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Original Research Article

Variations of the pulmonary valve

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Abstract

Objectives: The human pulmonary valve is an important structure and knowledge about its normal anatomy is essential in detecting valve diseases and in valve replacement surgeries. The normal pulmonary valve has three semilunar cusps or leaflets. The valve is known to present anatomical variations with respect to number of cusps, circumference and presence of fenestrations. The aim of the study is to detect and record such variations. A prospective observational type of study on 213 randomly selected autopsy cases with age ranging from 2 ½ to 89 years was conducted at Government Medical College, Kozhikode, Kerala with consent from Institutional Ethics Committee.

Methods: Pulmonary valves from hearts dissected during autopsy were washed thoroughly and fixed in formalin. Each specimen was numbered systematically. Circumference was measured. Age, sex, height, weight, weight of the heart and other cardiac anomalies, if any, were recorded.

Results: 1) Out of 213 cases, 211 valves had 3 cusps (99.06%), one valve had two cusps (0.46%) and one had four cusps (0.46%). 2) Fenestrations were seen in 103 valves (48.35%). 3) Cusps were asymmetrical in 15 valves (7.04%). 4) Circumference of the valve had a significant relation to age and sex of the individual.

Conclusions: The anatomical variations occurring in the pulmonary valve have significant clinical relevance in the diagnosis, management and prevention of valve diseases, valve repair and valve replacement surgeries.

Keywords: Pulmonary valve, cusps, bicuspid valve, quadricuspid valve, fenestrations.

1. Introduction

Pulmonary valve and aortic valve are grouped as semilunar valves of the heart; each valve has three semilunar cusps or leaflets. The pulmonary valve is the most superior and anterior of the cardiac valves. Normally, it lies anterior and to the left of the aortic valve. Its cusps are named right anterior, left anterior and posterior. The free margin of each cusp has a central localized thickening called the nodule of Arantius and the thin margin called the lunule. The pulmonary sinuses are the dilatations in the wall of the pulmonary trunk just above the cusps [1].

Each leaflet is an endocardial fold, with a fibrous core. Perforations or fenestrations within the leaflets close to the free margin and near the commissures are common.

During diastole, all three leaflets are tightly apposed. The valve opens passively during ventricular systole and closes rapidly at the end of systole. The pulmonary valve is difficult to visualize in echocardiography. Usually, only the posterior leaflet is visible when the valve is closed. [2]

1.1 Development

The pulmonary and aortic valves are derived from endocardial cushions that are formed at the junction of the truncus arteriosus and the conus. Two cushions, right and left, appear in the wall of the conus. They grow and fuse with each other. With the separation of the aortic and pulmonary orifices, the right and left cushions are each

subdivided into two parts, one part going to each orifice. Simultaneously two more endocardial cushions develop – anterior and posterior. As a result, the aortic and pulmonary openings each have three cushions, from which 3 cusps of the valve develop.

The pulmonary valve is at first ventral to the aortic valve. Subsequently there is a rotation so that the pulmonary valve comes to the ventral and to the left of the aortic valve. After this rotation, the cusps acquire their definitive relationships as follows: Aortic valve – 1 anterior (right) and two posterior (left posterior, right posterior), pulmonary valve – 1 posterior, 2 anterior [3].

The role of neural crest cells is well documented in the formation of the valves guarding the aorta and pulmonary trunk by Anna *et al* [4]. Since the neural crest cells take part in the development of several other structures in the body, anomalies of valves of the heart may be associated with anomalies at a different location in the body.

2. Materials and methods

The prospective observational study was conducted in the Department of Forensic Medicine, Government Medical College, Kozhikode, Kerala with consent from the Institutional Ethics Committee. The hearts were obtained from randomly selected autopsy cases conducted in the Department of Forensic Medicine over a period of 6 months. The age of the subjects (total 213 – 166 males and 47 females) ranged from 2 ½ to 89 years.

After autopsy, the hearts were washed thoroughly in running tap water. Weight of the heart and the thickness of ventricles were noted. All the four valves – aortic, pulmonary, mitral and tricuspid – were examined. The pulmonary valves were carefully removed by trimming the pulmonary trunk 1-2 cm above and below the valve. These valves were numbered systematically and immersed in 10% formalin. At a convenient time, they were studied in detail.

The valves were stretched and pinned to a wooden board with drawing pins. Circumference was measured using a ruler to the nearest millimeter. The horizontal and vertical length of each cusp was also measured. Other anatomical features like number of cusps, asymmetry, calcification, presence or absence of fenestrations and other associated cardiac anomalies were also noted. Photographs were taken.

An increase in the frequent use of conservative surgical techniques for repairing and replacing damaged valves has increased an interest in the anatomy of valves. Thrombo-embolism, ruptures, restenosis and calcifications in the prosthetic valves are the major causes of morbidity and mortality in patients undergoing valve replacement. This shows the necessity to develop a new valve, with minimum complications. The details regarding the anatomy of pulmonary valves will help in designing and manufacture of the prosthesis and also in aortic root replacement with pulmonary autograft.

3. Results

3.1 Number of cusps

Number of cusps in the pulmonary valves was variable. Normal valves with three cusps were present in 211 cases out of 213 (99.06%). Quadricuspid pulmonary valve was found in one heart (0.46%) and bicuspid, in one case (0.46%).

3.1.1 Quadricuspid pulmonary valve (Figure 1)

This rare variation was found in a 25 year old male. The cause of death was non-cardiac. The heart weighed 300gms. The thickness of the ventricles was normal. The circumference of the pulmonary valve was 6.5cm. The measurements of the cusps are given in Table 1.

Table 1: Measurements of cusps of quadricuspid valve (cm)

Measurement	Cusp 1	Cusp 2	Cusp 3	Cusp 4
Horizontal	2.1	2.1	0.2	2.1
Vertical	0.7	1	0.3	1.3

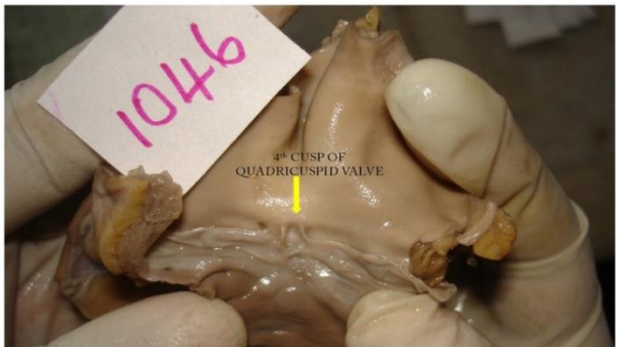

Figure 1: Quadricuspid pulmonary valve

From the table, it is evident that there were 3 large cusps with slight variation in measurements and a small cusp. No other cardiac anomalies were detected.

3.1.2 Bicuspid Pulmonary Valve (Figure 2)

This rare variant was found in a 43 year old male. Cause of death was non cardiac. Each cusp was 3.1cm horizontally and 1.2cm vertically. All the other valves, ventricles and septa were normal.

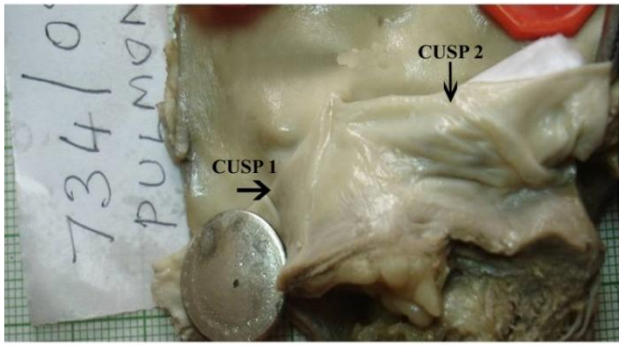

Figure 2: Bicuspid pulmonary valve

1) Asymmetry of cusps

In the present study, 15 valves (7.04%) showed asymmetrical cusps, the details of which are given in Table 2.

Table 2: Asymmetry of Cusps

No. of valves	Size of cusps	Percentage
198	Three equal size	92.95
6	Three different sizes	2.84
8	2 equal large, 1 small	3.75
1	4 unequal	0.46

2) Fenestrations: (Figure 3)

One hundred and three valves (48.35%) showed fenestrations. The size, shape and number of fenestrations varied. One, two or all the cusps showed minute openings, close to the margins. Lowest age in which the valve showed fenestrations was 2 ½ years – both aortic and pulmonary valves showed a small hole in one cusp each. But in an 88 year old male, no fenestrations were detected in the pulmonary or aortic valve. Only one cusp showed fenestrations in 59 valves; 2 cusps in 31 valves and all the three cusps in 13 valves.

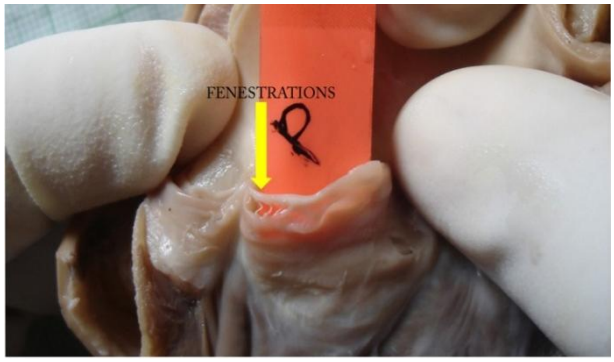

Figure 3: Pulmonary valve with fenestrations

3) Pulmonary valve circumference

With regard to age, there was a progressive increase in the circumference. The measurement was more in males than in females of the same age group, as shown in Table 3.

Table 3: Pulmonary Valve Circumference

Age group	Males		Females	
	Average (range) Cms	No. of cases	Average (range) cms	No. of cases
01-09	3.2(3.2)	1	3.6(3.6)	1
10-19	5.2(4.5-6.6)	7	5.46(4.2-6)	3
20-29	5.81(4.5-7.5)	28	4.71(3.3-6)	16
30-39	6.3(4.8-8)	27	5.47(4.5-6.9)	7
40-49	6.04(4.5-8.4)	35	4.8(4.8-7.5)	7
50-59	6.21(4.5-7.9)	20	6.02(6.3-7.5)	7
60-69	7.07(5.6-9)	23	7.02(6.3-7.5)	4
70-79	6.72(5.6-7.8)	15	7.8(7.5-8.1)	2
80-89	6.86(5.1-8.1)	6	6.6	1

In adult males (more than 21 years) the values ranged from 4.5 - 9cm and average value was 6.43cm. In adult females, the values ranged from 3.3 – 8.1cm and the average measurement was 6.2cm.

4. Discussion

4.1 Variations in the number of cusps

4.1.1 The quadricuspid pulmonary valve (QPV)

In the present study, QPV was detected in a 25 year old male. No associated cardiac anomalies were detected. Cause of death was non cardiac.

Quadricuspid pulmonary valve is a rare congenital anomaly, which is usually detected during an autopsy [5]. Incidence of QPV is 1 in 20,000 necropsies [6]. It is more frequent in males with a ratio of 2:1. Exact cause is unknown.

As isolated QPV is very rare .Anthony Enoch [7] has reported four cases of isolated congenital QPV and one case of QPV with rheumatic mitral valve disease. In each instance, the anomaly was an incidental finding at autopsy. None of the deceased had any clinical evidence of pulmonary valve lesion during life. In all the four cases, there were three large equal cusps and one small fenestrated cusp. He also detected a white patch of endocardial thickening about 0.5cm diameter beneath the small cusp in two cases. These white patches of endocardial fibrosis are well recognized at sites of abnormal blood flow 19(Hudson 65) and are described as “jet lesions”.

QPV can be associated with structural or functional alterations of the heart such as ASD or VSD, aortic valve abnormalities or patent ductus arteriosus [7] Nollen, Kodde *et al*[8] have reported QPV with secondary anomalies such as valvular stenosis and regurgitation. In one case, they had observed pulmonary artery aneurysm due to valve regurgitation.

Classification of quadricuspid semilunar valves:

In 1973, Hurwitz and Roberts [9] classified quadricuspid semilunar valves into 7 types (A-G).

A: Four equal cusps
B: Three equal and one smaller cusp
C: Two equal larger and two equal smaller
D: One large, two intermediate and 1 small
E: Three equal cusps and one larger cusp
F: Two equal larger and two unequal smaller
G: Four unequal cusps.

In the present study, there were 2 larger cusps with same horizontal length (2.1cm each). The fourth cusp was only 0.2cm long. The three larger cusps had different vertical measurements – 0.7, 1 and 1.3cm each. The smallest cusp was only 0.3cm vertically. Thus, we could say that the QPV observed in the present study belonged to ‘G’ type of Hurwitz and Roberts classification. None of the cusps had fenestrations and there were no endocardial fibrosis beneath the small cusp as reported by Anthony Enoch.

Carlos *et al* [10] have reported a case of QPV associated with ASD and pulmonary stenosis.

Hedayat *et al* [11] have detected a quadricuspid pulmonary valve in a live newborn by two dimensional echocardiography.

4.1.2 Bicuspid Pulmonary Valve (BPV)

In the present study, the bicuspid valve was detected in a 43 year old male. There were no other cardiac anomalies. Cause of death was non-cardiac. Both cusps were of equal size. There was no calcification. There were no fenestrations.

Mahato NK [12] has reported a case of isolated BPV without associated cardiac anomalies.

Emura *et al* [13] reported a case of congenital bicuspid pulmonary valve in a 95 years old male Japanese cadaver during dissection at Gifu University in 1990. No evidence of heart failure was detected in the cadaver. The left cusp was bigger than the right cusp.

Brinton and Campbell[14], based on a series of necropsy studies at the Guy's Hospital, are of the opinion that pulmonary malformations are associated predominantly with Fallot's Tetralogy or aortic stenosis.

Gerlis [15], Nair *et al*[16], Campbell and Kauntze [17], Fernandez *et al*[18] opined that BPV are associated with severe cardiac malformations.

The present study and the findings of Emura *et al* emphasizes the fact that an isolated bicuspid anomaly of the pulmonary valve may be compatible with normal cardiac functions and it need not be always associated with other cardiac anomalies.

4.1.3 Asymmetry of Cusps

In 15 valves out of 213, the cusps were asymmetrical (7.04%) and in 198 valves (92.96%), the three cusps were equal in size. In 6 cases, the three cusps were of different size. In 8 valves, there were two large cusps and a smaller one. In a quadricuspid valve all the cusps were of different dimensions.

It has been reported by Lee *et al*[19] that the presence of asymmetry among aortic valve cusps is strongly associated with development of aortic stenosis in the future but we could not find any report suggesting the occurrence of pulmonary stenosis in asymmetric pulmonary cusps.

In the present study, there was no evidence of pulmonary stenosis and right ventricular hypertrophy in the hearts in which asymmetry of cusps were detected.

4.1.4 Fenestration of cusps

Fenestrations or small perforations were detected parallel to the free margin of the cusps in 103 valves (48.35%). The size, shape and number of fenestrations varied. Only one cusp was affected in 59 valves, 2 cusps in 31 valves and all the three cusps were showing fenestrations in 13 valves. Rokitansky [20] has given an accurate description of fenestrations of semilunar valves. He reported that these openings are almost always situated near the margin of leaflets. They may be oval, elliptical or fissure-like.

Foxe [21] detected fenestrations of semilunar valves in 82% cases in a study conducted on 300 hearts. He observed that with increasing age, fenestrations often form within the lunular regions, near the commissures. They may be single or multiple. Because they are situated above the line of closure, they do not cause regurgitation. But chronic valvular incompetence can occur if the fenestrations are large and extend below the closure line or if the valve annulus dilates and causes stretching of cusp and prolapse. As a result, proper coaptation of the lunular areas of two opposing cusps will be lost.

Ben and Hathway [22] noted well defined fenestrations of pulmonary cusps in two out of three patients with pulmonary hypertension and a Graham Steell Murmur. They detected fenestrations in 72% of 342 hearts. They opined that, in some instances, fenestrations may lead to regurgitation.

We had studied the aortic valves in the same hearts and had observed fenestrations in 110 valves. Almost in all hearts, fenestrations of aortic valve are associated with fenestration of pulmonary valve also, suggesting a common embryological origin and mechanism which has given rise to the fenestrations. None of the patients died of cardiac causes.

4.4.5 Circumference of the valve

In the present study, we came to the conclusion that there is a definite relationship with age and circumference of the pulmonary valve. It was also noted that the circumference of the valve in males is more than that of females of the same age group.

Various authors have reported the measurements of pulmonary valve, which is shown in Table 4.

Table 4: Circumference of Pulmonary Valve

Name(s) of author	Circumference (cm)
Kouji Chida *et al*	6.8±0.8
Westaby S	4.88 ± 1.25
Garg S	6.5 ± 0.59
Ilankathir S	6.82
Brandenberg	7
Friedrich *et al*	7.2
Kinare *et al*	6.66
Present study	Male : 6.43; Females: 6.2

The values obtained in the present study are almost similar to those of Garg,[23] Ilankathir [24] and Kouji Chida [25] and Kinare.[26] Our values are higher than Westaby's [27] and lower than Brandenberg [28] and Friedrich [29].

5. Conclusion

Knowledge about the dimensions and morphology of the pulmonary valve is important in the diagnosis,

prognosis and planning surgical procedures. It will also help in manufacturing prosthetic valves. By frequent follow up and timely intervention, morbidity and mortality rates can be reduced in patients with valvular lesions.

Quadricuspid and bicuspid pulmonary valves can be isolated anomalies. But they can be associated with other cardiac anomalies, some of which may be serious. So, if a QPV or BPV is detected, the patient should be thoroughly investigated to rule out the possibility of other cardiac anomalies.

Fenestrations of the semilunar valves may give rise to regurgitation in the future. The cusps may rupture. They can give rise to pulmonary hypertension also. So patients, in whom fenestrations are detected by echocardiography, should have regular follow up or a surgical repair.

Ross in 1986 has introduced the technique of aortic root replacement with a pulmonary autograft in pediatric age group [30] (Ross-Konno operation). A pulmonary autograft has the following advantages – it is not prone for structural degeneration, does not require anticoagulant therapy and has a growth potential which avoids a reoperation in a growing child. So, prior to aortic root replacement with a pulmonary autograft, a thorough knowledge about the measurements and anomalies of the pulmonary valve is essential, so that it will not give rise to any problems in the future.

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