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# Pulmonary atresia with intact ventricular septum: A brief overview of management strategies and rationale

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ABSTRACT

Pulmonary atresia with intact ventricular septum is a rare congenital cardiac anomaly characterized by complete obstruction of outflow from the right ventricle and pulmonary blood flow that is completely ductal dependent. Appropriate therapeutic decision making early in the neonatal period is crucial to immediate survival. Ultimately, definitive palliation or correction is very much dependent upon the adequacy of right-sided cardiac structures. This article will review management strategies designed to optimize both immediate and longer term survival.



## 1. Introduction

Pulmonary atresia with intact ventricular septum is a rare congenital lesion, occurring in approximately 2.4–2.5% of all patients presenting with congenital heart disease. It is characterized by morphologic heterogeneity of right-sided cardiac structures; specifically, the tricuspid valve, the right ventricle and the main pulmonary artery [1,2]. It is the adequacy of these right-sided structures which will determine the type of repair these patients may undergo; either single ventricle palliation, one and one half ventricle palliation, or biventricular correction.

Pulmonary atresia with intact ventricular septum is a congenital cardiac lesion characterized by complete obstruction of the right ventricular outflow tract occurring at the level of the atretic pulmonary valve, an intact ventricular septum with no ventricular septal defect, variable degrees of right ventricular and tricuspid valve hypoplasia, and variable coronary anatomy. The pulmonary valve, across which there is no forward flow, usually exists as a tough imperforate membrane. Because of relatively unimpeded inflow, and totally obstructed outflow, the right ventricle is frequently markedly hypertrophic, and may exhibit fibrosis and endocardial fibroelastosis. The right ventricle may be large and tripartite, but it is more frequently small because of hypoplasia or absence of the tribecular portion, the infundibular portion, or both.

Right ventricular morphology may be characterized as unipartite, bipartite, or tripartite depending upon the concomitant degree of hypoplasia of the inflow, trabecular, and infundibular portions of the right ventricle [2]. The tricuspid valve is usually proportionate to the size of the right ventricle and may exhibit variable degrees of incompetence. Right atrial enlargement and some form of interatrial communication are always present. Although they may sometimes be small, the pulmonary arteries are frequently normal or nearly normal in size, receiving blood flow through a patent ductus arteriosus.

## 2. Initial medical management

Pulmonary atresia with intact ventricular septum is a ductal dependent lesion. The sole source of pulmonary blood flow is the patent ductus arteriosus and every effort must be made to maintain patency of the ductus. Consequently, as soon as the diagnosis is established echocardiographically in the newborn, prostaglandin E1 infusion is initiated to both establish and maintain ductal patency, increase pulmonary blood flow, and relieve hypoxemia and acidosis. Prostaglandin is typically infused at doses of 0.01 to 0.2 mg per kilogram per minute, and is adjusted according to systemic oxygen saturations. High doses of prostaglandin may cause mild hyperthermia and apnea, and one must be ready to endotrachealy intubate and mechanically ventilate the neonate if respiratory depression occurs. However, it is crucial that the prostaglandin infusion be maintained without interruption. More severely hypoxic or acidotic neonates may require endotracheal intubation and mechanical ventilation, coupled with reversal of acidosis with sodium bicarbonate until ductal dependency can be clearly established and maintained. In addition to establishing the diagnosis of pulmonary atresia with ductal dependent pulmonary blood flow, echocardiography is also used to define intracardiac structures, determine the adequacy of the atrial septal defect, and to determine the presence or absence of coronary sinusoids and right ventricular dependent coronary circulation.

Once the patient is stabilized and resuscitated, cardiac catheterization with cineangiography is then expeditiously performed, and is now both

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diagnostic and therapeutic. At the time of cardiac catheterization, balloon atrial septostomy may be performed to ensure unobstructed blood flow at the level of the atrial septum. If no coronary sinusoids with right ventricular dependent coronary circulation are found, the right ventricular outflow tract may be opened using radiofrequency perforation and balloon dilation of the atretic pulmonary valve. The technique we use was developed by M. Hubbell and S. Kaine at Children's Mercy Hospital. It is illustrated in Fig. 1 and described in detail in Appendix A. If extensive coronary sinusoids are found, the decision to proceed down a single ventricle pathway may be made in the neonatal period. Historically open pulmonary valvotomy has been used quite successfully [3,4]. Although reports of open pulmonary valvotomy still exist in the literature, radiofrequency perforation with balloon valvotomy of the pulmonary valve has completely displaced open pulmonary valvotomy at our institution. It has the advantage of being able to decompress the right ventricle without subjecting the patient to an open surgical procedure. If balloon valvotomy can be performed following radiofrequency perforation, the patient can then be slowly weaned off prostaglandins over a several day period of time to see if oxygen saturations can be maintained, or if additional palliation with a systemic to pulmonary arterial shunt is required. It should be noted that if radiofrequency perforation and balloon dilation is attempted, the cardiac surgical team should be notified and be on standby, as perforation of the right ventricular outflow tract with pericardial tamponade can occur.

## 3. Initial surgical management

Initial surgical management is palliative, and has two distinct goals. Goal one is to establish a reliable source of pulmonary blood flow through creation of a systemic to pulmonary arterial shunt. Goal two is to decompress the right ventricle by opening the atretic pulmonary valve. If radiofrequency perforation with balloon dilation is not available in an institution, open pulmonary valvotomy can be performed at the time the systemic to pulmonary arterial shunt is created. If there are extensive coronary sinusoids and right ventricular dependent coronary circulation, no attempt whatsoever should be made to open the right ventricular outflow tract, as acute myocardial infarction can occur in that setting. This cannot be emphasized strongly enough. In the past, attempts have been made to exclude the rudimentary right ventricular dependent portion of the coronary circulation through thromboexclusion with mixed results [5]. Other groups have chosen to ligate the right ventricular to coronary arterial connections [6]. We have chosen to leave the abnormal coronary connections alone, feeling that manipulation could result in myocardial infarction.

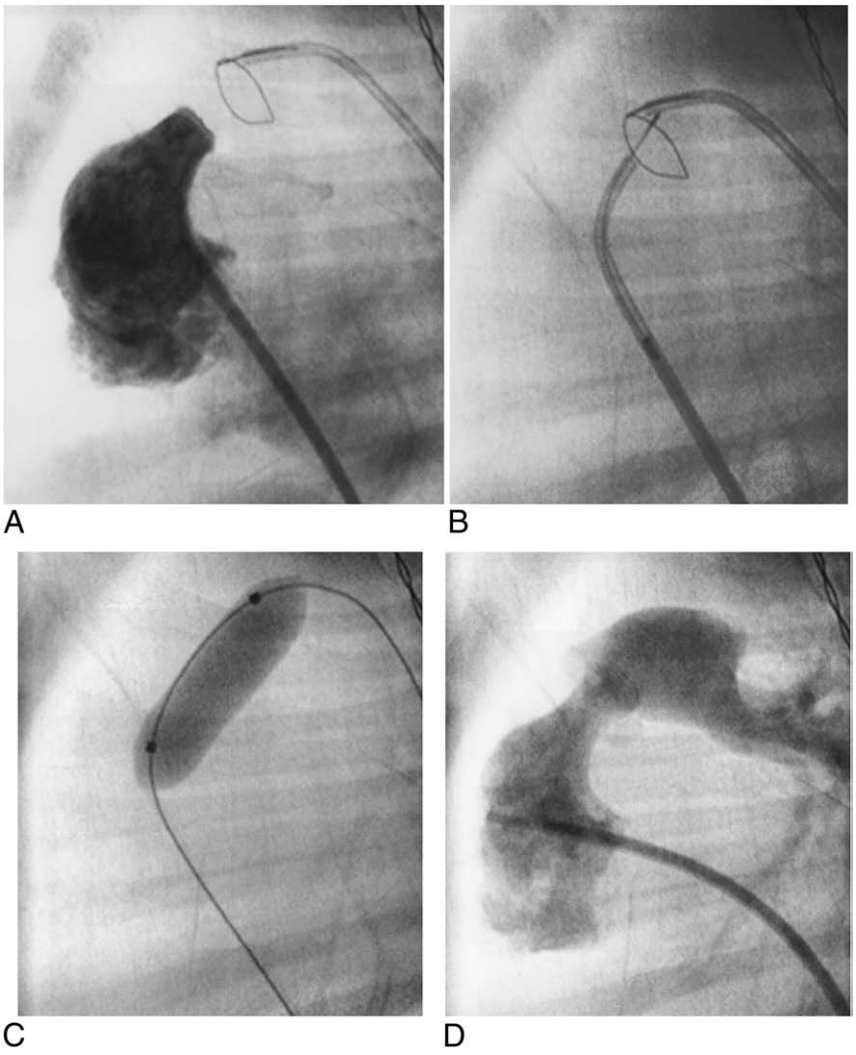


**Fig. 1.** Cardiac catheterization with radiofrequency perforation and balloon pulmonary valvotomy in a patient with pulmonary atresia. A. Injection is made into the right ventricle. Note the well developed infundibulum ending blindly. The catheter is in the patient's aorta, and has traversed the ductus arteriosus, with the tip of the catheter and snare lying in the proximal main pulmonary artery. A second catheter has been placed via right heart catheterization. B. The tip of the catheter is in the infundibulum and radiofrequency perforation of the atretic pulmonary valve has been accomplished. C. Balloon dilation of the atretic pulmonary valve has been accomplished. D. Right ventricular injection post balloon valvotomy shows adequate opening of the right ventricular outflow tract.