

Bicuspid pulmonary valve without associated cardiac anomalies: a case study of a rare occurrence

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Abstract

The present case study deals with a very rare occurrence of a bicuspid pulmonary valve without any other associated morphological anomaly in the heart. Bicuspid pulmonary valves have been rarely reported in isolation as this condition is very rare to occur alone and the reported cases are usually associated with other gross anomalies of the heart. The bifoliate valve in this particular instance is associated with a perfectly normal heart comprising of a) a normally placed, guarded and uncalcified aortic orifice; b) normally originating coronary arteries; c) a normal atrial and ventricular septa and musculature; and d) normal atrio-ventricular valves and papillary muscles. It may be true that the occurrence of asymptomatic bicuspid pulmonary valve is possible and can remain undetected till noticed incidentally.

Keywords: bicuspid, pulmonary, aortic, asymptomatic.

1 Introduction

The pulmonary and the aortic valves are carved out during development of the cardiovascular system at the junction of the conus arteriosus and the bulbus cordis. Both of these orifices are guarded by three semilunar cusps. These cusps guarding the two orifices are formed by sequential events out of existing four primitive cushions (ventral, dorsal, right and left) guarding the truncus-bulbar junction. The right and the left cushions each are divided into two separate cusps by a transversely oriented 'distal bulbar septum'. Each of these segmented parts from the right and the left cushions join with the ventral and the dorsal cushions. Thus six cusps are formed at these two orifices; the orientation of the cusps at these two openings being the mirror image of the other. The segment of the outflow tract anterior to the 'bulbar septum' develops as the pulmonary orifice whereas the part lying dorsal to the septum transforms into the aortic opening. The role of the neural crest cells is well documented as the predominating influence in shaping-up the formation of the valves guarding the great vessels (CREAZZO, GODT, LEATHERBURY et al., 1998; PAIGE, OLAOPA, FIRULLI et al., 2007; ROSENQUIST and FINNELL, 2007; WALDO, ZDANOWICZ, BURCH et al., 1999). Since the migration of the neural crest cells fashion the development of several other structures in their trail, it is perhaps true that an anomaly of function of these cells at a given location such as the truncus arteriosus may also affect a different anomaly at a different location within the heart or elsewhere in the body (HARAGUCHI, KENJI and YASUDA, 2008; TAN and MORRISS-KAY, 1986; CLOUTHIER, HOSODA, RICHARDSON et al., 1998). Deformities of the pulmonary valves have been previously reported. It has been noted that deformed pulmonary valves are usually associated with several other gross deformities of the heart and an isolated occurrence of a bicuspid pulmonary valve is quite a rarity and is usually associated with other cardiac anomalies (GERLIS,

1999; NAIR, THANGAROOPAN, CUNNINGHAM et al., 2006). On the other hand, it is found that bicuspid aortic valves are relatively common and may be congenital in origin as they cannot be linked with rheumatic infiltrations on histological examination as studied by Karsner and Koletsky (1947) or evidenced by clinical history; Campbell and Kauntze (1953). Afflictions of the pulmonary valves especially that of the bicuspid variety, on the contrary, are 'extremely uncommon and a healthy bicuspid valve in a heart that does not have some fairly gross abnormality of septa or origin of great vessels may well be unique'; as put forward by Enticknap (1956). A series of necropsy studies at the Guy's Hospital has revealed that pulmonary malformations are associated predominantly with the Fallot's tetralogy and one instance of Aortic stenosis (BRINTON and CAMPBELL, 1953).

2 Case report

The case reported here was detected in a routine undergraduate dissection session at the institute. The heart reported in the case was obtained from a 62 year old man who had died of cardio-respiratory arrest. On obtaining relevant details of the person, it was found that the person weighed about 54 kg and stood at 6' when assessed a couple of months before his death. The available documents related to the cadaver did not mention the presence of any co-existing cardiac anomaly in it or as the cause of death of the person. Examination of the pulmonary valves revealed its bicuspid nature (Figure 1). Further careful inspection discovered important features that were quite unique and in contrast to the usual anomalies associated with a bifoliate pulmonary valve. The rest of the heart was morphologically very normal. The heart weighed 290 g and looked healthy in its contours. The pulmonary orifice other than being guarded by a bicuspid valve was of normal dimensions measuring 11 cm

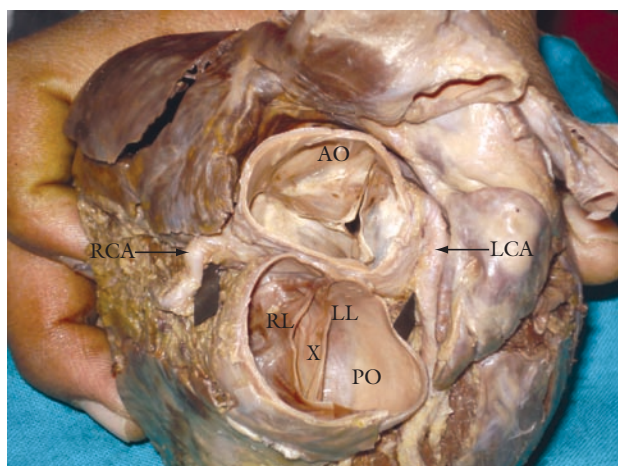


Figure 1. Pulmonary and Aortic openings. AO = Aortic orifice; PO = Pulmonary orifice. RCA = Right Coronary Artery; LCA = Left Coronary Artery; RL = Right Leaflet; LL = Left Leaflet; X = Pulmonary slit.

in circumference with the 4 cm long slit of the valve lying in an antero-posterior direction. Neither the aortic nor the pulmonary valves looked stenotic or felt calcified. The aortic orifice was guarded by three normally oriented cusps. The coronary arteries originated and branched normally. The atria or the ventricles showed no dilatation or hypertrophy on visual inspection. The thickness of the right ventricular wall was approximately 8 mm whereas the left ventricle had a thickness of 20 mm. The heart was without any septal defects or anomalous positioning of the great vessels. The papillary muscles, the chordate and the cusps of the confirmed to be normal by the department of Pathology of the institute.

3 Discussion

As stated earlier, the etiological factors leading to the formation of anomalous pulmonary valves include the abnormal functioning and migration of the neural crest cells. The involvement of such an etiology may be doubtful as the rest of the areas in the cadaver that are influenced by the migrating neural crest cells, seem morphologically normal on inspection. Fernandez et al. have extensively studied exclusively inbred strains of Syrian hamsters that have high propensity of acquiring congenital malformations affecting the pulmonary and the aortic valves (FERNÁNDEZ, FERNÁNDEZ, DURÁN et al., 1998) and have found them to be linked to several cardiac anomalies; very rare of the defective valves were reported as occurring independently and without an associated anomaly. The same researchers have concluded that an anomaly in the pulmonary valve more commonly present as a quadricuspid structure than a bicuspid one. Both the conditions are associated with gross structural anomalies. Innocuous presence of bifoliate pulmonary valves, as present in this case, is extremely rare. Some interesting experimental studies in chick embryos have indicated towards a haemodynamic etiology for these valvular anomalies. Colvee et al. (1983) have suggested a close relationship between the local haemo-dynamic patterns inside the ventricular cavity and the outflow tract that shape-up the morphology of the exit

valves of the great vessels. Almost all of the bicuspid valves mentioned in the previous two references possessed a transverse slit with the two cusps situated in a ventral and a dorsal orientation unlike that seen in this report. The incidence of quadricuspid pulmonary valves has been reported as common findings, both in asymptomatic necropsy specimen as well as in patients suffering from serious cardiac compromise. Bicuspid pulmonary valves are, on the contrary, associated with severe cardiac malformations (GERLIS, 1999; NAIR, THANGAROOPAN, CUNNINGHAM et al., 2006; CAMPBELL and KAUNTZE, 1953; FERNÁNDEZ, FERNÁNDEZ, DURÁN et al., 1998). The present report emphasizes the fact that an isolated bicuspid anomaly of the pulmonary valve may be compatible with normal cardiac anatomy and physiology. Though very unique and rare in its incidence, a bifoliate pulmonary valve can exist without other expected cardiac anomalies and remain uneventful even on its own accord. Such a rare condition of the pulmonary valve should be searched for as extensively as done for anomalies associated with the aortic valve (COLVEE and HURLE, 1983) for the rate of its prevalence and hereditary linkage (PAUPERIO, AZEVEDO and FERREIRA, 1999) as studied in aortic malformations. Establishment and acceptance of the fact that a bicuspid pulmonary valve can function normally and independent of any associated cardiac compromise can put to rest many an anxious situation.

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