Staged Correction of Pulmonary Atresia, Ventricular Septal Defect and Collateral Arteries

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Abstract

Objectives Pulmonary atresia (PA) with ventricular septal defect (VSD) and systemic-pulmonary collateral arteries (SPCA's) has a variable anatomy with regard to the pulmonary vasculature, asking for an individualized surgical treatment. A protocol was applied consisting of staged unifocalization and correction. Methods Since 1989 39 consecutive patients were included (median age at first operation 13 months). In selected cases a central aorto-pulmonary shunt was performed as first procedure. Unifocalization procedures were performed through a lateral thoracotomy. Correction consisted of shunt takedown, VSD closure and interposition of an allograft between the right ventricle and the reconstructed pulmonary artery. Postoperatively and at follow up echocardiographic data were obtained. Results In 39 patients 66 unifocalization procedures were performed. Early mortality was 5%. Seven patients were considered not suitable for correction, four of them died. One patient is awaiting further correction. Correction was done successfully in 28 patients. Operative mortality was 3% and late mortality 11%. Median follow-up after correction was 19 years. Eleven patients needed homograft replacement. Freedom from conduit replacement was 88%, 73% and 60% at 5, 10 and 15 years respectively. Right ventricular function was reasonable or good in 75 % of the patients. Conclusions After complete unifocalization 30/37 patients (81%) were considered correctable. The main reasons for palliative treatment without correction were pulmonary hypertension and/or inadequate outgrowth of pulmonary arteries. Staged approach of PA, VSD and SPCA's results in adequate correction and good functional capacity. RV function after correction remains reasonable or good in the majority of patients.

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Key words: Pulmonary atresia, Ventricular septal defect, Outcomes.

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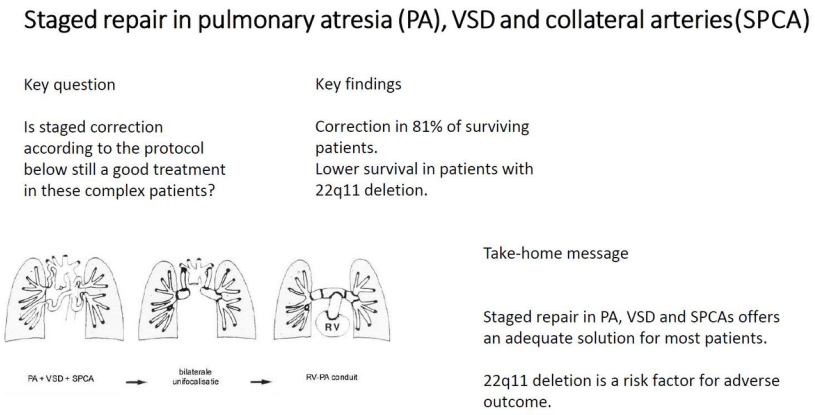
Visual abstract



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Methods

Since 1989 39 consecutive patients were included (median age at first operation 13 months). In selected cases a central aorto-pulmonary shunt was performed as first procedure. Unifocalization procedures were performed through a lateral thoracotomy. Correction consisted of shunt takedown, VSD closure and interposition of an allograft between the right ventricle and the reconstructed pulmonary artery. Postoperatively and at follow up echocardiographic data were obtained.

Results

In 39 patients 66 unifocalization procedures were performed. Early mortality was 5%. Seven patients were considered not suitable for correction, four of them died. One patient is awaiting further correction. Correction was done successfully in 28 patients. Operative mortality was 3% and late mortality 11%. Median follow-up after correction was 19 years. Eleven patients needed homograft replacement. Freedom from conduit replacement was 88%, 73% and 60% at 5, 10 and 15 years respectively. Right ventricular function was reasonable or good in 75 % of the patients.

Conclusions

After complete unifocalization 30/37 patients (81%) were considered correctable. The main reasons for palliative treatment without correction were pulmonary hypertension and/or inadequate outgrowth of pulmonary arteries. Staged approach of PA, VSD and SPCA's results in adequate correction and good functional capacity. RV function after correction remains reasonable or good in the majority of patients.

Introduction

During the last 30 years the surgical strategy for pulmonary atresia (PA) with ventricular septal defect (VSD) and systemic-pulmonary collateral arteries (SPCA's) is subject of ongoing discussion. There are several reports using the multi-staged approach as well as reports on two-staged approach or one-stage midline primary repair. [1-6] Furthermore it has been reported that unifocalization possibly does not bring long-term benefit in terms of late survival.[7]

Historically patients with PA, VSD and SPCA's were treated in our hospital with various surgical interventions depending on the clinical condition and previous cardio-surgical procedures on presentation in our clinic. Since 1989 a protocol as described previously was followed in all our patients presenting with PA, VSD and SPCA's.[8] This protocol consisted of staged unifocalization procedures with a subsequent total correction, with closure of the VSD and placing a pulmonary homograft between the right ventricular outflow tract (RVOT) and the pulmonary bifurcation. We report our results of 30 year experience with this staged protocol applied to all consecutive patients.

Material and methods

Since 1989 thirty-nine consecutive patients (21 boys, 18 girls) were included in this protocol. A 22Q11 deletion was diagnosed in 10 patients. The median age at the time of the first unifocalization was 13 months (range 2 weeks to 189 months). Diagnostic catheterization was performed to assess the pulmonary vascularity and perfusion. In 13 patients with a hypoplastic central confluent pulmonary artery a central aorto-pulmonary shunt was performed, intended to allow the confluent pulmonary artery to grow to improve the starting point for unifocalization.

Unifocalization was performed through a lateral thoracotomy with identification of all collateral arteries at that side. When adequate intrapulmonary connection was confirmed on preoperative angiography or intra-operatively, the dual supply SPCA could be closed. When such connection was not established, the collateral artery was anastomosed to the native pulmonary artery as close as possible to the hilar pulmonary vasculature. In case of an absent confluent pulmonary artery and in cases where further augmentation was indicated a modified Blalock-Taussig shunt was constructed to the ipsilateral subclavian artery. In 27 patients an additional unifocalization procedure on the contra-lateral side was performed to augment the blood supply to that lung. Before and after each procedure an angiogram was made.

To evaluate the growth of the pulmonary arterial system we retrospectively measured the pre- and post-unifocalization Nakata-index.[9]

The change in lung perfusion on angiogram pre- and post-unifocalization was studied retrospectively to evaluate the result of the procedure. We studied the total lung perfusion including perfusion by the SPCA's versus the lung perfusion by flow through the confluent pulmonary artery alone.

Based on the angiographic findings and data from catheterization measurements patients were selected for total correction. Pulmonary hypertension or unfavorable anatomic result of unifocalization at angiogram were contraindications for total correction. The total correction was performed through a median sternotomy with the use of extracorporeal circulation and moderate hypothermia. The modified Blalock-Taussig and central shunts were divided. The VSD was closed with a Gore-Tex® patch and a cryopreserved pulmonary homograft was interposed between the RVOT and the proximal pulmonary arterial system. Postoperative recovery and hospital or 30-day mortality is reported.

Long-term follow up was derived from the records. In 24 survivors, with a complete repair, echocardiographic data were available except in one patient who was lost to follow up. From 17 patients after successful correction MR imaging was available for analysis of the right ventricular function.

This study was approved by the Ethical Committee with no need for informed consent.

Statistical analysis was performed using Statistical Package for the Social Sciences (SPSS) software, version 24.0 (SPSS Inc., Chicago, IL). Frequencies were given as absolute numbers and percentages. The data were expressed as median with range. The paired t-test was performed for statistical analysis. We applied the χ^2 test to compare frequencies in the two groups. The Kaplan-Meier method was applied to estimate freedom from reintervention and for survival. A P value less than 0.05 was considered to indicate statistical significance.

Results

Unifocalization

After initiation of the program a catch up of patients to be treated was seen, reflected by a higher age at first surgery and the age at correction. During the first 5 years the mean age at first unifocalization was 5.9 years (range 3 months -14 years) and after this period the mean age was 1.8 years (range 1 month -16 years with only one older patient of 16 who came to our hospital from abroad). Thirty-nine consecutive patients with PA, VSD and SPCA's were entered in the protocol. In these 39 patients, in 13 patients a central shunt was the first procedure followed by 66 unifocalization procedures in which 129 collateral arteries were treated either with unifocalization or ligation. In 7 patients a collateral artery was closed by a percutaneously placed device.

In 50 unifocalization procedures a modified Blalock Taussig shunt was placed with a tube diameter $5 - 10$ mm, depending on the size of the patient, in later procedures a 5 mm tube prosthesis was most commonly used.

Based on several reports and our own clinical experience of the behavior of SPCA's we tend towards more intrapulmonary anastomoses to avoid long segments of remaining SPCA tissue which could lead to stenosis or dilatation.[10, 11]

In one patient the modified Blalock-Taussig shunt was obstructed two weeks after unifocalization and the patient died after a reoperation in which the modified Blalock-Taussig shunt was replaced. One other patient died in hospital 3 months after unifocalization with an unknown cause.

The median time between the first and second unifocalization was 8 months (range 2 weeks - 48 months), this second unifocalization was considered indicated in 27 patients. In twelve patients a second surgical unifocalization was not necessary. Four patients had SPCA's mainly on one side. Five patients received a coil closing a SPCA at the contralateral side after unifocalization at the other side. After the first unifocalization on one side one patient had still incomplete vascularization of both lungs and progressive pulmonary hypertension and was consequently treated with a central aorto-pulmonary shunt at the other side. One patient died before the second unifocalization probably due to pulmonary infection with an RS virus 3 months after unifocalization.

After unifocalization 20 additional procedures were needed. Apart from the coil closure of SPCA's several other interventions were performed. In 4 patients a perigraft seroma around the modified Blalock-Taussig shunt was removed.[12] In one patient the modified Blalock-Taussig shunt was incorrectly placed on the pulmonary vein, this was revised 2 weeks later. In 2 patients a modified Blalock-Taussig shunt and in 2 patients a central shunt was placed additionally because of cyanosis after the second unifocalization. In one patient a transannular patch was placed between the RVOT and the pulmonary trunk later followed by balloon dilatation and stent implantation in the left pulmonary artery. In two patients balloon dilatation of the modified Blalock-Taussig shunt was performed in one also with stent implantation. Two patients underwent revision of a central shunt with augmentation of the pulmonary artery, one with stent implantation

and the other with balloon dilatation of the left pulmonary artery. One patient underwent revision of the modified Blalock-Taussig shunt because of shunt occlusion. One patient underwent a sliding plasty of the distal trachea for stenosis with complete tracheal rings.

Nakata-index and lung perfusion

The median Nakata-indices (n=22), indicating the relative size of the pulmonary arteries were 143 (range 49-345) before and 190 (range 49-401) mm^2/BSA after unifocalization (p=0,097). The normal value of this PA-index is 330 mm^2/BSA [9].

From 30 patients we obtained detailed information about their lung perfusion by studying there cardiac catheterization images before and after their unifocalization procedure(s). The total lung perfusion was 99% before and 97% after unifocalization. Lung perfusion through the confluent pulmonary artery was 52% before and 89% after unifocalization. Although there seems to be an increase in perfusion through the confluent pulmonary artery, this difference was not significant probably due to the small groups.

Corrective surgery

30 of the 37 patients (81%) were considered correctable. One patient is awaiting correction after complete unifocalization.Twenty-nine patients underwent a total correction as described earlier which was successful in 28 patients (97%). Median age at correction was 2.6 years (range 1 to 17 years) and median interval between unifocalization and correction was 8 months (range 1 to 48 months). In all patients the VSD was closed. One patient needed a takedown during operation because of suprasystemic pressures of the right ventricle and impossibility to wean the patient from the extracorporeal circulation. One patient had a resternotomy for persistent bleeding the same day and recovered without complications.

Operative mortality was 3% (n=1). This patient had a reoperation for leakage of the proximal suture of the homograft at the same day of the correction. Because of hypoperfusion a major CVA developed and the patient died. Seven patients were not eligible for correction. In three patients there was pulmonary hypertension. One patient had malperfusion of the complete left lung, one patient had a hypoplasia of the right ventricle not suitable for repair, one patient had hypoplasia of the lungs and airways in combination with neuro-cognitive disorders and one patient was considered a too high risk operation due to developmental disorders. Figure 1 shows a flowchart of our patient population.

In the ten patients with 22q11 deletion, 1 is awaiting correction, 3 were ineligible (2 died, 1 still alive) for final correction and 6 patients had a definitive correction. One patient died post-operatively due to cerebral-vascular accident. Compared to non-syndromic patients, a lower percentage of the 22q11 deletion patients were reaching their final correction (23 out of 29 (79%)) versus 6 out of 9 (67%) respectively), although not significant (p=0.55).

Follow-up

Median follow up time after correction was 19 years (range 1 to 27 years). Overall survival after definitive correction was 96% at 20 years (Figure 2). One patient died four years after successful correction of unknown cause, probably of cardiac arrhythmia. Two patients died 20 years after correction due to progressive heart failure. Two of these had 22q11 deletion. From the survivors all but one are in NYHA class I or II. Four of the seven patients who were not suitable for correction died. One patient died 2 years after the last unifocalization due to respiratory failure and infection. One patient died of unknown cause 9 years after the last unifocalization procedure. One patient died 14 years after the last unifocalization of multi-organ failure and sepsis and one patient died of massive intracranial bleeding 9 years after the last unifocalization.

Among the 5 survivors with 22q11 deletion, one is awaiting correction, one is palliated in a reasonable condition, 3 had a definitive correction (one in reasonable condition and 2 in good condition). The overall survival in the 22q11 deletion patient was significantly (p=0.041) lower compared to non-syndromic patients, (5 out of 10 (50%)) versus 25 out of 29 (67%) respectively).

Pulmonary valve replacement

After final correction in 21 patients other interventions were performed. They are listed in Table 1 and consist mostly of pulmonary valve replacement either surgically or percutaneously and dilatation or stenting of pulmonary branches. The modified Blalock Taussig shunt mentioned in the Table was placed in a patient with stenosis of a hypoplastic left pulmonary artery. Freedom from pulmonary valve replacement was 88%, 73%, 60% and 27%% at 5, 10, 15 and 20 years respectively (Figure 3).

Echocardiography

Echocardiographic data after correction at the last check, showed in 18 patients (75%) a reasonable or good right ventricular function (RVF). Four patients (17%) had a moderate RVF. Only 2 patients (8%) had a severely impaired RVF.

The tricuspid regurgitation was trivial, mild, and moderate in 10 (42%), 10 (42%) and 4 patients (16%), respectively. The pulmonary regurgitation was absent/trivial, mild, moderate or severe in 6 (26%), 8 (35%), 6 (26%) and 3 (13%), respectively. The right ventricular dilatation was absent, mild, moderate and severe in 2 (8%), 4(17%), 12 (50%) and 6 (25%), respectively. If measurable, the median right ventricular (RV) pressure was estimated at median of 54 (25-108) mmHg. The median estimated pressure across the homograft is 19 (7-49) mmHg. Inherent thereto, the calculated pressure differences of 32 (0-95) mmHg suggests increased pulmonary artery pressures.

In 7 patients a small residual VSD was present, without hemodynamic significance in terms of flow.

MR imaging

From 17 patients after correction we obtained detailed MR imaging with a median interval between correction and MR image of 15,6 years (range 9-22 years). Based on the calculations the median right ventricular ejection fraction (RVEF) was 44% (range 13-62%), the median left ventricular ejection fraction (LVEF) was 52% (range 29-64%), the median RV end diastolic volume was 190 ml (range 94-339 ml), indexed 105 ml/m2 (range 76-176 ml/m2) and the median pulmonary regurgitation fraction was 19% (range 0-50%).

Discussion

In patients with PA, VSD and SPCAs several strategies are reported during the last 30 years indicating the challenging management [2, 13-16]. In 1989 we choose to follow one strategy for all consecutive patients and have sticked to this protocol since then. This allows to present after 30 years our long term results showing that 81% of the patients surviving after unifocalization are suitable for biventricular correction with a conduit between RVOT and the unifocalized pulmonary artery system. We consider this an adequate success rate in the complete and unselected cohort of these complex patients also compared to other series and strategies [2, 14, 17-21]. The use of a central aorto-pulmonary shunt to promote growth in diminutive pulmonary arteries is still subject of several studies [15, 22]. In a preliminary report of our first six cases we concluded that central shunts did not preclude the need for unifocalization with no significant growth of the pulmonary arteries [23].

Over the years of working with the staged approach several aspects evolved. We reduced the diameter of the MBT shunts during our experience. We learned to avoid as much tissue of the SPCAs as possible. Closure of unnecessary collateral arteries and intrapulmonary anastomoses are two factors to prevent later problems with stenosis or dilatation. Nevertheless these problems do occur due to the histological characteristics of the collateral arteries [10]. During follow up several dilatation and stenting procedures may be necessary and in some cases adequate relieve of stenosis cannot be maintained at the long term.

After our initial experience with central shunts with disappointing results with regard to growth and increase of the pulmonary artery size [23] we use these procedure for diminutive pulmonary arteries to promote outgrowth as mentioned by other authors [24, 25]. In our opinion rehabilitation of the pulmonary artery as promoted by several authors [15, 26, 27] may be useful, but still does not preclude further unifocalization in most of our cases.

During long term follow up allograft degeneration and replacement occurred as expected. An explanation for this could be that these allografts are placed in a more extra-anatomic position and in patients with a younger mean age at correction. Also elevated right ventricular pressures, probably due to an abnormal pulmonary vascular bed, can contribute to this degeneration process [28].

Considering the complex patient group we realize that there life expectancy is below normal, but we lack long term data comparable with other with right-sided allograft placement i.e. tetralogy of Fallot [29].

A limitation of this study is the relatively small group of patients. However we treated all cases consecutively and without selection during a long period of time with the same strategy by the same team. Because of these numbers we did not perform sub analysis for example for patients operated later on after our learning curve.

In conclusion staged repair of PA, VSD and SPCAs offers an adequate solution for most patients with a high correction rate and low operative mortality. Surgical or catheter interventions are necessary during follow up for pulmonary artery stenosis and allograft degeneration. Long term survival seems to be diminished.

We continue to follow these patients to achieve more data on long term outcome. The function of the right ventricle is a point of concern Echocardiography and cardiac MRI showed moderate and severe dilatation of the right ventricle (75%) and impairment of the function in 25% of the patients. Additional genetic abnormalities, in particular 22q11 deletion seems to be risk factor for adverse outcome[30].

Another limitation is the retrospective collection of data which resulting in incomplete data on exercise testing and quality of life studies. We are also looking forward to compare our long term results to those of other strategies, to make better comparison between different methods for this challenging patient group.

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Figure Legends

Figure 1

Flowchart of our patient population. Subsequent series of patients following the staged protocol of unifocalization and correction for pulmonary atresia, ventricular septal defect and systemic-pulmonary collateral arteries.

Figure 2

Kaplan-Meier curve of the survival after complete correction.

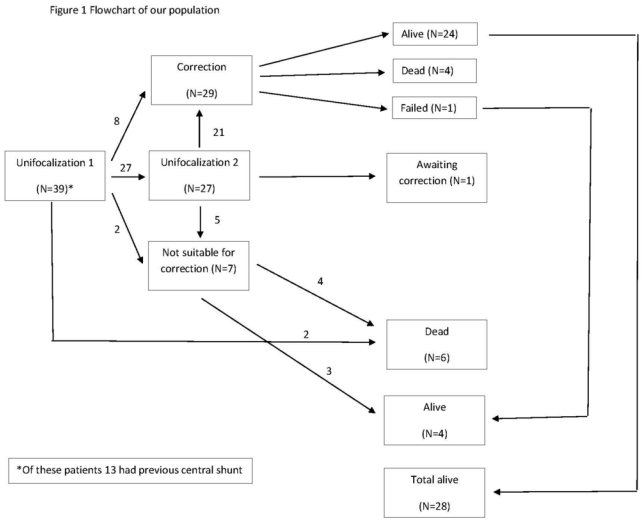
Figure 3

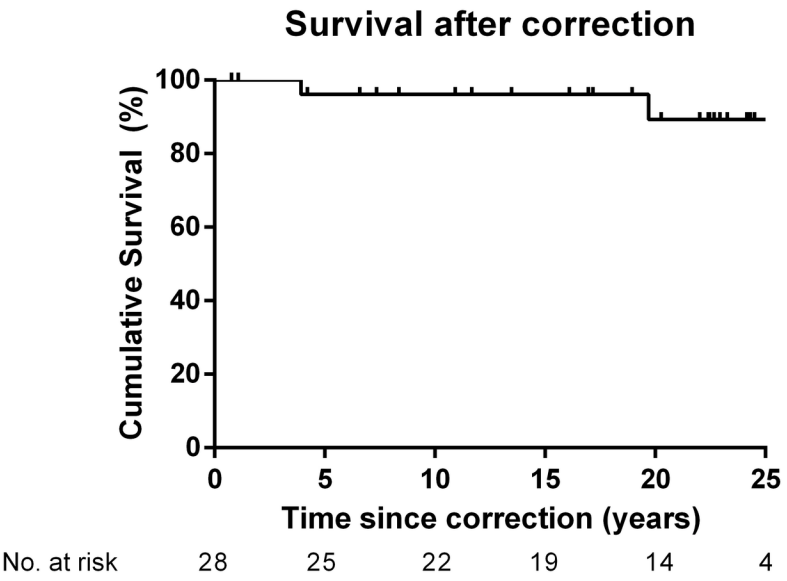
Kaplan-Meier curve of the freedom from pulmonary valve replacement after correction.

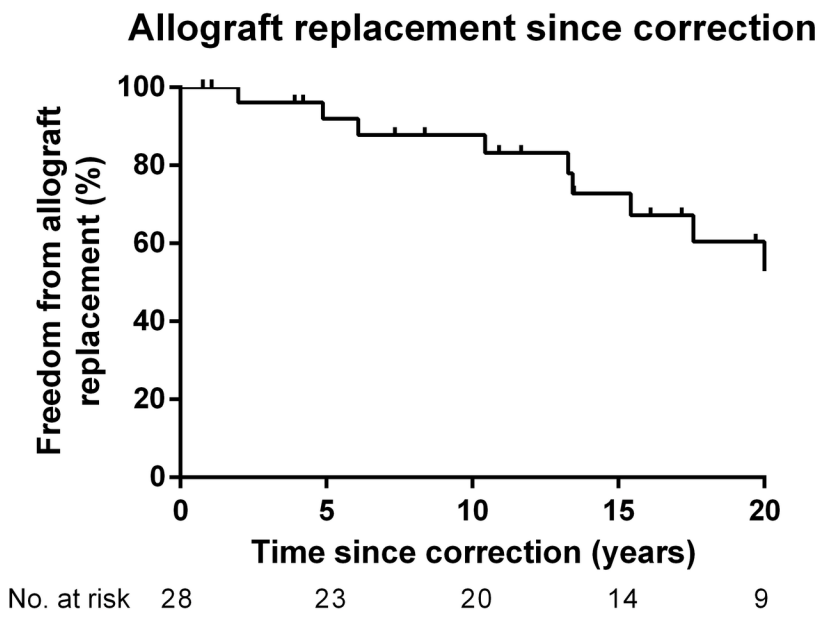
Table 1

Patient	Intervention
1	Melody valve implantation
2	Closure residual VSD, patch augmentation AP, allograft replacement
3	Relief of RVOT obstruction
4	Balloondilatation APS, allograft replacement, ascending aortic replacement, fenestrating ASD
5	MBT left, Melody valve implantation
6	Allograft replacement
7	Balloondilatation and stent APD
8	Allograft replacement
9	Closure residual VSD, allograft replacement
10	Balloondilatation APD, stenting APD and APS, Residual VSD, allograft implantation
11	Closure residual VSD, unifocalization right, Melody valve implantation
12	Allograft replacement
13	Stenting APS
14	Closure residual VSD
15	Balloondilatation and stent APD
16	Resternotomy for bleeding, stent APD and APS, allograft replacement and augmentation APD
17	Allograft replacement
18	Stenting APS, balloondilatation stent
19	Stenting APS and APD
20	Coil closure SPCA left

Table 1. Surgical procedures and catheter interventions during follow-up after successful correction.







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