

A Rare Case of Unicuspid Aortic Valve with Postoperative Heart Block

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Abstract

Unicuspid aortic valve (UAV) is an extremely rare cause of aortic stenosis, accounting for about 0.02% of the adult population. We present a rare case of UAV in a young woman who presented with dyspnea on exertion. She underwent extensive work-up, including a transthoracic echocardiogram, which was notable for critical aortic stenosis with significant pulmonary hypertension. A subsequent transesophageal echocardiogram confirmed the severe degree of aortic stenosis and revealed the possibility of a UAV. She was referred to a cardiothoracic surgeon and underwent bioprosthetic aortic valve replacement. Intraoperative evaluation confirmed the rare occurrence of a UAV. Postoperative course was complicated by complete heart block necessitating pacemaker placement.

Categories: Cardiac/Thoracic/Vascular Surgery, Cardiology

Keywords: congenital, aortic stenosis, unicuspid aortic valve, pacemaker

Introduction

Aortic stenosis is the abnormal narrowing of the aortic valve outlet. It is one of the most common valvular lesions, with an estimated prevalence of 2%-9% in the general adult population above the age of 65 years [1]. Unicuspid aortic valve (UAV) is an extremely rare cause of aortic stenosis, accounting for 0.02% of the adult population [2]. We present a case of UAV in a young woman who eventually underwent successful bioprosthetic aortic valve replacement.

Case Presentation

We report a case of a 28-year-old woman who presented to the office with complaints of progressive exertional dyspnea of three months duration. Of note, she was diagnosed with congenital aortic valve stenosis at birth and underwent balloon valvuloplasty as a neonate. She was followed by a pediatric cardiologist through her childhood and reported no difficulty with moderate-to-strenuous activity as an adolescent. However, as an adult, she spent several years of her life outside the country and was unable to follow up with a cardiologist during that interim period. On return to the States, she re-established care in light of her ongoing symptoms. She reported severe dyspnea on exertion with occasional lightheadedness and palpitations. A review of systems was negative except the aforementioned symptoms. On exam, she had an ejection systolic murmur with a click, best heard at the apex, and radiating to her neck.

Electrocardiography (Figure 1) was unremarkable while a transthoracic echocardiogram (TTE) (Figures 2 and 3) was notable for critical aortic stenosis (valve area 0.5 cm², mean gradient 40 mm of Hg) with significant pulmonary hypertension (pulmonary artery pressure 80-84 mm Hg systolic). Both ventricles appeared grossly normal with an estimated ejection fraction 55%. We consulted an adult congenital heart disease specialist and performed a transesophageal echocardiogram (TEE). The TEE confirmed the degree of aortic stenosis and indicated the possibility of a UAV.

She was referred to a cardiothoracic surgeon and underwent bioprosthetic aortic valve replacement. The intraoperative evaluation confirmed the rare occurrence of a UAV. However, the postoperative course was complicated by complete heart block with pacemaker placement.

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Human subjects: Consent was obtained by all participants in this study. The Wright Center for Graduate Medical Education Institutional Review Board (WCGME-IRB) issued approval n/a. DATE: May 30, 2019 TO: Divya Ravi, MD MPH FROM: The Wright Center for Graduate Medical Education Institutional Review Board (WCGME-IRB) PROJECT TITLE: [1442756-1] A Rare Case of Unicuspid Aortic Valve SUBMISSION TYPE: New Project ACTION: DETERMINATION OF EXEMPT STATUS DECISION DATE: May 30, 2019 REVIEW CATEGORY: Exemption category # 4 Thank you for your submission of New Project materials for this project. The Wright Center for Graduate Medical Education Institutional Review Board (WCGME-IRB) has determined this project is EXEMPT FROM IRB REVIEW according to federal regulations. We will retain a copy of this correspondence within our records. If you have any questions, please contact Kathleen Uhranowsky at 570-591-5197 or uhranowskyk@thewrightcenter.org. Please include your project title and reference number in all correspondence with this committee. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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