

Case Report

A CASE REPORT OF BICUSPID AORTIC VALVE WITH REFERENCES TO ANATOMICAL AND EMBRYOLOGICAL ASPECTS

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ABSTRACT

Bicuspid aortic valve (BAV) is the most common malformation in congenital cardiac disease with the prevalence of 1-2%. In Bicuspid aortic valve (BAV), the aortic valve has only two cusps instead of three cusps. In this paper, we are presenting a case of BAV associated with mild to moderate aortic stenosis and regurgitation, with no valvular incompetence. A 18 years old male student presented with the chief complaints of exertional dyspnea associated with occasional chest pain. Color Doppler echocardiogram revealed congenital bicuspid aortic valve with mild to moderate aortic stenosis. It showed mild bicuspid aortic regurgitation having normal ejection fraction and without valvular incompetence (aortic root dilatation). The patients with BAV are advisable to have a regular follow up with cardiologist or cardiac surgeons. In case of severe valvular dysfunction, aortic valve replacement is a treatment of choice.

KEY WORDS: Bicuspid Aortic valve, Embryology, Aortic stenosis, Aortic Regurgitations.

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INTRODUCTION

The aortic valve acts as a unidirectional channel between the heart and the aorta. Anatomically, the aortic valve has three small cusps or leaflets, which are surrounded by a sinus. Each cusp is crescent shaped, open fully in systole and close in diastole to regulate blood flow in a one-way direction, i.e., from the heart to aorta and preventing the backflow of blood to heart from aorta. In Bicuspid aortic valve (BAV), the aortic valve has only two cusps instead of three cusps. Many hypotheses are revolving around

this pathogenesis, however exact pathophysiology still remains unclear. BAV is the most common malformation in congenital cardiac disease with the prevalence of 1-2% [1, 2].

The prevalence of BAV is three times more common in males than females.

The BAV individual may be totally asymptomatic clinically and compatible with normal function until the third and fourth decades when the valve becomes stenosis and dysfunctional [3, 4]. BAV has been describe in both an isolated form and in associated with other

congenital cardiac disease such as coarctation of aorta, aortic stenosis, aortic dilatation, Tetralogy of Fallot [1]. This case report aims to further characterize this uncommon pathogenesis and to discuss BAV in term of anatomical and embryological aspects.

CASE REPORT

A 18 years old male student, who is a non-smoker, non-diabetic and non-hypertensive presented with the chief complaints of exertional dyspnea associated with occasional chest pain. A detailed history was taken. He is a single child born of non consanguineous parents. He was diagnosed to have a valvular heart disease when he had a history of fever for more than one week at an age of 3 years.

On physical examination, the patient was in stable condition with a short and average body built. There is no cyanosis and clubbing. Pulse rate was normal and regular (76 beats/ min). Blood pressure was 130/80 mmHg in right upper limb and 128/80mmHg in the left upper limb. On cardiac examination, apex beat was felt at 5th intercostal space and systolic thrill was felt in the aortic area. On auscultation, S1 and S2 with Ejection systolic murmur (EJM) were heard at the aortic area. No significant abnormality was found on CBC and chest X-ray. A baseline ECG was taken and found the normal sinus rhythm. Color Doppler echocardiogram revealed congenital bicuspid aortic valve with mild to moderate aortic stenosis (Fig 1). It showed mild bicuspid aortic regurgitation having normal ejection fraction (EF) i.e. left ventricular ejection fraction (LVEF) -65% with no valvular incompetence (aortic root dilatation).

Fig. 1: Transthoracic echocardiogram shows bicuspid aortic valve in the parasternal short axis view.



DISCUSSION

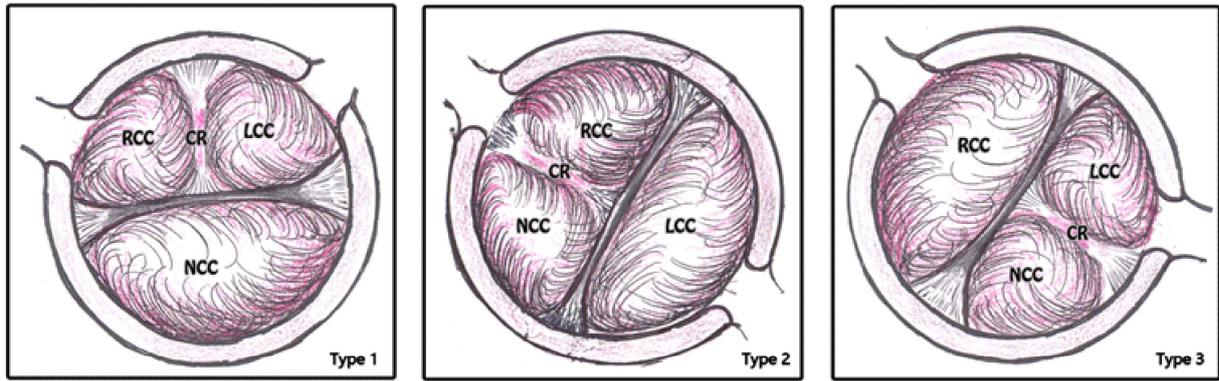
The fetal cardiac structure attains its definitive state by 8 weeks. The development of aortic and

pulmonary valves takes place simultaneously as semilunar valves at the junction of the truncus arteriosus and conus. The two truncal ridges or cushions develop in the right and left wall of truncus arteriosus. Simultaneously two more cushions anterior and posterior appear. With the separation of aortic and pulmonary openings by an aorticopulmonary septum, the right and left cushions divide into two halves. One part of cushions goes to aortic opening and other part goes to pulmonary opening. In the normal aortic valve, the right and left leaflets are formed from the right and left half of the cushions while the posterior leaflet is formed from the posterior cushions [2,5].

Adult aortic valve has three small cusps or leaflets (Right, Left and Posterior), which are surrounded by an aortic sinus. The right coronary artery arises from the right aortic sinus, and the left coronary artery arises from the left aortic sinus. The posterior aortic sinus does not give rise to the coronary artery, hence it referred as noncoronary cusp or sinus. In Bicuspid aortic valve (BAV), the aortic valve has only two cusps instead of three cusps. The etiology behind the Aortic valves with two leaflets may be due to the fusion of two of the three cusps [6]. Based on the fusion of the cusps, the bicuspid valve has classified into three types (Figure 2). Fusion of right cusps with the left cusps is the most common type of bicuspid aortic valve (79.6%). Second most common type is fusion of right cusps with noncoronary cusp (19.4%). The rarest, is fusion of the left cusps and non-coronary cusps (1%) [7]. In this paper, we are presenting a case report of bicuspid aortic valve with type I variety i.e. fusion of right cusps with left cusps.

The most common clinical presentations or conditions associated with BAV are aortic regurgitation (AR), aortic stenosis (AS), mixed AS and AR, aortic dilation, endocarditis and aortopathy. The BAV individual may be unaware of this pathology and compatible with normal function until when the valve becomes stenosis and dysfunctional. As BAV progress, it is often associated with different degrees of aortic stenosis or aortic regurgitation because of its calcification. The incidence of aortic stenosis associated with BAV was found to be 72% [4]. The main

Fig. 2: The classification of bicuspid aortic valves according to site of cusp fusion.



RCC- Right coronary cusp, LCC- Left coronary cusp, NCC- Non coronary cusp, CR- Conjoint Cusp

symptoms of aortic stenotic valve include exertional dyspnea, dizziness or syncope and chest pain. In our reported case, there was a limitation of patient's physical activity i.e. exertional dyspnea and echocardiogram of BAV reveals mild to moderate aortic stenosis and aortic regurgitation, with no valvular incompetence (aortic root dilatation).

Periodic and routine follow up of the patient with BAV by echocardiogram is an essential tool for establishing the diagnosis, quantifying the severity of the valve, to evaluate the functional status of the valve and to assess the left ventricular function.

CONCLUSION

In summary, Bicuspid aortic valve is a commonest entity of congenital heart disease with an etiology that remains unclear. In this paper, we are presenting a case of BAV associated with mild to moderate aortic stenosis and regurgitation. The patients with BAV are advisable to have a regular follow up with cardiologist or cardiac surgeons. In case of severe valvular dysfunction, aortic valve replacement is a treatment of choice.

Conflicts of Interests: None

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