A Case Report on Decreased Exercise Tolerance is not Related to a Sedentary Lifestyle

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Abstract
The acute onset of chest pain, dyspnea, and decreased exercise tolerance is of significant concern. When this occurs in a teenage patient who was previously considered healthy requires further extensive evaluation. It is important to restrict any and all physical activity until a full cardiorespiratory evaluation has been completed along with laboratory evaluation. The aforementioned symptoms in an adolescent patient requires evaluation for congenital heart disease with thorough evaluation of the patient’s family history. An electrocardiogram may identify non-specific pathology that would guide and confirm the need for further diagnostic testing and evaluation. It is imperative to discuss and explain with the patient and parents what the concerns are, and the need for restricted physical activity over the course of the evaluation. Young athletes will have the desire to continue with their sport, but as the healthcare provider you must ensure that the patient and parents understand the danger that may be associated including sudden cardiac death.

Keywords: Ebstein’s anomaly; Tricuspid regurgitation; Tricuspid valve annuloplasty; Atrioplasty; Foramen ovale; Atrialized right ventricle

Introduction
Ebstein’s anomaly is a rare congenital abnormality that affects the tricuspid valve and right ventricle of the heart, with consequential functional and structural disturbances of the right atrium. This can cause cyanosis (a blue tint to the skin caused by oxygen-poor blood). Lithium or benzodiazepines are the drugs which can be useful for the treatment of cyanosis.

Case Report
A 17-year-old female high school basketball player presented to her primary care physician with complaints of dyspnea, fatigue, and chest pain of sudden onset while playing basketball. She had also experienced lightheadedness with exertion. She has noticed decreased exercise tolerance with episodes of pre-syncope. A similar episode occurred two years prior, but was never evaluated. Due to these symptoms she has noticed a decrease in her level of performance. ECG in the clinic identified a sinus bradycardia at 57 with T-wave inversion in leads III and aVF, with an incomplete right bundle branch block (Figure 1). Patient was restricted from any further physical activity until echocardiogram and pediatric cardiology evaluation was completed. Echocardiogram demonstrated an Ebstein’s Anomaly which was initially treated conservatively with metoprolol. Exercise stress test was unremarkable.

Cardiac MRI was completed to further characterize the nature and extent of the anomaly. The MRI clearly identified an enlarged right atrium measuring 50 × 41 mm, a moderately dilated right ventricle with an end diastolic volume of 160 ml/m², and apical insertion of the tricuspid leaflet (Figure 2).

Unfortunately, she continued to experience symptoms with conservative management and was referred to a cardiothoracic surgeon. It was determined that surgical intervention was needed resulting in a tricuspid valve repair with patch augmentation of the anterior leaflet using autologous pericardium, plication of the atrialized right ventricle, inferior tricuspid valve annuloplasty, subtotal closure of patent foramen ovale, and a right reduction atriotomy. She did recover well with no complications. Warfarin was started and metoprolol was discontinued. She was cleared to return to physical activity including playing basketball.

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Discussion

Ebstein’s anomaly is a congenital disorder that affects 1 in 200,000 live births each year. This anomaly leads to malformation of the tricuspid valve and right ventricle. The septal and posterior leaflets of the tricuspid valve become adherent to the myocardium leading to tricuspid regurgitation. Downward displacement of the functional annulus with resulting atrialization of a portion of the right ventricle is observed. Dilatation of the right atrioventricular junction is seen with tethering of the anterior leaflet of the tricuspid valve. This may be the result of a poorly formed anterior leaflet, or short and poorly formed chordae tendineae. The end result being a leaflet that is displaced into the right ventricular outflow tract.

Ebstein’s anomaly leads to a functional disturbance of the right ventricle with right atrial dilatation as a result of tricuspid regurgitation. The atrialized portion of the right ventricle acts as a reservoir leading to decreased right ventricular outflow. These patients often present with cyanosis, right side heart failure, chest pain, dyspnea, arrhythmias, and sudden cardiac death.

Echocardiography is the diagnostic test of choice demonstrating apical displacement of the septal leaflet, tethering of the anterior leaflet, dilatation of the right atrium, and arterialization of the right ventricle. Echocardiogram may demonstrate a peaked and wide P-wave indicating right atrial enlargement along with evidence of an incomplete or complete right bundle branch block. Chest radiograph may demonstrate a normal cardiac silhouette versus a globe-shaped heart [1-3].

Asymptomatic patients with New York Heart Classification I and II can be treated conservatively. Right heart failure requires a low salt diet, preload reduction, adjusting heart rate and restricting exercise. This may be accomplished through use of diuretics, low dose angiotensin converting enzyme inhibitors, and beta-blockers. Surgery is indicated in the setting of right heart dilation and progressive right ventricular impairment. Surgical repair includes biventricular repair consisting of reconstruction of a competent monocuspid tricuspid valve, right ventriculorraphy, subtotal ASD closure, and reduction atrioplasty. A 1.5 ventricular repair is indicated in patients with poor right ventricular function. Severe left ventricular dysfunction with Ebstein’s anomaly requires heart transplantation [4,5].

Conclusion

An electrocardiogram may identify non-specific pathology that would guide and confirm the need for further diagnostic testing and evaluation. It is imperative to discuss and explain with the patient and parents what the concerns are and the need for restricted physical activity over the course of the evaluation. Young athletes will have the desire to continue with their sport, but as the healthcare provider you must ensure that the patient and parents understand the danger that may be associated including sudden cardiac death.

References