Anomalous Fusion of Great Vessels of Heart – A Rare and Morbid Cardiac Congenital Malformation

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ABSTRACT

From a developmental point of view, the abnormal connections between great arteries might be related to a deficient lengthening of the outflow tract during development. They are an important causal factor for cyanotic heart disease. This paper presents a report of a rare arterio-venous cardiac malformation case in which the ascending aorta was fused with the trunk of the pulmonary artery. Even though the perfusion related morbidity is undeniable in this case, the exact cause leading to mortality remains uniquely dubious. Exactly how the subject reported here escaped diagnoses and managed to survive until late adulthood remains a biological wonder. Such variations hold implications for cardiothoracic surgeons and clinicians dealing with cyanotic and circulatory disorders.

Keywords: Cyanotic Heart Disease, Aorta, Pulmonary Trunk, Arteriovenous Anastomoses, Cardiac Anomaly.

CASE REPORT:

This case reports a 65 year old female who upon routine dissection in the department dissection hall revealed a cardiac congenital malformation, seen as a rare variant of a common arterial trunk, in which the ascending aorta was in connection with the trunk of the pulmonary artery (Figure 1, 2 3). The ascending aorta showed hypoplasia, while the coronary arteries were free of any pathological findings. The atrial septum showed a closed foramen ovale and the ventricular septum did not show any defect. Only an isolated mild right ventricular hypertrophy and dilation with no other cardiac abnormalities was found. The left ventricle showed normal muscle mass and volume when compared with another healthy cadaver of comparable age, body mass and gender. The finger nails showed evidence of clubbing indicative of perfusion defects.

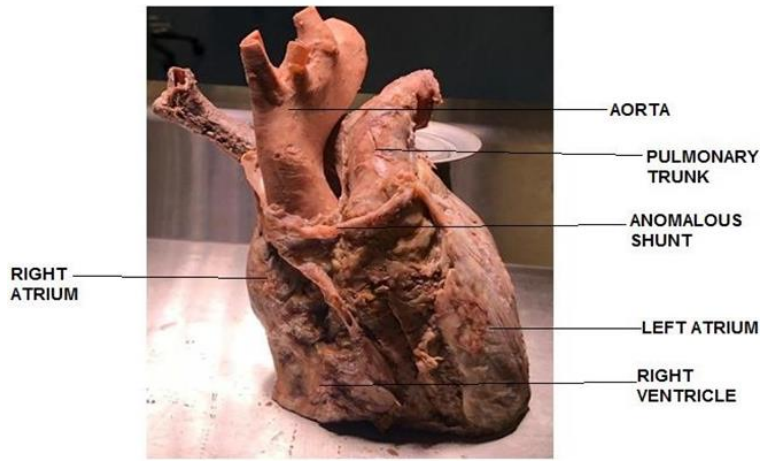

Figure 1: Anomalous aorto-arterial shunt between aorta and pulmonary trunk in spatial relation to other cardiac structures.

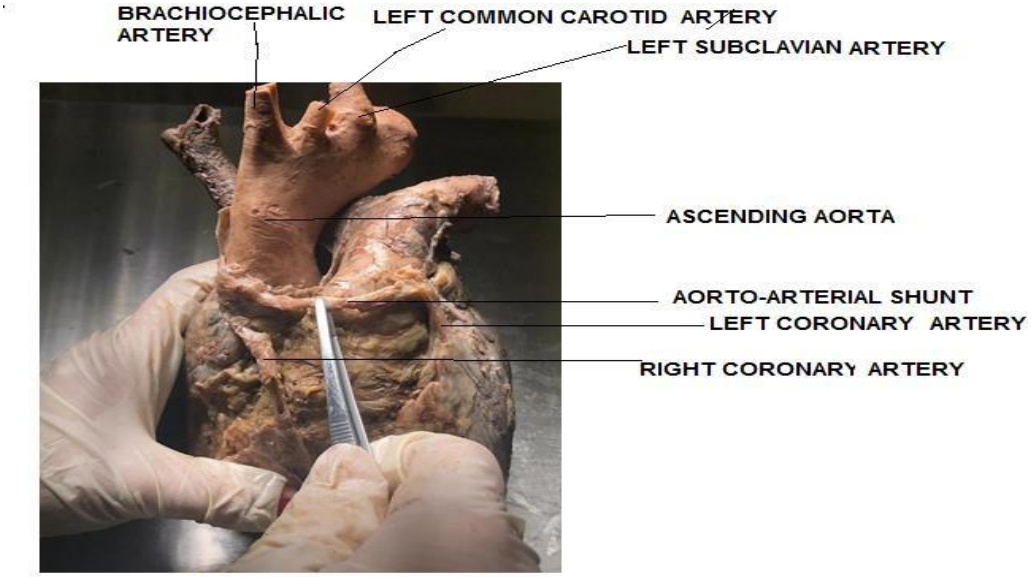

Figure 2: Aorto-arterial shunt with coronary vessels and great vessels

Figure 3: Base of aorta and base of pulmonary artery zoomed view to show origin and termination of aorto-arterial shunt (held with forceps).

Discussion:

The ductus arteriosus is the connecting vessel between the pulmonary trunk and the descending aorta during fetal life and it serves to bypass the immature pulmonary circulation (1). The connection closes after birth and can be seen as a fibrosed ligamentum arteriosum during adult life

(2). Developmentally the root and the proximal portion of the aorta and pulmary artery are from the truncus arteriosus, the distal portion of bulbus (Fig.4,5). By 5th week two opposing ridges, the right superior truncus swelling and the left inferior truncus swelling appear within the truncus, grow into a spiral manner, fuse to form the aorticopulmary septum (3) thus the truncus will be divided into aortic and pulmonary channels.

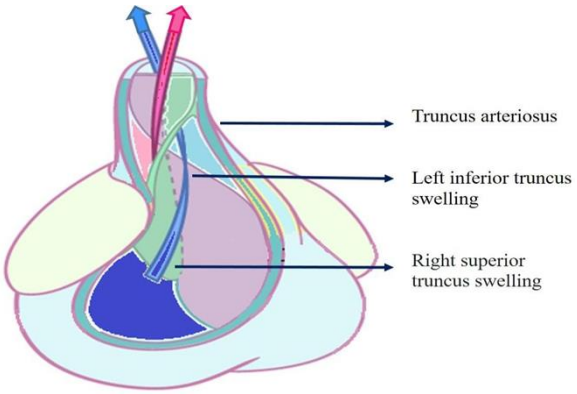

Figure 4: diagram showing the formation of septum within the truncus arteriosus.

Figure 5: Series of diagrams showing the possible formation of an anamolous connection between the developing aorta and pulmonary trunk.

Pulmonary and systemic circulation must be ideally separate in adulthood. Survival is dependent upon separate, parallel circulations (4). In the above discussed case it can be concluded that during separation the aorta and pulmonary trunk was not got separated entirely but an anamolous connection might have existed. Bidirectional shunting resulting from such anamolous conditions as described in this report can lead to severe systemic acidosis and hypoxia (5). The subject reported here might most likely have suffered from chronic fatigue syndrome due to mixing of oxygenated and deoxygenated blood (6). There was evidence of clubbing to support this speculation (7). The fetal circulation like state persisted in this individual though ductus arteriosus was not patent but fibrosed. It is also possible that an additional horizontally oriented prominent blood vessel had developed as an analogue of the umblical vein during fetal life, or it might be some errant branch of a latent aortic arch system of embryonic origins. The exact cause still remains unknown. The mild right ventricular hypertrophy indicates that with every systole, blood was flowing from aorta back to the pulmonary artery and from there to the right ventricle. This created a negative suction mechanism leading to an overworked right ventricle. The pulmonary valves appeared normal but we speculate that they might have been functionally deficient to augment this backflow cycle that finally led to increased muscle mass of the ventricle.

Though there are reports of right pulmonary artery coming out from aorta (8,9), such bridging branches between aorta proper and pulmonary trunk are unreported, to the best of authors knowledge (10).

From a developmental point of view, the abnormal connections between great arteries might be related to a deficient lengthening of the outflow tract during development, for which contributions from the so-called second heart field are necessary. They are an important causal factor for cyanotic heart disease and more exploration into gender preponderance is needed.

Conclusion:

These findings underline the value of 3-dimensional/4-dimensional ultrasound imaging when added to a cardiology screening program, and the need for improvements in screening routines by using pulse oximetry in order to discover isolated vascular defects before circulatory collapse occurs, as well as to reduce the medico-legal disputes in cases of missed diagnosis. We found the relevant literature search lacked a description of this congenital malformation, which supports our deeper perinatal investigation. Surgical treatment is the arterial switch procedure which is usually performed in childhood. Even though the morbidity is undeniable in this case, the mortality remains uniquely and surprisingly questionable. Exactly how the subject reported here escaped diagnoses and managed to survive until late adulthood remains a biological wonder. Such variations hold implications for cardiothoracic

surgeons and clinicians dealing with cyanotic and circulatory disorders.

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