Angiographic Morphologic Characteristics in Pulmonary Atresia with Intact Ventricular Septum

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Objective
To determine the morphological and functional changes in the right ventricle in pulmonary atresia with intact ventricular septum (PAIVS) for assessing the candidates for the different therapeutic procedures currently available.

Methods
Thirty-one patients underwent cineangiocardiographic study with axial projections. Their ages ranged from 1 to 50 days (x = 9.6), and 28 of them were studied during the first month of life. The statistical analysis comprised the following: chi-square test, Kruskal-Wallis test for the mean and standard deviation, multiple regression, and the 95% confidence interval (95%CI) was calculated. The significance level adopted was alpha ≤ 0.05.

Results
The patients were divided into 3 groups according to the angiographic morphology of the right ventricle (RV): group A – tripartite RV (n=16); group B – bipartite RV (n=9); and group C – unipartite RV (n=6). The diameter of the tricuspid valve was 10.28 ± 2.67 mm (A); 7.82 ± 3.41 (B); and 5.27 ± 0.57 (C) (P=0.0005). Pulmonary atresia was of the valvular type in all group A patients and of the infundibular type in all group C patients (P=0.0001). Coronary-cavitary connections were rare (2/16) in group A patients, but occurred in all group C patients (P=0.0006), with retrograde opacity of the aorta (flow from the RV to the aorta) in 2 group A patients and in all group C patients (P=0.0003). Three patients (2 in group C and 1 in group A) had right-ventricular-dependent coronary circulation. Isolated moderate/severe tricuspid regurgitation showed a tendency towards being more frequent in group A (P=0.0525). The angle of the ductus arteriosus with the descending aorta was as follows: 104.06 ± 8.98 in group A; 79.17 ± 33.08 in group B; and 39.0 ± 6.52 in group C (P=0.0016). The correlation between the diameter of the tricuspid valve and the angle of the ductus arteriosus with the descending aorta was 0.6568 (P=0.0002).

Conclusion
Because of the heterogeneity of the morphology of the RV in patients with pulmonary atresia with intact ventricular septum, knowledge about all these data is required for selecting candidates for the different therapeutic procedures.

Key words
pulmonary atresia, ductus arteriosus, congenital heart disease, prognostic factors, pulmonary valvotomy

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In 1839, Peacock had already reported 7 cases of pulmonary atresia with intact ventricular septum; however, Peacock attributed the first case reported to John Hunter in 1783. Pulmonary atresia with intact ventricular septum accounts for 1 to 3% of the cardiac malformations; in the neonatal period, however, it accounts for 1/3 of the cyanotic heart diseases. Its etiology has not yet been totally clarified, although some researchers claim inflammatory and infectious causes, as well as a certain degree of familial incidence. Extracardiac anomalies are rare, although communications between the coronary arteries and the right ventricle may occur in approximately 50% of the cases.

The first surgical approach to pulmonary atresia with intact ventricular septum was, according to Kirklin and Barrat-Boyes, attributed to Greenwell et al, who proposed a strategy of treatment based on ventricular morphology. This treatment aimed at performing a pulmonary valvotomy in infants with a right ventricular cavity of normal dimensions. The palliative procedures developed by Glenn, Blalock-Taussig, and Waterston were later replaced by corrective techniques that aimed at clearing the right ventricular outflow tract and pulmonary valve, increasing pulmonary blood flow, and, therefore, reestablishing the continuity of the right ventricle with the pulmonary artery. Since the beginning, all surgical efforts and attempts have undergone several changes; however, the selection of patients for surgery has always been based on the size of the right ventricular cavity, anatomy and function of the tricuspid valve, and the presence of coronary-cavitary connections. The following surgical techniques have been used: valvotomy with hypothermia or cardioplegia with cardiopulmonary by-pass; pulmonary valvectomy; and transannular patch.

With the development of interventional techniques for the pediatric patient, a new alternative for the treatment of this malformation appeared with cardiac catheterization. Due to its effectiveness and low incidence of complications, it has been increasingly regarded as a very attractive form of treatment for a great number of investigators.

Through angiographic analysis, our study aimed at providing indispensable information for the selection of candidates to the different alternatives of treatment currently available for this malformation.

Methods
Over a period of 10 years, 31 patients with pulmonary atresia and intact ventricular septum were studied from the angiographic point of view. The patients’ ages ranged from 1 to 50 days (x=9.6), 28 being studied in the first month of life. Nineteen patients were males.

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Right ventriculography performed in the caudocranial and lateral projections aimed at evidencing: 1) the degree of tricuspid regurgitation; 2) the diameter of the tricuspid valve 42; 3) presence of coronary-cavitary connections and right ventricular-dependent coronary circulation (severe proximal stenoses in the native coronary territory, mainly anterior descending and circumflex arteries — complete obstructions in proximal points of the native coronary circulation — fistula between the native coronary territory and the right ventricle — absence of connection of the left coronary artery with the aorta, and the origin of the pulmonary artery); and 4) right ventricular morphology, according to Goor and Lillehei 43 and Bull et al. 44.

Left ventriculography performed in the elongated left anterior oblique projection aimed at the following: 1) assessment of the size and configuration of the left ventricle; 2) opacities of the aorta and, based on its visualization, determination of the angle formed by the ductus arteriosus and the descending aorta; 3) visualization of the right and left coronary arteries and establishment of the type of ventricular perfusion (anterograde: from the aorta to the right ventricle during diastole).

The dichotomic data were assessed with the x² (chi-square) test and calculating the 95% confidence interval (95%CI). The descriptive data were expressed as mean ± standard deviation, and the Kruskal-Wallis test was used. The continuous variables were correlated using multiple regression. The significance level alpha = 0.05 was adopted.

Results

Age at the time of cardiac catheterization was as follows: up to 7 days of life, 23 patients; between 8 and 30 days of life, 5 patients; between 31 and 50 days of life, only 3 patients.

According to the type of right ventricular morphology on the angiocardiographic study 44, the 31 patients were divided into 3 groups: group A comprised 16 patients with tripartite right ventricular morphology (inlet portion, trabecular portion, and outlet portion). In 1 patient, the tricuspid valve was dysplastic (fig. 1-A). Only 1 patient in group A did not have tricuspid regurgitation (fig. 2-A). The only patients in whom coronary-cavitary connections were observed had a tricuspid valve diameter smaller (7.2 and 8.7 mm) than the mean in the group. In 1 patient, opacity of the aorta had a tricuspid valve diameter smaller (7.2 and 8.7 mm). In 1 patient, the tricuspid valve was dysplastic (fig. 1-C). Only 1 patient in group A did not have tricuspid regurgitation (fig. 2-A). In 2 patients, the ductus arteriosus had a completely distinct morphology: it was thin, tortuous, and formed an acute angle with the descending aorta (fig. 3-B). In the 3rd patient, the opacities of the aorta did not allow visualization of the ductus arteriosus. Of the 9 patients, 6 were using prostaglandin. Group C) this group was characterized in the angiocardiographic study by nonopacity of the infundibular and trabecular portions, the right ventricle being reduced to only its inlet portion (unipartite). Two patients had moderate to severe tricuspid regurgitation (fig. 2-C). Retrograde coronary perfusion occurred as follows: through both coronary arteries in 2 patients; through the left coronary artery in 2 patients; and, through the right coronary artery in 2 patients; and, through the right coronary artery in 2 patients.
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artery in the 2 remaining patients. Anterograde coronary perfusion could not be identified in 1 patient. In another patient (Fig. 3-C), opacity of the right ventricle was possible starting at the aorta, due to the great dimension of the coronary-cavitary connection, forming a fistula that allowed deviation of the flow of this vessel to the right ventricular cavity. This observation was confirmed by the finding of an oximetric difference between the right ventricle and the right atrium. This and another patient had right ventricle-dependent coronary circulation. The morphology of ductus arteriosus could not be shown in 1 patient. In all patients, the ductus arteriosus formed an acute angle with the descending aorta (fig. 3-C). All patients in this group used prostaglandin.

Discussion

The greatest morphofunctional changes in pulmonary atresia with intact septum are related to the tricuspid valve, trabecular portion, and right ventricular outlet. Because in pulmonary atresia with intact septum no communication between the right ventricle and the pulmonary trunk exists, the possibility of a patient surviving after birth is very small. Therefore, pulmonary blood is supplied through the ductus arteriosus, and, occasionally, through systemic pulmonary collaterals. The problem is that, in almost all cases, the ductus arteriosus begins the closure process right after birth, and, occasionally, it may remain functional after the 30th day of extrauterine life.

In recent years, several investigators 45,46 have emphasized...
the importance of the development of ductus arteriosus in congenital heart diseases, and, particularly, in the critical obstructions of the right ventricular outflow tract, in which the persistence of ductus arteriosus is an indispensable condition for survival \cite{46,47}. Ductus arteriosus has been described as a small-caliber, tortuous vascular structure, which forms with the descending aorta an angle smaller than 90° \cite{48}. These characteristics differ completely from those observed in the normal newborn, who, due to the direction of the fetal flow, has a large-caliber ductus arteriosus, which, in its junction with the descending aorta, forms an angle greater than 90°.

Contrary to that which has been reported in the literature, we have observed patients with pulmonary atresia with intact septum, who, on the angiocardioGraphic study, have a normally developed ductus arteriosus, forming an obtuse angle with the descending aorta \cite{49}.

In our case series, 5 of the 6 patients in group C had a ductus arteriosus with the classic morphology reported in the literature. When pulmonary obstruction is completed during fetal life, the ductus arteriosus may change its usual morphology, reduce its caliber, increase its tortuosity, and the angle between the pulmonary artery and the descending aorta may change, such that it is no longer obtuse. This change in its configuration could be explained by the absence of normal anterograde flow from the pulmonary artery to the aorta since the earliest phases of fetal life. The flow would occur only from the aorta to the pulmonary artery. This abnormal ductal morphology is also observed in patients with pulmonary atresia with ventricular communication, in whom pulmonary atresia is postulated to occur in even earlier phases of fetal life. On the other hand, all the 16 group A patients had ductus arteriosus of larger caliber, which formed an obtuse angle with the descending aorta.

Therefore, pulmonary atresia of the membranous type with imperforate valve is more frequently found in patients with pulmonary atresia with intact septum, who have a ductus arteriosus of usual morphology, forming an obtuse angle with the descending aorta. On the other hand, in patients with ductus arteriosus of anomalous morphology, atresia of the infundibular muscular type is more frequently found. The difference in age between patients in group A (older) and those in groups B and C is noteworthy, and may be due to the greater severity of the clinical findings in the patients of the latter groups, who had less favorable anatomies.

As all neonates with congenital heart diseases with pulmonary atresia in whom the pulmonary flow depends on the permeability of the ductus arteriosus, patients with pulmonary atresia with intact septum should be maintained under continuous infusion of prostaglandin until definition of the therapeutic strategy \cite{50-53}. The effects of the action of prostaglandins on the angiographic morphology are little known and cannot be ruled out. Due to the important vasodilating effect of prostaglandins on the pulmonary artery and ductus arteriosus, it is speculated that a tendency exists towards a reduction in the angle formed by the ductus arteriosus
and the descending aorta, consequent to reorientation of the increased flow from the aorta to the pulmonary artery.

In pulmonary atresia with intact septum, the major objective of the treatment is to promote clearance of the right ventricular outflow tract, either by thoracotomy or by the percutaneous route, aiming at stimulating the intracavitary flow, reducing the degree of hypertrophy, improving ventricular compliance, and enabling enlargement of the ventricular cavity to make biventricular connection feasible. In actual practice, however, the entire current literature agrees that these procedures are not indicated for all patients. This limitation is justified by the morphofunctional spectrum of these patients, thus the need for knowing the heterogeneity existing among patients with this disease.

In the selection of patients, the entire initial attention should be directed to the size or dimensions of the right ventricle. However, the difficulty resides in the importance given to the right ventricular size, because, unlike the left ventricle, the right ventricle does not have a geometrical model with which its volume can be reliably calculated. In addition, in pulmonary atresia with intact septum, the degree of opacity of the trabecular portion is variable, and, therefore, all measures used to calculate its real volume will certainly have an error in its correct estimate. Because of the distortions in right ventricular morphology, the assessment of its volume is accepted with some restrictions, as the formula by Simpson or Dodge cannot be correctly applied to hearts with a normal or 70% of the diameter of the mitral ring represents a risk factor or failure factor for biventricular connection19-22.

In our material, a strong correlation was observed between the diameter of the tricuspid valve ring or a diameter of the tricuspid valve < 70% of the normal or 70% of the diameter of the mitral valve ring represents a risk factor or failure factor for biventricular connection19-22.

In conclusion, pulmonary atresia with intact septum represents an important group of patients with cyanotic congenital heart disease in the neonatal period. The alternatives of treatment of this heterogeneous group require meticulous assessment of the sizes of the right ventricle and the tricuspid valve, and, mainly, the study of the coronary circulation in the form of right ventricle-dependent coronary circulation or other coronary anomalies. Although better results, not only surgical but also of interventional catheterization, have been recently reported in the literature, mortality still remains elevated.

References


