Development of the Ductus Arteriosus in Right Ventricular Outflow Tract Obstruction

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SUMMARY We studied the morphology of the ductus arteriosus in 14 infants, ages 2–90 days. Eight (group 1) had pulmonary atresia (structural and functional) with an intact interventricular septum; six (group 2) had pulmonary atresia with a ventricular septal defect. The inferior angle of the ductus arteriosus at the aortic junction was measured in each patient. In group 1, this angle was obtuse in all but one patient. In group 2, the angle was acute in all. Further study of intracardiac anatomy suggested that in group 1, the obtuse inferior angle of the ductus arteriosus was the result of a late and progressive obstructive phenomenon that allowed normal right-to-left flow through the ductus arteriosus during much of fetal life. In group 2, the direction of ductus arteriosus flow (normally from the pulmonary trunk to the aorta) was reversed, and flowed from the aorta to the pulmonary trunk. This reversal of flow was probably of early onset in the fetus, the aorta receiving the total combined ventricular output, and produced a small ductus arteriosus with an acute inferior angle. It is extremely important not to rule out pulmonary atresia with an intact interventricular septum when aortography in the newborn shows a normal-sized ductus arteriosus with an obtuse inferior angle. Despite existing pulmonary atresia, these patients have neither a hypoplastic right ventricle nor discontinuity of the right ventricle with the pulmonary artery.

THE IMPORTANCE of the development of the ductus arteriosus in congenital heart disease, particularly in right ventricular outflow tract obstruction in which patency of the ductus is essential to life, has recently been stressed. Although correct evaluation of the size of the ductus arteriosus in infants with pulmonary atresia is often difficult because constriction occurs shortly after birth, the ductus arteriosus has been described as a small structure that forms an acute inferior angle at its connection with the descending aorta. However, these features differ from those we observed in a select group of patients who, despite having pulmonary atresia with an intact interventricular septum, on cineangiography had a normal-sized ductus arteriosus forming a normal obtuse inferior angle with the descending aorta. The angiographic features and surgical prognosis of these patients must be differentiated from those of patients with an underdeveloped ductus arteriosus, pulmonary atresia and an intact interventricular septum.

In this report, we discuss the fetal development of the ductus arteriosus based on the time during fetal life at which the right ventricular outflow tract obstruction occurs, and on the consequent changes produced in the fetal circulation. This knowledge is important in the correct management of this group of high-risk patients.

Material and Methods

Fourteen patients, 2–90 days of age, underwent diagnostic hemodynamic and cineangiographic study (table 1). The interventricular septum was intact in eight (group 1) and six had a ventricular septal defect (group 2). In group 1, the pulmonary valve was atretic in five (structural atresia) and three had evidence of only pinpoint patency of the pulmonary valve (functional atresia). There was right-to-left atrial shunting in all. In group 2, all patients had pulmonary atresia and a right-to-left shunt at the ventricular level. The diagnosis was confirmed in all patients at necropsy or at surgery.

We determined the inferior angle between the ductus arteriosus and the descending aorta from the angiograms performed in the left anterior oblique projection, and compared these with angiograms obtained in full-term human newborn infants catheterized either to confirm the diagnosis of patent ductus arteriosus or to exclude congenital malformations suspected clinically.

Results

Group 1

One patient (fig. 1) had a small, competent tricuspid valve, a hypoplastic right ventricle and pulmonary atresia. Myocardial sinusoids were well developed, and opacification of the aorta occurred through an anastomosis of the sinusoids with the right and left coronary arteries. The inferior angle of the ductus arteriosus was 30° (fig. 2). This patient had type I pulmonary atresia by criteria of Davignon et al. In the other seven patients (fig. 2), the inferior angle of the ductus exceeded 90°. In four (fig. 3) the pulmonary valve was atretic and in three (fig. 4) there was evidence of pinpoint patency of the pulmonary valve with very faint opacification of the main pulmonary artery. All had well-formed tricuspid valves with mild-to-severe regurgitation. The right ventricular cavities were large, and myocardial sinusoids were absent. These patients had type II pulmonary atresia by the criteria of Davignon et al.
GROUP 2

In all six patients in group 2 (fig. 5), the right ventricular cavity was not hypoplastic. Only one patient had tricuspid regurgitation. The inferior angle of the ductus arteriosus was less than 50° in all.

Discussion

A narrow, tortuous ductus arteriosus with an acute inferior angle at the junction of the aorta and the ductus arteriosus and a wide aortic isthmus are common features in patients with pulmonary atresia or severe pulmonary stenosis.1 These features were observed in only one of eight patients with pulmonary atresia and intact ventricular septum (group 1), and in all those with a ventricular septal defect (group 2). We therefore considered that variations in intracardiac...
anatomy could lead to major differences in the growth and development of the great arteries and ductus arteriosus in fetuses with pulmonary atresia.

Group I included not only patients with structural pulmonary atresia, but also those who showed evidence of pinpoint patency of the pulmonary valve with only faint or no opacification of the pulmonary artery. We classified these patients as having functional pulmonary atresia because all had essentially no forward flow from the right ventricle and significant right-to-left shunting at the atrial level.

Although we did not classify the patients in this group according to the size of the ventricular cavity,* only one of the eight patients had a hypoplastic right ventricle and a small tricuspid valve without evidence of regurgitation. This, together with the development of large myocardial sinusoids, indicates the likelihood that blood did not pass freely from the right ventricle to the pulmonary artery during the greater part of this infant's fetal life. When obstruction to pulmonary flow has been critical or complete from early in fetal life, the size and configuration of the ductus arteriosus should reflect the small amount of aortic-to-pulmonary artery flow across the ductus arteriosus, and will differ from the normal ductus arteriosus, which acts as a continuous arch between the

![Figure 4. Patient 6. (A and B) Right ventricular cineangiograms (anteroposterior projection) showing the right ventricle (RV) in systole and diastole. The infundibulum (I) is markedly stenotic and there is severe tricuspid regurgitation. (C) Right ventricular cineangiogram (left anterior oblique projection) showing faint early opacification of the pulmonary artery, indicating pinpoint patency of the pulmonary valve (arrow). (D) Left ventricular cineangiogram (left anterior oblique projection) showing a constricted ductus arteriosus (D) with an obtuse inferior angle (arrow). AAO = ascending aorta; DAO = descending aorta; IS = aortic isthmus; LV = left ventricle; RA = right atrium.](image-url)
Table 1. (Continued)

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Pulmonary artery and the descending aorta. Further, the aorta will not show the normal isthmic narrowing that is a consequence of the reduced flow across the isthmus and the large right-to-left flow through the ductus arteriosus during normal fetal life.1, 3, 8 This infant had just such anatomy, confirming this theory.

The other seven patients in this group did not have hypoplastic right ventricular cavities, and myocardial sinusoids were absent in all of them. Four of the seven only had mild tricuspid insufficiency. These findings suggest that the normal or nearly normal size of the right ventricle, and certainly of the tricuspid orifice, reflected significant blood flow passing through the right ventricle during much of fetal life.

In pulmonary atresia with an intact interventricular septum, it is possible that the obstruction at the level of the pulmonary valve becomes progressively more severe during advancing fetal life, culminating at birth, in a pulmonary valve that is either a minute pinhole or even atretic. The presence of a normally developed and normally situated ductus arteriosus with an obtuse inferior angle in these patients suggests that during the larger part of fetal life there has been normally directed flow from the pulmonary artery to the descending aorta. However, the progressive obstructive phenomenon reorients flow into the right ventricle, and therefore, through the foramen ovale. Progressively more of systemic venous return enters the left atrium and is ejected by the left ventricle, thereby making the isthmic portion of the aorta wider than normal.

Although we have not encountered any reports of pulmonary atresia with these angiographic features, Bharati et al.9 describe the presence of a normal-sized ductus arteriosus in a morphologic study, but did not make any reference to the inferior angle.

All six patients in group 2 had large right ventricular cavities and tricuspid valves; only one patient

Figure 5. Patient 13. (A and B) Left ventricular cineangiogram (posteroanterior and lateral projections) showing a narrow, tortuous ductus arteriosus (D) with an acute inferior angle (arrow). The right ventricular cavity, opacified through the ventricular septal defect is of normal size but the infundibulum (I) is small and there is discontinuity between the right ventricle (RV) and pulmonary artery. AAO = ascending aorta; DAO = descending aorta; IS = aortic isthmus; LV = left ventricle.
had regurgitation. The aortic diameters had the same proportions as in group 1, but the ductus arteriosus was not normally developed, and the inferior angle was acute in all patients.8 These features suggest that the right ventricular outflow tract obstruction occurred early in fetal life, but the ventricular septal defect permitted the flow of blood from the right ventricle to the left ventricle and aorta, and therefore did not significantly affect right ventricular development. The configuration of the ductus arteriosus indicated flow from the aorta to the pulmonary trunk for a considerable portion of gestation.

These observations are important for correct diagnosis and surgical management of these patients and for understanding the developmental process. Although none of our patients underwent aortography, this approach is being used more and more frequently in the diagnostic investigation of cyanotic newborn infants.9 The angiographic features of a normal-sized ductus arteriosus with an obtuse inferior angle do not exclude the possibility of pulmonary atresia.

The size of the right ventricular cavity and the level of atresia, whether valvar or infundibular or both, may have important surgical and prognostic significance, although this is still a controversial subject.9,11,12 Freedom et al.10 used a technique that permits simultaneous opacification of the right ventricle and the pulmonary artery, thereby obtaining better definition of the level and site of the atretic segment. The patients who had a normally developed ductus arteriosus almost certainly maintained blood flow from the right ventricle to the pulmonary artery during a large part of fetal life. Consequently, they show neither a hypoplastic right ventricle nor discontinuity between this cavity and the pulmonary artery, because the atretic segment is localized at the valvar level.

In conclusion, patients with right ventricular outflow tract obstruction do not always have an underdeveloped ductus arteriosus with an acute inferior angle. We have described infants with pulmonary atresia with intact interventricular septum and a normally developed ductus arteriosus. In these infants, the obstructive phenomenon probably developed late in fetal life.

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