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Outcomes of palliative right ventricle to pulmonary artery connection for pulmonary atresia with ventricular septal defect†

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Abstract

OBJECTIVES: To determine the early, intermediate and long-term outcomes of pulmonary atresia with ventricular septal defect (PA/VSD) Types I, II and III initially palliated by a right ventricle to pulmonary artery (RVPA) connection.

METHODS: We performed a retrospective study from 2000 to 2014 that included 109 patients with PA/VSD who had undergone an RVPA connection (tetralogy of Fallot and PA/VSD Type IV excluded). The end-points of this strategy were adequate pulmonary artery tree post-palliation, second palliation, biventricular repair, right ventricular pressure post-biventricular repair and late reoperation. Mean follow-up was 5.4 years (1 day to 14–78 years).

RESULTS: Early mortality after an RVPA connection was 2.7% (3 of 109). The interstage mortality rate was 6.6% (7 of 106). Eighty-four (77%) patients had a biventricular repair and 8 patients (7%) are awaiting repair. Overall survival was 90% at 1 year and 81% at 10 years. The RVPA connection allowed significant growth of the native pulmonary artery with a Nakata index of 101 mm^2/m^2 before the RVPA connection and 274 mm^2/m^2 after ($P = 0.001$). Twenty-nine reinterventions for restrictive pulmonary blood flow have been done (9 before 2 months and 20 after 2 months). Of the 84 patients who had a repair, 22 patients (26%) initially had a right ventricular pressure greater than 40 mmHg. Twenty-eight patients (33%) required late reoperation.

CONCLUSIONS: Hospital deaths after the RVPA connection were low. The procedure allowed good growth of the native pulmonary artery. Biventricular repair was possible in a large number of cases. The late morbidity rate remains significant. Early reinterventions could be avoided by appropriate calibration. This technique appears to be suitable for any type of PA/VSD with central pulmonary arteries.

Keywords: Pulmonary atresia with ventricular septal defect • Major aortopulmonary collaterals • Palliative surgery • RVPA connection

INTRODUCTION

Pulmonary atresia with ventricular septal defect (PA/VSD) is an uncommon congenital heart condition. The anatomy of the pulmonary artery (PA) is complex and heterogeneous. Surgical management differs according to the specific type of PA/VSD [1, 2]. To close the ventricular septal defect (VSD), one must have a non-restrictive pulmonary tree and no pulmonary hypertension [3]. To increase the size of the PA, one needs to create an additional source of pulmonary blood flow. The ideal palliative procedure to increase the PA tree and maintain a low early mortality rate is still under debate. Construction of a systemic pulmonary artery shunt is one alternative. Either a modified Blalock–Taussig (MBT) shunt or a central shunt between the ascending aorta and the pulmonary arterial confluence may be performed, though perioperative mortality, in particular for the MBT, remains high [4, 5].

In contrast, construction of a right ventricle to pulmonary artery (RVPA) connection allows for an increase in the pulmonary blood flow without pulmonary hypertension restores an anterograde flow with a route for subsequent catheterization and improves survival due to a lower incidence of obstruction or thrombosis. Where possible, we create a posterior autologous RVPA connection. We believe that the flow this connection provides competes with that derived from the major aortopulmonary collaterals, with a resultant regression of major aortopulmonary collaterals and growth of a PA tree adequate for biventricular repair.

The objective of this study was to evaluate the outcomes of a surgical RVPA connection in a single institution regarding operative mortality and late survival, morbidity, PA tree growth, eligibility for subsequent biventricular repair and right ventricular pressure following biventricular repair.

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MATERIALS AND METHODS

Patient population

From January 2000 to February 2014, 109 consecutive patients diagnosed with PA/VSD underwent a right ventricle to pulmonary artery (RVPA) connection at Necker Sick Children Hospital (Paris). Thirty-three patients, 41 patients and 35 patients had, respectively, Type I, II or III PA/VSD according to the Castaneda classification. The preoperative patient characteristics are given in Table 1.

Exclusion criteria were PA/VSD Type IV of the Castaneda classification and any patients whose initial palliative procedure was not an RVPA connection (e.g. MBT shunts, central systemic pulmonary shunts or stented arterial ducts). Patients with tetralogy of Fallot, defined as any right ventricle outflow tract opening, were not included in this study; patients with PA/VSD and a univentricular heart, atrioventricular and ventriculo-arterial discordance or an atrioventricular septal defect were similarly excluded.

The health database records were reviewed in a retrospective study. Permission to undertake this study was obtained from the Paris V University ethics committee. The need for individual consent was waived.

Evaluation of the pulmonary anatomy was based on angiographic measurements. If these data were not available, data from echocardiographic or computed tomographic scans were analysed. Right and left PA diameters were measured and the Nakata index [6] was calculated. This analysis was performed before any palliative procedure and just before complete repair evaluation. ΔNakata was defined as the difference between the Nakata index prior to biventricular repair and before the RVPA connection. We defined an index of growth, which is the ratio between ΔNakata and the time between the 2 measurements (index of growth = ΔNakata index/Δtime).

The right ventricular pressure was obtained intraoperatively by direct RV puncture or by initial echocardiographic assessment in the intensive care unit (ICU) based on the tricuspid regurgitation velocity. We defined patients as having high or low RV pressure using a threshold of 40 mmHg and/or an RV pressure/systemic pressure ratio of 0.45.

Surgical technique for right ventricle to pulmonary artery connection

This surgical technique (technical points, safeguards and pitfalls) has been extensively described previously [7], including by our group.

Briefly, patients were operated on via a median sternotomy and under normothermic cardiopulmonary bypass (CPB). The heart was arrested with anterograde warm blood cardioplegia. The PA and their branches were extensively mobilized. Based on anatomical configuration and the preference of the surgeon, 3 different techniques were used and are briefly described below.

Direct posterior right ventricle to pulmonary artery autologous connection with anterior heterologous patch

This technique was used for PA/VSD Types I and III and comprised a vertical incision of the main PA in continuity with a limited infundibular incision, together with limited resection of the right ventricular outflow tract muscle bundles. The anterior wall was reconstructed using a heterologous pericardial patch, initially sized around a Hegar dilator and according to the patient's weight. Our strategy was initially to perform a large RVPA connection rather than a restrictive one because it was extremely easy to reduce the caliber.

Left atrial appendage

In the absence of the pulmonary trunk (PA/VSD Types II and III), the pulmonary outflow tract was reconstructed using the left atrial appendage [8]. 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 right ventricular outflow tract was thus made of autologous vascularized tissue. The anterior wall was reconstructed with a heterologous pericardial patch.

Table 1: Demographic data and morphological features before performing the right ventricle to pulmonary artery connection

Characteristics	PA/VSD Type I ($n = 33$)	PA/VSD Type II ($n = 41$)	PA/VSD Type III ($n = 35$)	Total ($n = 109$)	P-value
Sex, female/male	16/17	22/19	14/21	52/57	0.49
Mean age (days)	70 (213)	79 (251)	446 (874)	194 (555)	0.004
Mean weight (kg)	3.19 (1.5)	3.5 (1.75)	7.29 (6.8)	4.6 (4.46)	<0.001
Mean oxygen saturation	80 (12)	73 (16)	77 (14)	77 (14)	0.14
Cardiac or non-cardiac anomalies					
Chromosome 22q11 deletion	2 (6%)	10 (24%)	9 (25%)	21 (19%)	0.084
Right aortic arch	5 (15%)	10 (24%)	16 (45%)	31 (28%)	0.016
Bilateral superior vena cava	2 (6%)	3 (7%)	3 (8%)	8 (7%)	0.92
Coronary anomalies	10 (30%)	26 (63%)	2 (6%)	38 (35%)	<0.001
Anatomy					
Duct dependent	33 (100%)	40 (97.5%)	5 (14%)	78 (71%)	0.000
Mean diameter of the right PA (mm)	4 (0.85)	4 (0.81)	3.3 (1.6)	3.8 (1.19)	0.012
Mean diameter of the left PA (mm)	3.8 (0.89)	3.95 (0,95)	3.3 (1.6)	3.7 (1.2)	0.065
Mean Nakata index (mm^2/m^2)	120 (36)	123 (44)	58 (47)	101 (52)	<0.001

PA/VSD: pulmonary atresia with ventricular septal defect; PA: pulmonary artery.

Polytetrafluoroethylene conduit

The third reconstruction technique used a polytetrafluoroethylene tube graft to connect the RV to the pulmonary bifurcation. This procedure 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 left atrial appendage reconstructive method in case of PA/VSD with no pulmonary main trunk.

Strategy for major aortopulmonary collaterals

We feel that it is mandatory to create a complete diagram of the major aortopulmonary collaterals. It is achieved by taking an initial computed tomography scan followed by angiography. This diagram allows one to determinate the following parameters for each major aortopulmonary collateral and pulmonary segment perfusion:

a. The origin and 3D anatomy.
b. The closest distance between the major aortopulmonary collateral and the intrapericardial ipsilateral PA.
c. The size ratio between the major aortopulmonary collateral and the native PA.
d. The presence of a retrotracheal or a retro-oesophageal course.
e. A communication between the major aortopulmonary collateral and the true PA system and, if present, the relative size of the communication compared with the diameter of the largest major aortopulmonary collateral.
f. The number and localization of bronchopulmonary segments without any vascularization or with fixed pulmonary hypertension.

The general strategy for dealing with the major aortopulmonary collaterals was to avoid any unifocalization before 4 months of age, even when severe cyanosis led to an early palliative procedure (usually seen when small or severely stenosed aortopulmonary collaterals were present).

For patients who were up to 4 months old at the initial palliation, we tried to unifocalize non-communicating major aortopulmonary collaterals when the diameter was greater than 2 mm. For the last 5 years, we strongly considered unifocalization for restrictive major aortopulmonary collateral to PA communications (i.e. smaller communication than the largest major aortopulmonary collateral diameter) or for major aortopulmonary collaterals that were significantly larger than the native PAs. For large communicating major aortopulmonary collaterals, occlusion was achieved during the operation or by a catheter-based procedure, before or after the RVPA connection was made.

Biventricular repair

Based on clinical status, pulmonary angiography, the anatomical pulmonary vascular tree, pulmonary vascular resistance and the Nakata index (usually superior at 250 mm^2/m^2), we performed a biventricular repair. The VSD was closed, and right ventricular outflow tract reconstruction was achieved either with a transannular pericardial patch or with a valved conduit.

Follow-up

We conducted a retrospective single-institution study. Follow-up after the RVPA connection involved patient records and correspondence or structured telephone interviews with local referring cardiologists. Patient follow-up information was updated in June 2015. Twelve patients were lost to follow-up.

Statistical analysis

All values are given as mean ± standard deviation; categorical variables are reported as proportion (%). Associations between categorical variables were tested by Pearson's χ^2 test; however, when cell frequencies were less than or equal to 5, we used the Fisher's exact test. We compared continuous variables and categorical variables using the Mann–Whitney test but when we compared all three groups, we used the Kruskal-Wallis test. Survival after the RVAP connection was analysed using the Kaplan–Meier method. The log rank test was used to explore the significance of the difference between two groups. All data were analysed using statistical software SPSS version 20 (IBM, Armonk, NY).

RESULTS

Operative data

The median age and weight at the time of the RVPA connection were 12 days (range 1 day-13.6 years) and 3.1 kg (range 1.9-43 kg), respectively. Significant differences were observed between Group III and the others. Patients were older ($P = 0.004$) and heavier ($P < 0.001$) and the Nakata index was lower ($P < 0.001$) in Group III compared with Groups I and II.

For the entire cohort, the RVPA connection was achieved by pericardial patch enlargement in 66 (61%), left atrial appendage reconstruction in 12 (11%) and polytetrafluoroethylene conduit in 31 (28%). Being in Group II ($n = 41$) was strongly associated with the use of the left atrial appendage interposition ($P = 0.016$). Operative data are shown in Table 2.

In-hospital deaths

There were 3 in-hospital deaths (2.7%). Two patients died within the first 24 h. One patient died in the operating room with signs of unilateral lung hyperaemia, followed by secondary severe hypoxaemia. No extracorporeal life support was attempted. The other patient had a cardiac arrest from unknown aetiology immediately after extubation. The last patient died on Day 4 of pulmonary hypoxia with the diagnosis of an extended tracheal stenosis. No slide tracheoplasty was attempted. The hospital deaths are shown in Table 3.

Morbidity

The sternum was left open in 23 cases (21%). No significant differences were seen among the 3 groups related to delayed sternal closure ($P = 0.41$), mean mechanical ventilation time (73 ± 143 h, 91 ± 122 h, 91 ± 84 h, $P = 0.84$) or length of stay in the ICU (137 ± 143 h, 175 ± 165 h, 156 ± 113 h, $P = 0.5$). Data are shown in Table 2. Significant pulmonary over-circulation was

Table 2: Operative and postoperative characteristics after performing the right ventricle to pulmonary artery connection

Characteristics	PA/VSD Type I ($n = 33$)	PA/VSD Type II ($n = 41$)	PA/VSD Type III ($n = 35$)	Total ($n = 109$)	*P*-value
Surgical procedure					
Patch enlargement	25 (76)	22 (53)	19 (54)	66 (61%)	0.1
PTFE conduit	6 (18)	10 (24)	15 (42)	31 (28%)	0.06
Interposition left appendage	2 (6)	9 (22)	1 (3)	12 (11%)	0.023
Mean open size (mm)	5.1 (0.9)	5.2 (1.2)	5.9 (1.1)	5.4 (1.1)	0.002
Unifocalization	0	0	12 (34)	12 (11)	<0.001
Confluence PA plasty	6 (18)	12 (29)	8 (22)	26 (23)	0.53
LPA plasty	7 (21)	9 (21)	6 (17)	22 (20)	0.86
RPA plasty	1 (3)	7 (17)	7 (20)	15 (13)	0.079
CPB duration (min)	70 (26)	89 (38)	96 (56)	85 (43)	0.06
Cross-clamping duration (min)	34 (15)	36 (20)	29 (18)	33 (18)	0.05
Postoperative characteristics					
Delayed sternal closure (days)	7 (21)	11 (26)	5 (14)	23 (21%)	0.41
Mean oxygen saturation after surgery, %	91 (6)	90 (6)	88 (11)	90 (8)	0.29
Mechanical ventilation (h)	73 (92)	91 (122)	91 (84)	86 (102)	0.84
Length of ICU stay (h)	137 (143)	175 (165)	156 (113)	158 (142)	0.5
HFO	1 (3)	5 (12)	3 (8)	9 (8)	0.41

PA/VSD: pulmonary atresia with ventricular septal defect; PTFE: polytetrafluoroethylene; PA: pulmonary artery; LPA: left pulmonary artery; RPA: right pulmonary artery; CPB: cardiopulmonary bypass; ICU: intensive care unit; HFO: high-frequency oscillations.

Table 3: Complications after performing the right ventricle to pulmonary artery connection

Characteristics	PA/VSD Type I ($n = 33$)	PA/VSD Type II ($n = 41$)	PA/VSD Type III ($n = 35$)	Total ($n = 109$)	*P*-value
In-hospital deaths	1 (3%)	1 (2.5%)	1 (3%)	3 (2.8%)	1
Ventricular aneurysm	2 (6%)	1 (2.5%)	5 (14%)	8 (7%)	0.18
Excessive pulmonary blood flow	0	0	1 (2.9%)	1 (1%)	0.34
Restrictive pulmonary blood flow before 2 months	2 (6%)	6 (14.6%)	1 (3%)	9 (8%)	0.14
BT shunt	2	5	0	7 (6.4%)	
Enlargement of RVPA connection	0	1	1	2 (2%)	
Unifocalization	0	0	0	0	
Pulmonary artery plasty	0	1	0	1 (1%)	
Angioplasty	0	0	0	0	
Restrictive pulmonary blood flow between 2 months and biventricular repair	1 (3%)	4 (10%)	15 (42%)	20 (18%)	<0.001
BT shunt	0	2	3	5	
Enlargement of RVPA connection	1	0	4	5	
Unifocalization	0	0	4	4	
Pulmonary artery plasty	0	0	1	1	
Angioplasty	0	2	3	5	
BT shunt + unifocalization	0	0	2	2	
Enlargement of RVPA connection + unifocalization	0	0	1	1	
Enlargement of RVPA connection + pulmonary artery plasty	0	0	1	1	
Pericardial drainage	1 (3%)	0	3 (8.5%)	4 (4%)	0.10
Phrenic paralysis	0	1 (2.5%)	1 (3%)	2 (2%)	0.64

PA/VSD: pulmonary atresia with ventricular septal defect; RVPA: right ventricle to pulmonary artery; BT: Blalock–Taussig; RV: right ventricular; PA: pulmonary artery.

diagnosed in only 1 patient who needed reoperation on Day 1 to reduce the size of the RVPA connection .

Patients with restrictive pulmonary blood flow, defined as the need for palliative reinterventions, and subsequent augmentation of pulmonary blood flow (with the use of MBT shunts, enlargement of the RVPA connection, unifocalization, pulmonary arterioplasty and PA angioplasty) were divided into early (before 2 months) and late (after 2 months) reoperations.

Nine patients (8%) underwent reoperations for early restrictive blood flow within the first 2 months.

Twenty patients needed a late redo operation as a second stage to increase the pulmonary blood flow after 2 months at a median interval of 8.3 months (range 4.3 months-8 years). The risk for late redo operation was higher in the major aortopulmonary collateral group (Group III: 8/35; 51%) compared with the non-major aortopulmonary collateral group (Groups I and II: 15/74; 20%).

There were 8 cases of ventricular aneurysm: early reoperation was mandatory in 2 and anticipated complete repair in 6. The postoperative morbidities associated with the RVPA connection are shown in Table 3.

Pulmonary arterial growth

The mean initial Nakata index was 101 ± 52 mm^2/m^2. Patients with major aortopulmonary collaterals (Type III) had a significantly lower mean Nakata index before the RVPA connection (58.7 mm^2/m^2); $P < 0.001$) compared with PA/VSD Types I and II. The mean Nakata index before complete repair and the Δmean Nakata were not statistically different between the 3 groups ($P = 0.26$ and $P = 0.916$, respectively). The time between the initial Nakata measurement and the last evaluation was 17 ± 21 months with a significantly longer delay ($P = 0.003$) for the major aortopulmonary collateral group. Indeed, the delay between the RVPA connection and complete repair was longer for patients with major aortopulmonary collaterals than for the groups (Groups I and II) with non-major aortopulmonary collaterals. The mean index of growth (mm^2/m^2/month), representing the speed of growth of the PA, was 40.8 ± 71 mm^2/m^2/month. This growth was not different among the 3 groups. Data for PA growth are shown in Table 4.

Late interstage deaths

Seven patients died after the RVPA connection and prior to biventricular repair (6.6%). One patient died of RV pseudoaneurysm rupture, 1 patient died of gastrointestinal bleeding and 2 patients died of respiratory failure. Three sudden deaths were reported.

Complete surgical repair

Eighty-four patients (77%) have undergone complete repair, and 8 patients (7%) with optimal PA growth are awaiting repair. Four patients from abroad were lost to follow-up. Three patients were definitively considered unsuitable for septation. These 3 patients were PA/VSD Type III with severe hypoplastic PA and a Nakata index below 50 mm^2/m^2. Ten patients died, with 3 early deaths and 7 late deaths.

Patient characteristics at complete repair

The mean age of the 84 patients who underwent complete repair was 20.4 ± 29 months (range 2.7 months-14.5 years). There was a statistically significant difference between the operating age in the groups without major aortopulmonary collaterals (Groups I and II; 12.9 and 13.4 months, respectively) and the group with major aortopulmonary collaterals (43.4 months). The patients who underwent complete repair had a mean Nakata index of 307 mm^2/m^2. The differences in the Nakata index between the 3 groups were not statistically significant. Patients in Group I had predominantly undergone patch enlargement (83%). Patients in Group III had predominantly received prosthetic valved conduits (60%). Immediately after the biventricular repair, elevated RV pressure ≥40 mmHg was observed in 22 patients (26%). Data for biventricular repair are shown in Table 5.

The population of patients without biventricular repair comprised 60% with Type III, 32% with chromosome 22q11 deletion and 44% with a history of restrictive pulmonary blood flow following the RVPA connection. Patients without complete repair had a statistically lower Nakata index before the RVPA connection than patients with complete repair (70 vs 111 mm^2/m^2 respectively; $P = 0.001$). Patients without biventricular repair had a statistically lower index of growth than patients with biventricular repair (34 vs 42 mm^2/m^2/month, respectively; $P = 0.001$). Data for biventricular repair versus no biventricular repair are shown in Table 6.

Late outcomes

There were 6 late deaths after complete repair. Four of these patients had chromosome 22q11 deletion. The actuarial survival curve for the 109 patients undergoing RVPA connection is shown in Fig. 1. The survival was 90% at 1 year, 84% at 5 years and 81% at 10 years.

There was a significant difference in survival between patients with and without chromosome 22q11 deletion ($P = 0.005$). Survival is shown in Fig. 2.

DISCUSSION

This study represents the largest cohort of an initial RVPA connection for patients with PA/VSD. This palliation is part of a process of rehabilitation of the native pulmonary artery (NPA) (multistage approach) and has many advantages in the management of these patients with complex issues.

Mortality

The number of in-hospital deaths was low in our study (<3%) and compares with that reported by others. Carotti *et al.* reported a mortality rate of 13% [9], but they included patients without NPA. Metras *et al.* and Zhang *et al.* reported mortality rates of 10% and 2.7%, respectively [10, 11], but in Metras *et al.*'s experience, the mean Nakata index (20.6 mm^2/m^2) was lower than that in our study. This low initial mortality rate was emphasized by Zheng

Table 4: Pulmonary artery size characteristics before and after making the right ventricle to pulmonary artery connection

Characteristics	PA/VSD Type I	PA/VSD Type II	PA/VSD Type III	Total	*P*-value
Mean Nakata index before the RVPA connection (mm^2/m^2)	120 (36)	123 (44)	58.7 (47)	101 (52)	<0.001
Mean Nakata index before complete repair (mm^2/m^2)	346 (287)	301 (215)	223 (124)	274 (200)	0.26
ΔMean Nakata (mm^2/m^2) index	226 (290)	183 (200)	157 (99)	179 (183)	0.91
Time between 2 measures (months)	17 (28)	8 (6)	25 (24)	17 (21)	0.003
Mean index of growth (mm^2/m^2/month)	39.4 (54)	53.3 (85)	31.22 (65)	40.8 (71)	0.41

PA/VSD: pulmonary atresia with ventricular septal defect; RV: right ventricle; PA: pulmonary artery.

Table 5: Operative characteristics of biventricular repair

Characteristics	PA/VSD type I ($n = 30$)	PA/VSD type II ($n = 34$)	PA/VSD type III ($n = 20$)	Total ($n = 84$)	*P*-value
Mean weight (kg)	7.3 (4.4)	7.6 (2.7)	13.44 (10)	8.85 (6.2)	0.002
Mean age (months)	12.9 (24.2)	13.6 (14.3)	43.4 (42.5)	20.43 (29.4)	<0.001
Time between the RVPA connection and biventricular repair (months)	11.5 (18.6)	10.97 (8)	25.7 (21)	14.67 (17)	0.002
Mean Nakata index before biventricular repair (mm^2/m^2)	274 (204)	264 (202)	257 (130)	266 (186)	0.26
Surgical procedure					
Patch enlargement	25 (83%)	18 (53%)	4 (20%)	47 (56%)	<0.001
Prosthetic conduit valve	3 (10%)	13 (38%)	12 (60%)	28 (33%)	<0.001
Prosthetic conduit, no valve	0	1 (3%)	1 (5%)	2 (2%)	0.54
Patch enlargement with monocuspid valve	2 (6.6%)	2 (6%)	3 (15%)	7 (8%)	0.46
RPA plasty	8 (26%)	15 (44%)	2 (10%)	25 (30%)	0.03
LPA plasty	19 (63%)	17 (50%)	8 (40%)	44 (52%)	0.31
CPB duration (min)	139 (38)	146 (44)	151 (42)	144 (41)	0.44
Cross-clamping duration (min)	81 (28)	86 (24)	80 (30)	83 (27)	0.75
Right ventricular pressure superior at 40 mmHg	5 (17%)	7 (21%)	10 (50%)	22 (26%)	0.02

PA/VSD: pulmonary atresia with ventricular septal defect; RV: right ventricle; PA: pulmonary artery; LPA: left pulmonary artery; RPA: right pulmonary artery; CPB: cardiopulmonary bypass.

Table 6: Differences between patients with complete repair and without complete repair

Characteristics	No complete repair ($n = 25$)	Complete repair ($n = 84$)	*P*-value
Mean age during RVPA connection (months)	7.9 (2.2)	6 (2.2)	<0.001
Classification PA/VSD			
Type I	3 (12%)	30 (36%)	0.023
Type II	7 (28%)	34 (40%)	0.25
Type III	15 (60%)	20 (24%)	<0.001
Chromosome 22q11 deletion	8 (32%)	13 (15%)	0.067
RVPA connection procedure			
Patch enlargement	13 (52%)	53 (63%)	0.32
PTFE conduit	10 (40%)	21 (25%)	0.14
Interposition of left appendage	2 (8%)	10 (12%)	0.58
History of unifocalization	8 (32%)	10 (12%)	0.02
History of restrictive pulmonary blood flow after RVPA connection	11 (44%)	17 (20%)	0.001
Mean Nakata index before RVPA connection (mm^2/m^2)	70 (46)	111 (50)	<0.001
ΔNakata index (mm^2/m^2)	101 (98)	203 (196)	0.04
Index of growth (mm^2/m^2/month)	34 (92)	42 (64)	0.02

PA/VSD: pulmonary atresia with ventricular septal defect; PTFE: polytetrafluoroethylene; RV: right ventricle; PA: pulmonary artery.

et al. [12] who described a potential lower in-hospital mortality rate than with an MBT shunt (0% vs 12.5%, not significant). This low mortality rate was explained by a lower risk of thrombosis of the RVPA connection (no thromboses in our series). In addition, the RVPA connection does not have the disadvantages, such as diastolic run-off, impaired coronary perfusion and left heart volume overload, which can be encountered with MBT or central shunts. The risk of CPB and ventriculotomy seem to be acceptable, given the low in-hospital mortality rate that we observed. Although mortality rates for the MBT have been lower in recent decades, as shown by the historical series of Williams *et al.* [4], morbidity and mortality rates related to the MBT due to shunt thrombosis and the narrowing or distortion of the PA remain important. The only studies comparing the RVPA connection directly with the MBT shunt have been described in patients with hypoplastic left heart syndrome following the Norwood procedure. Establishing a conduit between the RV and the PA seems to result in an increase in in-hospital survival, better haemodynamic stability and a reduction in postoperative deaths [13].

The number of interstage deaths was 2.5 times higher than the number of early deaths (2.7% vs 6.6%) and is our key concern. Whatever the causes of death were, they illustrate the potential risk in this specific interstage period and lead us to an organized, close cardiologic follow-up schedule. We are concerned about the false aneurysms associated with the RVPA connection. A false aneurysm was identified as the cause of death in 1 case and suspected in 2 others (no post-mortem examination). We think that preventing infundibular pseudoaneurysm formation can reduce the number of deaths. This specific problem is discussed in the complication section below.

The only long-term risk factor associated with death was DiGeorge syndrome. The causes of the excess mortality rate are multifactorial. Patients with DiGeorge syndrome have a higher susceptibility to infectious and smaller native PAs.

The significant late mortality rate after repair reminds us that it is still a severe congenital heart condition, whatever the surgical programme be. Patients at risk are probably those with the worst arterial tree and high RV pressure after repair, though nothing can be asserted in this small subset population.

Pulmonary artery growth

The initial palliation leads to increased blood flow through the PA, which allows for growth of the NPA [2, 10, 14–16], favouring pulmonary vasculogenesis while creating competitive flow with the major aortopulmonary collaterals [17, 18]. In our series, the restrictive pulmonary blood flow enabled growth of the PA at a rate that is relatively constant. Our mean ΔNakata (179 mm^2/m^2) was similar to that in other series using the RVPA connection [2, 11, 12, 19, 20]. Amark *et al.* [2] found a ΔNakata of 74 mm^2/m^2 for the MBT shunt and 102 mm^2/m^2 for the RVPA connection, concluding that the RVPA connection seemed to promote better growth of the PA relative to an MBT shunt. Our index of growth, the ΔNakata/time ratio, appears to be a good predictive factor for biventricular repair (77% in this series, comparable to those from other studies [10, 11, 20]). As logically expected, there seemed to be a correlation between the initial size of the NPA and the duration of the interstage period. The growth rate of the PA, however, was similar among the patient groups, except for patients who had undergone unifocalisation of the major aorto-pulmonary collaterals.

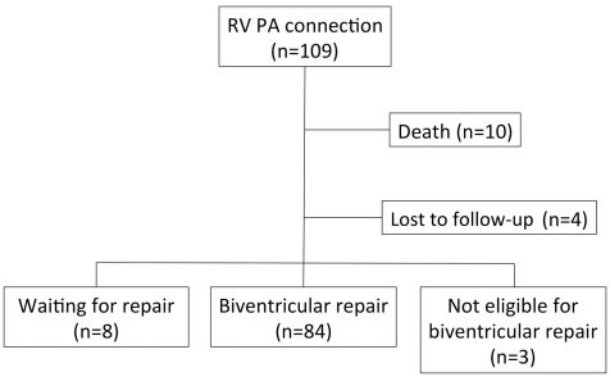

Figure 1: Flow diagram of outcomes in 109 patients (PA/VSD) with RV to PA connection. RVPA: right ventricular pulmonary artery; PA/VSD: pulmonary atresia with ventricular septal defect.

Complications

Morbidity remains high in our series but can probably be reduced by taking into consideration the results of this reported experience.

Inadequate initial pulmonary blood flow. The occurrence of pulmonary over-circulation was notably low (<1%) and easy to deal with by clipping the RVPA connection without CPB in order to reduce its diameter. On the other hand, reoperations were frequent for restrictive pulmonary blood flow. It is important to differentiate early reoperations (<2 months), which were for over-restrictive openings or due to extremely small branch PAs, and late reoperations (>2 months). These latter were due to an opening that became too restrictive as the child grew. Early reinterventions could be avoided by appropriate calibration and by achieving an RVPA connection that was not overly restrictive. Postoperatively, patients who have a VSD physiology rather than a very cyanotic situation are easier to manage in the ICU. This fact led us to enlarge somewhat our RVPA connections during the last years. For neonatal palliation, we do not hesitate to be

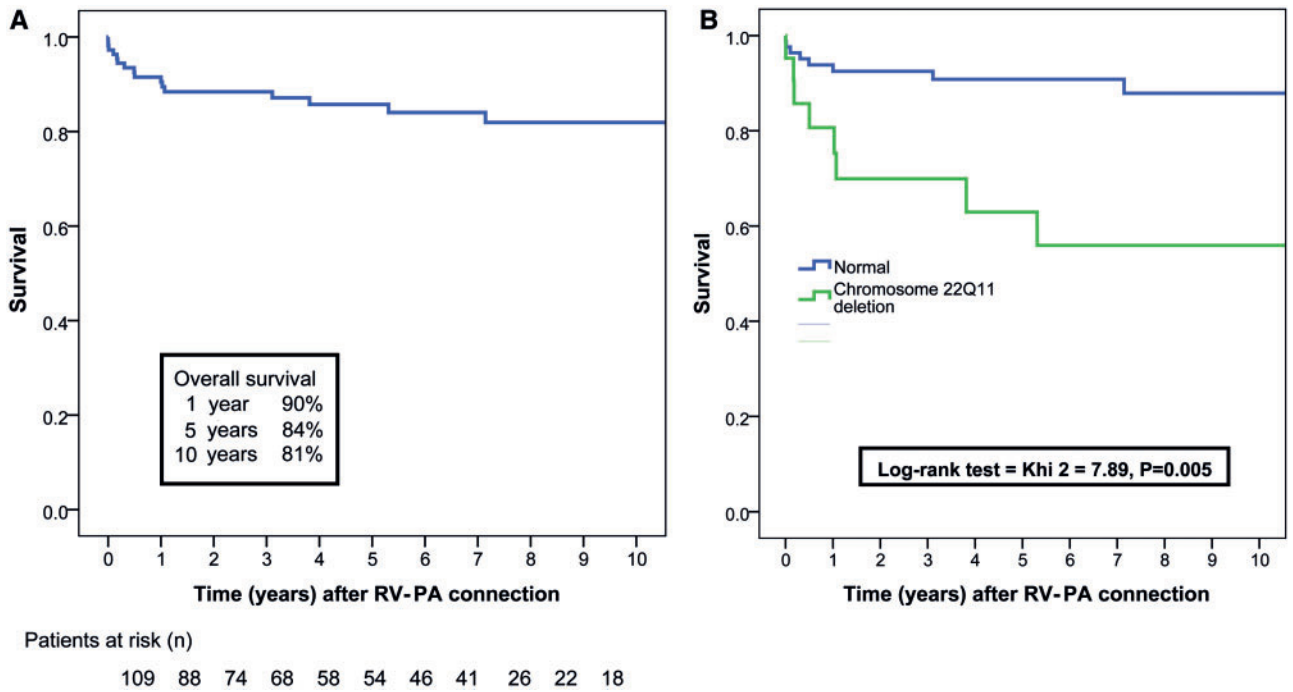

Figure 2: (**A**) Survival for the overall population for RVPA connection operation in 109 children with PA/VSD. (**B**) Survival for the overall population for RVPA connection operation comparing those with and without chromosome 22q11 deletion (green line). RVPA: right ventricular pulmonary artery; PA/VSD: pulmonary atresia with ventricular septal defect.

close to a diameter of 6 mm for 3.5 kg, especially if the PAs are hypoplastic. For older patients, it can be useful to insert a PA catheter in the distal RV connection to measure the pressure to be sure that we do not initiate pulmonary hypertension. Left atrium pressure too is important, in order to detect any pulmonary overflow. Finally, a transoesophageal echocardiogram can be used to evaluate the shunt at the level of the VSD, and a bidirectional shunt is an excellent sign of an appropriate RVPA connection, with a pulmonary/systemic ratio around 1. The need for inhaled nitric oxide, high fraction of inspired oxygen or the use of vasopressors in the operating room to achieve decent oxygen saturation is definitely a strong sign of low pulmonary blood flow.

In the case of a too restrictive RVPA connection diagnosed in the ICU, patients were reoperated using either an MBT shunt or refection/enlargement of the RVPA connection. The choice was left to the surgeon, based on his subjective evaluation of the anticipated difficulty to re-establish the RVPA connection (weakness of the underlying tissue, need for ventriculotomy enlargement due to a long conus, small PA bifurcation and previous use of the left atrial appendage). An MBT shunt was also an option.

False infundibular aneurysm. Another common complication (7%) was aneurysm formation at the ventriculotomy patch. These aneurysms are formed by a combination of systemic right ventricular pressure due to the unrestrictive VSD and weakness of the surgical patch suture in neonates. Eight patients had a false aneurysm during the follow-up period, of whom 2 required earlier reoperations; rupture of these aneurysms may have been responsible for some of the 3 sudden deaths. This surgical complication should be screened for at follow-up by a routine echocardiogram, a chest radiograph and even a computed tomography scan if indicated. Technically, in the case of low-weight babies, the left margin of the ventriculotomy can be fragile and close to the LAD coronary artery, leading to a weaker suture of the RVPA patch. We now do not hesitate to add pledgeted interrupted sutures to reinforce this area. Since we added this modification to our surgical protocol, no significant RV pseudoaneurysms have been reported.

Surgical strategy and general philosophy

Although the fine details of our surgical strategy have evolved slightly during these 15 years, our approach can be precisely described. The idea is to palliate by an RVPA connection all patients with PA/VSD Type I or II (with a duct-dependent pulmonary circulation) within the first few days after birth. For Type III patients, the operative indications are guided by the clinical status, the size of the native PAs and a qualitative analysis of the major aortopulmonary collaterals. If the patient is severely cyanotic before 4 months of age, we attempt the RVPA connection without unifocalization of non-communicant major aortopulmonary collaterals (especially for small diameter major aortopulmonary collaterals). On the other hand, if oxygen saturation is 75–80%, we prefer to perform the RVPA connection after 4 months of age, with complete unifocalization of all non-communicant major aortopulmonary collaterals through a sternotomy (diameter up to 1.5 mm) or for large communicant major aortopulmonary collaterals (especially when the maximum diameter of a major aortopulmonary collateral is greater than that of the major aortopulmonary collateral to PA communication and the central PAs are severely hypoplastic).

One could argue that this staged strategy leads to palliative operations, subsequent reinterventions and interstage deaths. Despite this negative aspect, our general philosophy is to afford the best pulmonary tree growth for optimal late repair but also to avoid the use of any valved conduit. This goal is achieved by having a posterior autologous connection close to the area of a repaired tetralogy of Fallot. This strategy probably reduces the incidence of reoperation even if patients will need pulmonary valvulation at some time in the future. Since 2014, based on our experience in the unvalved repair of the truncus arteriosus, we propose the early correction for Type I PA/VSD patients with a well-developed pulmonary tree without the use of any conduit.

Study limitations

The retrospective nature of this study is a limitation. The statistical results should be interpreted with caution because potential bias may have been introduced by the retrospective nature of the study and the absence of a control group.

CONCLUSION

A low rate of early death can be achieved in patients with PA/VSD with or without major aortopulmonary collaterals using the RVPA connection as an initial palliative strategy. This palliative procedure allows for adequate growth of the pulmonary tree and a large proportion of biventricular repairs and contributes to the involution of the aortopulmonary collaterals. Early reinterventions and interstage deaths are key concerns but should be reduced by the proposed surgical modifications and close follow-up. Despite all these considerations, repair with no pulmonary conduit, which is allowed by this strategy, is probably one of our first priorities.

Conflict of interest: none declared.

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