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# Combined Stress Echo Cardiopulmonary Exercise Testing in the Evaluation of Patient with Bicuspid Aortic Valve Disease: A Link that Might be Helpful

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# Abstract

Aortic stenosis (AS) is the most common valvular heart disease, and in younger population the most common cause of AS is bicuspid aortic valve (BAV). The optimal therapy in asymptomatic patients with severe AS and normal left ventricular ejection fraction (LVEF) is still a matter of debate. In these patients risk stratification is mostly based on exercise test results. Cardiopulmonary exercise testing (CPET) combined with stress-echocardiography is relatively novel diagnostic tool which might improve quantification of cardiopulmonary functional capacity and general assessment of asymptomatic patients with hemodinamically signifiant AS and normal LVEF and thus help in clinical decision making in these patients.

# **Keywords**

Asymptomatic aortic stenosis, Cardiopulmonary exercise testing, Left ventricular ejection fraction, Risk stratification

# Introduction

Bicuspid aortic valve (BAV) is one of the most common congenital heart abnormalities, occurring in about 1-2% of individuals [1,2], with strong male predominance. Likewise, in large-scale preparticipation cardiac screening programs for young, competitive athletes, BAV is one of the most commonly identified abnormalities [3]. Aside from the problems with aortic valve stenosis or regurgitation or with enlargement of the aorta, individuals with BAV can also suddenly develop the problem of aortic dissection, especially in the presence of dilated ascending aorta. BAV is compatible with regular sport activity when it is associated with mild valvular aortic stenosis and/or insufficiency. Eligibility to participate in competitive sports is withheld in patients with severe aortic valvular stenosis (AS) and/or moderate-severe aortic insufficiency [4]. In individuals who do not have significantly changed aortic valve, causing severe AS or AI, BAV does not, at present, represent an obstacle to obtaining eligibility to practice competitive sports. Risk stratification in asymptomatic patients with severe AS is predominantly based on exercise test results. Cardiopulmonary Exercise Testing (CPET) provides a global assessment of the integrated response to incremental exercise involving the cardiovascular, respiratory, neuropsychological and skeletal muscle systems. As such, CPET compared to conventional exercise testing (ET) could generate more conclusive results and differentiate patients with truly asymptomatic AS from symptomatic AS and functional limitations due to other causes [5].

### **Case Presentation**

A 24 year active recreational athlete was referred to routine examination. He had a history of asymptomatic BAV, and did not take any medications. He was sent to our clinic for complete cardiac check-up. During the physical examination his baseline blood pressure was 115/70 mmHg, cardiac auscultation revealed a normal first heart sound and soft second heart sound. Along the left sternal border a rough grade 3/6 ejection murmur with a mid-late systolic maximum was heard and a grade 3/6 diastolic decrescendo murmur. His resting ECG showed only early repolarization changes and signs of left ventricular (LV) hypertrophy (Figure 1). He reported excellent exercise tolerance and he wanted to continue with his high intensity training.

Echocardiographic examination before the exercise test showed moderate left ventricular LV concentric hypertrophy with excellent LV systolic function. Systolic gradient through BAV - maximal aortic gradient ( $P_{max}$ ) was 78 mmHg and the mean gradient ( $P_{mean}$ ) was 46 mmHg, obtained from right parasternal view (Figure 2), and aortic valve area (AVA) calculated by the continuity equation was 0.86 cm<sup>2</sup>. However, planimetric measured AVA was 2.4 cm<sup>2</sup>, and the valve itself did not look heavily calcified; (Figure 3). Echocardiographically there was no sign of subvalvular stenosis, or obstruction in left ventricular outflow tract. There was no dilatation in any segment of the aorta. The LV diastolic dimension (5.66cm) was at upper limit (in our centre the upper limit is considered as 5.6cm); (Figure 4). Echocardiographic images were analyzed by two experienced echocardiographers.

In order to analyze his exercise capacity and possible existence of AS related symptoms, as well as change in Doppler-derived parameters, we performed combined stress echo cardiopulmonary exercise testing (CPET, maximal Bruce protocol on treadmill). Echocardiography was repeated immediately after the test with patient returned at the same right lateral decubitus position for achieving the maximal gradient across the valve. Achieved heart rate was 182 beats/min (103% predicted) during 10 min of progressive exercise. Maximal achieved blood pressure was 195/95mmHg.

Myocardial oxygen consumption (VO<sub>2</sub>) at anaerobic threshold (AT) was 31 ml/kg/min, while peak VO<sub>2</sub> was 37.1 ml/kg/min (82% VO2<sub>max</sub> predicted). O2 pulse was 14.4 ml/beat (78% predicted). Breathing reserve and ventilatory equivalents were normal, VE/VCO2 slope was 25.52, and PETCO2 showed normal increase during progressive exercise. Aortic gradients at the end of the test were not significantly higher:  $P_{max}$  was 85 mmHg and  $P_{mean}$  53 mmHg, without wall motion abnormalities and without chest pain, dizziness and/or dyspnea. ECG showed horizontal ST segment depression 1 mV in V4-V6 ECG leads, which was, in the context of severe AS, not considered as significant; (Figure 5).

#### **Discussion**

In competitive athletes, or active recreational persons, sports activity is normally allowed if the valve dysfunction or the other complications associated are not severe [1,6]. CPET is a low risk, non-invasive investigation that allows accurate, dynamic assessment of cardiopulmonary performance during exercise [1,7]. By measuring dynamic gas exchange during graded exercise, CPET can identify potential limitations and can add more information to conventional echocardiographic and stress test parameters in identifying hemodynamically significant AS. These can be often underestimated by resting lung and cardiac function measurements and certainly are not obtainable during ET.

In our patient, although measuring high Doppler-echocardiographic gradient, we were not entirely sure how significant AS was, especially in the context of planimetric measured AVA and regular sports activity which our patient practiced. However, planimetric measured AVA is probably consequence of eccentric AV geometry and eccentrically placed cusps which, not rarely, can lead to underestimation of AS severity when analyzed with 2-D echocardiography. On the other hand, CPET parameters were normal, but for physically active young person one would expect to achieve higher % of  $VO_{2max}$  and better  $O_2$  pulse after maximal training. As  $VO_{2max}$  is limited by cardiovascular reserve (heart rate and stroke volume) rather than respiratory reserve, obtained cardiac functional capacity can certainly be explained with severe AS. The optimal duration of the test is around 10 minutes for proper assumption of the  $VO_{2max}$  [8], which was reached in our case (10.5min); (Figure 6).

In addition, in athletes with bicuspid aortic valve, besides the evaluation of the aortic valve, physiologic adaptations of the heart have to be differentiated from pathological changes, for which CPET testing is useful. Furthermore, the aorta deserves special attention, because in the case of a (probably genetically determined) dilated ascending aorta, an elevated risk for aortic rupture is present during intensive and competitive exercise, especially if contact sports are involved. A general judgement in athletes with BAV on their ability to participate in regular and competitive physical activity is, therefore, not possible, but have to made on case-by-case basis.

#### **Conclusion**

This patient has truly asymptomatic severe AS, and although he has preserved functional capacity one would expect somewhat higher percentages of VO2max and O2 pulse considering he is a highly physically active young individual. We have advised him to stop with the high intensity training but he may continue with moderate aerobic physical activity. However, precisely because he is so young, it is logical to expect that the aortic valve replacement is only a matter of time in his case.

# **Figures**



Figure 1: Resting Electrocardiogram

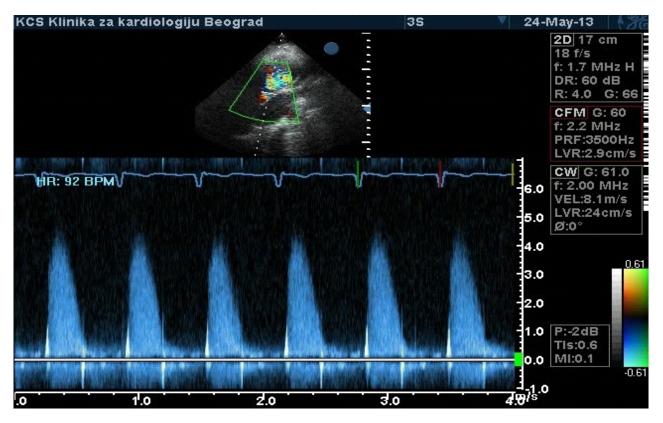


Figure 2: Maximal gradient at rest from right parasternal view



Figure 3: Aortic valve from the short-axis parasternal view at rest

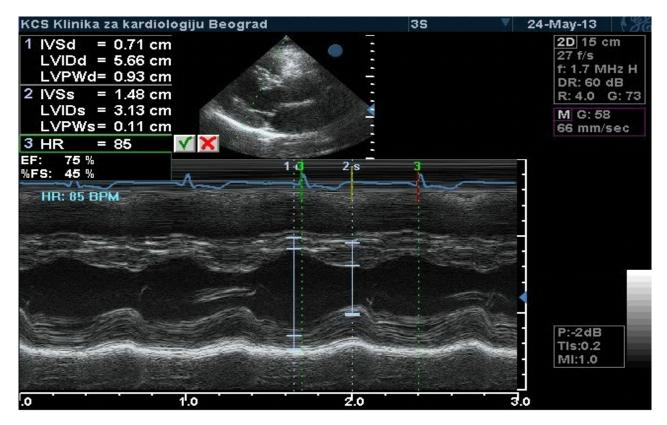


Figure 4: M-Mode long-axis parasternal view of the left ventricle

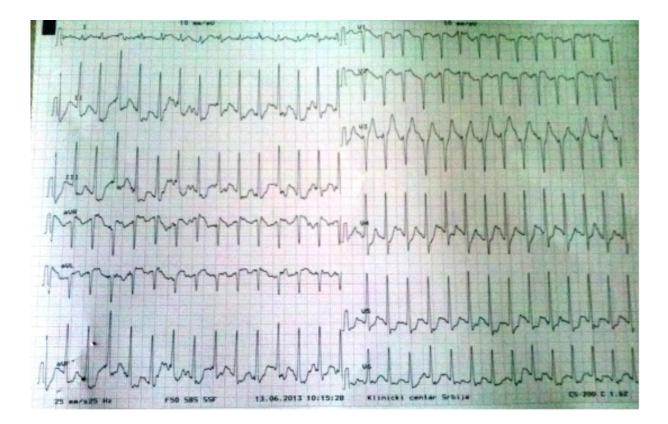


Figure 5: ECG changes at maximal effort during CPET

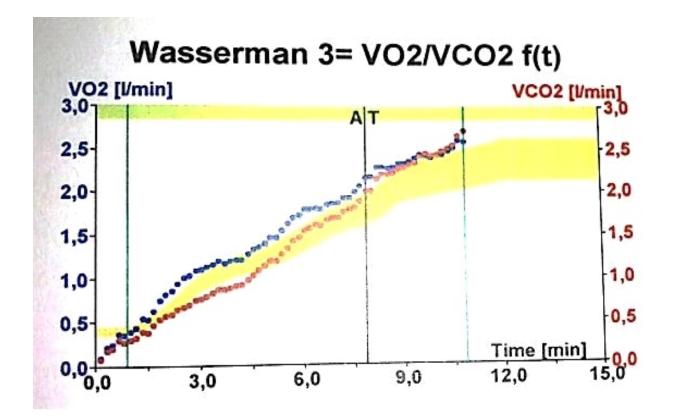


Figure 6: Wasserman's curve  $VO_2$  vs  $VCO_2$  and duration of the test

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