

Discordant atrioventricular connections with truncus arteriosus

Discordancia atrioventricular con tronco arterioso común

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Atrioventricular discordance occurs when the atria are connected to inappropriate ventricles. Indeed, the morphological right atrium is connected to the morphological left ventricle through the mitral valve, and the morphological left atrium is connected to the morphological right ventricle through the tricuspid valve. The purpose of this manuscript is to report a case of an anatomical specimen of atrioventricular discordance with a single outlet through the common truncus arteriosus from the collection of the department of embryology.

This specimen was studied with the morphological method of the segmental sequential system¹, and an anatomical-embryological correlation was made to explain this uncommon combination.

The anatomical specimen showed situs solitus; the ventricles were inverted: the morphological right ventricle was on the left and in an anterior position, while the morphologically left was right and in a posterior position. The right atrium was connected to a morphological left ventricle; both located on the right through a mitral valve. The left atrium was connected to a morphological right ventricle; both located on the left through a tricuspid valve (Fig. 1).

Both ventricles were externally delimited by the anterior descending artery. A single vessel emerged from the morphological right ventricle above an infundibulum that ran from front to back and from left to right; this vessel was divided into a left-sided aortic trunk and a

right-sided pulmonary trunk (Fig. 2). The truncal valve was tricuspid with thickened leaflets and without stenosis. In addition, there was a permeable foramen ovale.

In atrioventricular discordance, the ventricles are inverted both in situs solitus and situs inversus. Therefore, the spatial orientation of the atrioventricular orifices (the anterior mitral and the posterior tricuspid) is inverted. The atrioventricular valves are at the same level. In situs solitus, the morphological left ventricular outflow tract is located between the mitral and tricuspid valves with fibrous continuity. This feature produces a misalignment of the atrial and ventricular septa with the consequent formation of a prominent anterior recess in the morphological left ventricle. The incorrect alignment of the septa causes ventricular septal defect and infundibular pulmonary stenosis. Furthermore, it gives rise to a more extensive membranous septum. The anatomy of the outflow tract depends directly on the type of ventriculoarterial connection².

In this pathology, we can find all types of ventriculoarterial connection; however, the most frequent is the discordant connection, traditionally known as congenitally corrected transposition. Less frequent connections are the double outlet of the morphological right ventricle and the single outlet through the truncus arteriosus or the solitary aortic trunk due to pulmonary atresia. The presentation with concordance of ventriculoarterial connection and double outlet of the morphological left ventricle is uncommon².

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Figure 1. Internal view of the cardiac chambers located on the left showing the atrioventricular discordance. AI: left atrium; VD: right ventricle; VT: tricuspid valve.

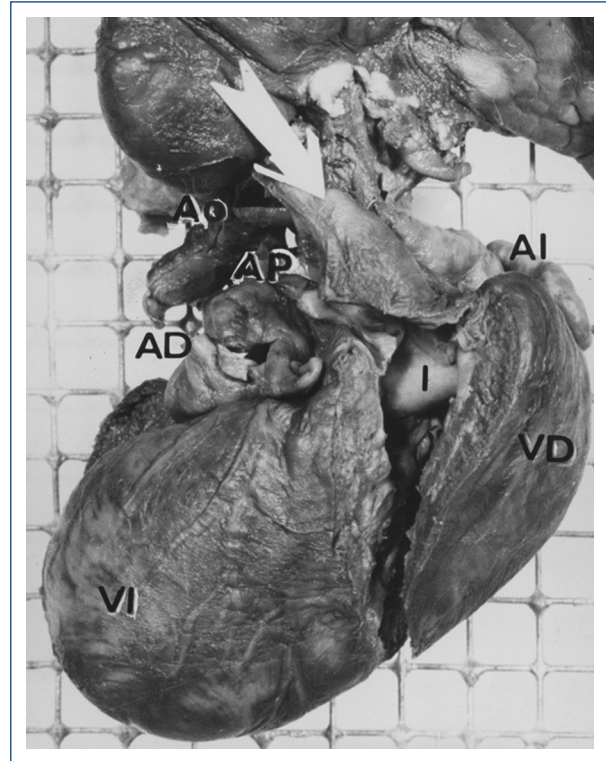


Figure 2. External frontal view of the heart with atrioventricular discordance and truncus arteriosus (white arrow). I: infundibulum; Ao: aorta; AP: pulmonary artery; AI: left atrium; AD: right atrium; VI: left ventricle; VD: right ventricle.

The conduction system is modified due to misalignment between the atrial and ventricular septa. In consequence, neither the node nor the His bundle makes contact with the ventricular conduction tissue or the trabeculated septum. Hence, in the corrected transposition, an atrioventricular branch is formed anterior to the pulmonary outflow tract, originating from an abnormal anterior atrioventricular node. In the presence of ventricular septal defect, the bundle descends in front of the septal defect. Moreover, the bundle descends anterior and lateral to the pulmonary outflow tract, directly related to the merging of the pulmonary valves to the interventricular septum².

In more than half of the cases, the most frequently associated lesion is ventricular septal defect (usually perimembranous). Pulmonary stenosis is present in just under half of the cases and can be valvular or subvalvular. Malformations of the atrioventricular valves, of which the most frequent is the Ebstein-type anomaly of the tricuspid valve, are found in up to 75% of cases³. Other associated lesions are atrial septal defect, mitral

cleft, right ventricular hypoplasia, mitral stenosis, pulmonary atresia, and ductus arteriosus.

De la Cruz et al.⁴ proposed an embryological explanation for atrioventricular discordance, in which they anticipated the existence of truncus arteriosus as a form of ventriculoarterial connection within this heart disease. Atrioventricular discordance is caused by the reversed torsion of the heart tube at the time of bulboventricular loop development. In situs solitus, the normal loop is twisted to the right (D-loop) resulting in concordant atrioventricular connections. On the other hand, when the torsion occurs to the left (L-loop), atrioventricular discordance occurs. Truncus arteriosus originates from the lack of development of the trunk septum that separates the aortic and pulmonary canals from the arterial trunk of the embryonic heart.

In the literature, there are some reports of cases of atrioventricular discordance with truncus arteriosus diagnosed by clinical data but without pathological specimen documentation⁵. This report is so far the third case corroborated by necropsy in the published research. In

the only two previous cases, the morphological description of the piece was not detailed, the segmental sequential system was not used as a reference, and there was no embryological correlation^{6,7}. In the first case reported in the literature, the specimen had a type II truncus arteriosus associated with ventricular septal defect. In the second report, the trunk valve was bivalve with a small ventricular septal defect. In our case, truncus arteriosus was type I associated with foramen ovale.

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Conflicts of interest

The authors declare that they have no conflicts of interest.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

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