CASE REPORTS

CARDIO-AORTIC FISTULA THROUGH ANOMALOUS CORONARY ARTERIES

BY

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Anomalous coronary arteries may bring the aorta into communication with the cardiac chambers or the pulmonary artery, creating various abnormalities comparable with arteriovenous fistulae that may be of clinical and surgical importance. Recently Edwards (1958) has summarized the field of coronary anomalies, contributing a functional and anatomical classification of them.

Among the specimens in the Department of Pathology of the Children’s Hospital of Buenos Aires, there are two in which coronary arteries bring the aorta into communication with the right ventricle. In both there is atresia of the pulmonary artery. The present case is of special interest because of the existence of an additional ventricular septal defect, absent in the heart of the patient of Kreutzer et al. (1953) where the right ventricle was normally developed.

Case Report. The patient who had been cyanotic since birth, was seen at the Department of Cardiology at the age of 20 days. The apical impulse was normal, and there was a systolic murmur of moderate intensity in the mid-præcordial, apical, and tricuspid areas, but no thrill. Radiological examination revealed slight cardiac enlargement with decreased pulmonary vascular markings, concavity of the pulmonary conus, and a convex right cardiac border. The cardiogram (Fig. 1) showed sinus tachycardia, a normal electrical axis, and normal conduction times: the P wave was peaked in lead II and right præcordial leads, with diphasic QRS in VI and positive T waves in the left præcordial leads.

At two months of age the child was readmitted to the hospital for further investigation. He weighed 4.1 kg. The cyanosis, which was permanent, had decreased slightly in intensity. Venous angiocardiography was performed one month later, and the child died suddenly 24 hours after this procedure. The first film of the angiocardiogram (Fig. 1b) shows the contrast medium in the superior vena cava, the right atrium and the inferior vena cava. Additionally, an anomalous blood vessel is seen extending across the cardiac silhouette from the right atrium to the region of the pulmonary conus. The degree of opacity of this anomalous blood vessel is equal to or greater than the opacity of the aorta. At necropsy it was found that this anomalous blood vessel corresponded to the fistulous right coronary artery. Contrast medium was not visualized within the right ventricle, the pulmonary trunk, or its branches.

Necropsy Findings. The heart was of approximately normal size. There was atresia of the pulmonary trunk but it was not transposed (Fig. 2a); there was a thin patent ductus arteriosus. The left ventricle made up two-thirds of the anterior surface of the heart, but the right ventricular mass was larger than is usual in simple pulmonary atresia. The right atrium was enlarged and hypertrophic. The vena cava and the patent foramen ovale were normal. The tricuspid orifice was hypoplastic, measuring at ring level 8 mm. in diameter. Its valves were proportionately small but fully developed, so that the anterior and posterior leaflets and both papillary muscles of the right ventricle could be easily recognized.

A striking feature of the specimen was the dilatation and tortuosity of both coronary arteries, which arose from the aorta at the normal sites of origin. Close to the apex of the heart the descending branches of both coronary arteries joined to form a whitish angiomatosus mass. A remarkable difference existed between the left circumflex coronary artery, which was normal, and the other tortuous arteries. The position within the thorax of the right coronary artery corresponded exactly to the blood vessel opacified in the first film of the angiocardiogram.

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FIG. 1.—(a) Electrocardiogram, described in the text.  (b) First film of the venous angiocardigram: S.V.C., superior vena cava; I.V.C., inferior vena cava; R.A., right atrium; A., aorta; R.V., right ventricle; L.V., left ventricle.  Across the cardiac silhouette, between the area shown by arrows, an anomalous blood vessel has been opacified.  At necropsy this vessel was shown to be the right coronary artery.

Water injected into the right ventricle immediately filled both coronary arteries, and the liquid was seen to spurt forth forcibly from the coronary ostia.  The right ventricular chamber was the size of a hazel-nut amidst thick muscular walls, and the pulmonary infundibulum was clearly recognizable as a blind pouch superior to the crista supraventricularis.  A typical ventricular septal defect 6 mm. in diameter was present, being located posterior to the undistorted papillary muscle of the conus. The aorta did not override the defect.

An ample connection was found between the right ventricular chamber and the aneurysmal mass at the apex, which in turn was in free communication with both descending coronary arteries.  Toward the apex the lumen of the right ventricle became narrowed to a muscular tract (Fig. 2b) which continued into an irregular sac with fibromuscular walls (Fig. 2c).  From this sac both descending coronary arteries could be entered with probes 3 mm. (anterior) and 2 mm. (posterior) in diameter.

The left ventricle and atrium were essentially normal but dilated.  No aortic abnormalities were present.  The coronary ostia were more than twice the normal size.  The lungs showed no edema or focal alveolar hemorrhage.  Microscopic examination revealed residual alveolitis; the arteries were thin walled, the lumen being large.  The rest of the pathological examination showed no other relevant abnormal findings.  The final diagnosis was atresia of the pulmonary trunk, tricuspid hypoplasia, a fistula between aorta and right ventricle through both coronary arteries, and a ventricular septal defect located posterior to the papillary muscle of the conus.
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Fig. 2.—(a) Anterior aspect of the specimen. The pulmonary trunk (P.T.) is atretic and the pulmonary circulation is maintained through a small patent ductus arteriosus (P.D.A.). Both the anterior descending coronary artery (A.D.C.A.) and the right coronary artery (R.C.A.) are enlarged and tortuous. The arrow points at the angiomatous aneurysmal mass present close to the apex of the heart. This aneurysm is formed by the junction of both descending coronary arteries, and an ample connection was present between it and the right ventricle which is somewhat smaller than the left.

(b) and (c). The apex of the right ventricle, opened by a section that passes close to the ventricular septum (V.S.) and includes the aneurysmal sac at the apex (a). A single polyethylene catheter has been threaded into both the anterior (A.D.C.) and the posterior (P.D.C.) descending coronary arteries, so that its mid-portion comes to lie within and through the aneurysmal sac at the apex, indicating the continuity of these structures. In (b) the specimen has been inclined to the right, showing the muscular tract continuing the lumen of the right ventricle into the aneurysmal sac (arrow). In (c) the specimen has been inclined to the left, showing the ventricular septum (V.S.) and the aneurysm at the apex (a).

Discussion

The second group of Edwards’ classification includes “coronary anomalies secondary to the effects of certain primary cardiac malformations, such as atresia of either the aortic or pulmonary valve”. Under these circumstances, and in the presence of an intact ventricular septum and a competent atroventricular valve, the anomalous vessel allows blood to be expelled from the ventricle during systole, and can be interpreted simply as a persistence of the normal vascular connections existing in the fetus by means of the Thebesian vessels. The anomalous increase of these connections would be secondary to the effects of obstruction to the ventricular outflow.

We believe that another interpretation can be considered. The coronary anomaly may be the primary malformation. In favour of this point of view are the following considerations: (i) the ventricular septum is a sponge-like structure, with multiple openings in it, till approximately the sixth week of development, at a time when the partitioning of the truncus is almost complete (Patten,
personal communication). Irrespective of the size of the pulmonary trunk, the right ventricle has apparently ample opportunity at this time to expel blood without the use of the normal Thebesian connections (Zanchi and Locatelli, 1958); (ii) the majority of aortic or pulmonary atresias are not associated with coronary anomalies; (iii) on the other hand, there are many cases on record of coronary fistulae with normal ventricular outflow tracts (Steinberg et al., 1958); (iv) in our present case there is an associated ventricular septal defect which, considered as an outlet to the right ventricular ejection, excludes the possibility that the coronary fistula is initially the consequence of mechanical outflow obstruction due to the pulmonary atresia. The same situation applies to the specimens described by Alexander and Green (1952) and Michel and Herbst (1957), which also had a ventricular septal defect with pulmonary atresia and coronary fistula.

Of main interest is the nature of the shunts established through coronary fistulae to the right ventricle. The pressure gradient between the connected chambers is responsible for significant left-to-right shunts after birth in patients in which the fistula is the only malformation (Brooks, 1885; Edwards et al., 1958). Surgical interruption of the shunt has been successfully accomplished in several such patients (Case et al., 1958). The circumstances of the shunt are different if the coronary fistula is associated with pulmonary atresia (Williams et al., 1951). Probably blood flows from the coronary arteries into the right ventricle during diastole, due to the gradient established by perfusion pressure. In systole, blood from this source plus that passed through the competent atrio-ventricular valve, must be ejected through the only possible outlet—the coronary fistula. Therefore, in these hearts the fistula carries a bidirectional shunt, alternating with the phases of the cardiac cycle.

In cases such as ours, the presence of an added ventricular septal defect makes it difficult to estimate the relative amount and direction of shunts mediated by both possible outlets to the right ventricle. However, we believe that the data in this case suggest that the shunt through the coronary fistula was predominantly right-to-left, and of more functional significance than the shunt through the septal defect. The degree of opacity of the right coronary artery can be interpreted as due to a large right-to-left shunt through it, since its opacity was at least equal to that of the aorta and greater than the opacity of the aortic branches. In our patient both ventricles probably retained throughout life the pressure relationships that existed before birth, because in the presence of pulmonary atresia the mechanisms that usually lower the right ventricular pressure are not operative. Furthermore, the size of the descending coronary arteries serving the fistula indicates, by comparison with the left circumflex of normal diameter, that the fistula functioned significantly in utero.

The function of the ventricular septal defect is not as clear. One would expect that, if it allowed a sizable shunt, the right ventricle would have developed normally. The final anatomical arrangement would then be that of a tetralogy of Fallot with pulmonary atresia and coronary fistula, a complex quite different from that existing in our specimen. The main difference appears to be the position of the septal defect in relation to the infundibular structures of the right ventricle (Becu et al., 1956). In the present case the defect is located posterior to the papillary muscle of the conus, and in the tetralogy of Fallot and its variants the defect is located characteristically anterior to a displaced papillary muscle of the conus, between it and the rotated crista supraventricularis. Assuming that ventricular pressures are essentially equal during fætal life, the amount of blood shunted through a ventricular defect will vary in relation to the position of the defect relative to the direction of linear flow.

For this reason a defect posterior to the papillary muscle of the conus with no overriding of the aorta may shunt less blood during fætal life than a defect of the same diameter but located anterior to the papillary muscle of the conus, that is, across the right ventricular outflow tract. A predominant shunt through the defect was not demonstrated in our patient, and the inference on anatomical grounds is that its importance was at least not greater than that of the shunt through the coronary fistula.

**Summary**

A case is described in which dilated and tortuous coronary arteries arose normally from the
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aorta giving descending branches that joined to form an angiomatous mass communicating freely with the right ventricle. This anomaly was associated with atresia of the pulmonary trunk, patency of the ductus arteriosus, enlargement with hypertrophy of the right atrium, and a hypoplastic tricuspid orifice. The significance of the anomaly is discussed.

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