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Pediatric Vascular Intervention: Pulmonary Valve Atresia

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Annotation. *Pulmonary atresia is a birth defect of the pulmonary valve. This valve controls blood flow from the right ventricle to the main pulmonary artery. The right ventricle is the lower right chamber of the heart. The pulmonary artery is the blood vessel that carries blood from the heart to the lungs. Pulmonary atresia is when this valve doesn't form at all. This means that no blood can go from the right ventricle of the heart out to the lungs. With pulmonary atresia, blood must use other routes to bypass the unformed pulmonary valve.*

Keywords. *pulmonary atresia, valve, ventricular septum, biventricular circulation, catheter, valvuloplasty.*

Perforation of an atretic pulmonary valve may be considered in patients with pulmonary atresia with intact ventricular septum (PA/IVS), depending on the size of the tricuspid valve and right ventricle, and the presence or absence of coronary artery abnormalities. Such patients are usually diagnosed either prenatally, or in the immediate neonatal period, and are dependent on a patent ductus arteriosus for pulmonary blood flow. Treatment aims to establish a stable source of pulmonary flow, as well as to relieve the right ventricular outflow obstruction and encourage growth of the right ventricle. Depending on variations in anatomy, some patients are able to achieve a biventricular circulation, while others can only proceed on the single-ventricle pathway. In the setting of RV-dependent coronary circulation defined as proximal coronary artery obstruction leading to inadequate antegrade coronary flow and dependence of myocardial perfusion on fistulous connections from the RV, RV decompression is contraindicated. Such cases may require cardiac transplantation.

Early catheter-based techniques to perforate the atretic pulmonary valve membrane, reported as early as 1991, included sharp instruments such as the stiff end of a coronary guidewire, and laser therapy via a fiber optic cable. These methods carry a high risk of complications, and so have been replaced by the use of radiofrequency perforation, most commonly with the Baylis Radiofrequency (RF) co-axial system (Baylis Medical, Montreal, Canada). More recently, chronic total occlusion (CTO) coronary wires have also been used, with good results. A femoral venous approach is preferred. Biplane imaging is crucial to performing the procedure safely. Hemodynamics typically show supra-systemic right ventricular pressure. Right ventricular angiography is used to assess the anatomy of the right ventricle and infundibulum, to measure the pulmonary valve size, and to exclude RV-dependent coronary circulation. Usual camera angles are straight lateral and slight cranial/right axial oblique to best define the right ventricular outflow tract and the atretic pulmonary valve membrane. If arterial access is obtained, an aortogram can demonstrate the patent ductus arteriosus, pulmonary artery anatomy, and the relationship of the main pulmonary artery to the infundibulum. Alternatively, an antegrade left ventricular angiogram may be performed.

If the anatomy appears favorable to proceed with intervention, then an end-hole catheter is positioned in the infundibulum perpendicular to the atretic membrane. Typically, a Judkins right coronary catheter or a cobra catheter will provide the best orientation, but this may vary on a case-by-case basis. Precise and stable positioning of the catheter tip against the membrane, directed toward the main pulmonary artery, is critical, as the RF system will then be advanced through this catheter. A gooseneck snare may be placed from the aortic side, via the patent ductus arteriosus, into the main pulmonary artery to act as a target for the RF perforation, even though this carries some risk of inducing ductal spasm. After confirmation of proper positioning, RF energy is applied and the RF wire advanced through the membrane into the main pulmonary artery. Once the wire crosses the membrane, RF energy is discontinued, and again correct position should be confirmed with biplane fluoroscopy and angiography. The co-axial catheter can then be advanced over the wire and used to exchange for a guidewire that is then used to facilitate valvuloplasty. If a CTO wire is used instead of RF, a similar sequence can be performed. Guidewire position across the PDA into the descending aorta is preferred, though a distal pulmonary artery branch can also be used. If a snare was placed in the main pulmonary artery to guide RF perforation, it can be used to gently pull the guidewire into the aorta, creating an arteriovenous loop. Care must be taken with this technique to avoid any tension on the wire. Once the wire is in stable position, valvuloplasty is performed. As with any pulmonary valvuloplasty, a final balloon diameter of 1.2 to 1.4 times the size of the valve annulus is recommended. As the membrane may be difficult to cross with a balloon, a small coronary balloon may be used to perform a partial dilation first, before attempting to deliver a larger balloon. Post-valvuloplasty, hemodynamics are obtained. A pressure wire may aid in this process to identify subvalvar versus valvar residual gradients. A final right ventricular angiogram is performed to assess the results of the procedure.

Transcatheter perforation of an atretic pulmonary valve carries a relatively high risk of procedural complications. In addition to the risks of pulmonary balloon valvuloplasty, as described in the previous section, there is the potential for inadvertent perforation of the right ventricular outflow tract or main pulmonary artery. The risk of complications has been reported as high as 15–20% in some series. A recent multi-center study from the Congenital Catheterization Research Collaborative showed a 10% risk of cardiac perforation. Even after successful initial decompression of the right ventricle, subsequent re-interventions are very common. Re-intervention may include repeat balloon pulmonary valvuloplasty, surgical augmentation of the right ventricular outflow tract, and/or the addition of a second source of pulmonary blood flow (either ductal stenting or surgical aorto-pulmonary shunt). Stenting of the ductus arteriosus, for those patients who do require a second source of pulmonary flow, may be performed at the same time as the initial valve perforation, or as a separate procedure. Predicting which patients will require an additional source of pulmonary blood flow remains difficult. Identified risk factors for re-intervention and/or inability to achieve a biventricular circulation include: residual pulmonary valve gradient after initial valvuloplasty, tricuspid valve hypoplasia, mild or lesser degree of tricuspid valve regurgitation, and a non-tripartite right ventricle.

All in all, treatment for pulmonary atresia depends on its severity. In some cases, blood flow can be improved by using cardiac catheterization. During this procedure, doctors can expand the valve using a balloon to keep the ductus arteriosus open. In some cases, they may need to place a stent (a small tube) to keep it open. In most cases of pulmonary atresia, a baby may need surgery soon after birth. During surgery, doctors widen or replace the pulmonary valve and enlarge the passage to the pulmonary artery. If a baby has a VSD, the doctor will use a patch. The patch closes the hole between the lower chambers of the heart. These actions will improve blood flow to the lungs and the rest of the body. If a baby has an underdeveloped right ventricle, staged surgical procedures might be needed**.**

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