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## Total Right Ventricular Dependent Coronary Circulation in Pulmonary Atresia With Intact Ventricular Septum

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This report describes the case of a full-term gestational female with a prenatal diagnosis of pulmonary atresia with intact ventricular septum. Cardiac ultrasound at birth confirmed the diagnosis with no evidence of coronary artery fistulas. The patient died 6 hours after a central aortic to pulmonary artery shunt had been created with bypass support. Postmortem examination showed a coronary artery fistula rising from the right ventricle and a complete absence of both coronary ostia. This rare finding has been reported sporadically in the English literature.

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**I**solated atresia of the pulmonary valve in a heart with an otherwise normal segmental anatomy is also known as pulmonary atresia with intact ventricular septum (PA/IVS). The wide spectrum of malformations within this entity variably affects to a different degree the right-sided structures (tricuspid valve [TV], right ventricle [RV], pulmonary artery) and may or may not involve anomalies of the coronary circulation.

Echocardiography corroborated the fetal diagnosis of pulmonary atresia with intact ventricular septum in a 39-week gestational female. The RV was small, with a TV

annulus diameter of 4 mm (mitral valve annulus = 10 mm). We found no evidence of dilated coronary arteries. A coronary artery supplying the RV appeared small and originating from the anterior aortic cusp. The left coronary artery also appeared to arise from the aorta. Color Doppler demonstrated a large patent ductus with left-to-right shunting and a patent foramen ovale with also left-to-right shunt. The pulmonary arteries were 2 mm in diameter and supplied by the ductus arteriosus.

The patient's blood oxygen saturation remained between 80% and 85% after being placed on prostaglandin to prevent premature closure of the ductus arteriosus. On hospital day 2, the patient was taken to the surgical theater where we used a simplified version of the venous-venous bypass technique published by Asou and colleagues [1]. After heparinization a 12F right angle venous cannula (Medtronic, Minneapolis, MN) was placed in the inferior vena cava while a 6F arterial cannula (Medtronic) was placed in the wall of the right atrium. By using this type of support, the patient continued to eject oxygenated blood while blood flow to the lungs was interrupted. Ligation of the ductus arteriosus was followed by placement of a 3.5-mm shunt from the ascending aorta to the bifurcation of the pulmonary artery (W.L. Gore and Assoc, Flagstaff, AZ). The main pulmonary artery was absent. The proximal anastomosis was completed with the help of a partial aortic clamp in the ascending aorta.

Discontinuation of bypass was uneventful. The patient's saturation oscillated between 75% and 80% on a 30% oxygen concentration in the respiratory mix. The patient was sent on minimal inotropic support and intubated to the pediatric intensive care unit for recovery. Approximately 6 hours postoperatively she suddenly became bradycardic and hypotensive. The chest was opened to rule out tamponade. Although no evidence of tamponade was present the patient arrested and died despite cardiopulmonary resuscitation efforts.

Autopsy confirmed the diagnosis of PA/IVS with an extremely hypoplastic right heart, a rudimentary TV, a patent foramen ovale, and a dilated left ventricle. A single coronary artery originating from a fistula to the small RV followed the distribution of the proximal right and the complete left coronary system. The transition between right and left coronary system was situated millimeters from the right coronary cusp. Coronary ostia were absent from the ascending aorta and the shunt was patent at the time of autopsy (Fig 1).

## Comment

The athypophysiology of PA/IVS is one of cyanosis at birth. Infants require patency of the ductus arteriosus to survive. Frequently, right ventricular pressures are supra-systemic and tricuspid regurgitation with diminished right ventricular compliance is common.

A gamut of right ventricular hypoplasia can be seen with this diagnosis. The right cavity hypoplasia is probably related to the lack of development of the trabecular and outlet portion of the RV. Lack of ventricular compliance is almost always part of the picture, mostly due to massive ventricular hypertrophy [2].

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The size of the TV annulus has direct implications in the development of the RV. In turn, there is an equally close relationship between the severity of RV underdevelopment and the presence of coronary artery fistulas. Different degrees of coronary artery abnormalities (ie, stenosis, dilatation) may be present in as many as 93% of patients with PA/IVS [3]. The dimensions of the TV and its degree of insufficiency combined with the size and compliance of the RV and the presence (or absence) of coronary fistula will help determine the different surgical options.

Relief of the RV by outlet enlargement and anatomic restitution between the RV and pulmonary artery is only possible when the coronary circulation is not fistulas dependent. There is a potentially significant amount of myocardium at risk of ischemia after RV decompression when abnormal coronary artery perfusion is present.

Management of PA/IVS with a small RV and TV annular diameter presenting with fistulas to the coronary circulation is essentially based on the creation of a systemic to pulmonary shunt.

It is difficult to discern from the English literature the exact incidence of total RV-dependent coronary circulation, although it may be as high as 3% to 8% [3, 4]. The few reports of total RV-dependent coronary circulation involved fatal outcomes [3-5]. Lenox and Briner [5] described in their case report the interesting circulatory pattern in the coronaries. Unlike similar cases with coronary ostia present, a total RV-dependent coronary system may only perfuse myocardium during systole [5]. This may explain the absence of typical fistula-type flow in the preoperative echocardiogram.

Our use of a modified venous-venous cardiopulmonary bypass masked the existence of this anomaly. Had we used regular bypass, decompression of the right-sided cavities would have exposed early global ischemia with the consequent failure to discontinue bypass support.

Aortic angiogram is still today the only reliable means

to define the coronary anatomy. The decision to take a patient diagnosed with PA/IVS to the operating room without the benefit of an angiogram was controversial. The close proximity between the aorta and the transition between right and left coronary systems gave the false impression of aortocoronary continuity on the transthoracic echocardiogram. Regardless, the question remains as to what approach should be taken when confronted with the absence of aortic coronary ostia and a complete RV-dependent coronary circulation. Since this unusual type of myocardial perfusion seems to have an inescapable fatal outcome, the treating physician may confront the quandary between supportive therapy and the exceptional alternative of heart transplantation.

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## A Case of Thymoma Protruding Into the Superior Vena Cava Through the Thymic Vein

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We describe an unusual case of an invasive thymoma protruding into the superior vena cava and left brachiocephalic vein through the thymic veins in a 64-year-old patient. The tumor was resected with a bypass of the right brachiocephalic vein and right atrium. Although this type of growth form is rare for an invasive thymoma, this case suggests that in surgical procedures for thymomas, meticulous examination of the thymic veins is necessary to avoid leaving residual tumor.

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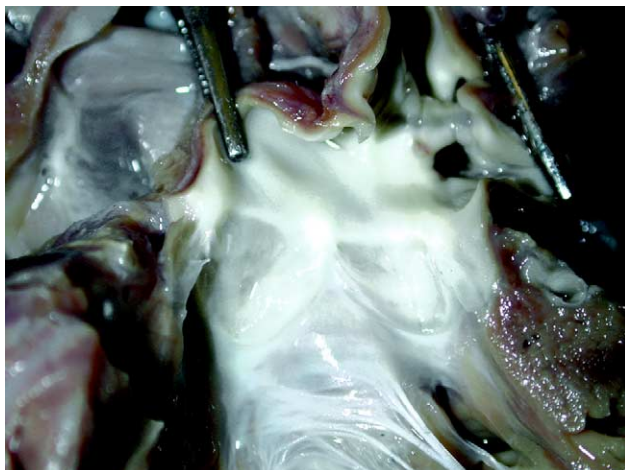


Fig 1. Longitudinal opening of the aortic root. The left and noncoronary cusps can be seen intact. Coronary ostia are absent in all three sinuses. The proximal end of the shunt can be seen in the ascending aorta.

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