Diagnosis of Aortic Atresia by Retrograde Aortography

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VENOUS angiocardiographic demonstration of a single arterial vessel leaving the heart and supplying both systemic and pulmonary circulations, suggests the diagnosis of persistent truncus arteriosus. Such was the initial impression in the case of a dyspneic, cyanotic, newborn male infant without a cardiac murmur. The electrocardiogram showed a normal sinus rhythm. The mean manifest electrical axis of the QRS complex was plus 110°. Lead V1 showed a qR pattern and flattened T waves. In lead V6 an Rs complex with an R wave of 17 mm and an S wave of 9 mm were present.

Since a murmur is commonly heard in persistent truncus arteriosus, absence of such a sign in this case led to this diagnosis being questioned. Accordingly, following the venous angiocardiography, a retrograde aortogram was performed. Demonstration by the latter study of a hypoplastic ascending aorta excluded the diagnosis of persistent truncus arteriosus and was strongly supportive for atresia of the aortic valve. After death, which occurred when the infant was 7 weeks old, the necropsy revealed this condition. It is of further interest that the patient was male and that death occurred in the neonatal period, both being usual features in this anomaly.

The case demonstrates the point that with venous angiocardiography demonstration of a large, seemingly single artery leaving the heart is not necessarily diagnostic of persistent truncus arteriosus since, as demonstrated by retrograde aortography in this case, a second vessel may be present which excludes this diagnosis. At the same time demonstration that the ascending aorta is hypoplastic while the pulmonary trunk is wide is considered strong evidence for aortic atresia.

Addendum
Since the preparation of this manuscript it was brought to our attention that a similar observation was experienced by Elliott, L. P. and associates: Aortic atresia. A case report and a review. Am. Heart J. In press.

Figure 2
Retrograde aortogram. Wide descending aorta and a hypoplastic ascending aorta (AA). At the base of the hypoplastic ascending aorta are seen the origins of the two coronary arteries (R.C.; L.C.). Demonstration of two great arteries excludes the diagnosis of persistent truncus arteriosus and the presence of a wide pulmonary trunk and a hypoplastic aorta is considered to be strong evidence for aortic valvular atresia.
Figure 1
Lateral perspective of the venous angiogram. Opaque material enters the right atrium from the inferior vena cava and is carried through the right atrium and ventricle and into a large arterial vessel. Arising from the latter trunk are the two pulmonary arteries and, beyond, the descending aorta is filled. Initially, the large arterial vessel was considered possibly a persistent truncus arteriosus. This vessel is now known to be the pulmonary trunk.

Figure 2

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Diagrammatic representation of the heart and great vessels in the case presented. Atresia at the level of the aortic valve above which arise the two coronary arteries. The pulmonary trunk (P.A.) is wide and branches into the two pulmonary arteries. A patent ductus arteriosus carries blood into the aorta and is the only channel for supply of the aorta. In the arch and in the ascending portions the blood flows in retrograde direction. The ascending aorta is hypoplastic and functionally is essentially a common coronary artery. R.C. = Right coronary artery. A.D. and L.C. = Anterior descending and left circumflex coronary arteries respectively arising from left coronary artery. R.A. = Right atrial cavity. R.V. = Right ventricular cavity. P.V. = Pulmonary vein. Illustration modified from Edwards and associates "An Atlas of Congenital Anomalies of the Heart and Great Vessels." Springfield, Illinois, Charles C Thomas, 1954, 202 pp.

The specimen from the case of which the angiocardiogram and retrograde aortogram are illustrated. The wall of the right ventricle (R.V.) is hypertrophied. The cavity is dilated. Although a left ventricular cavity was present (not illustrated in this view) it was nonfunctional because of the aortic atresia. Functionally, the right ventricle represents a common ventricular chamber. The pulmonary trunk (P.T.) gives rise to the left and right pulmonary arteries (L.P.; R.P.). By way of the patent ductus arteriosus (D.) the aorta is supplied from the right ventricle. The ascending aorta (A.A.) is hypoplastic and represents the narrow arterial vessel identified in the retrograde aortogram from which a diagnosis of atresia of the aortic valve could be supported.
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