Pulmonary Artery Reconstruction After Failed Pulmonary Artery Stents

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Background. **Pulmonary artery stents are widely deployed in patients with stenoses in the branch pulmonary arteries. However, stents do not address more peripheral sites of stenosis and invariably develop in-stent restenosis. The purpose of this study was to review our experience with pulmonary artery reconstruction after failed pulmonary artery stents.**

Methods. **This was a retrospective study of 56 patients who underwent pulmonary artery reconstruction after failed pulmonary artery stents. These patients had undergone a median of 2 (range, 0-5) previous surgical procedures and 2 (range, 1-4) pulmonary artery stents.**

Results. **The median age at stent surgery was 5 (range, 0.3-23.6) years. The majority of stents (79%) were completely removed and patch augmented. The minority of stents (21%) were felt to be unremovable and thus were split longitudinally and reconstructed using a pulmonary artery homograft. There was 1 (1.8%) operative mortality. The mean pulmonary artery-to-aortic pressure ratio decreased from a preoperative value of 0.91 ± 0.21 to a postoperative value of 0.31 ± 0.07 ($P < .001$). The median hospital length of stay was 10 days. The median duration of follow-up was 1.8 years. There has been no midterm mortality. Six patients have undergone balloon dilation postoperatively for residual pulmonary artery stenosis.**

Conclusions. **Pulmonary artery reconstruction resulted in a significant decrease in pulmonary artery-to-aortic pressure ratios. The subsequent need for reintervention on the pulmonary arteries has been relatively low (11% to date). These results suggest that patients with pulmonary artery stents can be successfully treated with surgical reconstruction.**

(Ann Thorac Surg 2020;110:949-55)

There is a wide variety of congenital heart defects that may be associated with abnormalities of the pulmonary artery architecture. This includes all of the conotruncal defects,[1-4] as well as peripheral pulmonary artery stenosis associated with Williams and Alagille syndromes.[5-7] Pulmonary arterial stenoses result in an increase in right ventricular pressure, which is dependent on the number and severity of the stenoses. These pulmonary arterial stenoses also result in a maldistribution of pulmonary blood flow by directing flow away from the areas downstream from the stenoses and thereby concentrating blood flow to the areas supplied by unobstructed vessels. When severe, abnormalities in pulmonary artery architecture may result in severe right ventricular hypertension, leading to right heart failure and death.[8,9]

Historically, the treatment of pulmonary arterial stenoses has been divided into 2 anatomic domains. Stenoses located within the hilum have been viewed as amenable to a surgical treatment, whereas lesions outside the hilum were viewed as beyond the boundaries of surgery.[10,11] The pericardium served as the metaphysical boundary between lesions that could or could not be treated surgically. This surgical principle has become widely accepted and has been passed down decade by decade without a significant challenge to its veracity.

As a consequence of this theoretical boundary between what can and cannot be treated surgically, pulmonary artery stenoses located outside the hilum have been viewed as the domain for interventional catheterization techniques.[12-14] Pulmonary artery balloon dilation and pulmonary artery stents have become widely employed and accepted as the standard of care by the American Heart Association. It is estimated that more than 1000 pulmonary artery stenting procedures are performed in the United States each year. However, the efficacy of pulmonary artery balloon dilation and stenting for peripheral pulmonary artery stenosis has not been proven.

We have developed a systematic surgical approach to peripheral pulmonary artery stenosis.[15] The original application of this surgical approach was to address

Accepted for publication Jan 2, 2020.

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Published by Elsevier Inc.

0003-4975/$36.00
https://doi.org/10.1016/j.athoracsur.2020.01.007

pulmonary arterial stenoses associated with Williams, Alagille, and other nonsyndromic patients.[16-18]

The techniques that we have developed have also been applicable to many other forms of peripheral pulmonary artery stenosis.[19] These same techniques for pulmonary artery reconstruction can be applied to patients referred to our institution after failed pulmonary artery stents. The purpose of this study was to summarize our experience with pulmonary artery reconstruction in these patients.

Material and Methods

This was a retrospective review of patients who had a pulmonary stent placed during an interventional catheterization procedure and were subsequently referred to Stanford University for pulmonary artery reconstruction. The study was approved by the institutional review board at Stanford University (Protocol ID #50154, approved on March 7, 2019).

From January 2014 to August 2019, 56 patients were identified who met the previously mentioned entry criteria. Twenty-two (41%) patients had identifiable syndromes, including 8 with Alagille syndrome, 9 with DiGeorge syndrome, 3 with Williams syndrome, 1 with Down syndrome, and 1 with a partial deletion of chromosome 19.

The 56 patients in this study had a variety of underlying congenital heart defects, including pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries (n = 26), peripheral pulmonary artery stenosis (n = 11), tetralogy of Fallot (n = 10), truncus arteriosus (n = 3), transposition of the great arteries (n = 3), double outlet right ventricle (n = 2), and hypoplastic left heart syndrome (n = 1).

Fifty-four (96%) of the 56 patients had undergone 1 or more prior cardiac surgical procedure. The median number of prior cardiac surgeries was 2 (range, 0-5). This included 23 patients who had 1 previous cardiac operation, 19 patients who had 2, 7 patients who had 3, 5 patients who had 4, and 1 patient who had 5 (for a total of 107 previous surgical procedures).

The 56 patients underwent a median of 4 (range, 1-9) cardiac catheterizations before referral for pulmonary artery reconstruction (a total of 218 previous cardiac catheterizations). This included a median of 2 (range, 1-4) pulmonary artery stents. For the 56 patients, there were a total of 104 pulmonary artery stents deployed, including 25 patients who had 1 pulmonary stent, 19 patients who had 2, 7 patients who had 3, and 5 patients who had 4.

A histogram demonstrating the preoperative peak systolic pressure ratio values for the individual patients is shown in Figure 1. The mean preoperative central pulmonary to aortic pressure ratio was 0.91 ± 0.21.

The surgical technique that we have developed for pulmonary artery reconstruction in the presence of failed pulmonary artery stents is an adaption of many techniques that we have described previously for unifocalization revision surgery, peripheral pulmonary artery stenosis surgery, and other complex pulmonary artery reconstructions.[15-22] The procedures are performed through a median sternotomy. Ninety-six percent of the patients had undergone prior cardiac surgical procedures thus necessitating a redo sternotomy. The phrenic nerves are marked with fine monofilament suture to identify the location and lift up the nerve as the dissection continues more distally.[23]

The pulmonary arteries (or conduit) are dissected out to the level of the segmental branches before institution of bypass. This goal can usually be completely accomplished

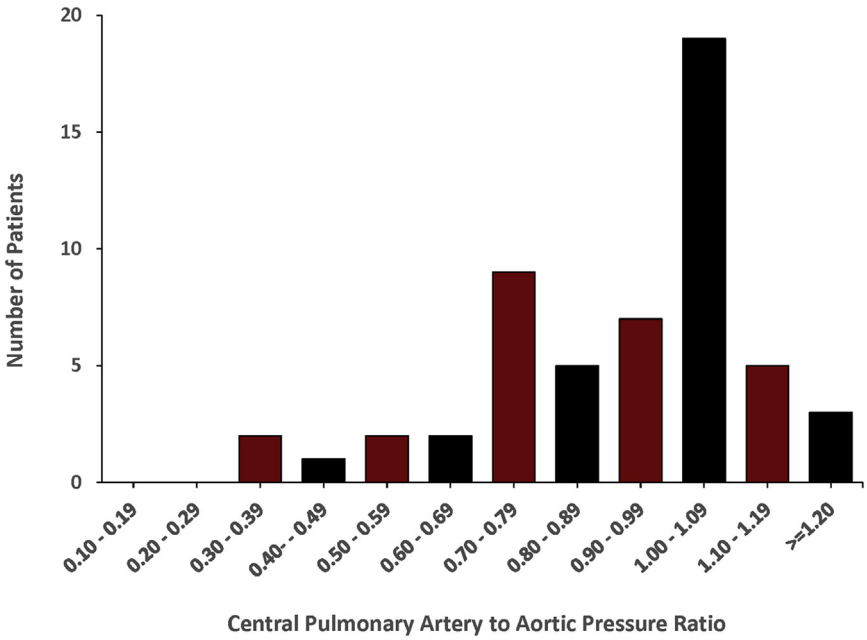

Figure 1. Histogram demonstrating the preoperative central pulmonary artery-to-aortic peak systolic pressure ratios for the individual patients.

on the right side and slightly less completely on the left because the presence of the main pulmonary artery or conduit and more posterior angle of take-off of the left pulmonary artery. It is important to perform as much of this dissection as possible before heparinization in order to prevent intraparenchymal hemorrhage. A typical case demonstrating bilateral pulmonary artery stents and peripheral pulmonary artery stenosis is shown in Figure 2A.

After going on bypass, the main pulmonary artery (or conduit) is divided, as are the right and left branch pulmonary arteries (Figure 2B). The individual branch pulmonary arteries are then incised along the inferior and medial border through the pulmonary artery stent into the lower lobe segmental vessels (Figure 2C). The site of the pulmonary artery stent is inspected and a decision is made regarding the feasibility of removing the entirety of the stent vs leaving a portion of the stent in place. This decision is based in part on whether the stent can be peeled away from the intima without destroying too much of the integrity of the artery (Figure 2D). The feasibility of peeling the stent out is often influenced by the duration of time that the stent has been in place. Stents that have resulted in "jailed" lobar origins (eg, the left pulmonary artery stent shown in the figures) carry a

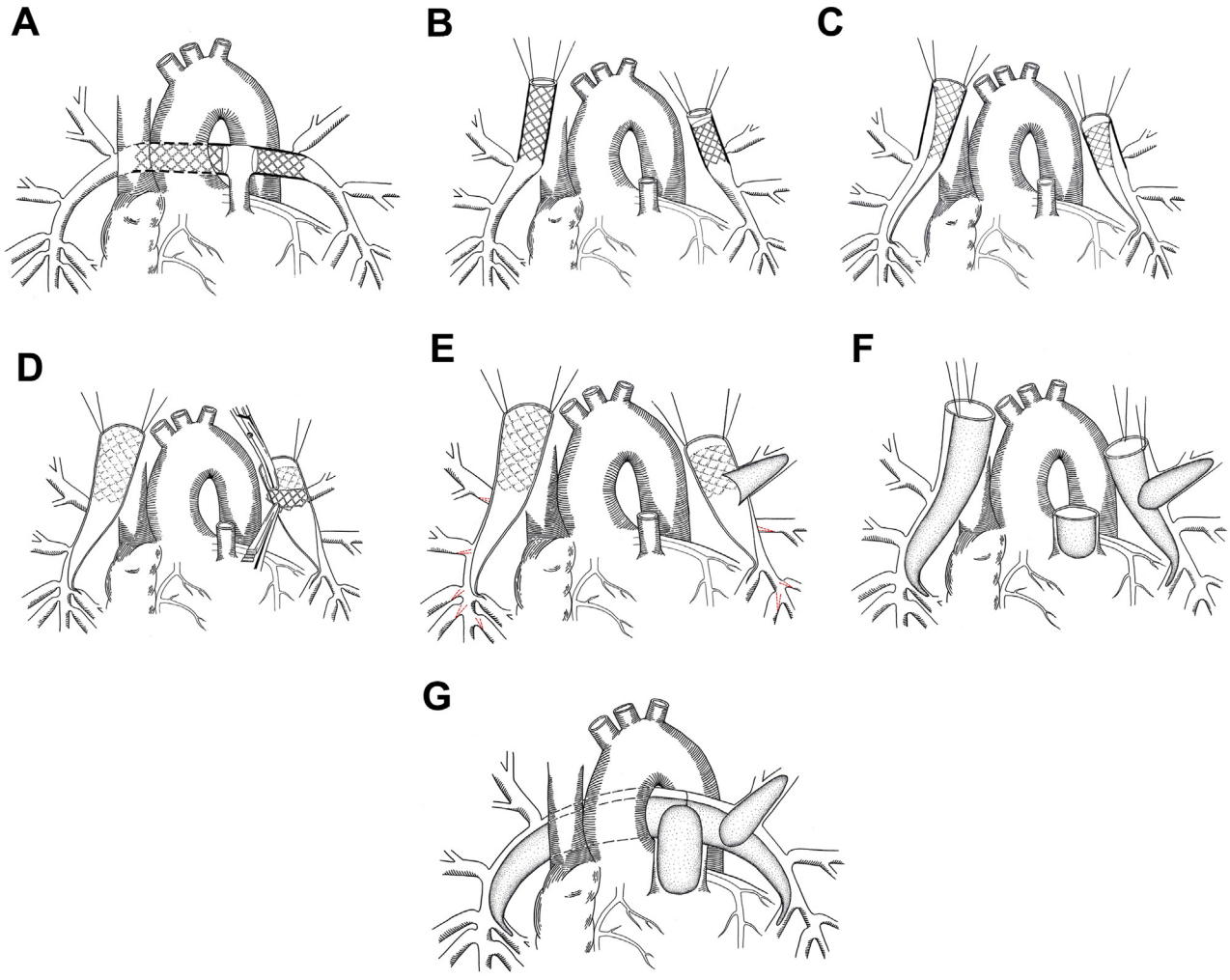

Figure 2. (A) Illustration demonstrating the typical anatomy of a patient who had peripheral pulmonary artery stenosis and previously underwent a hilum-to-hilum patch and placement of stents into the right branch pulmonary artery and left branch pulmonary artery . The left upper lobe orifice is "jailed" by the stent. (B) Illustration demonstrating division of the main pulmonary artery and division of the right and left pulmonary arteries. (C) Illustration demonstrating incisions along the inferior and medial aspect of the branch pulmonary arteries. These incisions cut through the stents and are then continued into the lower lobe medial segmental orifices. (D) Illustration demonstrating removal of the left pulmonary artery stent. This leaves a denuded surface without a true intima. By removing the stent, access is obtained to the previously jailed orifice of the left upper lobe. (E) Illustration demonstrating reconstruction of the multiple peripheral pulmonary arterial stenoses using the "V-plasty" ostial repair technique. The jailed left upper lobe branch is augmented with a pulmonary artery homograft patch. (F) The branch pulmonary arteries are augmented with a long pulmonary artery homograft patch. The main pulmonary artery has been augmented with an additional homograft patch. (G) Illustration demonstrating restoration of continuity between the right and left branch pulmonary arteries and reconnection to the main pulmonary artery. The illustration demonstrates the branch pulmonary arteries in the anatomic position. However, in some circumstances the pulmonary arteries are reconstructed in front of the aorta (LeCompte procedure).

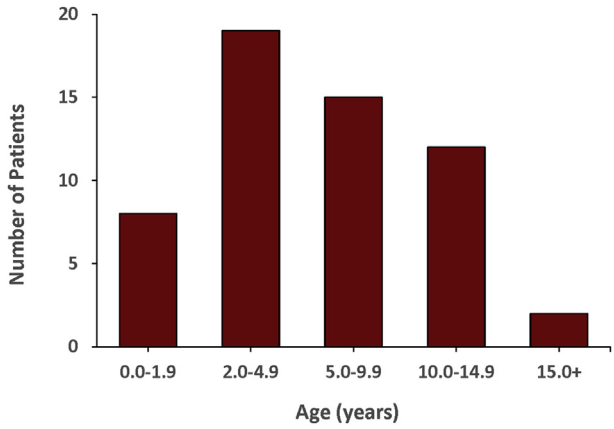

Figure 3. Histogram demonstrating the distribution of ages at the time of surgical pulmonary artery reconstruction. The median age was 5 years.

mandate for removal in order to facilitate reconstruction of the obstructed artery.

Peripheral pulmonary arterial stenoses are typically addressed by performing V-shaped ostioplasties (Figure 2E).[16-20] Pulmonary artery homograft is then used to patch augment the pulmonary artery including the previous stent site (Figure 2F). The branch pulmonary arteries are subsequently brought together and the main pulmonary artery (or aortic homograft conduit) reconnected to the reconstructed pulmonary arteries (Figure 2G).

Results are reported as mean ± SD or as median and range, where appropriate. A comparison of the preoperative to postoperative values was performed using Pearson's chi-square analysis.

Results

The median age of the patients at the time of pulmonary artery reconstruction was 5 (range, 0.3-23.6) years. The distribution of ages at the time of pulmonary artery reconstruction is shown in Figure 3.

Table 1. Concomitant Procedures Performed During the Pulmonary Artery Reconstruction

Procedure	Count
Conduit replacement	26
VSD repair	20
ASD repair	10
Conduit placement	6
Main pulmonary artery patch	6
Tricuspid valve repair	3
Infundibular resection	3
Aortic valve replacement	2
Supravalve aortic stenosis repair	2
Partial anomalous pulmonary drainage	2
Other	4

ASD, atrial septal defect; VSD, ventricular septal defect.

The median duration of cardiopulmonary bypass was 402 (range, 60-844) minutes. Twenty-three (41%) patients had a period of aortic cross-clamp, with a median duration of 34 (range, 12 to 162) minutes. A list of the concomitant procedures performed is summarized in Table 1.

During the surgical repair, 81 of the 104 (79%) pulmonary artery stents were completely removed, whereas 23 (21%) were deemed unremovable and therefore were split and subsequently patch-augmented. Fifty-one of the 56 patients underwent repair of peripheral pulmonary stenoses distal to the stents, with a median of 14 (range, 3-28) peripheral repairs performed.

A histogram demonstrating the postoperative central pulmonary artery to aortic peak systolic pressure ratios for individual patients is shown in Figure 4. The mean central pulmonary artery-to-aortic pressure ratio decreased from a preoperative mean value of 0.91 ± 0.21 to a postoperative mean value of 0.31 ± 0.07 ($P < .001$). This comparison is shown in Figure 5.

The median hospital length of stay was 10 (range, 4-59) days. Median duration of follow-up was 1.8 years (range, 1 month to 5.3 years). There has been no midterm mortality. None of the patients have to date undergone any reoperations on the branch or peripheral pulmonary arteries. One patient has undergone reoperation for repair of a pseudoaneurysm.

Fourteen (25%) patients have subsequently had a cardiac catheterization procedure. Six of these patients underwent balloon dilation of a residual pulmonary artery stenosis. These balloon dilations occurred at 7, 11, 22, 28, 36, and 48 months postoperatively. The Kaplan-Meier curve demonstrating freedom from death or reintervention on the distal pulmonary arterial bed is shown in Figure 6. These 6 patients underwent a total of 10 balloon dilations of the reconstructed pulmonary arterial bed. Six of the 10 balloon dilations were performed at the site of a previously stented pulmonary artery. Four of these 6 sites had residual stents that had been deemed unremovable and thus had been split and patch augmented. Two of the 6 sites were where stents had been completely removed. Finally, there were 3 patients who underwent 4 balloon dilations of the pulmonary arteries that were remote from any previous stent. In addition to these balloon procedures, there was 1 patient who underwent transcatheter pulmonary valve replacement.

Comment

This study was performed to evaluate our experience with pulmonary artery reconstruction in patients who had failed pulmonary artery stents. The data demonstrate that pulmonary artery reconstruction resulted in a significant decrease in pulmonary artery-to-aortic pressure ratios. At midterm follow-up, no patient has required surgical reintervention on the reconstructed pulmonary artery bed, and relatively few patients have undergone catheter-based interventions. These results suggest that surgical reconstruction can be highly effective in treating peripheral pulmonary stenoses and repair of the stent site after deployment of pulmonary artery stents.

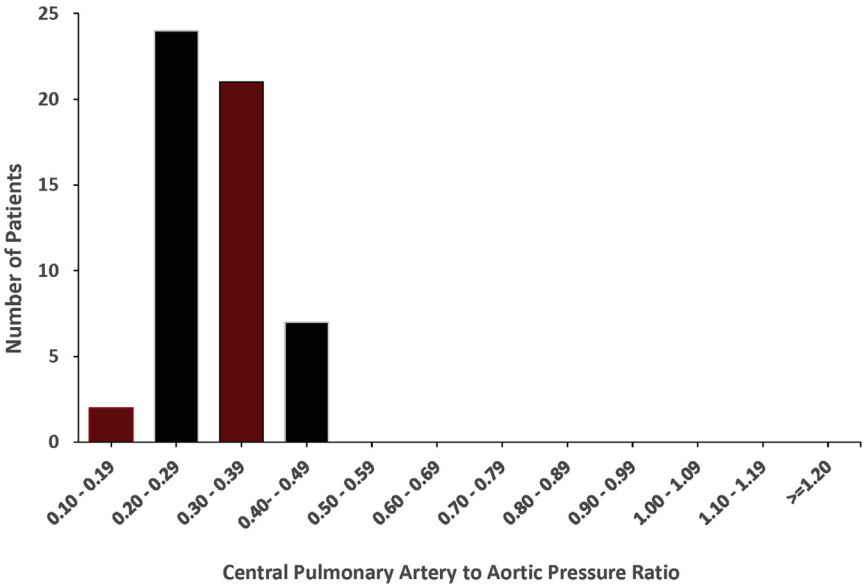

Figure 4. Histogram demonstrating the postoperative central pulmonary artery-to-aortic peak systolic pressure ratios for the individual patients.

We have developed a systematic surgical approach to the treatment of peripheral pulmonary artery stenosis.[15-18] This approach has evolved substantially over a relatively short time as more and more experience has been garnered at our center. One important modification is dividing the right and left branch pulmonary arteries. This allows the surgeon to pull up on the individual branch pulmonary arteries and reach the basilar segments of the lower lobes. This visualization cannot be achieved without division of the branch pulmonary arteries. By dividing the branch pulmonary arteries, each branch can be reflected laterally, which means that the incision down to the basilar segment origins is oriented immediately in front of the surgeon. In contrast, if continuity of the branch pulmonary arteries is maintained, the incision is on the underside of the vessel and makes it much more awkward to perform this work.

The development of the V-shaped ostioplasty for ostial stenoses has been another important modification to our armamentarium for peripheral pulmonary artery reconstruction.[19-23] This technique is the application of the Heineke-Mikulicz principle to a vascular reconstruction of pulmonary artery ostial stenoses. The technique can be applied to very small vessels, including segmental and even subsegmental branches. It has another theoretical advantage, in that it is an entirely tissue-to-tissue anastomosis. Finally, it is also much quicker to perform than a patch augmentation, in addition to being applicable to

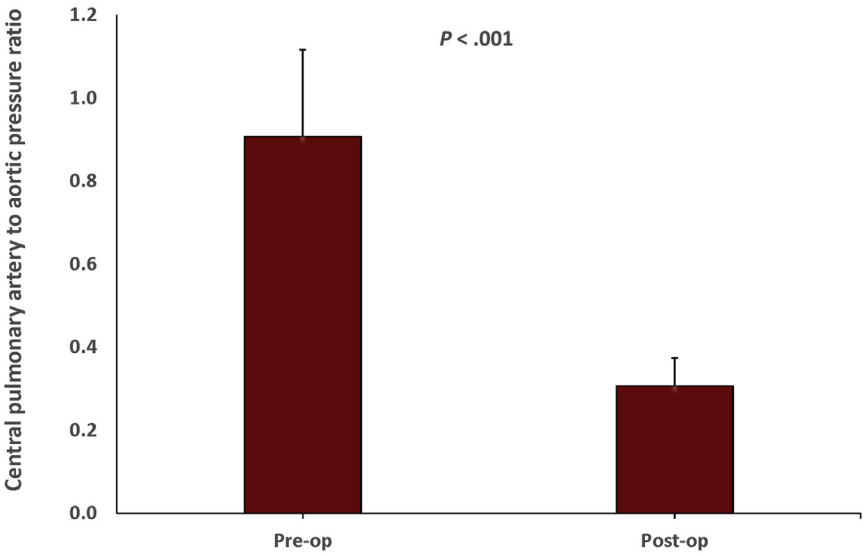

Figure 5. Bar graph demonstrating the pulmonary artery to aortic peak systolic pressure ratio preoperatively (pre-op) and immediately postoperatively (post-op). The mean pressure decreased from 0.91 ± 0.21 preoperatively to a postoperative mean value of 0.31 ± 0.07 ($P < .001$).

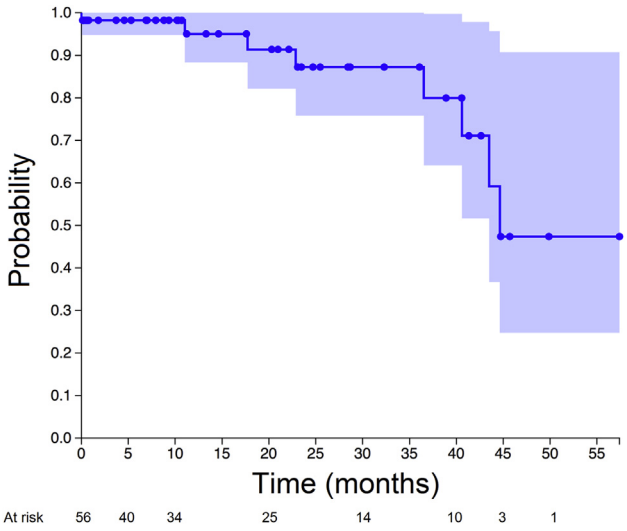

Figure 6. Kaplan-Meier curve demonstrating freedom from death or reintervention on the branch pulmonary arteries. This analysis predicts a freedom from reintervention of 87% at 2 years.

considerably smaller vessels. As a consequence, our group has been able to treat more of the peripheral disease than we could when we used an entirely patch-based repair.

The presence of pulmonary artery stents adds an extra layer of complexity to these procedures. From the outside, there is always an intense inflammatory reaction to the stent, and it is not uncommon that some of the tines have penetrated through the pulmonary artery wall. Cutting through stents is analogous to cutting through a chain-link fence or strands of barbed wire. On the inside, the stents are embedded in the pulmonary artery wall and covered by a fibrous neointima. The exposed portions of the stent are cut off, and an attempt is made to develop a plane and peel the stent away from the pulmonary artery wall. There are times when the entirety of the stent can be removed, as illustrated in Figure 2D. However, this is sometimes not feasible, particularly when the stents have penetrated through the arterial wall or have been in place for a lengthy period of time. In this event, the stent is bent open along its longitudinal axis, as shown in Figure 2E. After addressing the peripheral pulmonary artery stenoses, the branch pulmonary arteries are augmented with a homograft patch (as depicted in Figure 2F).

It is our recommendation that all patients undergo close surveillance after pulmonary artery reconstruction.[22] This surveillance includes an echocardiogram every 3 months, a lung perfusion scan at 6 months, and a cardiac catheterization at 1 year. Patients who have an indication that the right ventricular pressures have increased (based on the tricuspid regurgitation jet) or who have a significant change in lung perfusion scan distribution should undergo earlier cardiac catheterization to identify the underlying cause of the problem. Four of the 6 patients in this study who have undergone balloon dilation of their pulmonary arteries had some indication of a change in the pulmonary artery pressures before the procedure. These interventions occurred at 7, 11, 22, and 28 months after pulmonary artery reconstruction.

Conversely, there were 2 patients who underwent a routine surveillance cardiac catheterization at 36 and 48 months after pulmonary artery reconstruction and were found to have a site of stenosis amenable to balloon dilation. It is not surprising that the 4 patients who had an indication of increased right ventricular pressures underwent more interventions than did patients who were undergoing routine surveillance (median of 2 vs 1).

There have been 8 additional patients who have undergone a surveillance cardiac catheterization and were not found to be in need of any intervention. Thus, 2 of 10 (20%) patients undergoing routine surveillance cardiac catheterization were found to have a site of distal stenosis. It is worth noting that many patients are well past due for their postoperative cardiac catheterization. It is probable that more patients will eventually undergo interventions, given the relatively short follow-up for some patients.

It is interesting that a disproportionate number of balloon dilations occurred at the site of a stent that could not be removed and therefore were split and patch-augmented. Specifically, 4 of 23 (17%) stents that were not removed subsequently required balloon dilation, compared with 2 of 81 (3%) for stents that were completely removed. It is not particularly surprising that an area comprising old stent and patch material would be prone to the development of restenosis. To date, the majority of patients (89%) who underwent pulmonary artery reconstruction in the setting of a previous stent have not required any reintervention, and no patients have required surgical reconstruction of the pulmonary arterial tree.

The patients included in this study are reasonably representative of the anatomic diagnoses and syndromes associated with abnormalities of the pulmonary artery architecture. Specifically, 44% of the patients had the diagnosis of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries, and 26% had peripheral pulmonary artery stenosis. Consistent with these anatomic diagnoses, 17% of the patients had DiGeorge syndrome[18,23] and an additional 15% had Alagille syndrome.[24] While the literature suggests that these syndromes may be associated with increased operative risk,[2-7] our experience in this cohort indicates that these theoretical obstacles can be successfully navigated.

The most common application of pulmonary artery stents is placement in the proximal branch pulmonary arteries. As this is the same domain as conventional surgical techniques, there are dual options for treatment of lesions within the hilum. The decision to recommend surgery vs pulmonary artery stenting is most often dictated by the presence or absence of concomitant lesions. Patients who require surgical treatment of intracardiac lesions can readily undergo repair of the branch pulmonary arteries.[25] Conversely, in the absence of a surgical indication, pulmonary artery stents are often favored based on the less invasive nature of this intervention. One important caveat is that the majority of

patients in the current series had near-systemic level pulmonary artery pressures, indicative of widespread and extensive peripheral pulmonary artery stenoses. The application of either surgical techniques or stents in the central pulmonary vasculature in patients whose primary problem is peripheral stenoses is certainly destined to fail.

Interventional catheterization techniques applied to peripheral pulmonary arterial stenoses have demonstrated disappointing results.[12-14] The literature on this subject consistently reveals an inability to relieve gradients at the lobar and segmental level.[26] This is, in part, attributable to the fact that the stenoses are invariably located at the ostia, a location that does not respond to ballooning and is not amenable to stenting. Furthermore, attempts to balloon or stent these areas have demonstrated a high risk of rupture or perforation owing to the fact that the vessels distal to obstruction are extremely thin-walled. Thus, the very lesions that historically were in most need of treatment were found not to be amenable to interventional techniques. This is the fundamental reason why we developed a surgical approach to peripheral pulmonary artery stenosis.

In summary, this study was performed to evaluate the surgical results of pulmonary artery reconstruction in patients who previously had undergone pulmonary artery stenting. The majority of patients included in this study had high residual pulmonary artery pressures despite multiple interventional procedures. This physiology is indicative of widespread peripheral pulmonary stenoses, which by definition do not respond to proximal interventions. Surgical pulmonary artery reconstructions resulted in significant decreases in the central pulmonary artery pressures and thus far have had a relatively low reintervention rate. These results suggest that pulmonary artery reconstruction can be very successful in the setting of prior pulmonary artery stenting.

The illustrations demonstrating the surgical technique for pulmonary artery stent surgery (Figures 2A-2G) were drawn by Erin Anne Mainwaring.

References

1. Kirklin JW, Blackstone EH, Jonas RA, et al. Morphologic and surgical determinants of outcome events after repair of tetralogy of Fallot and pulmonary stenosis: a two-institution study. *J Thorac Cardiovasc Surg*. 1992;103:706-723.
2. Michielon G, Marino B, Formigari R, et al. Genetic syndromes and outcome after surgical correction of tetralogy of Fallot. *Ann Thorac Surg*. 2006;81:968-975.
3. Carotti A, Digilio MC, Piacentini G, Saffirio C, DiDonato RM, Marino B. Cardiac defects and results of cardiac Surgery in 22q11.2 deletion syndrome. *Dev Disabil Res Rev*. 2008;14:35-42.
4. Formigari R, Michielon G, Digilio MC, et al. Genetic syndromes and congenital heart defects: how is surgical management affected? *Eur J Cardiothorac Surg*. 2009;35:606-614.
5. Collins RT 2nd. Cardiovascular disease in Williams syndrome. *Circulation*. 2013;127:2125-2134.
6. Collins RT 2nd, Kaplan PB, Somes GW, Rome JJ. Cardiovascular abnormalities, interventions, and long-term outcomes in infantile Williams syndrome. *J Pediatr*. 2010;156:253-258.e1.
7. Blue GM, Mah JM, Cole AD, et al. The negative impact of Alagille syndrome on survival of infants with pulmonary atresia. *J Thorac Cardiovasc Surg*. 2007;133:1094-1096.
8. Wren C, Oslizok P, Bull C. Natural history of supravalvar aortic stenosis and pulmonary artery stenosis. *J Am Coll Cardiol*. 1990;15:1625-1630.
9. Wessel A, Pankau R, Kececioglu D, et al. Three decades of follow-up of aortic and pulmonary vascular lesions in the Williams-Beuren syndrome. *Am J Med Genet*. 1994;52:297-301.
10. Gandy KL, Tweddell JS, Petech AN. How we approach peripheral pulmonary stenosis in Williams-Beuren syndrome. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu*. 2009;12:118-121.
11. Stamm C, Friehs I, Moran AM, et al. Surgery for bilateral outflow tract obstruction in elastin arteriopathy. *J Thorac Cardiovasc Surg*. 2000;120:755-763.
12. Trivedi KR, Benson LN. Interventional strategies in the management of peripheral pulmonary artery stenosis. *J Interv Cardiol*. 2003;16:171-188.
13. Geggel RL, Gauvreau K, Lock JE. Balloon dilation angioplasty of peripheral pulmonary stenosis associated with Williams syndrome. *Circulation*. 2001;103:2165-2170.
14. Cunningham JW, McElhinney DB, Gauvreau K, et al. Outcomes after primary transcatheter therapy in infants and young children with severe bilateral peripheral pulmonary artery stenosis. *Circ Cardiovasc Interv*. 2013;6:460-467.
15. Monge M, Mainwaring RD, Sheikh AY, Punn R, Reddy VM, Hanley FL. Surgical reconstruction of peripheral pulmonary artery stenosis in Williams and Alagille syndromes. *J Thorac Cardiovasc Surg*. 2013;145:476-481.
16. Mainwaring RD, Hanley FL. Surgical reconstruction of peripheral pulmonary artery stenosis. *Semin Thorac Cardiovasc Surg*. 2017;29:198-205.
17. Mainwaring RD, Ibrahimiye AN, Hanley FL. Surgical technique for reconstruction of peripheral pulmonary artery stenosis and other complex peripheral reconstructions. *Ann Thorac Surg*. 2016;102:e181-e183.
18. Collins RT 2nd, Mainwaring RD, MacMillen KL, Hanley FL. Outcomes of pulmonary artery reconstruction in Williams syndrome. *Ann Thorac Surg*. 2019;108:146-153.
19. Martin E, Mainwaring RD, Collins RT II, MacMillen KL, Hanley FL. Surgical repair of peripheral pulmonary artery stenosis in patients without Williams or Alagille syndromes [e-pub ahead of print]. *Semin Thorac Cardiovasc Surg*. https://doi.org/10.1053/j.semtcvs.2020.01.003, accessed January 17, 2020.
20. Ma M, Reinhartz O, Mainwaring RD, Hanley FL. Surgical reconstruction of peripheral pulmonary artery stenosis. *Oper Tech Thorac Cardiovasc Surg*. 2019;24:163-175.
21. Mainwaring RD, Patrick WL, Ma M, Hanley FL. An Analysis of patients requiring unifocalization revision following midline unifocalization for pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals. *Eur J Cardiothorac Surg*. 2018;54:63-70.
22. Mainwaring RD, Patrick WL, Rosenblatt TR, et al. Surgical results of unifocalization revision. *J Thorac Cardiovasc Surg*. 2019;158:534-545.
23. Greene CL, Mainwaring RD, Sidell D, Yarlagadda V, Patrick WL, Hanley FL. Impact of phrenic nerve palsy and the need for diaphragm plication following surgery for pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals. *Semin Thorac Cardiovasc Surg*. 2018;30:318-324.
24. Mainwaring RD, Sheikh AY, Reddy VM, Punn R, Hanley FL. Surgical results in patients with pulmonary atresia/major aortopulmonary collaterals and Alagille syndrome. *Eur J Cardiothorac Surg*. 2012;42:235-241.
25. Cresalia NM, Armstrong AK, Romano JC, et al. Long-term outcomes after surgical pulmonary arterioplasty and risk factors for reintervention. *Ann Thorac Surg*. 2018;105:622-628.
26. Collins RT 2nd. Cardiovascular disease in Williams syndrome. *Curr Opin Pediatr*. 2018;30:609-615.