Outcomes of Pulmonary Artery Reconstruction in Williams Syndrome

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Background. **The study sought to evaluate the short-term and midterm outcomes of surgical pulmonary artery reconstruction in patients with Williams syndrome (WS).**

Methods. **We performed a retrospective cohort study of all patients with WS who underwent surgical pulmonary artery reconstruction at Lucile Packard Children's Hospital between January 2001 and May 2018.**

Results. **There were 25 WS patients (52% female) who underwent pulmonary artery reconstruction during the study period. Median age at surgery was 2.4 (interquartile range [IQR], 0.9 to 4.5) years. Median preoperative right ventricular (RV) pressure was 80 (IQR, 70 to 90) mm Hg and aortic pressure was 96 (IQR, 90 to 107) mm Hg, with an RV-to-aortic pressure ratio of 0.8 (IQR, 0.7 to 1.0). The median number of pulmonary arterioplasty patches was 16.5 (IQR, 6.5 to 24). Median postoperative RV pressure was 27 (IQR 20 to 31) mm Hg and aortic pressure was 90 (IQR, 87 to 105) mm Hg, with an RV-to-aortic pressure ratio of 0.27 (IQR, 0.22 to 0.35). The postoperative RV pressure and RV-to-aortic pressure ratio were significantly lower than preoperative RV pressure and RV-to-aortic pressure ratio ($p < 0.0001$ for both). There was 1 (4%) postoperative death. In a median follow-up of 2.6 (IQR, 0.94 to 3.4) years, 1 (4.2%) patient has undergone RV outflow tract aneurysm repair and 2 (8.3%) patients have undergone balloon dilation of the pulmonary arteries.**

Conclusions. **Multilevel, surgical pulmonary artery reconstruction addressing severe extrapericardial stenoses is highly effective in patients with WS. This technique results in immediate normalization of RV pressure and has a low rate of reintervention in midterm follow-up.**

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Williams syndrome (WS), also known as Williams-Beuren syndrome, is a multisystem disorder occurring in approximately 1 in 10,000 live births [1] and resulting from a deletion of approximately 26 to 28 genes on chromosome 7q11.23 [2]. Cardiovascular pathology, largely comprising arterial stenoses, occurs in about 80% of cases [3] and results from the deletion of the *ELN* gene, which codes for the protein elastin [4]. Approximately one-third of patients with WS require intervention for arterial stenoses, with supravalvar aortic stenosis and peripheral pulmonary artery stenosis (PPS) comprising the large majority of those operative lesions [3].

PPS in patients with WS was first reported in 1964 by Beuren and colleagues [5]. Subsequently, PPS of at least a mild degree has been shown to occur in approximately 40% of all patients with WS [6]; however, for those who present in the first year of life, PPS is present in almost two-thirds of cases [7]. While these data are consistent with the long-held concept that PPS in WS generally tends to improve over time [6], a subgroup of patients with WS and PPS has severe disease with systemic or suprasystemic right ventricular (RV) pressures [8] that appear not to demonstrate the same spontaneous disease regression.

Data on surgical interventions for PPS in patients with WS are limited, having been reported in very few series. We have previously reported excellent results from our experience with surgical reconstruction of PPS in a mixed cohort of 16 patients with WS and Alagille syndrome [9]. Because our experience with WS has continued to grow and because there were only 7 patients with WS in our prior report, we sought to reevaluate our experience with pulmonary artery reconstruction in patients with WS.

Patients and Methods

Patients

With the approval of the Institutional Review Board of Stanford University, data were obtained for all patients with WS who had undergone pulmonary artery reconstruction at Lucile Packard Children's Hospital Stanford between January 2001 and May 2018. Data culled from the patients' clinical and surgical records included demographics, baseline clinical data, preoperative and postoperative hemodynamic data acquired at catheterization, intraoperative data from surgical and anesthesia

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reports, and echocardiographic data. Additionally, each patient's primary caregiver was contacted via telephone and current follow-up data were obtained, including any subsequent cardiac surgical or interventional catheterization procedures since the time of pulmonary artery reconstruction. In the cases when subsequent interventions had been performed, surgical or catheterization reports were obtained from the treating institution. Additionally, copies of the most recent echocardiograms were obtained from each patient's primary cardiologist, and these images were reviewed by a cardiologist (R.T.C.). All patients with WS who are evaluated at our program and have evidence of RV hypertension, as defined by a high pressure estimate based on tricuspid regurgitation velocity, RV hypertrophy, or an abnormal ventricular septal position (flattening in systole), undergo cardiac catheterization to assess pressures and characterize the pulmonary arterial tree angiographically.

Surgical Technique

We have previously reported the details of the surgical technique used for pulmonary artery reconstruction in PPS at our institution [10, 11]; however, owing to the surgical complexity of this population, we reiterate our approach here. Additionally, our surgical technique has undergone progressive evolution. Specifically, the early experience relied on a limited number of patches aimed at addressing major areas of obstruction. As outlined subsequently, in more recent years, we have begun dividing the main and branch pulmonary arteries, thus allowing greater exposure and improvement in the reconstruction. Further, we have developed the technique of the V-plasty for ostial stenosis repair [10]. This has represented a significant advancement in our surgical technique.

To achieve the most optimal postoperative hemodynamics, we address all hemodynamic abnormalities concomitantly, and the procedures are performed through a median sternotomy. Even in the setting of significant coronary artery stenosis, the pulmonary artery reconstruction is addressed first for 2 reasons. If the left-sided lesions were addressed first, there would be a significant amount of bleeding from the aortic and coronary artery suture lines, thus creating difficulty with the pulmonary artery reconstruction. Additionally, our experience has shown that the combination of cardiopulmonary bypass, hypothermia, and left ventricular venting reduces myocardial oxygen consumption sufficiently, such that any effect of coronary stenosis on myocardial perfusion is rendered insignificant.

In addressing the pulmonary arteries, the main pulmonary artery and the right and left branch pulmonary arteries are dissected free from the surrounding structures (Fig 1A). Before the extra-pericardial portion of the pulmonary artery dissection, both pleural spaces are opened, and the course of both phrenic nerves is marked with fine Prolene sutures (Ethicon, Somerville, NJ). The lobar and segmental branches are sequentially identified and the external anatomic appearance is compared with the preoperative angiogram. The entire surgical dissection is performed before administration of heparin to achieve complete hemostasis.

After dissection of the pulmonary arteries, cardiopulmonary bypass is instituted at a flow rate of 100 $mL \cdot kg \cdot min$ and the patient is cooled to 25°C. The entirety of the pulmonary artery reconstruction is performed with the heart beating and a vent catheter inserted through the right superior pulmonary vein into the left ventricle. The main pulmonary artery is then divided distally and the right and left branch pulmonary arteries are separated. Neuroclips are placed on each of the lobar branches to control back-bleeding.

The right and left branch pulmonary arteries are incised along the inferior and medical aspect of each. The incision is continued distally after the medial border of the artery and extended into the medial basal segment of the lower lobe. This exposure provides direct visualization of the lobar and segmental ostia. The ostia are probed with metal dilators to assess the degree of stenosis. While this direct assessment usually corresponds with the angiographic appearance, it is not uncommon to realize additional stenoses not visualized on the prior angiograms due to a concatenation of shadows or the angle of projection. Stay sutures are placed at the upper edges of the branch pulmonary arteries to facilitate exposure (Fig 1B). The orientation of the incision is critical, as the opening of the artery should face into the free area and away from the superior pulmonary veins and bronchi. The patients we have operated with WS and PPS typically have systemic or near-systemic RV pressures, and therefore have ostial stenoses of nearly every lobar and segmental orifice.

Stenoses at each of the 23 potential branch points are addressed sequentially, and any subsegmental ostial stenoses are addressed as well. As a result, the total number of sites of augmentation may exceed 25 or 30 to address completely the PPS. The segmental and subsegmental stenoses are often amenable to surgical ostioplasty using a Heineke-Mikulicz technique (V-plasty), particularly in the presence of ostial stenosis with a normal distal vessel. The V-plasty technique is ideally suited to smaller vessels arising at acute angles of less than 30 degrees. V-plasties are quite amenable to the majority of the ostial stenoses, as these are typically short and can be repaired quite efficiently with this technique. The V-plasty is performed by incising the carina perpendicular to the carinal axis and extending this incision along both vessels beyond the area of stenosis. The "V" that is created is then sutured together with 8-0 Prolene. The final result is a suture line that is rotated 90 degrees from the original orientation of the carina. Two major advantages of the V-plasty technique include a tissue-to-tissue anastomosis and the fact that a V-plasty can be performed in one-third to one-fourth the amount of time compared with patch augmentation.

There are other circumstances where patch augmentation is required. Indications for patch augmentation include long-segment stenoses of larger vessels (eg, the intermedius artery leading to the basilar segments) or when a smaller vessel comes off at a perpendicular angle.

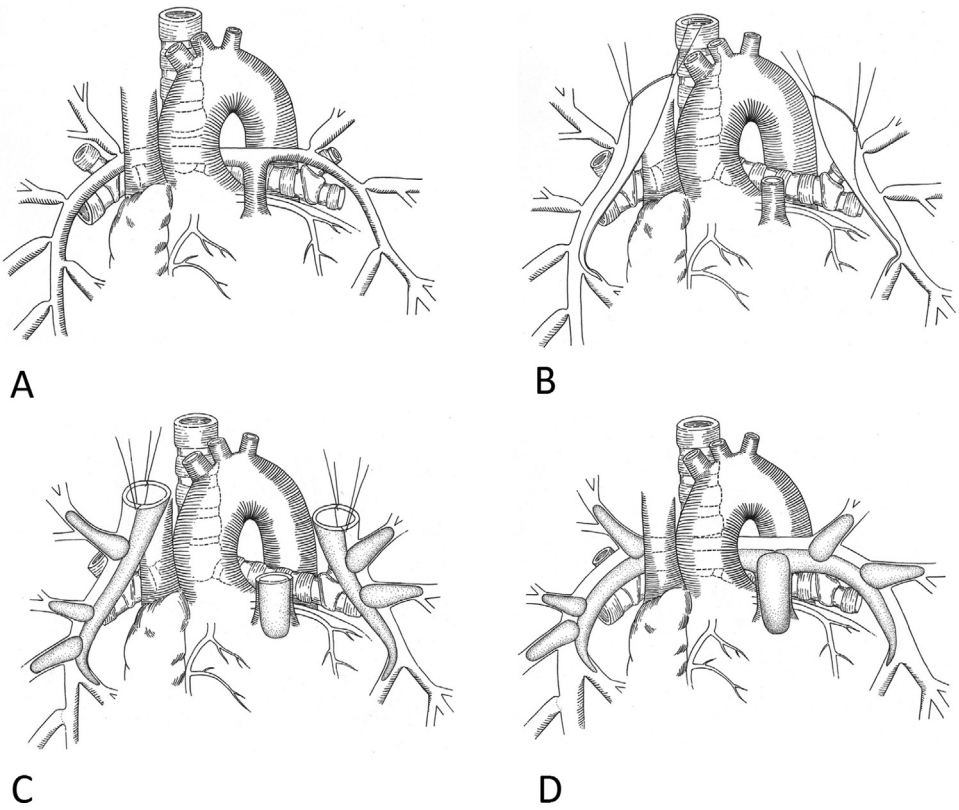

Fig 1. Illustration demonstrating the pulmonary artery anatomy and surgical technique in patients with Williams syndrome and peripheral pulmonary artery stenosis undergoing pulmonary artery reconstruction. (A) Typical anatomy with multiple lobar and segmental stenoses. (B) The initial step in pulmonary artery reconstruction. The main pulmonary artery is divided, as are the right and left branch pulmonary arteries. An incision is made along the inferior aspect of the proximal branch pulmonary arteries and continues along the medial border of the lower lobe artery. (C) The surgical reconstruction of the peripheral pulmonary artery stenoses. An ostioplasty has been performed to the basilar segment of the right lower lobe. Separate homograft patches have been performed to augment the segmental and lobar branch stenoses. A long, homograft patch has been used to augment the branch pulmonary arteries. (D) Reestablishment of branch pulmonary artery continuity and anastomosis of the branch pulmonary arteries to the reconstructed main pulmonary artery. (Reproduced from Mainwaring and colleagues [11] with permission from The Annals of Thoracic Surgery; illustrations by Erin Anne Mainwaring.)

The patch augmentations are performed with pulmonary artery homograft and are sutured in place with 8-0 Prolene. After all lobar and segmental branches have been addressed, the branch pulmonary artery is augmented with a long, pulmonary homograft patch, as shown in Figure 1C.

At this juncture, attention is directed to repair of the left-sided stenoses. In the presence of supravalvar aortic stenosis, we utilize direct cannulation of the innominate artery to facilitate the repair. The aorta is cross-clamped and cardioplegia is administered to achieve electromechanical quiescence. We have performed either a 2- or 3-sinus repair of the aorta using aortic homograft patch, depending on the nature and severity of the supravalvar aortic stenosis. The coronary ostia are inspected under direct vision and probed with metal dilators. Coronary stenoses are repaired by making an incision in the aorta in line with the direction of the coronary artery starting approximately 1 cm from the coronary ostium. The incision is carried out onto the coronary artery through the area of stenosis and beyond for approximately 5 to 6 mm of the nonstenotic segment. The incision is then augmented with a homograft patch. After completion of the coronary and aortic reconstruction, the aortic cross-clamp is removed and rewarming is initiated. Finally, the pulmonary arteries are anastomosed back together (Fig 1D); this step is often accompanied by a LeCompte maneuver due to space limitations through the aortopulmonary window. The limited space of the aortopulmonary window may be related the stenotic branch pulmonary arteries typical in patients with WS, but it may also be an acquired issue after the repair of supravalvar aortic stenosis. In addition to addressing space limitations, the LeCompte maneuver also moves the reconstructed and enlarged branch pulmonary arteries away from the airway, thereby potentially obviating postoperative airway compression.

As a result of preoperative chronically elevated RV pressures in our patients with WS and PPS, the RV is typically markedly hypertrophied. For this reason, to prevent the exacerbation of postoperative dynamic RV outflow tract obstruction, we avoid the use of epinephrine, preferring instead low-dose dopamine and milrinone. Two transthoracic pressure lines are placed to

monitor left atrial and RV pressure. The patient is weaned from cardiopulmonary bypass. Modified ultrafiltration is not used; however, all cases require a large amount of blood and blood products. Ventricular function and presence or absence of a gradient across the RV outflow tract are assessed with transesophageal echocardiography. The RV-to-aortic peak systolic pressure ratios are recorded in the operative note at the conclusion of the procedure. The chest is rarely left open, with the exception of when there is significant pulmonary dysfunction.

Statistical Methods

Descriptive statistics were expressed as median (interquartile range [IQR]) for continuous variables and percentage and frequency for categorical variables. Freedom from death or reintervention was analyzed using Kaplan-Meier analysis.

Results

Twenty-five patients underwent surgical repair of PPS during the study period. Of those, 14 underwent concomitant supravalvar aortic stenosis repair, with patch augmentation of the coronary arteries in 4. Eleven of the 25 (44%) patients had undergone prior surgical interventions at outside centers, including repair of supravalvar aortic stenosis and interhilar branch pulmonary artery augmentation. The preoperative characteristics of the 25 study patients are reported in Table 1. There was 1 (4%) operative mortality in a patient with severe PPS and supravalvar aortic stenosis. This patient underwent repair of both the PPS and ascending aorta, but postoperatively had depressed left ventricular function, developed multisystem organ dysfunction, and subsequently expired. Notably, this patient did not undergo the type of multilevel pulmonary artery reconstruction typically employed in the remainder of the study cohort. Another patient had a cardiac arrest with induction of anesthesia in the operating room but was resuscitated uneventfully. A different patient received postoperative mechanical circulatory support due to severe, reperfusion injury of the lungs, but subsequently recovered uneventfully. These 3 events represent a rate of major adverse cardiovascular events of 12% (n = 3 of 25).

Table 1. Characteristics of Patients With Williams Syndrome Who Underwent Pulmonary Artery Reconstruction

Variable	Result
Age at surgery, years	2.4 (0.9–4.5)
Female	13 (52)
Height, cm	83 (66–101)
Mass, kg	10.5 (7.2–13.6)
Preoperative right ventricular pressure, mm Hg	80 (70–90)
Preoperative left ventricular pressure, mm Hg	96 (90–107)
Preoperative right ventricle-to-left ventricle pressure ratio	0.84 (0.7–1.0)

Values are median (interquartile range) or n (%).

As reported in Table 2, the patients underwent a median of 16.5 (IQR, 6 to 24) homograft patch augmentations or Heineke-Mikulicz osteoplasties, with a median cardiopulmonary bypass time of 419 (IQR, 240 to 568) minutes. As shown in Figure 2, the median postoperative RV-to-aortic peak systolic pressure ratio was significantly lower than the preoperative ratio (0.27 [IQR, 0.22 to 0.35] versus 0.84 [IQR, 0.7 to 1.0]; $p < 0.0001$). This represents a 68% reduction in the RV-to-aortic pressure ratio. Figure 3 demonstrates typical pre and postoperative angiograms in a patient with WS who underwent surgical pulmonary artery reconstruction.

As shown in Table 3, the majority of patients underwent procedures to address bilateral outflow obstruction. The large majority of those patients underwent repair of supravalvar aortic stenosis, while 2 underwent procedures to address arch obstruction.

Of the 24 patients who survived to hospital discharge, 23 had documented follow-up within the last year, and 1 patient had been lost to follow-up immediately after hospital discharge. During a median duration of follow-up of 2.6 (IQR, 0.9 to 3.41) years, there has been no late mortality in the 24 operative survivors. One (4.2%) patient has subsequently had a repair of a RV outflow tract aneurysm 2.9 years after pulmonary artery reconstruction. Two (8.3%) other patients underwent balloon dilation angioplasty during follow-up after pulmonary artery reconstruction, one 9 months postoperative and the other at 12 months. There have been no reinterventions undertaken in the remaining 21 patients. Kaplan-Meier analysis demonstrated 96% survival for all patients with WS operated for PPS during the study period. Overall, in the 25 patients in the cohort, freedom from death or reintervention was 76% in follow-up to 15 years (Fig 4). In the 23 patients in whom recent follow-up could be attained, all were doing well clinically without the need for reintervention, cardiac-related hospitalization, or concern for RV hypertension.

Comment

This study demonstrates that peripheral pulmonary artery reconstruction addressing severe extrapericardial

Table 2. Operative Characteristics of Patients With Williams Syndrome Who Underwent Pulmonary Artery Reconstruction (N = 25)

Variable	Result
Bypass time, minutes	419 (240–568)
Cross-clamp time, minutes	44 (0–60)
Total number of pulmonary artery patches	16.5 (7–24)
Delayed sternal closure	5 (20)
Total hospital length of stay, days	13 (9–21)
Mortality	1 (4)
Postoperative right ventricular pressure, mm Hg	27 (20–31)
Postoperative left ventricular pressure, mm Hg	90 (87–105)
Postoperative right ventricle-to-left ventricle pressure ratio	0.27 (0.22–0.35)

Values are median (interquartile range) or n (%).

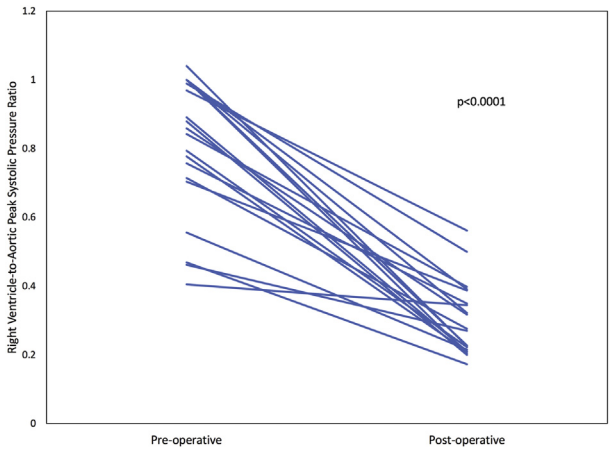

Fig 2. Line graph comparison of preoperative and postoperative right ventricle-to-aortic peak systolic pressure ratios. The right ventricle-to-aortic pressure ratios were significantly lower postoperatively ($p < 0.0001$).

stenoses is highly effective in patients with WS. The method yields significant, immediate decreases in the RV-to-aortic pressure ratio, returning the RV pressure to near-normal levels. Further, the improvement in the RV pressure is maintained over multiple years.

Surgical series of pulmonary arterioplasty in patients with WS are lacking. Other than our paper describing results in 7 patients with WS in a mixed cohort, Collins and colleagues [3] have reported some data on interventions for PPS in a relatively large cohort of 270 patients with WS. In that study, of the 99 patients with PPS, interventions for PPS were undertaken in only 17. Of those, surgical pulmonary arterioplasty was undertaken in only 8, whereas the majority of PPS interventions were catheter based. Specific reintervention data were not included in that study. However, recently Cresalia and colleagues [12] have reported their results of surgical pulmonary arterioplasty in a cohort that explicitly excluded patients with WS. Reinterventions were undertaken in one-third of the patients in their study, as compared with our total reintervention rate of 13%. Additionally, the majority of our patients underwent combined procedures for bilateral outflow tract obstruction, and a significant number had undergone prior surgical procedures at other institutions. These results suggest our surgical technique for pulmonary artery reconstruction is superior to the standard, interhilar surgical pulmonary arterioplasty.

Our mortality rate of 4% is consistent with that of Cresalia and colleagues [12], as well as that in a recent study of patients with WS from the Society of Thoracic Surgeons database. Hornik and colleagues [13] reported an overall mortality rate of 4.5% in patients with WS who underwent cardiac surgery. Additionally, 9.2% of patients suffered a major adverse cardiovascular event (defined as

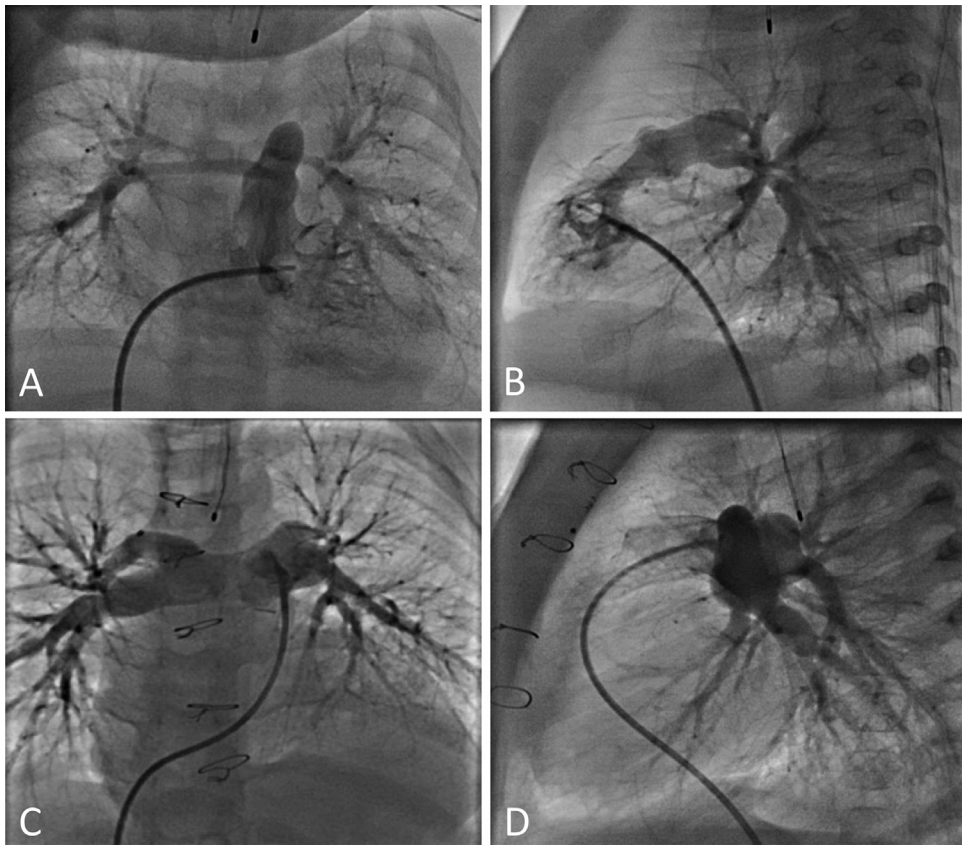

Fig 3. Typical angiograms in a patient with Williams syndrome before and after surgical pulmonary artery reconstruction. (A) A preoperative, anteroposterior projection of a Berman catheter in the right ventricle. Contrast injection demonstrates severe bilateral pulmonary artery stenoses of the branch, segmental, and subsegmental arteries. (B) The lateral projection of the same contrast injection in panel A. (C) A postoperative anteroposterior projection of a Berman catheter in the main pulmonary artery. The multilevel, bilateral pulmonary artery stenoses are almost entirely resolved. (D) The lateral projection of the same contrast injection in panel C. Notably, this patient's preoperative right ventricular-to-aortic pressure ratio was 1, and the postoperative right ventricular-to-aortic pressure ratio was 0.37.

Table 3. Surgical Procedure Data in Patients With Williams Syndrome Who Underwent Repair of Peripheral Pulmonary Artery Stenosis

Patient	Age (Years)	Sex	Concomitant Surgical Procedure(s)
1	2.5	M	None
2	1.0	F	SVAS repair, ASD closure, coronary artery augmentation
3	7.4	M	SVAS repair, long-segment patch augmentation of the aortic arch
4	2.5	M	Pulmonary artery stent explantation, main pulmonary artery augmentation
5	2.4	F	VSD closure, long-segment patch augmentation of the aortic arch
6	3.5	F	SVAS repair, tricuspid valvuloplasty, patch augmentation of SVC obstruction
7	7.3	F	None
8[a]	3.1	M	SVAS repair, coronary artery augmentation
9	7.0	F	None
10	0.2	F	SVAS repair, main pulmonary artery augmentation, coronary artery augmentation
11[b]	12.1	F	Stent explantation, pulmonary artery augmentation, pulmonary valvuloplasty, resection of subpulmonary infundibular obstruction, ASD closure
12	6.5	M	Main pulmonary artery augmentation, pulmonary valve commissurotomy, ASD closure
13	1.5	F	SVAS repair, long-segment patch augmentation of the aortic arch, main pulmonary artery augmentation
14	0.7	M	SVAS repair, main pulmonary artery augmentation
15	0.5	F	SVAS repair, coronary artery augmentation, repair of double aortic arch, pulmonary valve commissurotomy, ASD closure
16	0.9	M	SVAS repair, aortopexy
17[c]	0.5	M	SVAS repair, main pulmonary artery augmentation
18[a]	1.3	F	SVAS repair, repair of right pulmonary artery aneurysm
19	0.03	F	SVAS repair, main pulmonary artery augmentation, VSD closure
20	1.9	F	Main pulmonary artery augmentation, ASD closure
21	0.2	M	SVAS repair, main pulmonary artery augmentation, repair of right pulmonary artery dissection
22	1.6	M	Main pulmonary artery augmentation, aortic arch-to-descending aorta interposition graft
23	4.5	M	Main pulmonary artery augmentation
24	5.4	F	None
25	3.1	M	ASD closure

[a] Underwent pulmonary artery balloon dilation angioplasty during follow-up. [b] Underwent right ventricular outflow tract aneurysm during follow-up. [c] Postoperative mortality.

ASD = secundum atrial septal defect; F = female; M = male; SVAS = supravalvar aortic stenosis; SVC = superior vena cava; VSD = ventricular septal defect.

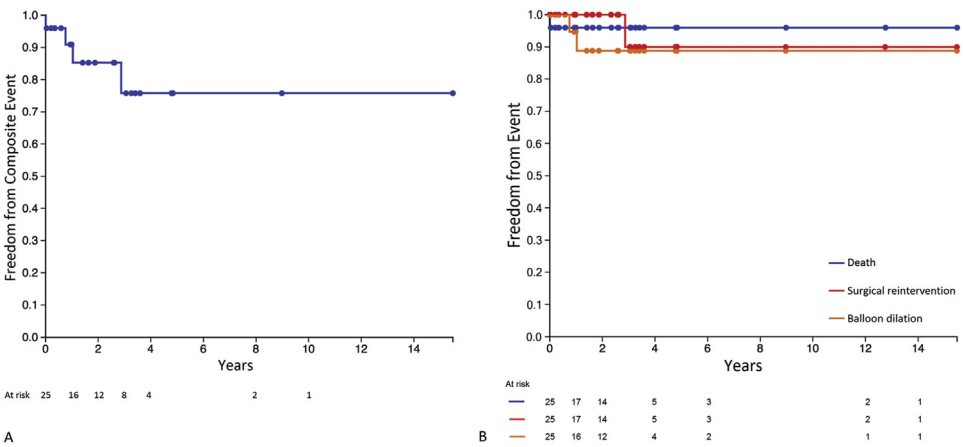

Fig 4. Kaplan-Meier analysis in 25 patients with Williams syndrome who underwent pulmonary artery reconstruction during the study period. (A) Freedom from the composite event of death, surgical reintervention, and transcatheter reintervention (balloon dilation). The freedom from death or reintervention was 76% at up to 15 years of follow-up. (B) Individual curves for freedom from death, surgical reintervention, and transcatheter reintervention.

death, cardiac arrest, or mechanical circulatory support). However, in that study, the large majority of patients (78%) underwent procedures addressing unilateral outflow tract obstruction. The rate of a major cardiovascular event was significantly higher in those with complex left ventricular outflow tract obstruction (9%) and bilateral outflow tract obstruction (22%). In the light of those results from Hornik and colleagues [13], our results in patients with WS, the majority of whom underwent procedures to address bilateral outflow tract obstruction, have been excellent.

While not our management approach, transcatheter balloon dilation and stenting of PPS have been the primary mode of intervention in patients with WS [3]. This approach in patients with WS was first reported by Geggel and colleagues [8]. However, the literature reporting transcatheter dilation of PPS is not particularly convincing with regard to benefit in WS. In their original study in 25 patients, Geggel and colleagues [8] reported that while there was angiographic improvement in the size of the proximal pulmonary arteries, the mean RV pressure did not change as a result of balloon dilation. Additionally, the difference in the RV-to-aortic pressure ratio only decreased greater than or equal to 20% in 4 of the 39 procedures. Thereafter, Cunningham and colleagues [14] reported on transcatheter outcomes in 69 patients (23 of whom had WS) who underwent transcatheter dilation of PPS. In that study, the RV-to-aortic pressure ratio did not change in patients with WS. Additionally, the need for reintervention was 75% at 5 years for a transcatheter reintervention and 23% at 5 years for surgical reintervention, with the majority of surgical reintervention undertaken in the first postoperative year. Further, pulmonary artery aneurysms occur in 18% of cases who have undergone balloon dilation [8], and rupture of those aneurysms has been reported [14]. While the present study did not seek to compare the outcomes of multilevel surgical pulmonary artery reconstruction to those of transcatheter interventions, a straightforward assessment of the published data on transcatheter interventions in the light of our surgical results suggests a multilevel surgical approach to PPS in WS has better outcomes, not only with regard to the immediate postoperative RV-to-aortic ratio, but also with regard to the need for further reintervention. Continued follow-up in our cohort over time will allow for a better assessment of the long-term durability of our surgical approach.

The risk of sudden death in WS has been estimated to be 25 to 100 times higher than that of the general population [15]. The majority of reported cases of sudden cardiovascular collapse have occurred in the setting of anesthetic administration. The etiology has most often been attributed to the concomitant presence of supravalvar aortic stenosis and coronary ostial stenosis leading to decreased myocardial perfusion when the coronary perfusion pressure decreases in the setting of anesthetic-induced hypotension [16], though this may be an incomplete explanation [17]. However, with the institution of detailed periprocedural planning and anesthetic management plans, the risk of adverse events can be mitigated greatly [18]. Our program uses a detailed risk stratification algorithm for patients with WS to direct periprocedural management, and no adverse events have occurred since its institution [19].

This study has several limitations. It is a retrospective analysis and is subject to the limitations inherent in such a design. Not all of the patients in the study underwent the same multilevel pulmonary artery reconstruction technique by the same surgeon. This did impact our results, as the 1 postoperative mortality was an outlier in regard to the surgical technique and surgeon. The cohort of patients is small and the excellence of our results could be impacted by the small sample size. While a straightforward review of our results compared with published transcatheter results favors our approach, this study was not designed to specifically address this question and no definitive statement can thus be made. Owing to a relatively recent increase in our surgical volume of patients with WS, the present study truly represents a midterm length of follow-up. It is possible the low reintervention rate in our study may be, in part, explained by our follow-up duration. However, the prior work of Cunningham and colleagues [14] suggests that, in patients with WS who have severe PPS such as ours, if reintervention is going to be needed, the large majority will need it within the first year after the primary intervention.

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ABTS 2019 Congenital Cardiac Surgery Subspecialty Written Examination

The 2019 Congenital Cardiac Surgery subspecialty written examination will be held Monday, December 9, 2019, at multiple sites throughout the United States using an electronic format. The closing date for applications is August 15, 2019. Those wishing to be considered for examination must apply online at www.abts.org.

A candidate applying for admission to the Congenital Cardiac Surgery subspecialty written examination must fulfill all the requirements of the Board in force at the time the application is received.

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