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Case Report

Tetracuspid Right Atrioventricular Valve: A Case Report

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ABSTRACT

The right atrioventricular valve (tricuspid valve) allows blood to flow from the right atrium down into the right ventricle. Tricuspid valve disease is rare, but includes tricuspid valve regurgitation, stenosis, and Ebstein anomaly. During regular dissections for undergraduate medical students, in the department of Anatomy, Kasturba Medical College, Manipal, we observed the following variations in the heart in an adult male cadaver. The right atrioventricular valve showed four cusps instead of usual three cusps, anterior, posterior, septal. The extra cusp was anterolateral in position. This variation is important for the cardiothoracic surgeon.

Key words: Tricuspid valve, conus arteriosus, chordae tendineae, septal cusps, papillary muscle

INTRODUCTION

The right atrioventricular valve (tricuspid valve) allows blood to flow from the right atrium down into the right ventricle. Tricuspid valve disease is rare, but includes tricuspid valve regurgitation, stenosis, and Ebstein anomaly. The right AV valve has three, roughly triangular shaped, cusps that project into the ventricle: the anterior (superior), posterior (inferior) and septal. The anterior is the largest cusp interposed between the AV orifice and the conus arteriosus. The posterior is connected to the right margin of the ventricle and the septal cusp to the ventricular septum. The bases of the cusps are attached to a fibrous ring at the AV orifice, where they are continuous with one another. The ventricular surfaces of the cusps are divided into three

zones: the distal rough zone, the basal zone and the proximal, thin and translucent clear zone. (1) The rough zone serves as points of insertion for the chordae tendineae, which arise from the apices of conical muscular projections of the ventricle wall, called papillary muscle. (2)

CASE REPORT

During regular dissections for undergraduate medical students, in the department of Anatomy, Kasturba Medical College. Manipal. we observed following variations in the heart in an adult male cadaver. The right atrioventricular valve showed four cusps instead of usual three cusps, anterior, posterior, septal. The extra cusp was anterolateral in position as shown in figure 1.



Fig 1. Showing the Tetracuspid right AV valve from the atrial side. 1- Septal cusp, 2- Anterior cusp, 3- Anterolateral cusp, 4- Posterior cusp, FO- Fossa ovalis, CO- Coronary ostium

DISCUSSION

Beginning in the 5th week of gestation, the AV valve develops from the excavation of the supporting ventricular myocardium. Cavitation of the ventricular walls forms a sponge work of muscular bundles. Some of these remain as the trabeculae carneae and others become the papillary muscles and chordae tendineae which run from the papillary muscles to the AV valve. By the 8th week, the heart presents a complete AV valvular structure.

Previous studies suggest that the number of leaflets may vary, or that accessory leaflets may be found between the leaflets. Morphological main morphometrical criteria to distinguish between supernumerary and commissural cusps have not been achieved. Commissural cusps are small accessory cusps occurring at the site of junction between adjacent cusps, i.e. at the site of the congenital fusion of the original commissures that do not reach the fibrous ring of the valve. (6)

In a post mortem study, the right AV valve was not consistently tricuspid, but was observed to present with 2, 4, 5 or 6 cusps in 72% of cases. In valves with 2 cusps, the septal cusp is larger than the anterior one. In valves with 3 cusps, the posterior cusp is

associated with a less prominent septal cusp. When the valve comprises 4 cusps, the anterior and posterior ones become less prominent and an anterolateral cusp emerges as observed in the present case report. (5)

A study by Gerola et al on AV valve in children showed that the commonest finding was 3 cusps, while a fourth cusp, if present, was classified as anterolateral in location. The number of tendinous cords was greater for the anterior and septal cusps than for the posterior and anterolateral cusps. (7)

According to Lang et al another morphologic feature which causes malformation of the tricuspid valve is Ebstein's anomaly. In this rare and complex disorder, the primary lesion is downward displacement of the basal attachment of the posterior and septal leaflets. (8)

Ebstein's malformation is. essence, a manifestation of abnormal development of both the myocardium and the valve components. Ebstein's anomaly is characterized by adhesion of the septal and posterior tricuspid valve leaflets to the underlying myocardium due to failure of delamination. This leads to displacement of the annulus of the tricuspid valve. The anterior leaflet may be severely deformed and may form a large 'sail-like' intracavitary curtain, which can even lead to RV outflow tract obstruction. (9)

According to Yoon et al only a few cases are asymptomatic. In most cases, accessory valve tissue is associated with other cardiac malformations. Chordal replacement or augmentation with expanded polytetrafluoroethylene suture is a useful technique in the repair of congenitally dysplastic tricuspid valves with abnormal chordal structures. Hence this variation is important for the cardiothoracic surgeon. (10)

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