Neonatal right ventricle to pulmonary connection as a palliative procedure for pulmonary atresia with ventricular septal defect or severe tetralogy of Fallot[†]

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Abstract

OBJECTIVES: Right ventricle to pulmonary artery connection (RVPA connection) without prosthetic material has been our ideal strategy to palliate pulmonary atresia with ventricular septal defect (VSD) or severe tetralogy of Fallot for the last decade. We speculate that RVPA connection ensures adequate postoperative haemodynamics for symptomatic neonates and promotes pulmonary artery rehabilitation. The present study was undertaken to assess the outcome of this strategy.

METHODS: Between 2000 and 2010, among 107 patients who benefited from an RVPA connection, 57 were neonates. Forty-eight of these underwent autologous tissue reconstruction, 5 using left atrial appendage. Median weight was 2.9 kg (range 1.8–4.4). Median Nakata index was 100 mm^2/m^2 (range 17–185 mm^2/m^2); 12% had major aortopulmonary collaterals. All patients were reviewed retrospectively. End-points were death or complete repair; reintervention for restrictive pulmonary blood flow was considered as failure. At follow-up, we evaluated reintervention after complete repair, and quality of life.

RESULTS: There were 2 early deaths (RV hypoplasia and RV failure) and 3 late sudden deaths (range 3–6 months). Pulmonary blood flow required to be increased in 8 patients: 4 underwent shunt after a median delay of 1 month; RVPA connection enlargement was needed in 3; 1 patient had percutaneous angioplasty. Finally, 47 patients (81%) had a complete repair, of which 70% were performed without prosthetic material at a median age of 7 months (range 2–53), with a median Nakata index of 221 mm^2/m^2 (range 102–891). One patient died early and 1 was a failure with opening of the VSD after intracardiac repair. At last follow-up, 4 patients were still awaiting repair, with 1 late death and 5 who had required reintervention after intracardiac repair; there were 3 conduit replacements and 2 balloon dilatation patch enlargements.

CONCLUSIONS: The neonatal RVPA connection approach (i) provides an acceptable survival rate with a satisfactory haemodynamic adaptation, (ii) facilitates rehabilitation of PAs and (iii) avoids the use of prosthetic graft at correction.

Keywords: Neonatal · Pulmonary atresia with ventricular septal defect · Major aortopulmonary collaterals · Tetralogy of Fallot with pulmonary atresia · Surgical management · Palliative surgery

INTRODUCTION

Symptomatic neonatal patients (severe cyanosis or cyanotic spell) or ductus arteriosus circulation-dependant patients with pulmonary atresia (PA) and ventricular septal defect (VSD) or tetralogy of Fallot (TOF) need a surgically created additional source of pulmonary blood flow in the first weeks of life. For this population, two problems still emerge. First, the identification of patients whose pulmonary arteries are too small to permit a complete repair is imprecise [1]. Some surgical teams have even proposed early primary complete repair for patients with neonatal TOF [2–5]. Different results have suggested some limitations (high mortality or reintervention) of this strategy for these specific neonatal symptomatic groups of patients with too-small pulmonary arteries [3, 6, 7]. Secondly, the ideal palliative procedure to increase homogeneously and efficaciously the size of pulmonary arteries with low early postoperative mortality, is still debated. Palliative Blalock–Taussig (BT) shunting and right ventricle to pulmonary artery (RVPA) connection are actually the most utilized solutions [8–14]; even stenting of the patent ductus seems to be prevalent in some centres.

Irrespective of the size of the native pulmonary arteries, the philosophy of our team is to construct a palliative RVPA

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connection [5, 10–12, 15]. When anatomy is favourable, we preferentially use autologous tissue.

We believe that this strategy ensures a stable haemodynamic postoperative period, low mortality at palliation and homogeneous growth of pulmonary arteries. It further provides easy access for potential pulmonary artery percutaneous procedures and avoids the use of a prosthetic conduit at repair. The present study was undertaken to assess the outcome of this strategy in the neonatal period.

MATERIALS AND METHODS

Study design

Between 2000 and 2010, 107 patients with PA/VSD or severe pulmonary hypoplasia with TOF were enrolled in a programme of pulmonary rehabilitation with an initial palliation by RVPA connection while 64 had a BT shunt during the same period. The option between RVPA connection and BT shunt was left to the choice of the surgeon with a general strategy to prefer BT shunt for the largest pulmonary arteries. Of the 107 RVPA connection patients, only 57 were neonates, and included in the present study to evaluate the results of this approach in a carefully selected cohort. Inclusion criteria were PA/VSD or TOF with extreme cyanosis or cyanotic spells, despite medical therapy or a duct-dependent circulation. Patients with absent central native pulmonary arteries (PA/VSD type IV; Castaneda classification [16]) were excluded. The success of this strategy was defined by the subsequent completion of definitive two-ventricle repair (VSD closure with subsystemic RV pressure) confirming the satisfactory rehabilitation of the pulmonary arteries. Permission to perform health records review was obtained from the Paris V University Ethics Committee. The need for individual consent was waived.

Study population

A group of 57 patients was selected between the period 2000 and 2010, according to our inclusion and exclusion criteria. Figure 1A shows the number of patients operated on by year during the study period. The populations and their anatomical characteristics are reported in Table 1 together with the type of RVPA connection. The median age of operation was of 10 days (range 3–29). The median weight was 2.9 kg (range 1.8–4.2 kg). The population was comprised as follows: 24 (42%), 14 (25%), 7 (12%) and 12 (21%) patients as PA/VSD type I; II; III (according to the classification of Castaneda [16] and TOF, respectively. Nine patients (21%) had genetic anomalies with significant representation of Di-George syndrome (89%). The patients with Di-George syndrome were preferentially associated with PA/VSD type III (P = 0.05).

The majority of the study population had cyanotic spells (54%), 25% had saturations <70% and 10 patients (18%) underwent mechanical ventilation before surgery. Three patients (5%) presented with severe ventricular dysfunction. Co-existent anatomical anomalies were of coronary artery anomalies (left coronary artery from the right ostium) in 4 patients (7%) and hypoplastic aortic arch in 1 patient with TOF.

In this population, the median Nakata index was 100 mm^2/m^2, (range 17–185 mm^2/m^2). Twenty-five percent of the cohort had a Nakata index <77 mm^2/m^2. Patients with PA/VSD type III had a
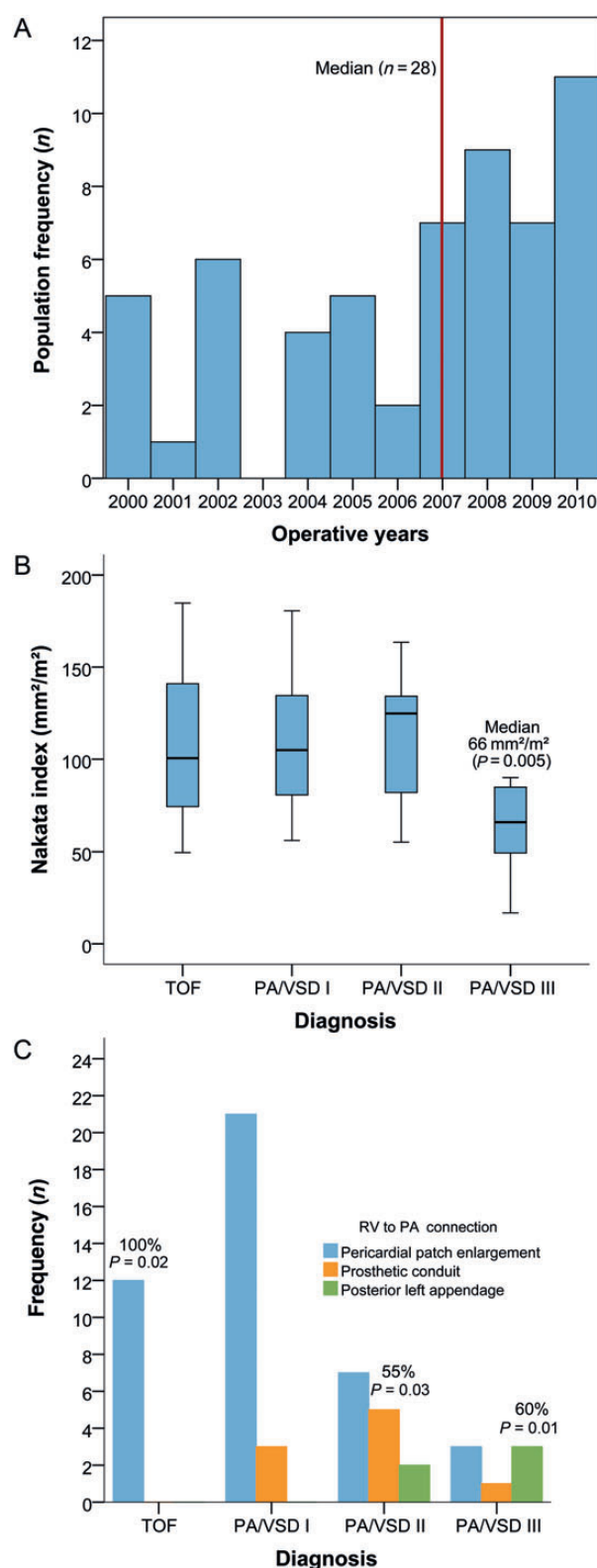

Figure 1: (**A**) Number of patients operated by year during the study period. (**B**) Distribution of Nakata index in the various diagnostic groups. (**C**) Distribution of the three different surgical procedures by diagnostic.

significantly lower median Nakata index (66 mm^2/m^2; P = 0.005) compared with the remaining patients. Figure 1B represents the distribution of Nakata index in the various diagnostic groups.

Table 1: Population and anatomic characteristics before the initial palliation

Variables	PA/VSD I (*n* = 24)	PA/VSD II (*n* = 14)	PA/VSD III (*n* = 7)	TOF (*n* = 12)	Total (*n* = 57)
Age (day)	8 [5–25]	9 [6–23]	10 [3–24]	21 [3–29]	10 [3–29]
Sex (male)	14 (58)	7 (50)	3 (43)	8 (66)	32 (56)
Weight (kg)	2.7 [2–4.4]	2.8 [1.9–3.9]	3.1 [2.6–4]	2.9 [1.8–4.2]	2.9 [1.8–4.4]
BSA (m^2)	0.2 [0.2–0.4]	0.2 [0.2–0.3]	0.22 [0.2–0.5]	0.2 [0.2–0.3]	0.2 [0.2–0.5]
History prior to surgery					
Genetic syndrome	4 (12.5)	5 (35)	3 (43)	–	12 (21)
Di-George syndrome	4 (17)	1 (7)	3 (43)	–	8 (14)
Prematurity/hypotrophy	2 (8)	4 (29)	–	–	6 (11)
Arterial saturation (%)	77 [50–94]	81 [55–93]	80 [40–89]	82 [60–90]	80 [40–94]
Ventricular dysfunction	2 (8)	–	–	1 (8)	3 (5)
Cyanotic spell	11 (46)	6 (43)	4 (57)	10 (83)	31 (54)
Saturation <70%	8 (33)	7 (50)	2 (29)	3 (25)	20 (35)
Mechanical ventilation	2 (8)	4 (29)	2 (29	2 (17)	10 (18)
Anatomy					
Coronary anomalies	1 (4)	3 (21)	–	–	4 (7)
Nakata index (mm^2/m^2)	104 [56–181]	124 [55–164]	66 [17–90]	100 [50–185]	100 [17–185]
Diameter of the right PA (mm)	4 [3–5]	4 [2.5–5]	3 [1.5–3.5]	4 [2.5–5]	4 [1.5–5]
Diameter of the left PA (mm)	3.7 [2–5]	3.7 [2–6]	3 [1.5–4]	3.5 [2.5–5]	3.5 [1.5–6]
Surgical procedure					
Patch enlargement	21 (88)	7 (50)	3 (43)	12 (100)	43 (75)
PTFE conduit	3 (12)	5 (36)	1 (14)	–	9 (16)
Interposition left appendage	–	2 (14)	3 (43)	–	5 (9)
Open size (mm)	5 [3.5–6]	5 [4–6]	5 [4–5]	5 [4–5.5]	5 [3.5–6]
LPA plasty	–	1 (7)	2 (29)	1 (8)	4 (7)
RPA plasty	–	1 (7)	–	–	1 (2)
Confluence PA plasty	1 (4)	5 (36)	1 (14)	1 (8)	8 (14)
PA plasty	1 (4)	5 (36)	2 (27)	1 (8)	9 (16)
Correction hypoplastic arch	–	–	–	1 (8)	1 (2)

Data are shown as medians and minus, maximal interval or as numbers and percentages.
PA/VSD: pulmonary atresia with ventricular septal defect; TOF: tetralogy of Fallot; BSA: body surface area; LPA: left pulmonary artery; RV: right ventricle; PA: pulmonary artery; RPA: right pulmonary artery.

Surgical technique

All procedures were performed through a median sternotomy under normothermic cardiopulmonary bypass (CPB) using bicaval cannulation and left ventricle (LV) venting. The heart was arrested with antegrade warm blood cardioplegia, which was repeated every 10 min. The proximal branch pulmonary arteries together with the bifurcation were mobilized extensively and the arterial duct ligated and divided if present. Right atrial and left atrial pressures were monitored at the end of the procedure.

Strategically there were three techniques for surgical reconstruction depending on anatomical configuration and surgeon preference. In general, we preferred to use autologous tissue to achieve reconstruction of the posterior wall of the RVPA connection to minimize the use of heterologous materials, in particular prosthetic conduits, at the time of complete repair. The first technique used was in cases of TOF and PA type I and III, which comprised a vertical incision of the main PA in continuity with an infundibular incision of 5–8 mm, together with limited resection of right ventricle outflow tract (RVOT) muscle bundles. Great care was taken to avoid injury to the coronary arteries when opening the infundibulum, particularly in low birth-weight babies with PA where the distal infundibulum is in close proximity to the left anterior descending artery (LAD), aorta and right coronary ostium. In cases with marked hypoplasia of the pulmonary trunk or atresia, posterior continuity of the RVPA connection was augmented using interrupted sutures to approximate the distal margin of

the ventriculotomy with the proximal end of the pulmonary arteriotomy. The anterior wall was reconstructed using a heterologous pericardial patch initially sized at 5–6 mm (depending on the patient weight), around a Hegar dilator; this was subsequently adjusted off-CPB according to systemic arterial blood oxygen saturations, distal systolic pulmonary artery pressure and magnitude of the shunt through the VSD if necessary. In particular, where the central pulmonary arteries were of adequate calibre with no significant peripheral stenoses, care was taken to ensure the RVPA connection was sufficiently restrictive to pulmonary blood flow. The size adjustment was performed by means of a fine 'U'-stitch in the anterior heterologous patch. Our strategy was to initially perform a 'large' RV to PA connection rather than a very restrictive one, due to the fact that it was extremely easy to reduce the calibre. On the contrary, enlargement of the connection would need the use of iterative bypass and aortic cross clamp.

Heterologous pericardium was initially used to reconstruct the anterior walls of all patients. This choice was made to avoid any risk of aneurysm or calcifications of the patch itself. For the patients <3 kg, we now prefer to use a glutaraldehyde autologous pericardium that is easier to handle with the smallest pulmonary arteries. The growth potential of the autologous treated patch is also probably very limited.

The second method of reconstruction used (called 'autologous tissue reconstruction' due to the reconstruction of a posterior wall made of viable autologous tissue) was in cases of PA (type II or III) with no main PA trunk. In this method, RVPA continuity was

established without any extracardiac conduit; the pulmonary outflow tract was reconstructed using the left atrial appendage (LAA). After extensive mobilization of the pulmonary arteries and bifurcation, the left appendage was sutured to the distal end of the right ventriculotomy and to the pulmonary confluence that was left in its anatomical position. The entire posterior wall of the RVOT was thus made of autologous vascularized tissue. The anterior wall was reconstructed with a bovine pericardial patch, again sized at 6 mm diameter and adjusted if necessary.

The third technique for reconstruction employed a 5–6-mm polytetrafluoroethylene (PTFE) tube graft to connect the RV to the pulmonary bifurcation. This has been used in the presence of an anomalous left coronary artery crossing the infundibulum or if the surgeon was not familiar with the use of the LAA reconstructive method in case of no PA with no pulmonary main trunk.

Follow-up and data analysis

Follow-up data were obtained during a 4-month closing interval for the study (October 2011–January 2012). Data were obtained for 55 patients (96% complete).

Primary end-points included survival and complete repair (success of pulmonary artery rehabilitation). At last follow-up, we assessed survival, reintervention rates and quality of life after complete repair. For patients who were still awaiting complete repair, our evaluation was focused on pulmonary artery rehabilitation.

We considered our strategy as having failed for the following reasons: death, failure to achieve complete repair, if a Blalock–Taussig (BT) shunt procedure or surgical enlargement of the RVPA connection was necessary prior to complete repair.

Continuous data were tested for normality using the Kolmogorov–Smirnov test. Data were described as frequencies, medians with stated ranges or means with standard deviation, as appropriate. Comparisons were performed using the χ^2 or Fisher's exact tests for categorical variables and the Mann–Whitney U-test or Kruskal–Wallis test for continuous variables. Estimates of time-related survival and freedom from adverse events were calculated using the Kaplan–Meier method. All analyses were performed using SPSS for Windows version 17.0 (SPSS, Inc., Chicago, IL, USA).

RESULTS

Early results

Operative data. The median times for CPB and cardioplegic cardiac arrest were 64 min (range 26–188 min) and 25 min (range 13–76 min), respectively.

The distribution of the three different surgical procedures was: 43 patients (76%) underwent RVPA connection with patch enlargement; 9 patients (16%) had a PTFE conduit and 5 (9%) had a LAA interposition to establish the posterior aspect of the RVPA continuity. The pericardial patch technique for RVPA connection was used for all the patients with TOF ($P = 0.02$). Fifty-six percent of conduits was used for patients with PA/VSD type II, ($P = 0.03$) and 60% of the LAA technique was used for patients with PA/VSD type III ($P = 0.01$; Fig. 1C).

Hospital mortality. There were 2 operative deaths (3.5%), defined as death within 30 days of operation or during the same hospital admission. Early deaths occurred within the first 24 h. One patient had an uncommon morphological anatomy with a small RV and tricuspid valve that was essentially closer to PA with intact ventricular septum, though a small VSD was present. The other patient had a TOF with a Nakata index of 130 mm^2/m^2. He died 3 h postoperatively due to pulmonary overcirculation. The type of RVOT reconstruction (direct connection, LAA or PTFE conduit) was not significantly associated with hospital death. No predictive factors (weight, PA type, preoperative ventilation, size of preoperative pulmonary arteries and cross-clamp time) were identified for hospital mortality in the univariate model.

Early morbidity. Median duration of mechanical ventilation was 48 h (range 5–600 h) and median intensive care unit (ICU) length of stay was 4 days (range 1–35 days; Table 2). Inotropic support was used in all patients, at least until extubation.

Table 2: Operative and postoperative characteristics after RV to PA connection

Variable	PA/VSD I (n = 24)	PA/VSD II (n = 14)	PA/VSD III (n = 7)	TOF (n = 12)	Total (n = 57)
CPB duration (min)	69 [26–188]	83 [45–163]	77 [38–96]	54 [34–110]	64 [26–188]
Cross-clamping duration (min)	26 [13–57]	29 [16–76]	30 [18–32]	23 [14–59]	25 [13–76]
Postoperative characteristics					
Mechanical ventilation (h)	41 [17–465]	48 [12–312]	45 [19–96]	42 [5–288]	48 [5–600]
Length of ICU stay (days)	3 [1–35]	5 [2–18]	4 [3–14]	6 [1–16]	4 [1–35]
Arterial saturation (%)	90 [85–95]	95 [92–95]	94 [82.5–95]	92 [89–95]	92 [70–99]
Delayed sternal closure	4 (16.7)	3 (21.4)	1 (14.3)	8 (66.7)	16 (28)
Ventricular dysfunction	5 (20.8)	3 (21.4)	–	3 (25)	11 (19.3)
Excessive pulmonary blood flow	2 (8.3)	4 (28.6)	–	2 (16.7)	8 (14)
Restrictive pulmonary blood flow	3 (12.5)	1 (7.1)	1 (14.3)		5 (9)
BT shunt	3 (12.5)	1 (7.1)	–	–	4 (7)
Enlargement RV to PA	–	–	1 (14.3)	–	1 (2)
Sepsis	3 (12.5)	2 (14.3)	–	–	5 (8.8)
Prolonged pleuro-pericardic effusion	2 (8.3)	1 (7.1)	–	2 (16.7)	5 (8.8)
Phrenic paralysis	–	1 (7.1)	–	–	1 (1.8)
Kidney failure requiring dialysis	1 (4.2)	–	–	–	1 (1.7)
Hospital death	1 (4.2)	–	–	1 (8.3)	2 (3.5)

Data are shown as medians and range (minus, major) or as numbers and percentages.
CPB: cardiopulmonary bypass; ICU: intensive care unit; BT: Blalock–Taussig.

Ventricular dysfunction (right or left) occurred in 11 patients and the sternum was left open in 16 cases (28%) to allow for haemodynamic stability. In patients with delayed sternal closure, the sternum was closed in the ICU after a median duration of 1.5 days (range 1–3 days). Sixty-seven percent of patients with a TOF had delayed sternal closure ($P = 0.002$).

Pulmonary overcirculation was diagnosed in 8 patients (7 patch enlargement and 1 PTFE conduit) with no statistical correlation to initial surgical procedure. Diagnosis was based on poor peripheral tissue perfusion (metabolic acidosis, increase of lactate levels and oliguria). Only 1 of these patients needed reoperation to reduce the size of the RVPA connection. The remaining pulmonary over-circulation patients were medically treated (high haematocrit, low inhaled O_2). The median duration of mechanical ventilation in this subgroup was higher, 108 h (range 24–465 h) compared with other patients, 39 h (range 5–600 h) ($P = 0.03$). The median ICU stay was also higher in this former group, though not significantly different. The Nakata index was higher in comparison with the remaining patients with, respectively, 120 mm^2/m^2 (range 56–146 mm^2/m^2) vs 90 mm^2/m^2 (range 17–185 mm^2/m^2), though this difference was not statistically significant.

Excessive restrictive pulmonary blood flow (defined as failure or impossibility of extubation) led to immediate reoperations in 5 patients in whom 4 BT shunts and 1 enlargement of the RVPA connection were performed. Four of these 5 patients were operated on before 2005. The median duration of mechanical ventilation in these patients was 72 h (range 13–600 h) and median ICU stay 6 days (range 2–35 days). Durations of mechanical ventilation and ICU stay for this reoperated subgroup were not significantly different compared with the other patients. The median Nakata index was lower compared with the remaining non-reoperated patients: 87 mm^2/m (range 66–181 mm^2/m^2) vs 100 mm^2/m^2 (range 17–185 mm^2/m^2). The presence of unusual coronary artery anatomy was an incremental risk factor for early restrictive pulmonary blood flow ($P = 0.03$).

Late results

Of the 55 early survivors, 2 patients (3.5%) were lost to follow-up. The mean age at the last follow-up of the 53 remaining patients was 3 years (range 0.5–12 years).

Late survival. Four late deaths occurred after a median interval of 5 months (range 3–6 months).

Three patients died after palliation but before complete repair and 1 died early after complete repair.

During the interim period, 3 patients died suddenly at home. None of them had severe complications after initial palliation, or a prolonged ICU stay. For the first 5 months after initial palliation, haemodynamics and oxygen saturations were considered as appropriate. Only one post-mortem examination was conducted and rupture of an infundibular aneurysm was noted 6 months after palliation.

The patient who died after complete repair had a Di-George syndrome and situs inversus and was born with severe RV dilatation and LV compression. He died early at the age of 6 months and presented a biventricular dysfunction and severe pulmonary sepsis. The Di-George syndrome was an incremental risk factor for late and overall mortality ($P = 0.001$).

Reintervention for inadequate pulmonary blood flow. None of the patients who had excessive pulmonary blood flow

($n = 8$) were reoperated upon, though 1 of them had persistent overcirculation at 3 months of age. In this case, rehabilitation of pulmonary artery branches had been optimal and complete repair was performed immediately.

Two patients with secondary severe low pulmonary blood flow had late enlargement of the RVPA connection at the ages of 8 and 16 months, respectively (the pulmonary arterial tree had been unsuitable for complete repair). Another patient underwent percutaneous angioplasty with stenting of the RVPA connection at 5 months postoperatively.

Finally, 8 patients benefited from reintervention to increase pulmonary blood flow at some time between palliation and complete repair (5 early and 3 late reinterventions). Incremental risk factors for presenting with restrictive pulmonary blood flow in the period between initial palliation and complete repair were: presence of genetic anomaly ($P = 0.05$); need for early postoperative high frequency oscillated ventilation ($P = 0.03$); need for peritoneal dialysis ($P = 0.01$).

Of the 7 patients with PA/VSD type III, only 1 had undergone unifocalization of major aortopulmonary collaterals (MAPCAs) at the age of 24 months. Two patients had embolization of communicating MAPCAs for excessive pulmonary blood flow at 10 and 24 months.

Reintervention for disharmonious pulmonary arteries growth. Three patients underwent isolated reintervention for stenosis of the left pulmonary artery when complete repair was not feasible due to poor pulmonary artery growth. Two patients had percutaneous angioplasty with stenting and one, a surgical plasty. The use of LAA for RV to PA connection was the only incremental risk factor of left pulmonary stenosis (PS) ($P = 0.02$).

Infundibular aneurysm. Infundibular aneurysm was diagnosed in 6 patients (3 enlargements, 2 PTFE conduit and 1 LAA interposition) at a median age of 3 months range (1–7 months). False aneurysms developed on the left side of the infundibular patch or conduit, on the suture line itself. One patient died due to this complication (described above). Two patients underwent emergency reoperation 1 month after initial palliation. The other patients were kept under surveillance until time of complete repair since the false aneurysm was considered small. In the false aneurysm group, the median Nakata index was lower than in the remaining patients: 69 mm^2/m^2 (range 17–155 mm^2/m^2) vs 100 mm^2/m^2 (range 49–185 mm^2/m^2), respectively; $P = \text{NS}$. No incremental risk factor (including type of surgical procedure) for infundibular aneurysm was identified though all patients had PA.

Overall reinterventions after initial palliation. Late complications and reinterventions are shown according to the diagnostic group in Table 3. Freedom from reintervention before complete repair was $78 \pm 6\%$ and $73 \pm 7\%$ at 6 and 9 months, respectively (Fig. 2). There was no significant difference comparing patients with a PTFE conduit for RVOT reconstruction vs those in whom only autologous tissues had been used.

Complete repair: success or failure. Two patients (3.5%) were lost to follow-up. Figure 3 provides an overview of the patient population. Among the entire population, 7 patients were considered as clinical failures: 6 deaths (2 early, 3 late, 1 early after complete repair) and 1 complete repair failure. Forty-seven patients (85%) underwent complete repair; 1 of these was a failure. At the last follow-up, 1 patient had died late after complete repair; 48 patients were alive and well, with 3 of these due to undergo complete repair soon.

Table 3: Complication and reintervention between the palliation procedures and the complete repair in survivor

Late complication/reintervention	PA/VSD I (n = 22)	PA/VSD II (n = 13)	PA/VSD III (n = 7)	TOF (n = 11)	Overall (n = 53)
Over pulmonary circulation	–	1 (8)	–	–	1 (2)
Restrictive pulmonary flow	–	2 (15)	1 (14)	–	3 (6)
Disharmonious PA	1 (5)	1 (8)	1 (14)	–	3 (6)
Infundibular aneurysmal	1 (5)	3 (23)	2 (29)	–	6 (11)
MAPCAs unifocalization	–	–	1 (14)	–	1 (2)
MAPCAs embolization	–	–	2 (28)	–	2 (4)

Data are shown as medians and range (minus, major) or as numbers and percentages.
PA: pulmonary artery; MAPCAs: major aortopulmonary collaterals.
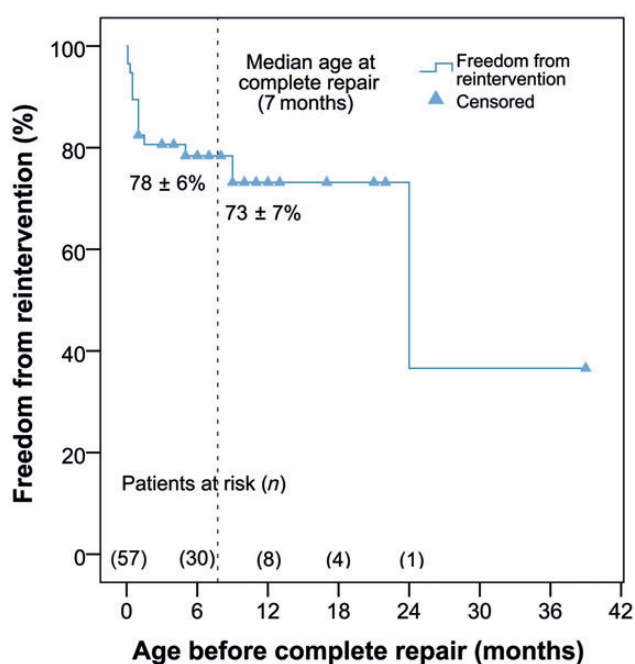
Figure 2: Actuarial freedom from reintervention before complete repair.

The 47 of 55 (85%) survivors who underwent complete repair (performed because of moderate or severe cyanosis, aneurysmal infundibular patch or pulmonary arteries stenosis) did so provided that the native pulmonary arteries had achieved an optimal size (subjective assessment by the surgical team or objective measurement of pulmonary artery size in 27 patients after percutaneous catheterization).

The Nakata index could be evaluated retrospectively for 19 patients, with a median of 233 mm^2/m^2 (range 102–891 mm^2/m^2). Median age at complete repair was 7 months (range 1–49 months).

In 37 patients, complete repair was performed as the first subsequent step after the initial palliation by RVPA connection (Fig. 3). Thirty-six of the 37 were alive following successful VSD closure without previous reintervention for excessive or restrictive pulmonary blood flow during the interim period. This subgroup represented an ideal outcome of this strategy and was made up of 9 TOF (75% of palliated TOF patients) and 28 PA/VSD (62% of palliated PA/VSD patients) patients. If we now consider the type of initial palliation for this success group, 28 patients (out of the 43, 65%) had a patch enlargement, 4 patients had a PTFE conduit (out of 9, 44%) and 5 (out of 5, 100%) benefited from a LAA

reconstruction. In this small series, there was no statistical difference for complete repair without intermediate reintervention, between TOF and PA/VSD or the types of surgical RV to PA connection.

In the 10 other patients, augmentation or restriction of pulmonary blood flow was performed during the interim period between RVPA connection and complete repair as described above and comprised: additional BT shunt (n = 4); enlargement of RVPA conduit (n = 4); unifocalization of MAPCA (n = 1); surgical restrictive RVPA conduit (n = 1); embolization of MAPCA (n = 1).

The reconstruction of the RVOT at the time of complete repair was carried out using a prosthetic valved conduit in 13 patients (28%), all were PA/VSD. Only 3 of 27 patients (11%) had a prosthetic conduit since 2006 at complete repair.

The 5 patients who had interposition of LAA at initial palliation all underwent subsequent complete repair. Two of these patients needed the use of a valved conduit at repair; 1 had developed a large aneurysm at the suture line of a PTFE conduit that had been inserted prior to definitive repair to enlarge severely hypoplastic pulmonary arteries. The other patient had been operated on at the time of complete repair by another team with a different philosophy, and a prosthetic conduit had been implanted.

Associated procedures were required at repair in 23 patients (left pulmonary angioplasty in 19; right pulmonary angioplasty in 5 and reconstruction of the pulmonary confluence in 5). No patients needed a new unifocalization or ligation of MAPCA.

Operative and postoperative data by diagnostic group are given in Table 4.

At complete repair, incremental risk factors for valved-conduit implantation instead of autologous tissue repair were: PA/VSD II (P = 0.01); coronary artery anomalies (P = 0.004); patients with PTFE conduit at initial palliation (P = 0.001); reintervention for restrictive pulmonary blood flow during the interim period (P = 0.02).

Two patients had failure of complete repair. One patient died early at the age of 5 months (PA/VSD II) with severe chronic pulmonary sepsis and ventricular dysfunction (described above). The other patient, at the age of 2 months, with PA/VSD type II (neonatal Nakata index 56 mm^2/m^2) had been operated on in an emergency for rupture of an infundibular aneurysm. Closure of the VSD was done at the same time as RVOT reconstruction. Severe RV dysfunction occurred in the operating theatre with a RV/LV pressure ratio of almost one; fenestration of the VSD patch was performed in this case.

Early complications after complete repair occurred in 9 patients (including one patient who died following failure of complete
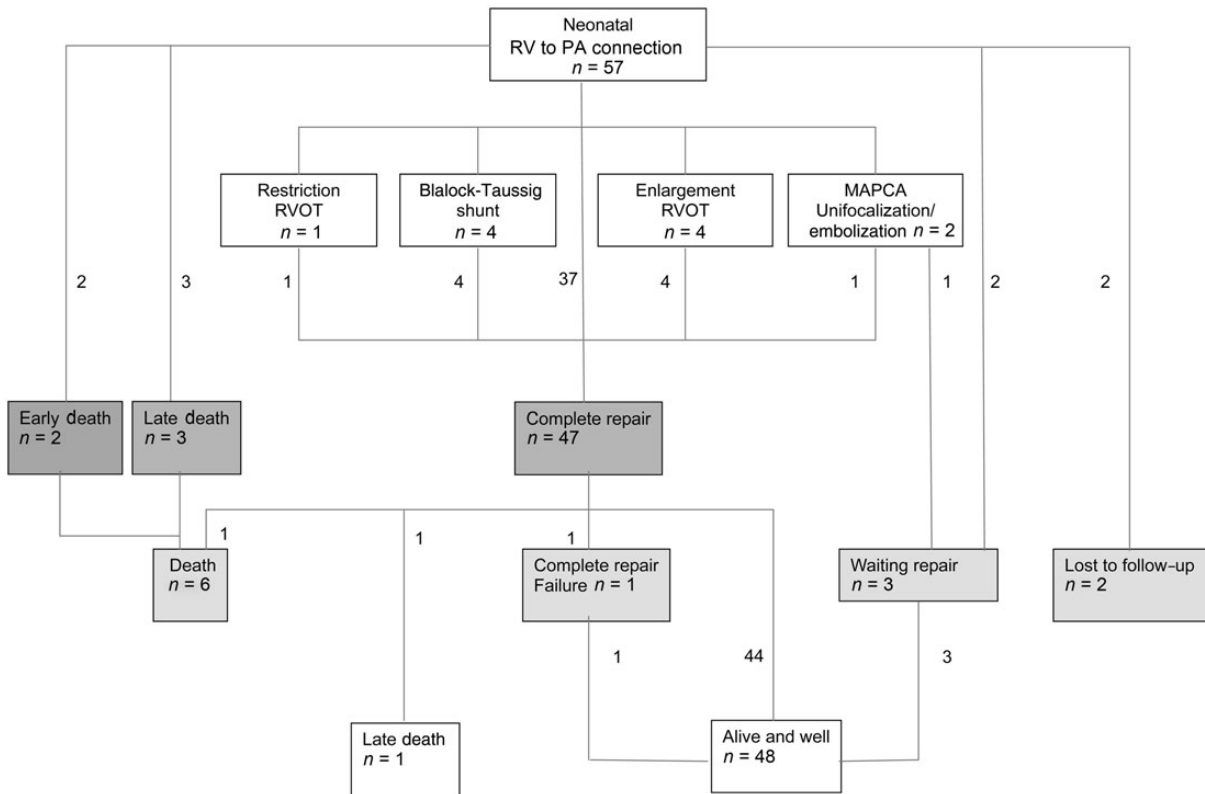
Figure 3: Overview of the study population who underwent the neonatal RV to PA connection.

Table 4: Intraoperative and postoperative characteristics after the complete repair (n = 47)

Variable	PA/VSD I (n = 19)	PA/VSD II (n = 12)	PA/VSD III (n = 6)	TOF (n = 10)	Total (n = 47)
Age	7 [1–53]	7.5 [3–36]	13.5 [5–49]	6 [4–21]	7 [1–53]
Nakata index (n = 20) (mm^2/m^2)	211 [113–388]	194 [102–891]	375 [320–430]	221 [116–289]	221 [102–891]
Right pulmonary size (n = 20) (mm)	6 [4–8.5]	6.5 [4–12]	8.5 [8–9]	7 [5–8]	7 [5–8]
Left pulmonary size (n = 20) (mm)	5 [4–8]	7 [3–13]	8.5 [8–9]	7 [3–8]	7 [5–8]
Prosthetic conduit valve	4 (21)	7 (58)	2 (33)	–	13 (28)
Enlargement of RVOT	15 (79)	5 (42)	4 (67)	10 (100)	34 (72)
With monocuspid valve	3 (16)	2 (17)	–	–	5 (11)
Left pulmonary angioplasty	9 (47)	5 (42)	1 (17)	3 (30)	18 (38)
Right pulmonary angioplasty	1 (6)	1 (8)	2 (33)	1 (10)	5 (11)
Reconstruction pulmonary confluence	–	2 (17)	3 (50)	–	5 (11)
CPB duration (min)	149 [117–315]	193 [118–249]	144 [100–210]	113 [85–124]	137 [85–315]
Cross-clamping duration (min)	97 [54–119]	92 [69–137]	96 [56–111]	61 [49–74]	88 [49–137]
Postoperative characteristics					
Early death	–	1 (8)	–	–	1 (2)
Failed closing VSD	–	1 (8)	–	–	1 (2)
Complications					
RV dysfunction	1 (5)	–	–	–	1 (2)
Residual VSD	1 (5)	1 (8)	–	–	2 (4)
Sepsis	1 (5)	2 (16)	–	–	3 (6)
Phrenic paralysis	–	–	–	1 (10)	1 (2)

Data are shown as medians and range minus, maximal or as numbers and percentages.
RVOT: right ventricular outflow tract; CPB: cardiopulmonary bypass; VSD: ventricular septal defect; RV: right ventricle.

repair). These complications were as follows (Table 4): right ventricular dysfunction (n = 1); residual VSD (n = 2); sepsis (n = 3); phrenic paralysis (n = 1); severe ventilatory disorder due to bronchial compression with subsequent death (n = 1) and opening of VSD required (n = 1).

At last follow-up. At late follow-up, there were 48 patients alive and well and 1 late death (Fig. 3). Of those surviving, 45 had undergone successful complete repair; 1 patient had a failed complete repair and 3 were still awaiting complete repair with a satisfactory pulmonary tree growth at last evaluation.

For the 45 survivors following complete repair, the median follow-up was 2.6 years (range 0.5–11 years). During this period, 1 patient with the Di-George syndrome died with respiratory distress in the context of pulmonary fibrosis 6 months after complete repair.

The actuarial survival for the overall population was 86.5% at 3 years (Fig. 4A).

Five patients (11%) had been reoperated on at a median of 30 months after complete repair (range 6–55 months). Three patients (7%) underwent a conduit replacement with pulmonary angioplasty due to stenosis at a median of 52 months (range 30–55 months), with a prosthetic graft implanted at the time of complete repair. One of these patients underwent a further conduit replacement 7 years later. Two patients (4%) with a pericardial patch enlargement had a percutaneous angioplasty, 1 at the site of RV to PA connection and 1 for the left pulmonary artery.

Figure 4B shows the reintervention rates for patients following complete repair according to whether they had received prosthetic conduits or only patch enlargement. For the survivors, clinical status was excellent and quality of life was deemed normal. Systolic RV pressures were available for 25 patients. Twenty-one patients had a pressure lower 40 mmHg. For the 4 other patients, the RV pressure was higher. One of these patients had a moderate stenosis of the prosthetic graft conduit, and 2 had a moderate residual VSD.
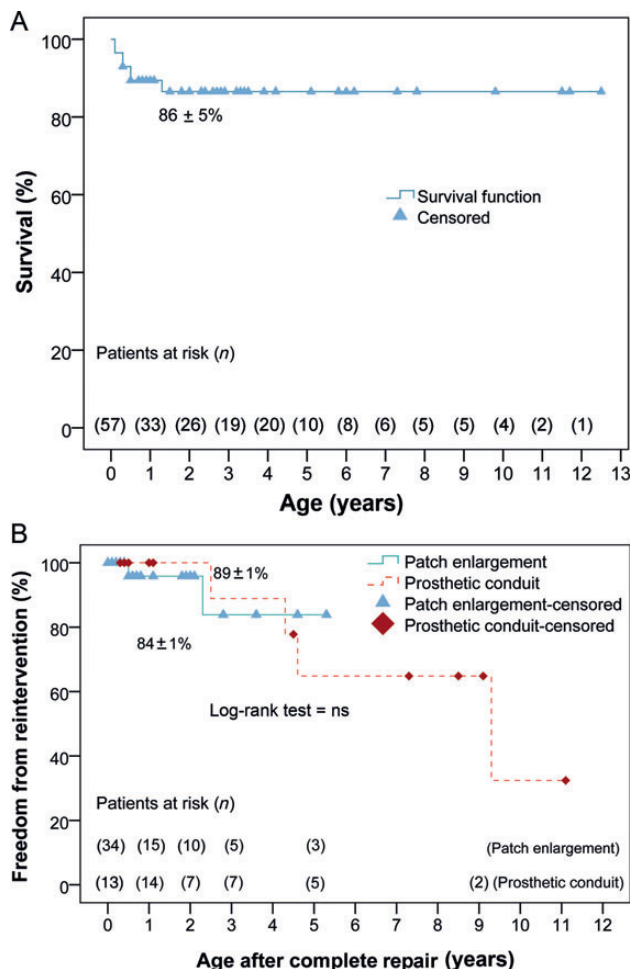

Figure 4: (**A**) Actuarial survival for the overall population at the last follow-up. (**B**) Actuarial freedom from reintervention for patients following complete repair according to whether they had received prosthetic conduit or only patch enlargement.

DISCUSSION

Taking into account all the considerations of the present series of cases, one should emphasize that our neonatal population represents the most vulnerable group of symptomatic patients with TOF or PA/VSD that must be dealt with. The management strategy that our group adopted, based on previous studies [1, 10–12, 17], was to undertake a neonatal RVPA connection in order to achieve cardiopulmonary stability, improvement in O_2 saturation or preservation of pulmonary blood flow (for ductus-dependant circulation). In addition, we aimed to optimize pulmonary artery growth and allow for MAPCAs involution, particularly in patients who would otherwise be considered too high risk for early complete repair [5, 14, 18, 19].

Evaluation of the suitability of this strategy has to be made by taking into account the potential morbidity and mortality at any given stage as well as the adequate growth of pulmonary arteries. One should further aim to avoid iterative palliative surgery and should also consider the expected rate of successful complete repair and any potential need for a valved conduit.

Mortality

The early mortality was low (3.5%) in our series of neonates which comprised mainly patients with symptomatic neonatal TOF (though one patient had an anatomy closer to PA with intact ventricular septum). In comparison, Kim *et al.* [20] reported the outcome for initial modified BT shunt palliation in TOF/PA and a ductus-dependent pulmonary circulation with 22% overall mortality. We are now particularly aware of the incidence of pulmonary overcirculation that can lead to acute LV dilatation, bradycardia and cardiac arrest. This must be the main concern in the first postoperative hours, particularly for adequately sized pulmonary artery beds. Nevertheless, we really think that our current strategy might reduce the early mortality when compared with neonatal BT shunts. These impressions seem confirmed by various series analysing the mortality after systemic to pulmonary artery shunt intervention in neonates with miscellaneous congenital heart diseases. The early mortality after systemic to pulmonary artery shunt was more than 8% [21, 22].

This may be at least in part due to the avoidance of shunt physiology (diastolic run-off, impaired coronary perfusion and left heart volume overload). These considerations have some parallels with the arguments for using Sano conduits vs BT shunts in Norwood patients [23]. This is in spite of the potential advantages of the BT shunt including avoidance of CPB, cross-clamp and ventriculotomy. Hence, BT shunts are extremely rare nowadays at our institution.

Pulmonary overcirculation

Eight patients presented with excessive pulmonary blood flow in the postoperative period. Their Nakata index was higher in comparison with the remaining patients, and the morbidity was higher (ventilation time and ICU stay). One patient needed to be reoperated on and 1 died before redo surgery. As a result, we now consider potential reduction of the RVPA connection when signs of systemic malperfusion appear, particularly for TOF. Flow-calibration of an excessively large RVPA connection is easy to perform using a haemostatic clip or a *U*-stitch. In cases with

pulmonary arterial trees of the largest capacity, the distal PA pressure can be measured by direct puncture, ideally aiming to achieve 1:2 for the systolic pulmonary artery:systemic pressure ratio. Further, based on our experience with infants, we think that early postoperative MAPCA catheter embolization may be recommended.

It should be noted, however, that the systemic arterial oxygen saturation is often not a good indicator of pulmonary blood flow as this may reach 100% even without pulmonary overcirculation if the RVPA restriction does not lead to a right-left shunt at VSD level. Following these guidelines in patients with TOF and PA with the highest Nakata indices should help reduce the incidence of pulmonary overcirculation and the need for delayed sternal closure.

Minimizing the infundibulotomy and muscular band resection is probably another key point in improving the tolerance to possible overcirculation in TOF patients.

Inter-stage mortality and ventricular aneurysm

The inter-stage mortality (all before the sixth postoperative month) is a matter of great concern, with 1 death due to rupture of a ventricular aneurysm and 2 sudden deaths that might have had the same underlying cause, as described above [14]. Six other patients developed a ventricular aneurysm at the left margin of the infundibular patch at earlier time-points up to 7 months (median 3 months), 2 of whom needed urgent reoperation. All were diagnosed with PA and a median Nakata index of 69 m^2/m^2, which was lower than the rest of the population. Our hypothesis is that, in this specific subset of patients with severe PA, the distal infundibulum was under-developed, resulting in a short distance between the aorta and the LAD. The left margin of the ventriculotomy patch might therefore have been susceptible to progressive rupture in the zone of fragile muscle. In these cases, particularly in low-weight babies, we now prefer to use treated autologous pericardial patches that are softer and easier to suture without tearing of the muscular margin. In all cases, we recommend a strict 1 month-interval follow-up to detect any ventricular aneurysm by echocardiography; chest X-ray, CT or MRI may further be performed to confirm the diagnosis.

Early excessively restrictive pulmonary blood flow

Five patients needed early reoperations for excessively restrictive pulmonary blood flow, all except one prior to 2005. Four BT shunts and one redo RVPA connection were performed. Three underlying aetiologies were identified in these cases: the creation of an inadequately sized surgical RVPA connection in cases with a low Nakata index; residual stenosis of branch pulmonary arteries; and finally, the presence of a coronary anomaly.

These patients had prolonged ventilation times and ICU length of stay, though they were not statistically significant. It is likely that most, if not all, of these patients could have avoided early reoperation had the initial palliative surgery been more effectively performed. We believe that the initial palliative procedure is a critical one for these patients and that this should be performed by senior surgeons appropriately trained in this technique. One might consider percutaneous stenting of the RVPA connection as an alternative to redo surgery. In practice, however, we found that later repair without a conduit was more difficult to achieve

due to extensive stent-induced fibrosis in the autologous posterior tissue wall.

PTFE conduit at initial palliation

Sixteen percent of palliative surgery required the use of a 5–6-mm PTFE conduit, essentially for PA/VSD type II and in cases with anomalous coronary artery. The need for PTFE conduits has dramatically decreased over the last 5 years (data not shown), mainly because all surgeons are now adequately trained in the use of autologous tissue for reconstruction of the RVOT in these instances (LAA reconstructive technique), including for cases of persistent truncus arteriosus. We do think that it is possible to avoid the use of conduits at initial palliation in all cases, except when a major coronary artery is crossing the infundibulum. Avoidance of conduit palliation is a key point (even if not correlated with a higher risk of inter-stage reintervention) to avoid valved conduits at the time of complete repair and potential fibro-intimal stenosis during the inter-stage period, something that we did not encounter in this small group of patients.

Anomalous coronary arteries

Origin of the LAD from the right coronary ostia or from the RCA with a subsequent course crossing the infundibulum is not rare in TOF or PA/VSD. In this series, this situation resulted in use of more conduits at palliation and more early reinterventions for excessively restrictive pulmonary blood flow. These results have influenced our initial strategy and we now carefully consider the option of a modified BT shunt in this particular anatomy.

Autologous tissue reconstruction

Avoidance of a valved conduit at surgical repair is an important strategy. This has been the philosophy in our unit for many different congenital heart defects including: Fallot with anomalous coronary anatomy, PA/VSD, transposition of the great vessels with PS, double discordance with PS or PA and truncus arteriosus. In our present series of neonatal patients, the LAA posterior wall reconstructive method was necessary in only 5 cases. Some additional 'palliative conduit' patients could have benefited from this technique (see above: PTFE conduit at palliation). Although it is more challenging with a higher risk of left pulmonary artery stenosis, it may help avoid the use of a valved conduit at subsequent repair, and additional reoperations; this has been the case in 2 of 5 patients (1 patient was reoperated upon at another centre).

Despite the increased technical difficulties and based on our larger experience of RVPA connection for older patients (1 month to >6 months), we are convinced that this autologous tissue strategy results in the use of fewer valved conduits at reoperation and fewer conduit exchanges in the long term, as we have already demonstrated in patients with truncus arteriosus [24].

Late reinterventions

Persistent pulmonary overcirculation was not a cause for late reoperation. In 8 cases of pulmonary overcirculation, the rehabilitation of pulmonary artery branches was optimal and complete

repair was performed. No persistent pulmonary hypertension was seen after repair at 5, 8 and 16 months. On the other hand, excessively restrictive pulmonary blood flow was particularly challenging, both in the immediate postoperative period as well as at the mid-term follow-up. Three late reinterventions were performed to increase pulmonary blood flow (two enlargements of an RVPA connection and one stenting).

Overall, severe pulmonary overcirculation was an important cause of morbidity in the first 24 postoperative hours. The real problem we had to face in this series, however, was the occurrence of excessive restriction of pulmonary blood flow by various mechanisms. These included: over-restrictive RVPA connection; persistent muscular obstruction at infundibular level; distortion of the pulmonary bifurcation and pulmonary branch stenosis. Measuring the systolic pulmonary artery pressure in the distal main PA reconstruction (which should be up to 30–40% of the systolic arterial pressure) and evaluating the harmonious anatomy of the PA pulmonary artery bifurcation should, however, help prevent most of these over-restrictive pulmonary blood flow situations. In cases of very late reoperation for restrictive flow, it is usually not an initial technical failure but a situation where the pulmonary artery growth was not sufficient to allow complete repair due to progressive relative stenosis of the initial RVPA connection as a result of somatic growth.

MAPCAs management

The management of MAPCAs comprised: avoidance of neonatal unifocalization; early catheter embolization of 'communicating MAPCAs' if pulmonary overcirculation is noted; and finally, unifocalization of only the few, most significant MAPCAs at the time of repair. In our study population, many MAPCAs were naturally small or stenotic, something that can probably be expected in this symptomatic neonatal population because in case of well-developed MAPCAs, patients do not require an early pulmonary blood increase. On the contrary, PA/VSD patients (with no ductal dependence) with a good initial saturation have probably larger MAPCAs that might need unifocalization later, but do not require neonatal surgery. If significant haemodynamic MAPCAs were present at surgery, CPB was conducted on moderate hypothermia with an oversized vent placed in the left atrium (14–16 mm venous cannulae) to control excessive pulmonary venous return. We believe, however, that antegrade pulmonary blood flow via a RVPA connection can result in restriction of blood flow within MAPCAs at mid-term, as described previously by others [8, 12].

Pulmonary artery growth

As mentioned, the other teams demonstrated that a palliative RVPA connection permits one to achieve repair with adequate growth and recruitment of segmental pulmonary arterial branches [10–12]. In our present series, the mean Nakata index increased to 230 mm^2/m^2 at the time of repair within a limited inter-stage period (mean 7 months). Finally, we achieved successful biventricular repair in 85% of patients surviving the neonatal palliative procedure. Our data suggest that pulmonary rehabilitation is possible after a short time of additional pulmonary blood flow established by a RVPA connection with a low mortality at subsequent repair.

Study limitations

The retrospective nature of the study without a control group is a limitation. The population was small and not necessarily representative of the majority of patients with this pathology. Our narrow inclusion and exclusion criteria may have resulted in a large number of patients being excluded. Finally, the statistical results should be interpreted with caution, as potential bias may have been introduced by these stringent selection criteria.

CONCLUSIONS

Our study has demonstrated that excellent early survival may be achieved in symptomatic neonatal patients with TOF/PA using the RVPA connection as an initial palliative strategy. Even though our patients exhibited a not-significant morbidity, they had a more stable early postoperative course and haemodynamic than one might expect in a similar population palliated by means of BT shunts. Our technique facilitates pulmonary artery rehabilitation and, importantly, we might limit the need for conduits at subsequent repair by primarily using autologous tissue-based reconstructive techniques. Further long-term follow-up will be needed to better clarify the late results of this strategy.

Conflict of interest: none declared.

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APPENDIX A. CONFERENCE DISCUSSION

Dr F. Fynn-Thompson (*Boston, MA, USA*): Our general philosophy at our centre in Boston regarding neonates with tetralogy of Fallot and pulmonary atresia, with confluent native branch pulmonary arteries, is quite similar to yours in that we attempt to establish early RV-to-PA continuity as the first step of a staged approach. We routinely perform a preoperative cath on most of our patients, and then use this initial roadmap to tailor our surgical approach based on what that shows.

In those patients with a main pulmonary artery segment, we also try to establish RV-to-PA continuity with autologous tissue much in the same way that you have described. However, in those with no discernible MPA but still with confluent branch pulmonary arteries, we believe that using a valved homograft conduit is preferable because it avoids possible distortion and undue tension on the confluence of the branch pulmonary arteries.

In reading your paper, I was a bit confused about your exact preoperative workup, so this brings me to my first question, which is to ask, how are you evaluating your patients preoperatively? And how is that information used to make decisions about your specific surgical approach?

Dr Gerelli: Excuse me, can you repeat the question please?

Dr Fynn-Thompson: How do you work up your patients? Do you cath every patient? Do you go by echo? How do you make decisions about which of those three surgical techniques to use?

Dr Gerelli: Yes. The surgical techniques have evolved over time. Actually, we prefer to use only reconstruction of right ventricular outflow tract with autologous tissue and just use prosthetic conduit for severe anomalies of the coronary artery.

Dr P. Vouhé (*Paris, France*): Excuse me, may I answer the question? All patients had cardiac catheterization.

Dr F. Fynn-Thompson: Okay. Very good. Then my follow-up question is with regard to your high incidence of infundibular aneurysms. From my calculation, this was about 15%, so 7 out of your 47 patients had an infundibular aneurysm, and a large part of your interstage mortality was due to rupture of these aneurysms. I believe this to be a function of the excessive tension on RV-PA connection.

So my question to you is, based on your retrospective analysis and your higher than usual incidence of these aneurysms, will you use this data to reconsider your approach in some patients, especially those with the smallest branch pulmonary arteries, and will you alter your surgical technique based on that?

Dr Gerelli: For the aneurysms, the problem was the severe pulmonary artery under-development, with a Nakata index of 69 mm²/m² in our series. An under-developed infundibulum with a short distance between the aorta and left coronary artery causes a weak zone around the left margin of the ventriculotomy patch. Sewing the patch to the left margin was very difficult, and actually, we first run the suture line at this left margin of the patch.

Dr V. Tsang (*London, UK*): I just want to make a comment about RV-to-PA connection at the time of palliation or at the time of repair. This is a personal experience.

As we know, the proximity of the left main coronary artery just behind the atretic segment of the PA can be a problem. Despite the best intentions with the patching, there is a small risk of distorting the left main coronary artery. You do not have to occlude it. You only have to distort it.

And if you see in LV function a loss of AV synchrony, look at your left main coronary immediately. You may have to do something about it quick. That is all I want to say.