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Persistent Left Fifth Aortic Arch Associated with Tetralogy of Fallot

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Abstract. A case of persistent left fifth aortic arch, forming a congenital ''double-lumen'' aortic arch, has been diagnosed on angiocardiography during life. It appeared as an unusual vascular structure running inferiorly and parallel to the ''real'' aortic arch from the innominate artery to the left subclavian artery superior to the pulmonary artery. This anomaly was found in the setting of tetralogy of Fallot, an association never described before, with patent ductus arteriosus (previously reported in most cases). The left aortic arch in this case was not a source of pulmonary circulation, as described in previous cases with pulmonary atresia and ventricular septal defect, but was a systemic-to-systemic connection without functional relevance.

Key words: Aortic arch — Embryology — Congenital heart malformation — Tetralogy of Fallot

Persistence of the embryonal fifth aortic arch is an unusual finding in humans. In the past the existence of this anatomic feature has been the object of research; in 1919 Huntington [9] described the presence of the fifth arch in the cat embryo; and in 1922 Buell [2] found a similar structure in the chick embryo. That same year Congdon [3] reported that endothelial sprouts arising from the aortic sac ventrally and from the descending thoracic aorta correspond to the upper and lower ends of the fifth arch in humans. Despite these findings the existence of the fifth arterial arch in mammals was denied, or if it did occur it was believed to be present only transiently [1].

Gerlis et al. [7] pointed out that two anatomic situations may be found depending on whether the persistent fifth aortic arch forms a systemic-to-systemic or a systemic-to-pulmonary connection. We describe a systemic-to-systemic connection due to a persistent fifth aortic arch in an unusual association with tetralogy of Fallot. The diagnosis was obtained by angiocardiography during life.

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Case Report

A 10-month-old female infant was admitted for a corrective operation of tetralogy of Fallot. She was born at 38 weeks' gestation weighing 2470 g. A cesarean section had been performed because of fetal distress. Tetralogy of Fallot was diagnosed within a few days of birth during a cardiologic evaluation for a systolic murmur and mild cyanosis. Thereafter the baby had normal growth, with persistent mild cyanosis and without anoxic spells.

At the time of admission she appeared in good general condition and was well grown (weight 9230 g, height 76 cm). Transcutaneous oxygen saturation was 85%. Peripheral arterial pulses were palpable and symmetric. The heart rate was regular, at 110/min; on auscultation a single second heart sound was heard together with a 3/6 systolic ejection murmur along the left sternal border.

On echocardiography, the following features were noted: situs solitus, levocardia, normal venoatrial connections, and atrioventricular and ventriculoarterial concordance. A large subaortic interventricular septal defect with anterocephalad displacement of the infundibular septum and an overriding aortic valve was also seen. An infundibular and valvar pulmonary stenosis with a peak instantaneous pressure gradient of about 75 mm Hg was revealed by a Doppler study. The pulmonary trunk and main branches were of normal size. A left-sided aortic arch, patent ductus arteriosus, and normal anatomy of the proximal portion of the coronary arteries were demonstrated (Fig. 1).

Cardiac catheterization confirmed the presence of structural and functional features inherent in tetralogy of Fallot with normal anatomy of the coronary arteries and normal size of the pulmonary arteries. At angiography the aortic arch appeared left-sided with normal branching of the branchiocephalic arteries and evidence of patent ductus arteriosus in the normal position. A separate channel, arising from the aorta just beyond the origin of the innominate artery, ran in parallel under the aortic arch, rejoining the aorta at the level of the left subclavian artery (Figs. 2, 3).

The baby underwent successful surgical correction of the tetralogy of Fallot. Direct inspection of the aortic arch revealed a single large structure without clear evidence of two separate vessels. The postoperative period was uneventful.

Discussion

Persistence of the fifth aortic arch in humans, although rare, is now considered possible, with an incidence of almost 1 in 330 autopsy studies [7]. It is likely, however,

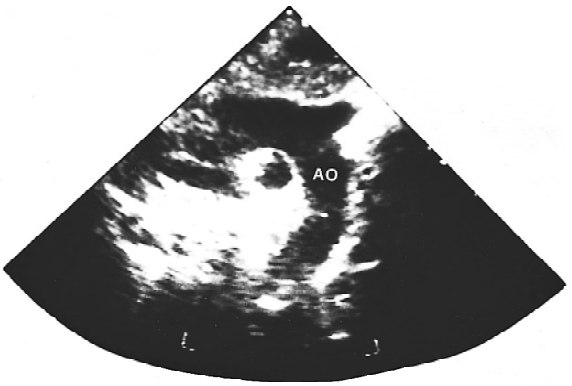

Fig. 1. Echocardiographic frame showing an apparently normal aortic arch. *AO,* aorta

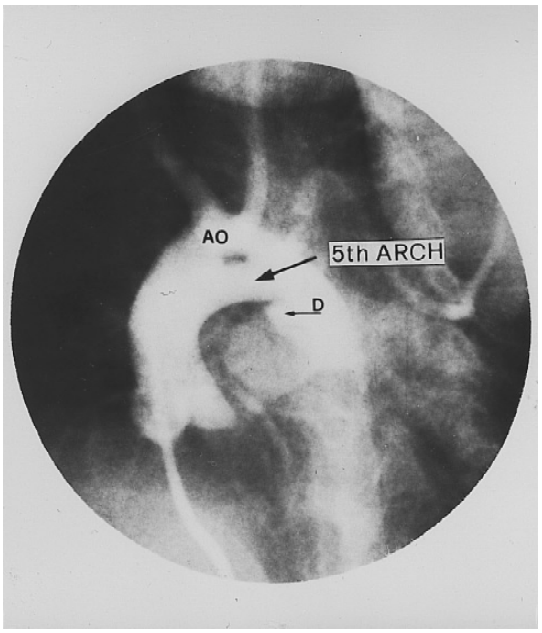

Fig. 2. Aortogram, long-axial view. There is evidence of two parallel horizontal channels forming the aortic arch. From the upper channel (*Ao*), the ''real'' aortic arch originates from the fourth embryonal arterial arch, and arteries arise as usual (from left to right: innominate, left common carotid, and left subclavian). The lower channel (*5th ARCH*) is the persistent left fifth arterial arch. A patent ductus arteriosus (*D*) is present, with evidence of main pulmonary branches

that persistent fifth aortic arches go unrecognized, rather than being rare. Van Praagh and Van Praagh [11] described the first autopsy finding of this congenital anomaly in association with tricuspid atresia, cor triatriatum, infundibular and valvular pulmonary stenosis, and patent ductus arteriosus (PDA). Izukawa et al. [10] described two other autopsy findings of persistent left fifth aortic arch; for the first time, two well separated, parallel, complete aortic arches of almost equal size were ob-

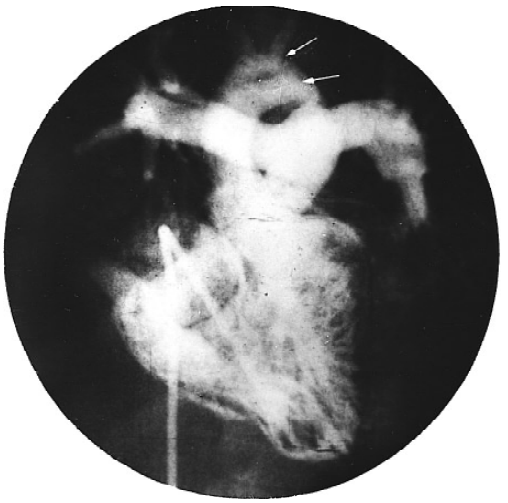

Fig. 3. Right ventriculogram, right oblique view. Right ventricle is dilated and hypertrophic. Pulmonary trunk and branches are of regular size. The contrast medium makes the overriding aorta evident. The wide subaortic septal defect and the valvular and infundibular pulmonary stenosis is not well shown here. There is evidence of a double-lumen aortic arch (*white arrows*) and patent ductus arteriosus

served anterior to the trachea. The lower vessel, extending from the level of the innominate artery to the level of the subclavian artery, was then considered to be the persistent left fifth aortic arch.

Currently, it is accepted that this anomaly presents in two forms [7]. With the first type, the fifth aortic arch connects the ascending and descending aorta, running underneath the (fourth) aortic arch from the innominate artery to the left subclavian artery and giving origin to a congenital double-lumen aortic arch. This systemic-to-systemic connection has no functional relevance and may be associated with various congenital cardiovascular anomalies, the most frequent being aortic coarctation, PDA, and bicuspid aortic valve [4, 10]. With the second type, the fifth aortic arch connects the ascending aorta and a derivative of the embryonic sixth arch, forming a systemic-to-pulmonary connection. This situation has usually been described in association with pulmonary atresia and VSD. Here the persistent fifth aortic arch is not only an anatomic finding but has an important functional role as a source of blood supply to the lung and may be misdiagnosed as the arterial duct. Only one case of pulmonary-to-systemic connection in which the fifth aortic arch was a source of systemic flow has been described in the setting of aortic atresia with interruption of the aortic arch [7]. No case of pulmonary atresia and VSD with ventriculoarterial concordance has been reported in association with a systemic-to-systemic connection; only two cases have been described in association with ventriculoarterial discordance [6, 8].

To our knowledge, our case is the first report of tetralogy of Fallot associated with a persistent fifth aortic

arch not being a source of pulmonary blood supply but a systemic-to-systemic connection. If surgical correction had been carried out based only on the echocardiographic findings, the persistent fifth aortic arch would have remained undiagnosed because neither echocardiography nor surgical inspection were diagnostic; in fact, it had no hemodynamic significance and would not have mattered.

In agreement with Gerlis et al. [7], we believe that the real incidence of this anomaly is underestimated, particularly when a systemic-to-systemic connection without functional relevance is associated with relatively simple congenital anomalies. In these cases surgical correction is more and more widely carried out based on echocardiographic evidence alone without angiography. Currently, a reliable diagnosis during life has been possible only by selective angiography of the aortic arch [5]. Magnetic resonance imaging may be a valid noninvasive alternative to angiography to delineate previously identified cases and discover new, unexpected ones, but to our knowledge no such study has been reported.

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