PULMONARY ATRESIA WITH BIVENTRICULAR AORTA

BY

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Among the cyanotic congenital heart malformations, there is a group characterized anatomically by the presence of an atretic pulmonary artery and a biventricular aorta which acts as the only functioning vessel arising from the heart. A mixture of oxygenated blood from the left ventricle and unsaturated blood from the right ventricle is injected into the aorta, and the pulmonary vessels are filled from the aorta either through a patent ductus arteriosus, bronchial collaterals, or both. The main clinical features of patients bearing this type of heart malformation are deep cyanosis, single second heart sound, and decreased lung vascularity. A systemic pulmonary anastomosis is the most generally accepted surgical procedure of choice.

The term “pseudotruncus arteriosus” or “truncus aorticus” has been used by some authors (Brinton and Campbell, 1953; Cooley, Bahnsen, and Hanlon, 1949; Humphreys, 1932; Kjellberg et al., 1959; Kugel, 1931; Taussig, 1960) to designate those cases of atretic pulmonary artery and overriding aorta with pulmonary artery normally placed in front of the aorta, i.e. extreme forms of Fallot’s tetralogy. In our opinion, this term should also include the much less frequent group of malformed hearts with atretic pulmonary artery and biventricular aorta but with transposition of the great arteries, since the basic hemodynamics, in many ways similar to that of a true truncus, is essentially the same regardless of the spatial relation between the two great arteries.

The expression pulmonary atresia with biventricular aorta defines the basic anatomical and hemodynamic features, and semantically separates these cases from those of pulmonary atresia with normal aortic root, which, anatomically, hemodynamically, and clinically, constitute a totally different group.

It is the purpose of this paper to describe the anatomical aspects of 18 necropsy cases fulfilling the preceding definition, and to outline some anatomical types. A review of the published material on combined pulmonary atresia, biventricular aorta, and transposition of the great arteries is also presented.

MATERIAL AND METHODS

Eighteen hearts with pulmonary atresia and biventricular aorta were studied: eight from the Museum of the Instituto Nacional de Cardiología de México, representing 3 per cent of the 252 specimens; and 10 from the Museum of Pathology of the Hospital J. M. de los Ríos of Caracas, representing 4 per cent of the 250 specimens of congenital heart disease.

In each specimen the infundibulum of the right ventricle was studied, as well as the pulmonary valve, the calibre and length of the pulmonary artery trunk and its spatial relation with the ascending portion of the aorta, the degree of aortic overriding, the size of the branches of the pulmonary artery, and the thickness of the walls of both ventricles. Other associations were looked for and noted.

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ANATOMICAL DESCRIPTION

For the purpose of anatomical description we will consider separately those cases of pulmonary atresia and overriding aorta without transposition of the great arteries—extreme Fallot’s tetralogy—and those with pulmonary atresia, overriding aorta, and transposed great arteries.

Group I: Pulmonary atresia and biventricular aorta without transposition of the great arteries (extreme Fallot’s tetralogy) (Fig. 1A). There were 15 specimens in this group (83%). The pulmonary artery of decreased calibre arose in every case to the right of the aorta in an anterior position, crossed it ventrally, and then turned back and leftward before dividing into the right and left pulmonary branches. The right anterior ventricular wall at the level of the infundibulum, immediately below the pulmonary artery, exhibits a prominent muscular bulge which gives the specimens a characteristic external aspect. They all showed marked hypertrophy of the right ventricle and an excessive length of the pulmonary artery trunk. The calibre of the pulmonary trunk and its branches was decreased in all cases. A ventricular septal defect was present in every case, behind the crista supraventricularis and immediately below the aorta, which overrode the interventricular septum in variable degree (33% overriding in 4 cases, 66% in 6 cases, and 100% in 5 cases).

In 7 cases the ductus arteriosus was patent; in 9 there was a patent foramen ovale, and in 2 there was an atrial septal defect in the region of the fossa ovalis.

Taking into account some important variations in the anatomy of the infundibulum of the right ventricle and of the pulmonary valve, 3 anatomical subgroups will be described, the recognition of which may prove useful for the diagnosis and helpful when surgical treatment is considered.

(A) Atresia of the pulmonary valve and infundibular pulmonary stenosis (Fig. 2A and B). There were 4 cases in this subgroup (26%). In all of them the crista supraventricularis was either of
normal thickness or thicker than normal. The infundibulum of the right ventricle was very narrow, but an infundibular chamber was discernible. The pulmonary sigmoid valve cusps were totally fused causing an atresia of the pulmonary orifice.

(B) *Valvular pulmonary stenosis and aplasia of the infundibulum* (Fig. 3A). Only one case in our series (6%) had these characteristics. The infundibular cavity was absent because of the fusion of the parietal band of the crista supraventricularis with the anterior infundibular wall, forming a muscular mass with a thickness of about 20 mm. The pulmonary artery showed valvular stenosis but the arterial orifice was not atretic.

(C) *Atresia of the pulmonary valve and aplasia of the infundibulum* (Fig. 3B). This was the most numerous subgroup (10 specimens=66%). The crista supraventricularis was always muscular and it was fused with the anterior infundibular wall, thus producing the aplasia of the infundibulum. Both structures together formed an anteriorly placed muscular mass whose thickness ranged from
PULMONARY ATRESIA WITH BIVENTRICULAR AORTA

Fig. 3.—Pulmonary atresia and biventricular aorta without transposition of the great arteries (extreme Fallot's tetralogy). (A) Specimen illustrating the subgroup with infundibular aplasia and stenosis of the pulmonary valve. A section has been made in the anterior wall of the right ventricle in order to show the stenotic pulmonary valve arising directly from the muscular mass formed by the fusion of the crista supraventricularis and the anterior infundibular wall. (B) Specimen illustrating the group with aplasia of the infundibulum and atresia of the pulmonary valve. The right ventricle has been opened to show these anatomical features. (1) pulmonary artery; (2) aorta.

8 to 20 mm. with a mean of 13.3 mm. The pulmonary sigmoid valve cusps were fused, and the pulmonary artery actually arose from the muscular mass previously described.

Group II: Pulmonary atresia, biventricular aorta, and transposed great arteries (Fig. 2C and D). Three cases in our series belonged to this group (16%). It was characterized by the fact that the aorta arose in front of the crista supraventricularis and ran ventral and parallel to the trunk of the atretic pulmonary artery. The latter was placed behind and to the left, and it emerged as a blind sac from an atretic orifice. In two cases there was levocardia with situs inversus and a persistent common atrio-ventricular canal of the complete form. In the third a ventricular septal defect behind the crista supraventricularis formed the pathway through which the right-sided aorta received the blood from the left ventricle.

Discussion

In 1956 one of us (de la Cruz) proposed an ontogenic theory for the explanation of congenital truncoconal malformations. An account of the embryological processes involved in the production of the anatomical heart alterations we are dealing with was contained in that publication. A brief discussion of those developmental deviations will be included herein for a better understanding of the anatomical variations found in this series.

An extreme unequal partition of the truncus-conus, at the expense of the pulmonary artery, and malformation of the primordia of the pulmonary sigmoid valve cusps, are the embryological alterations common to all cases of pulmonary atresia with biventricular aorta. The unequal partition determines the small calibre of the pulmonary artery trunk and prevents the normal alignment of the truncoconal septum with the primitive interventricular septum, thus originating a ventricular septal defect, above which the aorta overrides the interventricular septum.

In the cases without transposition of the great arteries (extreme Fallot's tetralogy) there is a normal rotation of the spiral truncoconal septum (Fig. 1A). The truncoconal ridges arise with a normal clockwise rotation of 180° and, when they fuse with one another, they determine the formation of a spiral septum with the same type and degree of rotation. This makes the pulmonary artery branches (sixth aortic arch) which, at the cephalic end of the truncus, have a sinistro-dorsal position, communicate with the anatomically right ventricle which, in the caudal end of the conus,
has a dextro-ventral position. For the same reason the aortic arch (fourth aortic arch), which at the cephalic end of the truncus has a dextro-ventral situation, will communicate with the anatomically left ventricle, which in the caudal end of the conus has a sinistro-dorsal position (Fig. 1A). Thus, the pulmonary artery arises anteriorly and to the right crossing the aorta ventrally, thence to direct itself backwards and leftwards before dividing into right and left pulmonary artery branches.

Variations in the degree of unequal partition of the truncus-conus at the level of the conal region and of malformations of the primordia of the pulmonary sigmoid valve cusps bring about the existence of anatomical varieties.

In the cases with pulmonary valve atresia and infundibular pulmonary stenosis, the unequal partition of the truncus-conus is not severe enough at the level of the conal region to produce infundibular aplasia. The annulus of the pulmonary valve is very narrow and the primordia of the pulmonary sigmoid valve cusps present extreme alterations with complete fusion of the valve cusps and consequent pulmonary valvular atresia.

In the cases with infundibular aplasia and valvular pulmonary stenosis the degree of unequal partition of the truncus-conus is extreme at the level of the conal region and produces a complete fusion of the parietal band of the crista supraventricularis with the anterior infundibular wall: this produces aplasia of the infundibulum of the pulmonary artery. The valvular pulmonary ring is smaller than normal, leaving little space for the primordia of the pulmonary sigmoid valve cusps to grow; the latter fuse partially, originating valvular pulmonary stenosis.

In the cases with valvular pulmonary atresia and aplasia of the infundibulum the degree of unequal partition of the truncus-conus at the level of the conal region is of the same magnitude as in the previous group. The distortion of the primordia of the pulmonary sigmoid valve cusps is also extreme and results in valvular pulmonary atresia.

In the cases with transposed great arteries, besides the unequal partition of the truncus-conus and the alterations of the primordia of the pulmonary sigmoid valve cusps there is an abnormal development of the truncocral septum. The truncocral ridges lack their normal clockwise rotation of 180°; they emerge in a straight manner and when they fuse with one another a straight truncocral septum is formed (Fig. 1B). This fact makes the pulmonary artery branches (sixth aortic arch), which at the cephalic end of the truncus have a sinistro-dorsal position, communicate with the anatomically left ventricle, which at the caudal end of the conus also has a sinistro-dorsal position. Similarly, the aortic arch (fourth aortic arch), which at the cephalic end of the truncus occupies a dextro-ventral position, will communicate with the anatomically right ventricle, which at the caudal end of the conus has a similar dextro-ventral situation (Fig. 1B). The aorta arises ventrally and to the right of the pulmonary artery, and both great arteries are parallel. The alterations of the primordia of the pulmonary sigmoid valve cusps are severe and lead to complete fusion and consequent atresia of the pulmonary valve.

Of our 18 cases of pulmonary atresia with biventricular aorta, 3 had transposed great arteries. This low incidence in comparison with the Fallot’s tetralogy group is also found in the published reports. In reviewing the reports we found 14 cases of this group, and in 5 of them there was a complete description of the specimens (Fontana and Edwards, 1962; Kelsey, Gilmore, and Edwards, 1953; Kugel, 1931; Rosman, 1942; Young and Griswold, 1951): 1 had transposition of the great arteries with pulmonary atresia associated with a single ventricle and a single atrium (Rossman, 1942); in 3 the ventricular septal defect was described as occupying an anterior position immediately below the ventrally-placed aorta (Kelsey et al., 1953; Kugel, 1931; Young and Griswold, 1951), and in a fifth case the exact situation of the ventricular septal defect was not defined (Fontana and Edwards, 1962). In 2 of our 3 cases there was an associated endocardial cushion defect and the ventricular septal defect was of the type commonly seen in this malformation, i.e. posteriorly placed, in the region of the atrio-ventricular canal. In our third case the ventricular septal defect was placed behind the crista supraventricularis, and the atretic pulmonary artery seemed to arise directly from the interventricular septum. This can be explained on the basis of a delayed disappearance of the conoventricular flange in this case, causing dextroposition of the truncus-conus,
PULMONARY ATRESIA WITH BIVENTRICULAR AORTA

TABLE I

PULMONARY ATRESIA WITH BIVENTRICULAR AORTA AND TRANSPOSITION OF THE GREAT ARTERIES

<table>
<thead>
<tr>
<th>Author</th>
<th>Age</th>
<th>Associated malformations</th>
<th>Observations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Breschet (1826)</td>
<td>1 mth</td>
<td>Dextrocardia, atrial septal defect, ventricular septal defect</td>
<td></td>
</tr>
<tr>
<td>Hickman (1869)</td>
<td>1½ mth</td>
<td>Lævocardia, with situs viscerum inversus abdominalis, atrial septal defect, ventricular septal defect</td>
<td>Corrected transposition of the great arteries</td>
</tr>
<tr>
<td>Northrup (1889)</td>
<td>31 dy</td>
<td>Dextrocardia, cor biloculare, patent ductus arteriosus</td>
<td></td>
</tr>
<tr>
<td>Baumgarth (1902)</td>
<td>1 yr</td>
<td>Dextrocardia, cor biloculare, patent ductus arteriosus</td>
<td></td>
</tr>
<tr>
<td>McCrae (1905)</td>
<td>7 wk</td>
<td>Lævocardia, with situs viscerum inversus abdominalis, atrial septal defect, patent ductus arteriosus</td>
<td>Corrected transposition of the great arteries</td>
</tr>
<tr>
<td>Beattie (1922)</td>
<td>Infant</td>
<td>Dextrocardia, atrio-ventricularis communis complete variety, ventricular septal defect, patent ductus arteriosus</td>
<td>Corrected transposition of the great arteries</td>
</tr>
<tr>
<td>Rösler (1930)</td>
<td>6 yr</td>
<td>Dextrocardia, atrial septal defect, ventricular septal defect, patent ductus arteriosus</td>
<td>Corrected transposition of the great arteries</td>
</tr>
<tr>
<td>Kugel (1931)</td>
<td>3 mth</td>
<td>Ventricular septal defect, patent ductus arteriosus, agenesis of the right pulmonary artery</td>
<td></td>
</tr>
<tr>
<td>Rossman (1942)</td>
<td>9½ mth</td>
<td>Cor biloculare, atrio-ventricularis communis, complete variety; patent ductus arteriosus</td>
<td></td>
</tr>
<tr>
<td>Miskall and Fraser</td>
<td>3½ dy</td>
<td>Dextrocardia, cor biloculare, patent ductus arteriosus</td>
<td>Corrected transposition of the great arteries</td>
</tr>
<tr>
<td>Mannheimer (1949)</td>
<td>7 yr</td>
<td>Dextrocardia, cor biloculare, patent ductus arteriosus</td>
<td></td>
</tr>
<tr>
<td>Young and Griswold</td>
<td>23 mth</td>
<td>Lævocardia, with situs viscerum inversus abdominalis, ventricular septal defect, patent ductus arteriosus</td>
<td></td>
</tr>
<tr>
<td>Kelsey et al. (1953)</td>
<td>10 mth</td>
<td>Dextro-rotation, patent foramen ovale, right-sided patent ductus arteriosus, single ventricle</td>
<td>Corrected transposition of the great arteries</td>
</tr>
<tr>
<td>Fontana and Edwards</td>
<td>6 mth</td>
<td>Lævocardia with situs inversus, atrial septal defect, ventricular septal defect, patent ductus arteriosus</td>
<td></td>
</tr>
<tr>
<td>Our Case (1)</td>
<td>1½ yr</td>
<td>Ventricular septal defect, patent ductus arteriosus, patent foramen ovale</td>
<td>Corrected transposition of the great arteries</td>
</tr>
<tr>
<td>Our Case (2)</td>
<td>7 mth</td>
<td>Lævocardia with situs inversus, atrio-ventricularis communis</td>
<td>Corrected transposition of the great arteries</td>
</tr>
<tr>
<td>Our Case (3)</td>
<td>3 yr</td>
<td>Lævocardia with situs inversus, atrio-ventricularis communis</td>
<td></td>
</tr>
</tbody>
</table>

which, in turn, makes the ventricular septal defect appear behind the crista supraventricularis, and the atretic pulmonary artery arise in close relation with the interventricular septum.

It is interesting to point out that the great majority of the published cases had abnormal cardiac positions: 7 had dextrocardia, 4 had levocardia, and 1 had dextro-rotation. In two of our cases there was levocardia with situs inversus (see Table I).

Some other embryological facts are worth pointing out. In the first place it is important to emphasize that the branches of the pulmonary artery and the main trunk of this vessel have a different embryological origin. In fact, the branches originate from the sixth aortic arch and the trunk from the division of the truncus arteriosus. For this reason, even in cases in which the pulmonary artery is atretic and its trunk of reduced calibre, the right and left pulmonary branches may be of middle size or even near-normal size. In these a successful systemic-pulmonary anastomosis (Cooley et al., 1949) can be performed even though the existence of pulmonary atresia could incline one to
expect a poor result. Of the 18 cases studied, 3 had practically normal-sized pulmonary branches; in 10 the pulmonary branches were smaller than normal, but even then larger than the pulmonary trunk; only 5 had very small pulmonary branches, too small to accept increased flow through a Blalock fistula or other type of systemic-pulmonary anastomosis.

In those cases with pulmonary atresia and biventricular aorta without transposition of the great arteries (extreme Fallot’s tetralogy), there is no direct correlation between the degree of unequal partition of the truncus-conus and the degree of dextroposition (de la Cruz and da Rocha, 1956). This is expressed anatomically by the fact that, despite the extreme unequal partition of the truncus-conus with a very small pulmonary trunk in all cases, the overriding of the aorta was quite variable from one case to the other.

The recognition of different anatomical subgroups is important since the surgical possibilities are different for each one of them. In the group without transposed great arteries (extreme Fallot’s tetralogy) the variations in the anatomy of the infundibulum of the pulmonary artery and the pulmonary sigmoid valve have important surgical implications. In those with a narrow infundibular chamber and valvar pulmonary atresia, a complete surgical repair may be indicated if other considerations do not contraindicate the procedure (Brinton and Campbell, 1953; Johns, Williams, and Blalock, 1953). Those cases with infundibular aplasia, with or without concomitant valvar atresia, are generally beyond the possibilities of a complete repair and they may only benefit from an anastomotic procedure. However, Lillehei (Lillehei et al., 1955) operated on a patient with infundibular aplasia and atresia of the pulmonary artery, and he was able to do a complete correction with good results.

In those cases of atretic pulmonary artery, biventricular aorta, and transposed great arteries, the only surgical possibility is an anastomotic procedure of the Blalock type. This operation in cases with transposition of the great arteries with stenosis or atresia of the pulmonary artery, although with less favourable results than in Fallot’s tetralogy, is followed by improvement in a good number of cases (Cleland et al., 1957; Young and Griswold, 1951). Sometimes, even after angiocardiographic studies, the presence or absence of transposition of the great arteries cannot be established with certainty (Cleland et al., 1957). In these, the diagnosis of pulmonary atresia with overriding aorta justifies the indication of the anastomotic operation regardless of the reciprocal position of the aorta and the pulmonary artery.

SUMMARY

The authors present an anatomical study of 18 specimens with pulmonary atresia and biventricular aorta. Two anatomical groups are distinguished according to the existence or absence of transposition of the great arteries. Three subgroups are outlined among the cases without transposition of the great arteries (extreme Fallot’s tetralogy), determined by variations in the anatomy of the infundibulum of the pulmonary artery and the pulmonary sigmoid valves. The embryological deviations involved are discussed and the surgical possibilities of each anatomical group are analysed.

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REFERENCES

PULMONARY ATRESIA WITH BIVENTRICULAR AORTA


