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Transportation of the Heart Valves-A Case Report of Rare Anatomical Anomaly

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ABSTRACT

Introduction: *The atrioventricular valve is present between the right atrium and right ventricle and the mitral valve between the left atrium and left ventricle. Papillary muscles are anchored to the cusps of the AV valve via the chordae tendineae and they contract to prevent prolapse of the valve. Observation: Herein, we present a case with reversal of valves where the mitral annulus had 3 instead of the usual 2 cusps and vice versa on the right side. The left ventricle was dissected as per Cunningham's dissector, the 3 cusps were attached via. chordae tendinae to 3 papillary muscles whereas the right ventricle was seen to have 2 papillary muscles.*

Keywords: Reversal, Mitral valves, Anomaly, Tricuspid valve

INTRODUCTION

Due to an increase in patient load requiring valve replacement surgeries, it is of paramount importance to understand the anatomy of the valvular complex. The tricuspid atrioventricular valve is present between the right atrium and right ventricle whereas the mitral valve or the bicuspid valve is present at the junction of the left atrium and left ventricle. Papillary muscles are placed in the ventricles attach to the cusps of the AV valve via the chordae tendineae and contract to prevent inversion or prolapse of the valve [1]. We present a rare case report of the variations of the heart valves. A brief anatomical basis and their developmental and embryological basis has been attempted to explain the phenomenon.

CASE REPORT

As a part of routine undergraduate dissection; in a cadaver of North Indian ethnicity, 70 years male, the heart was removed and emptied of blood inside, washed thoroughly in running tap water. As per Cunningham's dissector, the orientation and position of the heart were confirmed after which the left atrial wall was opened by an incision through the right and left inferior pulmonary veins and the mitral valve was inspected from above. The mitral annulus had 3 instead of the usual 2 cusps. The left ventricle was opened by an incision on the sternocostal surface of the heart extending from the apex, parallel and close to the interventricular septum up to the aortic orifice. The 3 cusps were attached via chordae tendinae to 3 papillary muscles. The left ventricle appeared normal (Figure 1).

Measurements were taken with the help of a divider, thread, and a millimeter scale. The Circumference of Mitral Annulus was taken by keeping a thread in the sulcal margin and the point is marked where it meets the beginning of the thread and this distance in the thread is measured as 10.2 cm. Using divider and millimeter-scale the height of each valve leaflet was measured by the perpendicular line from the annulus to the center of the free edge of the leaflet which was 2.7 cm, 3 cm, and 2.5 cm.

Similarly, the right atrium was opened by incising between the openings of superior and inferior vena cava, 2 cusps (Septal-larger and Anterior) were visualized rather than the usual 3. Further dissection of the right ventricle revealed the 2 cusps attached to 2 papillary muscles. The right ventricle appeared the usual. The circumference was 9.5 cm and the height of each valve leaflet was 2.5 cm and 3.1 cm (Figure 2 and Figure 3).

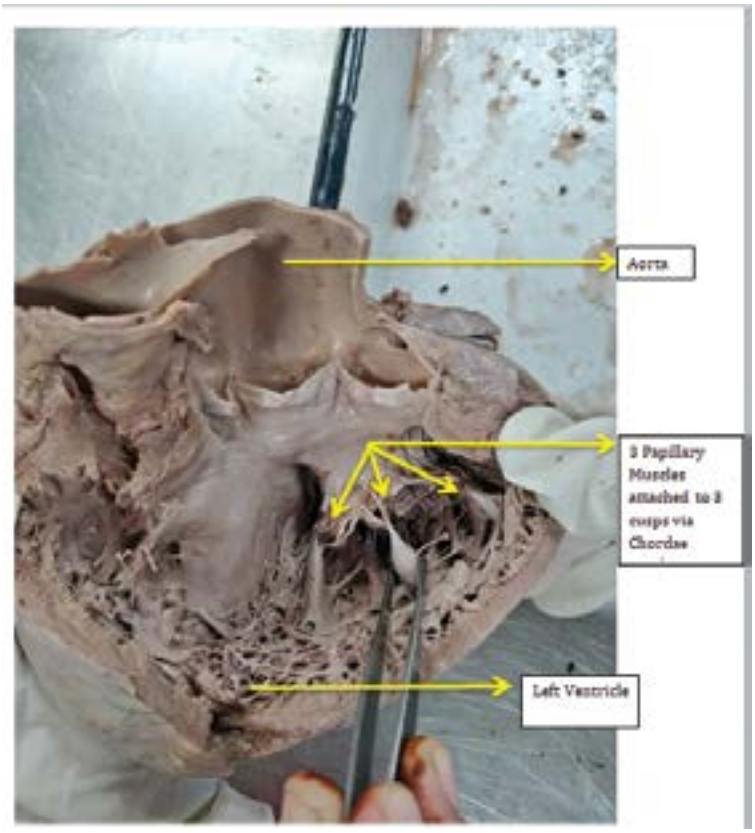

Figure 1 Left ventricle has been cut open showing 3 papillary muscles attached to the 3 cusps via. chordae tendenae

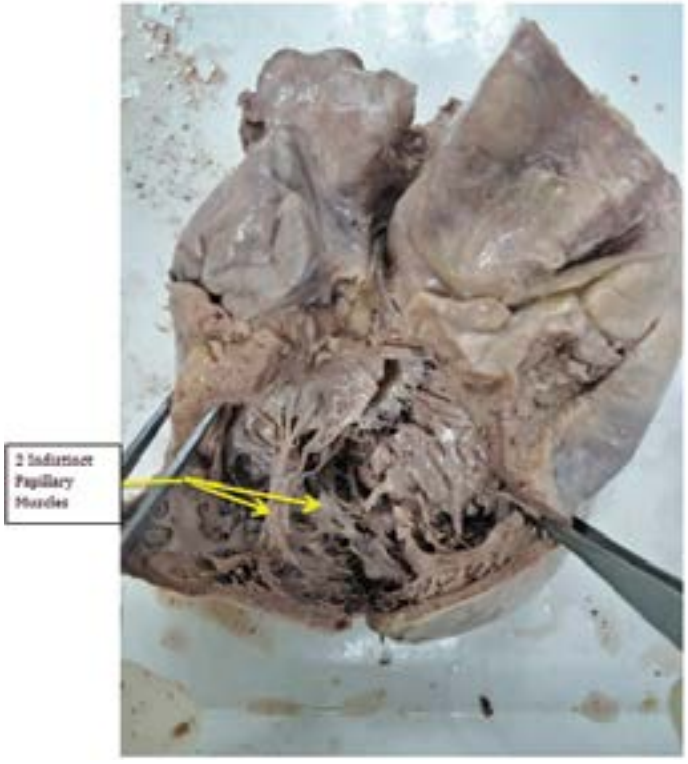

Figure 2 Right ventricle cut open showing 2 papillary muscles attached to 2 cusps via. chordae tendenae

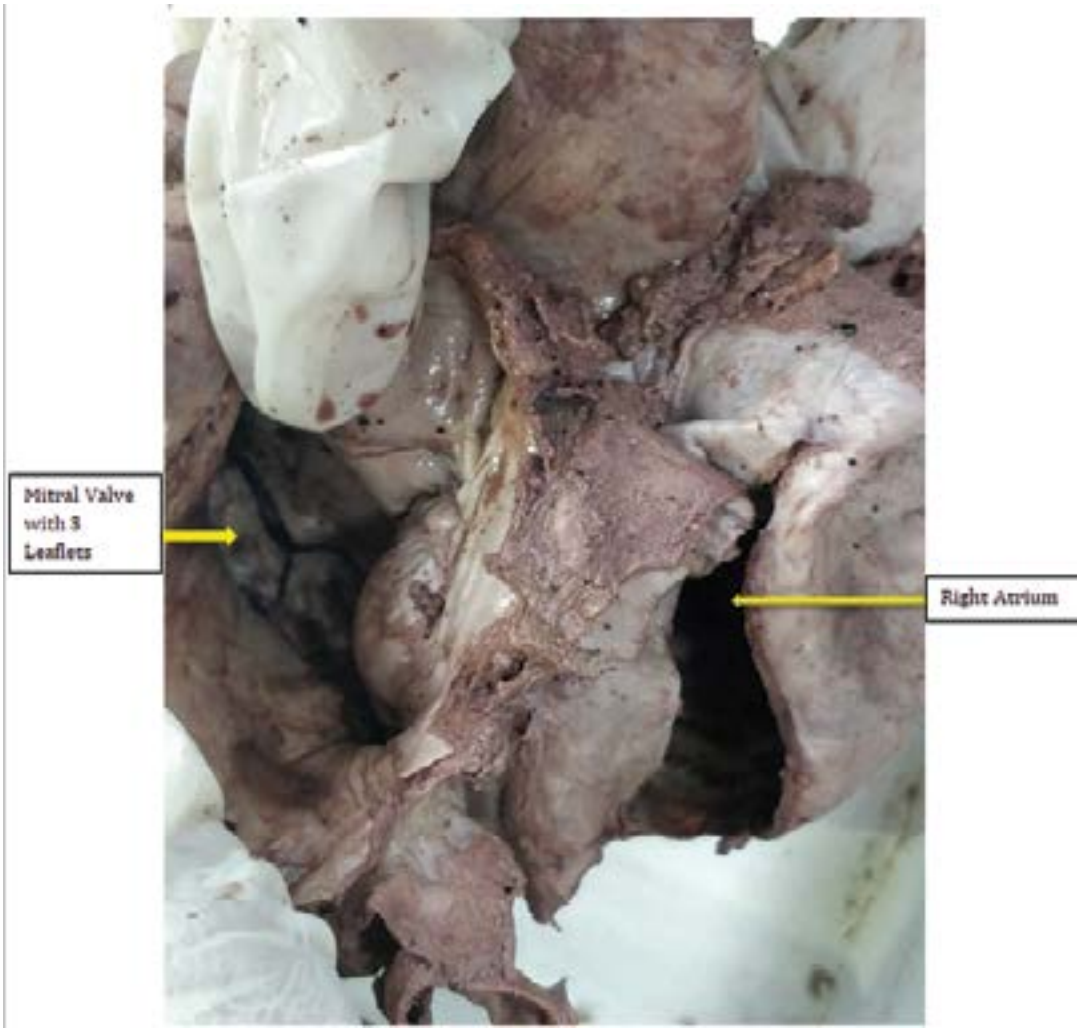

Figure 3 Right and left atrium cut open; Mitral valve with 3 leaflets can be seen

DISCUSSION

The heart is a fused pair of a valved muscular pumps circulating the blood in series through systemic and pulmonary circulations. The right atrium receives the superior and inferior venae cavae and the main myocardial venous inflow via the coronary sinus. This systemic venous blood traverses the right atrioventricular orifice, guarded by the tricuspid valve to enter the inlet component of the right ventricle.

The tricuspid valve leaflets are located anteroposteriorly, septally, and inferiorly, corresponding to the marginal sectors of the AV orifice named in co-junction. The inferior leaflet is described as being posterior but when assessed in the anatomical position, the leaflet is positioned inferiorly. A triangular zone of Koch is defined between the attachment of the septal leaflet of the tricuspid valve, the anteromedial margin of the ostium of the coronary sinus, and the palpable round, collagenous subendocardial tendon of Todaro. Koch's triangle is a landmark of particular surgical importance, indicating the site of the atrioventricular node and its atrial connections [1]. Each leaflet is a reduplication of endocardium enclosing a collagenous core, continuous marginally and on its ventricular aspect with diverging fascicles of chordae tendineae and basally confluent with the annular connective tissue. The anteroposterior leaflet is the largest component of the tricuspid valve attached chiefly to the AV junction on the postero-lateral aspect of the supraventricular crest and extending along its septal limb to the membranous septum ending at the anteroseptal commissure. The attachment of the septal leaflet passes from the inferoseptal commissure on the inferior ventricular wall across the muscular septum, then angling across the membranous septum to the anteroseptal commissure. The

septal leaflet defines one of the borders of the triangle of Koch, thereby aiding the location of the AV node at the apex of this triangle and ensuring avoidance during tricuspid valve surgery. The inferior leaflet is wholly mural in attachment and guards the diaphragmatic surface of the AV junction, its limits being the anteroseptal and anteroinferior commissures. Despite its name, the tricuspid valve acts more like a bicuspid valve because its smallest leaflet is fixed between the atrial and ventricular septa.

The left atrium receives all the pulmonary inflow of oxygenated blood and some coronary venous inflow. It contracts to fill the left ventricle through the left atrioventricular orifice guarded by its mitral valve. The mitral valve leaflets have long been described as paired structures but small accessory leaflets have almost always been found between the two major leaflets and so the mitral valve should be described as a continuous veil that is attached around the entire circumference of the mitral orifice. These anteromedial (inferoseptal) and posterolateral (super posterior) extremities may be regarded as two independent commissures. When the valve is laid open, the anterior leaflet (aortic, septal) is seen to guard one-third of the circumference of the orifice and to be semicircular or triangular with few or no indentations. Hinging on its annular attachment, and continue with its subaortic curtain, it is critically placed between the inlet and the outlet of the ventricle. The posterior leaflet (mural, ventricular) usually has two or more indentations. It has a wider attachment to the annulus than does the anterior leaflet, guarding two-thirds of the circumferential attachments.

Left ventricular contraction closes the mitral valve and opens the aortic valve, enabling the ventricle to eject via the left ventricular outflow tract into the aortic sinuses and the ascending aorta, and thence to the entire systemic arterial tree, including the coronary arteries.

Development of AV Valve

The formation of cardiac mesenchymal cells at the atrioventricular canal and the proximal myocardial outflow tract is followed by their migration into the cardiac extracellular matrix. These cells proliferate between the endocardium and myocardium and with local accumulation of extracellular matrix molecules, produce protrusions, termed endocardial or cardiac cushions, which bulge into the primary heart tube and initially provide the valvular mechanisms required in the atrioventricular canal and outflow tract. Their position corresponds to the future positions of the definitive cardiac valves. In the distal part of the outflow tract, which initially has myocardial walls, cells that are derived from the neural crest cells subsequently make significant contributions to the mesenchyme of the endocardial cushions. Although proper migration of the neural crest cells is crucial for normal development of the outflow tract and formation of the leaflets and sinuses of the arterial valves, their function is largely obscure. They are no longer found in leaflets of the arterial walls in the formed heart or in the muscular subpulmonary infundibulum, which is also derived from the outflow cushions. The endocardial cushions ultimately fuse with the myocardium and with local accumulation of extracellular matrix molecules, produce protrusions, termed endocardial or cardiac cushions, which bulge into the primary heart tube and initially provide the valvular mechanisms required in the atrioventricular canal and outflow tract. Their position corresponds to the future positions of the definitive cardiac valves. in the distal part of the outflow tract, which initially has myocardial walls, cells that are derived from the neural crest cells subsequently make significant contributions to the mesenchyme of the endocardial cushions. Although proper migration of the neural crest cells is crucial for normal development of the outflow tract and formation of the leaflets and sinuses of the arterial valves, their function is largely obscure. They are no longer found in leaflets of the arterial walls in the formed heart or in the muscular subpulmonary infundibulum, which is also derived from the outflow cushions. The endocardial cushions ultimately fuse. At the time of their fusion, the AV endocardial cushions are large, relative to the size of the AV orifice; they will provide the scaffold for the formation of the leaflets of the tricuspid and bicuspid valve [2].

Anatomic Variations of the AV Valve

Although the right AV valve has three leaflets, studies have suggested that the number of leaflets may vary, or that accessory leaflets may be found between the main leaflets; like the former, they consist of a fold of endocardium strengthened by fibrous tissue. Even though researchers have tried to establish morphological and morphometrical criteria to distinguish between supernumerary and commissural cusps, no consensus has been achieved [3]. Commissural cusps are small accessory cusps occurring at the site of the 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. 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; moreover, additional "commissural cusps" were found in 64% cases independent of the number of "supernumerary cusps" [3].

In valves with 2 cusps, the septal cusp is larger than the anterior one which is consistent with our case report. 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. In this case, the posterior cusp, besides a size reduction, undergoes medial dislocation assuming a more posterior position. In valves with 5 cusps, the posterior cusp does not change in size but undergoes lateral dislocation, assuming a position similar to that seen in valves with three cusps. In addition, the appearance of a posteromedial cusp leads to a reduction in the size of the septal one, while the anterior cusp undergoes little or no alteration. There is also some reduction in the size of the anterolateral cusp. When the valve consists of 6 cusps, the sixth one, named postero-lateral, is located between the anterolateral and posterior cusps. Size reduction is noted mainly in the septal and anterolateral cusps [4]. An anatomic study of the AV valve in children showed that the commonest finding was 3 cusps, whiles a fourth cusp, if present, and 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. In addition, the posterior region of the valve annulus in 35.7% of the cases was occupied by undeveloped valve tissue with the posterior valve located anteriorly in these cases [5]. An accessory, mobile or fixed, right AV valve leaflet not contributing to the commissure is a rare finding, most often reported in children with complex congenital cardiac malformations (Fallot's tetralogy, Ebstein's anomaly). The mobile type is a parachute-like leaflet floating freely in the RV. The fixed type is firmly anchored to the interventricular septum by short chordae. When associated with Ventricular Septum Defects (VSDs), both types can cause partial to near-complete obstruction of the VSD [6]. Only a few cases are asymptomatic. In most cases, accessory valve tissue is associated with other cardiac malformations, therefore the symptoms of this anomaly depend on coexisting ones [7]. Accessory tissue originating from the tricuspid valve and protruding into the Left Ventricle (LV) outflow tract through a VSD has also been described. This tissue can form a pouch, the walls of which are similar to the tissue of the normal tricuspid valve or can simply be a papillary-like mass of connective tissue. The result can vary from mild to severe LV outflow tract obstruction. Adhesions to the rims of the VSD obstructing the VSD can also be noted [8].

The available literature is scarce on anatomic anomalies of the mitral valve but we report an accessory leaflet in the mitral valve. The accessory mitral valve has been deemed a rare phenomenon and has been accountable for severe mitral regurgitation secondary to outflow tract obstruction [9].

CONCLUSION

This case report is a rarity as complete transposition of the valvular complex is seldomly observed. We attribute this variation as a consequence of its embryological formation. About our case report, in the case of 3 cusps in the mitral valve, there is a possibility of 3 endocardial cushion formation whereas underdevelopment of one of the endocardial cushions in the tricuspid valve might be responsible for 2 cusps formation.

As mentioned in the literature, reports of 2 cusps in the tricuspid valve have been reported but the tricuspid valve functionally works like a bicuspid valve leading to insignificant circulatory changes. On the other hand, the presence of supernumerary cusps in the mitral valve poses a threat for outflow tract obstruction leading to severe mitral regurgitation. Therefore, this variation reporting is of interest to cardiologists, cardiothoracic surgeons, and interventionists.

DECLARATIONS

Conflicts of Interest

The authors declared no potential conflicts of interest concerning the research, authorship, and/or publication of this article.

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