Surgical Technique for Repair of Peripheral Pulmonary Artery Stenosis and Other Complex Peripheral Reconstructions



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Surgical reconstruction of peripheral pulmonary artery stenosis is a technically challenging procedure due to the need to access all lobar and segmental branches. This paper describes our surgical approach that entails division of the main pulmonary and separation of the branch pulmonary

Peripheral pulmonary artery stenosis (PPAS) is a rare and complex form of congenital heart disease that is associated with Williams and Alagille syndromes. The natural history of PPAS is highly dependent on the degree of obstruction. Mild cases have a favorable prognosis and may demonstrate spontaneous regression of the peripheral stenoses. These mild cases will rarely need any type of intervention. In contrast, severe PPAS is characterized by multiple stenoses of lobar and segmental branches and tends to progress over time [1]. Severe PPAS results in right ventricular hypertension, right ventricular hypertrophy, and eventually in right ventricular failure. The severe cases of PPAS almost invariably require intervention in order to alleviate the adverse consequences of high right ventricular pressures.

The optimal management of patients with severe PPAS remains controversial. Many centers have advocated a multimodality approach, which includes surgical augmentation of the central pulmonary arteries and catheter-based treatment of the more distal stenoses [2, 3]. However, there is little evidence that this multimodality approach has any lasting efficacy for this disease. The Achilles' heel of catheter-based treatment has proven to be the ineffectiveness of relieving obstruction at the lobar and segmental level.

In 2012, we reported a series of 16 patients who underwent surgical reconstruction for severe PPAS [4]. This was the first report in the literature advocating an entirely surgical approach to this disease. The results of this study demonstrated a decrease in right ventricle to aortic peak systolic pressure ratios from 0.88 ± 0.07 to 0.40 ± 0.04 , representing a 55% decrement. There was no early or late mortality in this surgical cohort. Based on these results, we recommended that patients who require treatment for severe PPAS undergo surgical reconstruction. arteries. This surgical approach can also be utilized for other complex peripheral pulmonary artery reconstructions.

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The surgical technique for reconstruction of PPAS is a technically challenging procedure due to the need to access all lobar and segmental branches. Here we describe in detail our surgical approach for peripheral pulmonary artery reconstruction.

Technique

A median sternotomy incision is performed and the pericardium opened. The main pulmonary artery as well as the right and left branch pulmonary arteries are dissected free from the surrounding structures (Fig 1). This intrapericardial portion of this dissection is performed in a standard fashion. Prior to the extrapericardial portion of the pulmonary artery dissection, both pleural spaces are opened and the course of both phrenic nerves marked with fine Prolene (Ethicon, Somerville, NJ) sutures. The lobar and segmental branches are sequentially identified and the external anatomic appearance compared to the preoperative angiogram. It is imperative to perform the entire surgical dissection prior to administration of heparin to achieve complete hemostasis. In addition, we have noted a significant incidence of intraparenchymal hemorrhage if the dissection is performed after the patient is anticoagulated.

Cardiopulmonary bypass is instituted and the patient cooled to 25°C. This degree of hypothermia results in a significant reduction in metabolic rate, permitting the safe reduction in pump flow with the important physiologic effect of a commensurate decrease in collateral flow to the lung. A vent catheter is inserted through the right superior pulmonary vein into the left ventricle. The main pulmonary artery is divided distally and the right and left branch pulmonary arteries separated. Neuroclips are placed on each of the lobar branches to prevent backbleeding.

The right and left branch pulmonary arteries are incised starting along the inferior aspect of the artery. This incision is continued distally following the medial border of the artery and extended into the medial basal

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Fig 1. Artist's illustration demonstrating typical anatomy associated with peripheral pulmonary artery stenosis. There are multiple lobar and segmental stenoses.

segment of each lower lobe. Stay sutures are placed at the upper edges of the branch pulmonary arteries to facilitate exposure (Fig 2). The orientation of this incision is critical, as it results in the opening in each artery facing into the free space away from the superior pulmonary veins and bronchi. Other potential orientations of this incision uniformly result in a competition for space with adjoining structures. Specifically, an anterior incision would result in competing for space with the upper lobe pulmonary veins. A superior incision would result in competition with the distal bronchi as well as the origin of the lobar branches, while a posterior incision would compete with the proximal bronchus.



Fig 2. Illustration demonstrating the initial step in reconstruction of peripheral pulmonary artery stenosis. 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 continuing along the medial border of the lower lobe artery.



Fig 3. Illustration demonstrating surgical reconstruction of peripheral pulmonary artery stenosis. An ostioplasty has been performed to the basilar segment of the right lower lobe. Separate homograft patches are performed to augment the segmental and lobar branch stenoses. A long homograft patch is then sutured in place to augment the branch pulmonary arteries.

Each of the lobar or segmental stenoses are then addressed surgically. Segmental stenoses may be amenable to surgical ostioplasty using a Heineke-Mikulicz technique. Alternatively, if the anatomy is not amenable to ostioplasty, the artery is incised and augmented with a patch of pulmonary artery homograft. The primary longitudinal incisions in the branch pulmonary arteries are then augmented with long pulmonary homograft patches (Fig 3).

The reconstructed branch pulmonary arteries are then sewn together end to end. The main pulmonary artery is augmented with a patch of pulmonary artery homograft. The main pulmonary artery is then anastomosed to the



Fig 4. Illustration demonstrating reestablishment of continuity between the right and left branch pulmonary arteries, and reanatomosis to the reconstructed main pulmonary artery.

reconstructed branch pulmonary arteries to complete the operation (Fig 4).

The patient is rewarmed during the finishing stages of the reconstruction. Two transthoracic pressure lines are placed to monitor left atrial and right ventricular pressures. The patient is weaned from cardiopulmonary bypass and transesophageal echocardiography is performed to assess ventricular function.

Comment

Reconstruction of PPAS is technically quite challenging due to the need to address the extrapericardial stenoses to the middle and lower lobes. Many congenital heart surgeons have limited experience with this dissection and reconstruction. Thus, it is understandable why the surgical treatment of PPAS has not been widely adopted, and by default, has been considered to be a disease that could be best treated in the interventional catheterization lab.

Our center has acquired an extensive experience with extrapericardial pulmonary artery surgery in the context of patients with pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries (PA/ VSD/MAPCAs) [5]. It was this broad experience with peripheral pulmonary artery reconstruction that was subsequently applied to PPAS. The surgical technique described in this report of separating the right and left branch pulmonary arteries in order to access the distal branches and underside of the arteries is also frequently utilized in reoperations for PA/VSD/MAPCAs. The applicability of surgical ostioplasty for segmental stenoses and homograft patch augmentation for lobar stenoses is similar for both PPAS and PA/VSD/MAPCAs.

In summary, this report provides a description of our technical approach to the surgical reconstruction of PPAS. This technique is also applicable to reoperative surgery in patients with PA/VSD/MAPCAs and residual peripheral architectural stenoses.

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