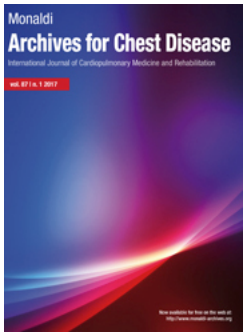

Monaldi Archives for Chest Disease

eISSN 2532-5264 https://www.monaldi-archives.org/

Monaldi Arch Chest Dis 2023 [Online ahead of print]

To cite this Article:
Avesani M, Sabatino J, Sirico D, Di Salvo G. **Unusual case of severe aortic regurgitation in a child with bicuspid aortic valve.** *Monaldi Arch Chest Dis* doi: 10.4081/monaldi.2023.2713

Unusual case of severe aortic regurgitation in a child with bicuspid aortic valve

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Abstract

Aortic regurgitation (AR) is common after aortic balloon valvuloplasty in children and it has been associated with large balloon/annulus ratio, abnormal valve morphology and aortic valve prolapse. We present a rare case of severe AR after aortic balloon valvuloplasty due to aortic strands rupture, causing prolapse of the co-joined cusp. These findings were identified by 3D echocardiography, highlighting the importance of this imaging technique even in pediatric patients.

Key words: aortic strands; 3D echocardiography; aortic regurgitation.

Introduction

Aortic regurgitation (AR) is one of the possible complications of aortic balloon valvuloplasty in pediatric patients. The progression from mild to moderate/severe AR seems to occur at a steady rate over time, with 65% freedom from moderate or severe AR at 5-year follow-up reported in some studies [1,2]. Valve morphology and intra-operative factors, such as large balloon/annulus ratio have been associated with the development of AR [3]. Here, we present a rare case of severe AR after aortic balloon valvuloplasty in a pediatric patient, due to aortic strands rupture.

Case Report

We describe the case of a child with severely stenotic bicuspid aortic valve (BAV) who underwent aortic balloon valvuloplasty and developed significant AR. She came to our attention because of an abnormal structure identified in the ascending aorta during an echocardiographic control. We performed two-D echocardiogram, which showed severe AR, with a mobile structure protruding into the left ventricle outflow tract (Figure 1A, green arrow) and confirmed the presence of an hyperechogenic structure at the level of the ascending aorta (Figure 1B, yellow arrow). To better investigate the nature of this finding, three-D echocardiography was performed, and it allowed the visualization of ruptured aortic strands, causing prolapse of the co-joined cusp (Figure 2, green arrows and yellow stars).

Discussion

The knowledge about aortic fibrous strands is sparse. They are described both in patients with BAV and tricuspid AV and are more common in Asian countries and males [4-6].

Since during embryogenesis aortic valve comes from bulges projecting towards the ventricle, it has been hypothesized that fibrous tissue persists between aortic valve and aortic wall [4,6], maybe with function of valve suspension [1]. In addition, autoptic data on fenestrations in aortic valves showed the presence of a network of strands also connecting fenestrations among semilunar cusps [7].

AR due to fibrous strands rupture is rare and it can involve strands connecting fenestrated cusps [8], or strands between the aortic wall and the cusps [6,9]. In the latter situation, fibrous strands rupture has been described in the right or right and noncoronary cusps mostly [10]. The cases reported in the literature occurred in adults or elderly patients, where hypertension and degeneration of tissues can contribute to the process [4-6,9,11].

AR is common after aortic balloon valvuloplasty in children and it has been associated with large balloon/annulus ratio, abnormal valve morphology and aortic valve prolapse [4,12]. By contrast, to the best of our knowledge, no case of AR post balloon valvuloplasty has been associated to strands rupture in pediatric patients.

Conclusions

We reported that, even if rare, post procedural rupture of fibrous stands can contribute to the mechanism of AR post balloon valvuloplasty. 3D echo could be a valid technique for the identification of these structures and to help clinicians to understand AR mechanisms, even in children.

Ethics approval and consent to participate: No ethical committee approval was required for this case report by the Department, because this article does not contain any studies with human participants or animals. Informed consent was obtained from the patient included in this study.

Consent for publication: The manuscript does not contain any individual person's data in any form.

Conflict of interest: The authors declare that they have no competing interests, and all authors confirm accuracy.

Authors' contribution: MA, JS: conceptualizing and writing; DS: images analysis and writing; GDS: supervision and final revision. All the authors read and approved the final version of the manuscript and agreed to be accountable for all aspects of the work.

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Figure 1. 2D echocardiogram showing severe aortic regurgitation (A), and the mobile structure protruding into the left ventricle outflow tract (green arrow), and confirming (B) the hyperechogenic structure at the level of the ascending aorta (yellow arrow).

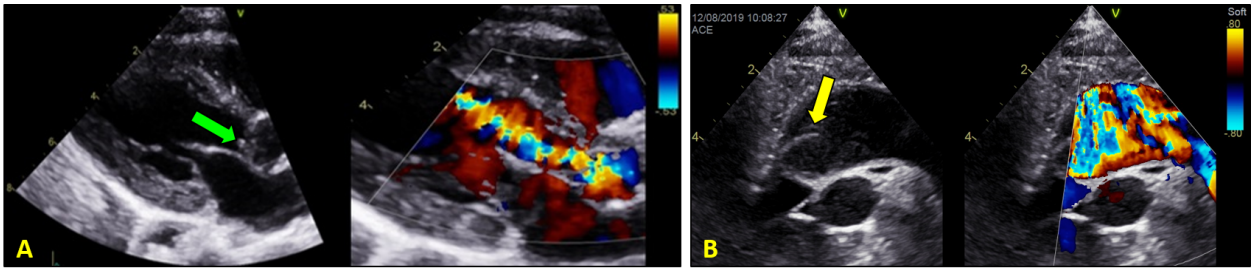

Figure 2. 3D echocardiography visualizing the ruptured aortic strands.