et al.* [17] reported that implantation of an RV-to-PA conduit significantly increases the risk of RVOT obstruction and reoperation in patients with ToF and ACA due to the lack of growth potential and the natural evolution of the currently used devices. The research to develop an ideal RV-to-PA conduit via tissue engineering [25, 26] might reduce this crucial problem in the future.

In our cohort, most of the patients were repaired without the use of a conduit, despite the presence of an ACA. Only 1 patient required valved conduit placement during the repair, and 2 required the insertion of an additional extracardiac polytetrafluoroethylene tube while the annulus was preserved. Neither of these 2 patients actually required reintervention, probably due to the natural growth of the preserved RVOT, allowing efficient pulmonary flow despite progressive restriction of the tube. The RV/LV pressure ratio did not significantly differ whether the valve was spared or not but was identified as a risk factor for reintervention and reoperation, especially for RVOT residual stenosis. The RV/LV pressure ratio had already been identified as a risk factor for mortality after ToF repair and/or for RVOT obstruction and reoperation in numerous publications [17, 27]. We recommend that the surgical team concentrate on this key point at repair by optimizing the muscular resection beneath the CA and always measure the right ventricular pressure after the patient is weaned from CPB, especially when the annulus is preserved. A ratio greater than 2/3 should raise the question of an immediate second look at the RVOT, depending on the perioperative findings.

An optimal repair without the use of a prosthetic conduit is correlated in this series with a low long-term reoperation rate,

probably minimizing the cumulative mortality risk due to multiple redo operations.

Limitations

The main limitations of our study are its retrospective nature and the limited number of patients included, although it is one of the largest cohorts of patients with ToF and abnormal CA that has been reported. The choice of the surgical technique was at the discretion of the surgeon, which could be considered a bias.

CONCLUSION

We can conclude that patients with ToF with abnormal CA can be effectively treated without the use of an RV-PA conduit. The association of aggressive infundibular resection, pulmonary valve commissurotomy (if the annulus is preserved) and infundibular and PA trunk patch enlargement resulted in an acceptable mortality rate and few reinterventions for residual pulmonary stenosis without coronary damage. If there is any doubt after repair, one should always measure RV pressure and relieve the right outflow tract obstruction if the RV/LV pressure ratio is inadequate.

Conflict of interest: none declared.

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