

Surgical Techniques for Repair of Peripheral Pulmonary Artery Stenosis

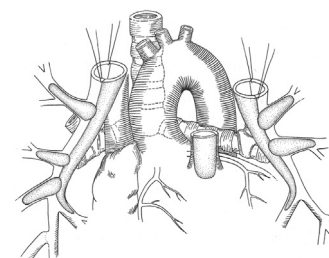


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Peripheral pulmonary artery stenosis (PPAS) is a rare form of congenital heart disease that is most frequently associated with Williams and Alagille syndromes. These patients typically have systemic level right ventricular pressures secondary to obstruction at the lobar, segmental, and subsegmental branches. The current management of patients with PPAS remains somewhat controversial. We have pioneered an entirely surgical approach for the reconstruction of PPAS. This approach initially entailed a surgical patch augmentation of all major lobar branches and effectively reduced the right ventricular pressures by more than half. This was the first report demonstrating an effective approach to this disease. Over the past 5 years, we have gradually evolved our technique of reconstruction to include segmental and subsegmental branch stenoses. An important technical aspect of this approach entails the division of the main pulmonary and separation of the branch pulmonary arteries to access the lower lobe branches. Pulmonary artery homograft patches are used to augment hypoplastic pulmonary artery branches. In addition, we perform a Heinecke-Miculicz-type ostioplasty for isolated ostial stenoses. The technical details of the surgical approach to PPAS are outlined in this article and can also be used for other complex peripheral pulmonary artery reconstructions.

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An illustration demonstrating key steps in the surgical technique for reconstruction of peripheral pulmonary artery stenosis.

Central Message

This article provides a detailed description of the surgical techniques that have been developed for the surgical reconstruction of peripheral pulmonary artery stenosis.

INTRODUCTION

Peripheral pulmonary artery stenosis (PPAS) is a rare and complex form of congenital heart disease that is associated with Williams and Alagille syndromes.^{1–3} The natural history of PPAS is highly dependent on the degree of obstruction.^{4–7} Mild cases have a favorable prognosis and may demonstrate spontaneous regression of the peripheral stenosis.⁸ These mild cases 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. Severe PPAS results in right ventricular hypertension, right ventricular hypertrophy, and eventually right ventricular failure. The severe cases of PPAS almost invariably require intervention to alleviate the adverse consequences of high right ventricular pressures.^{9,10}

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The optimal management of patients with severe PPAS remains controversial.¹¹ Many centers have advocated a “multimodality approach” that includes surgical augmentation of the central pulmonary arteries and catheter-based treatment of the more distal stenosis.^{12,13} 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.¹⁴ This was the first report in the literature advocating an entirely surgical approach to this disease. The surgical technique for reconstruction of PPAS is a technically challenging procedure because of the need to access all lobar and segmental branches. This article describes our current surgical approach for peripheral pulmonary artery reconstruction.

SURGICAL TECHNIQUE

A median sternotomy incision is performed, and the pericardium is opened. The main pulmonary artery as well as the right and left branch pulmonary arteries is dissected free

from the surrounding structures (Fig. 1). The intrapericardial portion of this dissection is performed in a standard fashion. Before the extrapericardial portion of the pulmonary artery dissection, both pleural spaces are opened, and the course of both phrenic nerves is marked with fine prolene sutures. The lobar and segmental branches are sequentially identified, and the external anatomical appearance is compared with the preoperative angiogram.

It is imperative to perform the entire surgical dissection before the 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 is cooled to 25°C. This degree of hypothermia results in a significant reduction in metabolic rate, permitting the safe reduction in pump flow with an important physiological 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 are 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 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, whereas a posterior incision would compete with the proximal bronchus.

Each of the peripheral stenoses is then addressed sequentially. Segmental and subsegmental stenoses are often amenable to surgical ostioplasty using a Heineke-Miculicz technique (Fig. 3A), particularly in the presence of ostial stenosis with a normal distal vessel (Fig. 3B). The vessel is probed with a metal dilator to get a “feel” for the severity and extent of the stenosis (Fig. 3C). The carina of the

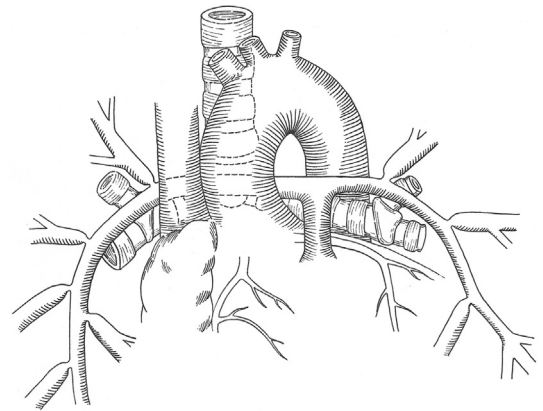


Figure 1. An illustration demonstrating typical anatomy associated with peripheral pulmonary artery stenosis (PPAS). There are multiple lobar and segmental stenoses. (Reproduced with permission from Mainwaring et al.¹⁷)

vessel is then incised with fine scissors to a point beyond the limits of the stenosis (Fig. 3D). The incision is then closed in a transverse direction (90° to the direction of the incision) with 8-0 prolene sutures (Fig. 3E).

There are circumstances when the anatomy of a peripheral pulmonary artery would not be amenable to ostioplasty. Specifically, in the setting of a longer stenosis, an ostioplasty would not adequately address the full extent of the narrowing leading to the 2 distal branches (Fig. 4A). This

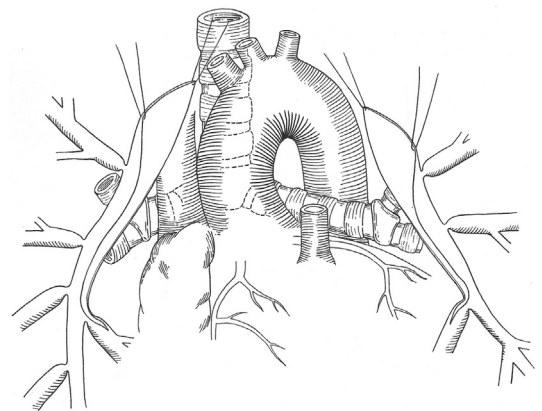


Figure 2. An illustration demonstrating the initial step in the reconstruction of PPAS. The main pulmonary artery is divided, as are the right and left branch pulmonary arteries. An incision is made along the inferior aspect of the proximal branch pulmonary arteries and continuing along the medial border of the lower lobe artery. (Reproduced with permission from Mainwaring et al.¹⁷)

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