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PRIMARY INSTRUMENTAL EXAMINATION IN THE ANOMALY OF PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM

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Annotation

Pulmonary atresia with intact ventricular septum is a cyanotic congenital heart disease characterized by complete obstruction of the right ventricular outflow tract, right ventricular hypoplasia, myocardial hypertrophy, and deranged coronary arteries. Historically, treatment strategies have resulted in high mortality rates. Advances in fetal imaging have led to improvements in our ability to prognosticate which patients are likely to have functionally single versus biventricular therapeutic pathways.

Keywords: Pulmonary atresia, ventricular septum, X-Ray, Ebstein's anomaly, coronary stenosis.

Pulmonary atresia with intact ventricular septum is a rare congenital cardiac lesion involving the structure of the right heart, with significant morphological heterogeneity. While atresia of the right ventricular outflow tract is central to this condition, significant variations in inlet, outlet, and apical ventricular characteristics exist between patients. Furthermore, developmental abnormalities in the formation of the coronary arteries present significant therapeutic challenges. Below are the instrumental examination methods that should be performed in this case:

Chest X-Ray. In patients with right ventricular hypoplasia, the cardiac silhouette may appear normal or mildly enlarged. The lungs will be oligemic from reduced pulmonary blood flow. In patients with right ventricular dilation, or Ebstein's malformation, the heart may occupy the entire thoracic cavity, making vascular marking difficult to delineate

Electrocardiography Patients usually maintain sinus rhythm, with a QRS axis in the normal orientation or with right axis deviation. Patients with small right ventricular cavities, and extensive mural hypertrophy, will typically have findings of left

ventricular hypertrophy from unopposed left-sided forces. In the subset of patients with dilated right ventricular cavities, right ventricular hypertrophy may be evident. Subtle ST segment changes may reflect a degree of subendocardial ischemia secondary to chronic right ventricular hypertension. Following placement of a shunt, it is important to monitor patients closely for electrocardiographic signs of ischemia secondary to coronary arterial steal. This would also be evident if right ventricular decompression was incorrectly performed in a patient with right ventricular-dependent coronary circulation and may result in rapid death.

Echocardiography. Providing a detailed assessment of the intra- and extracardiac anatomy is essential in order to optimize treatment strategy. Echocardiography will demonstrate the presence and size of right to left shunting across the oval fossa. Careful imaging of the right ventricular outflow tract must be performed to establish the presence of anterograde or retrograde flow. While structural atresia is common in this condition, functional atresia may also occur due to severe tricuspid regurgitation, or severe right ventricular dysfunction. The presence of a pulmonary insufficiency jet demonstrates a structural valve patency. Determining the degree of infundibular hypoplasia or atresia may guide the interventionist on their ability to establish forward flow. The pulmonary valvar diameter, and the diameter of the right and left pulmonary arteries, should be assessed. The right ventricular cavity dimension and tricuspid valvar diameter can be assessed from the apical four chamber and the parasternal long axis views. Measurements of the tricuspid valve are taken from hinge point to hinge point and the z-score measured. A z-score of lower than-3 is associated with a lower likelihood of biventricular repair. The arterial duct can be visualized from a high left parasternal view. It is often less tortuous than in other forms of pulmonary atresia. Careful assessment of the coronary arterial tree is of the utmost importance prior to intervention. This begins with a detailed echocardiographic exam. Fistulous communications may be demonstrated from the apical four chambers when sweeping anterior to posterior. Although not diagnostic, the presence of such connections should alert the physician to the risk of right ventricular-dependent coronary circulation. Careful review of the coronary arterial origins and proximal segments should be performed in a short axis parasternal view. Single or absent coronary arteries are extremely concerning. Color Doppler will

demonstrate the direction of coronary flow to evaluate antegrade or retrograde filling. Echocardiography, nonetheless, has significant limitations in delineation of the coronary arterial tree and the presence of coronary stenosis; therefore, further imaging modalities are required.

All in all, this disease is a pathological condition that manifests itself at birth and significantly negatively affects the quality of life of a child. Its early detection and correct diagnosis are very important in saving the child's life. A disease that is detected late or complicated can even cause the death of the child.

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