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An Odyssey of a Stenosed Bicuspid Aortic Valve

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Case Report

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ABSTRACT

Bicuspid aortic valve (BAV) is known to cause the heart's aortic valve narrow. This prevents the aortic valve from opening fully causing reduction and/or block flow of blood from the heart. The clinical presentation of BAV is highly variable. In many cases, BAV is asymptomatic, however in some it may become calcified later in life. Its major manifestations are aortopathy, valvular dysfunctions and endocarditis. We report a case of BAV with a follow-up for over 40 years to assess the temporal association and progression of various complications for better understanding of disease process.

Keywords: Bicuspid aortic valve; aortic stenosis; aortic regurgitation.

1. INTRODUCTION

Bicuspid aortic valve (BAV), being the most common congenital heart disease, it causes a spectrum of diseases, like aortic valve stenosis and severe heart failure (HF) in newborns, whereas aortic dissection in adults [1]. Previous studies revealed that 1 to 2% of the entire population has BAV disease [2,3]. However, BAV disease is present in up to 20% of patients over the age of 80 [4]. The factors determining the prognosis of BAV disease are older age, phenotype, flow dynamics, gender, elevated systolic blood pressure (SBP), smoking, and presence of valvular and ventricular dysfunctions, aortic dilatation, endocarditis, high total cholesterol levels, valve degeneration score, and genetic predisposition [5].

Several studies have reported significant disparities in the relationship between aorta dilation and BAV characteristics. Recently in an impressive study with 852 adult patients having BAV, showed phenotypic predictors of valve dysfunction and aortic root dilation [6,7]. However, with a near normal life expectancy, many twists and turns in the therapeutic process can often pose significant treatment challenges [8]. Early detection and continued surveillance are crucial to allow early intervention and to preserve cardiac function.

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2. CASE REPORT

A 64 year old man, whose medical history) revealed that, at the age of 22 years (in May 1975), presented with symptoms of exertional breathlessness of 6 months duration (NYHA-class-II: New York Heart Association class II) (documents well preserved all these years). On evaluation, he was found to have a grade 3/6 late peaking crescendo-decrescendo ejection systolic murmur in the right second intercostal space, radiating to the carotids with a ejection click. Chest X-ray was normal and ECG showed a normal axis, concentric left ventricular hypertrophy (LVH). Clinical suspicion of aortic stenosis was confirmed with aortogram, which showed a calcified bicuspid aortic valve (BAV) with peak gradient of 60 mm Hg across the aortic valve and normal sized aorta. Surgical aortic valvotomy (SAV) was performed which brought instant relief in his symptoms.

Patient was relatively asymptomatic for next 18 years. However, in January 1993 he again developed exertional breathlessness and symptoms of pre-syncope. On clinical evaluation, restenosis of aortic valve was suspected. Two-Dimensional Echocardiography (2D-ECHO) confirmed presence of calcific BAV with mean gradient of 40 mm Hg and moderate aortic valve stenosis (Indexed valve area of 0.7 cm^2/m^2) and moderate aortic valve regurgitation with LV ejection fraction of 60%. A2 decision to surgically replace the aortic valve was taken in December 1993; he underwent aortic valve replacement (AVR) with a Medtronic mechanical prosthetic valve of No.25.

Post AVR, on the second day patient developed complete heart block (CHB) (Fig. 1). He was observed in the hospital for next 15 days. Initially CHB was managed with a temporary pacemaker and a trial of steroids. Due to persistent nature of CHB, a permanent single chambered pacemaker (VVI type) was implanted at the right ventricular (RV) apex. Post pacemaker ECG showed left bundle branch block (LBBB) morphology with QRS duration of 150 ms with well visualized P waves (Fig. 2a, b). After 12 years, in 2005, he underwent a pacemaker generator (Battery) replacement. However, in 2010, he started developing symptoms of dyspnoea and fatigue (NYHA class II). ECG showed no 'P' waves with QRS complex duration of 160 ms with ventricular pacing rhythm suggestive of atrial fibrillation (AF) (Fig. 3a). 2D-ECHO showed normal aortic valve with no regional wall motion abnormalities (RWMA). LV ejection fraction was 50%.

Over the next seven years (2010 to 2017), there was a gradual fall in the LV ejection fraction from 50% to 25% with presence of diastolic MR. The mechanical prosthetic valve showed some areas of pannus formation with mean gradient of 17mm Hg across the aortic valve. Patient had progressed from NYHA class II to NYHA class III symptoms over this period. 2D-ECHO and dyssynchrony study was performed in view of progressive worsening of symptoms and broad QRS complex on ECG. The parameters suggested significant intra and inter ventricular dyssynchrony. Patient was subjected to up-gradation from a single chamber to biventricular pacemaker with a right atrial (RA) and LV leads in posterio-lateral branch of coronary sinus. Original unipolar right ventricular (RV) apical lead pacing was continued (Fig. 3b). Coronary angiogram was normal.

Post cardiac resynchronization therapy (CRT), ECG showed QRS width reduced to 130 ms with complete abolition of diastolic MR and significant improvement in LV systolic function with ejection fraction increasing to 40% on 2D-ECHO. There was also improvement in symptoms with 6 min walk distance increased from 235 meters to 340 meters. He is on regular follow-up and currently is asymptomatic.

3. DISCUSSION

The clinical course of BAV is highly unpredictable. BAV is not only associated with abnormal valve morphology, but also associated with disease of the ascending aorta leading to aneurysm formation and aortic dissection [8]. The natural history and management of BAV and severe aortic stenosis is well documented in medical literature [9]. However, the guidelines are not clear on indications for either surgical aortic valvoplasty (SAV) or balloon aortic dilatation (BAD). It has been reported that both procedures are reasonably safe, effective and can postpone the need for aortic valve replacement (AVR) [10]. A study done by Fratz group in 2008 showed aortic gradients at discharge were significantly better in patients who underwent SAV [11]. Kaplan-Meier analysis at 10 years, comparing SAV and BAD were as follows: freedom from re-intervention, 72% versus 53% and freedom from AVR, 80% versus 75% [12]. Hence, one can infer that both these procedures can be safely advocated in children

and young adults. The benefit derived by our patient who underwent SAV at the age of 22 years with an active lifestyle was for 17 years, before he needed an AVR, validates this point. However, BAD was not a well-established procedure at that time and hence the best option SAV was adopted.

AVR can result in the development of conduction abnormalities and increased risk of sudden death. The association of intra-ventricular conduction disorders and aortic valve surgery is often transient. However, persistent conduction defects, which need permanent pacemaker (PPM) implantation, are reported to occur in between 2% and 7% of patients undergoing AVR [13]. During 1960s, the incidence of CHB after AVR was 13% [14].

Dual chamber pacemaker is the device of choice implanted in patients developing heart block post AVR [14]. However in our patient, single chamber VVI pacemaker implanted post-operatively, either out of monetary constraints or by choice. Good atrial activity was seen in 12 lead ECG at time of first pacemaker implantation. He later developed worsening in NYHA class, reduced effort tolerance, LV dysfunction worsened by onset of AF.

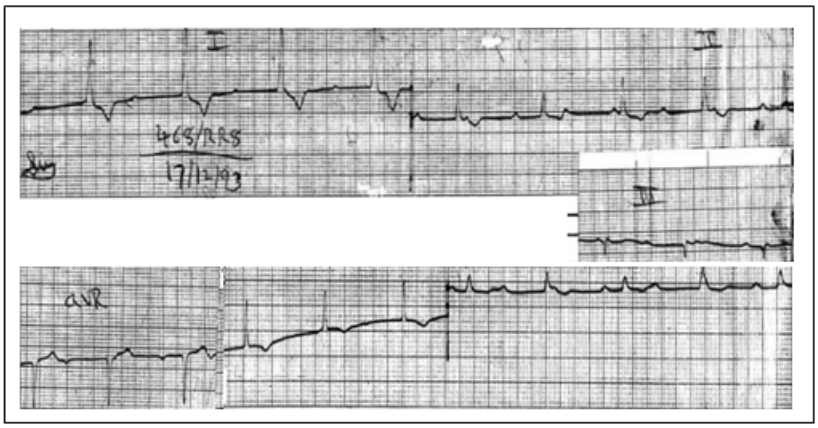

Fig. 1. ECG showing complete heart block (CHB)

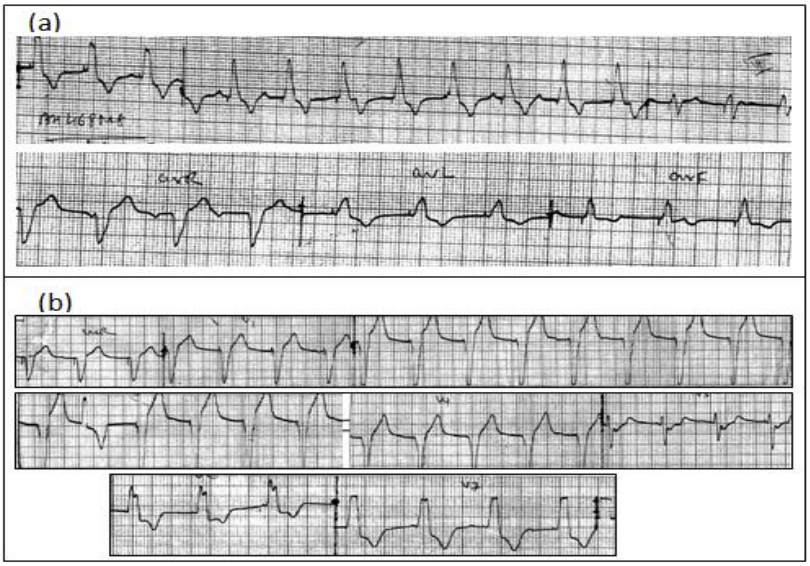

Fig. 2a-b. ECG showing single chamber Pacemaker (VVI) before implantation at the right ventricular (RV) apex (a). Post pacemaker ECG showing well visualized P waves (b)

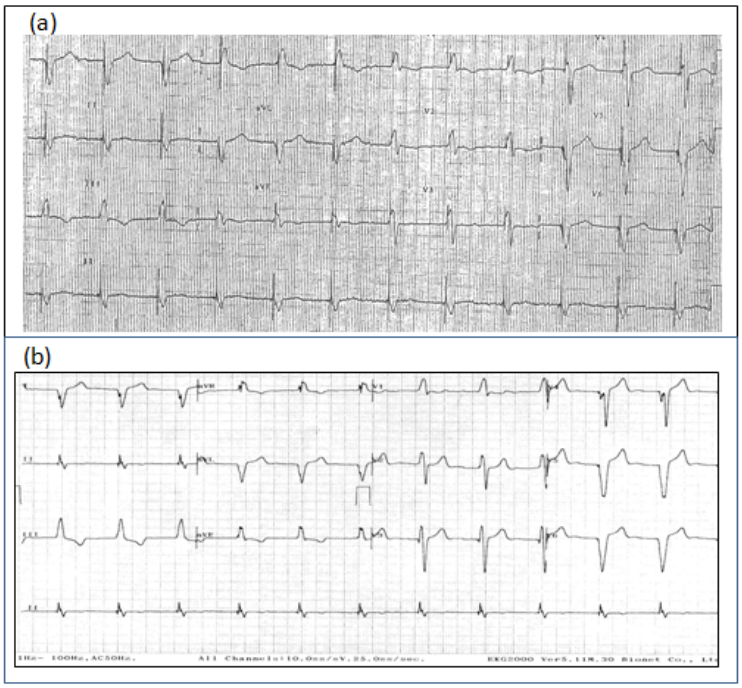

Fig. 3a-b. ECG showing a paced rhythm with AF (a). ECG post CRT implantation (b)

In 2006, Gasparini et al. [15] reported the results of a study conducted at two medical centers, which included 723 CRT system recipients, of whom 561 were in Sinus Rhythm and 162 were in permanent AF. Over a mean follow-up of 25 ± 18 months, the results were (i) increase in functional capacity, magnitude of reverse remodeling and LV systolic function; and (ii) decrease in NYHA functional class were observed in patients in sinus rhythm (SR), as well as patients in AF. The magnitude of increase in LV ejection fraction and LV end systolic volume, and decrease in NYHA functional class, was significantly greater in patients whose AV node had been ablated. Likewise, improvement in functional capacity in our patient was noted after change of pacing modality, as manifested by improvement in 6 min walk test from 235m to 340 m.

4. CONCLUSION

BAV is a congenital, phenotypically variable and genetically heterogeneous heart disease [5]. Patient education and emphasis on regular follow-up is essential to detect cardiac abnormalities for effective treatment. With constant up-gradation in medical devices, one has to keep abreast to the explosion in knowledge of various techniques and their drawbacks, which may help in better understanding of the disease process.

CONSENT AND ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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