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.

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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 the 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 a 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 a 2D 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 a hyperechogenic structure at the level of the ascending aorta (Figure 1B, yellow arrow). To better investigate the nature of this finding, 3D 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

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, the aortic valve comes from bulges projecting towards the ventricle, it has been hypothesized that fibrous tissue persists between the aortic valve and the aortic wall [4,6], maybe with the 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].

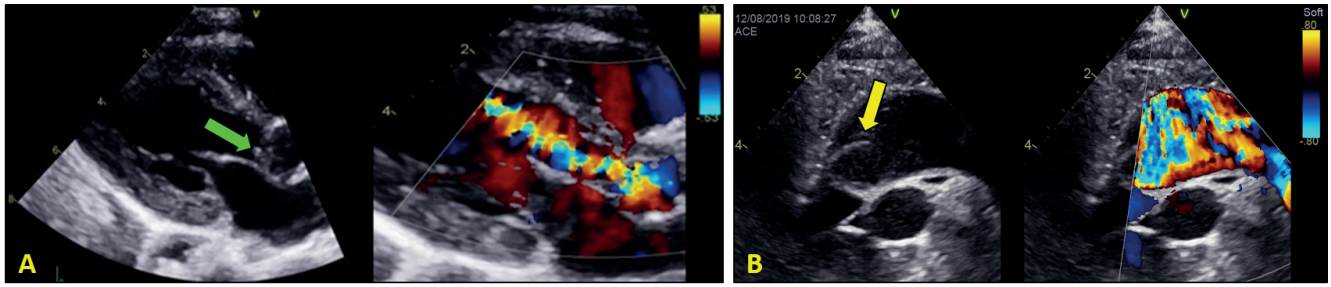

Figure 1. 2D echocardiogram showing severe aortic regurgitation (A), and the mobile structure protruding into the left ventricle outflow tract (green arrow) and confirming the hyperechogenic structure at the level of the ascending aorta (yellow arrow) (B).

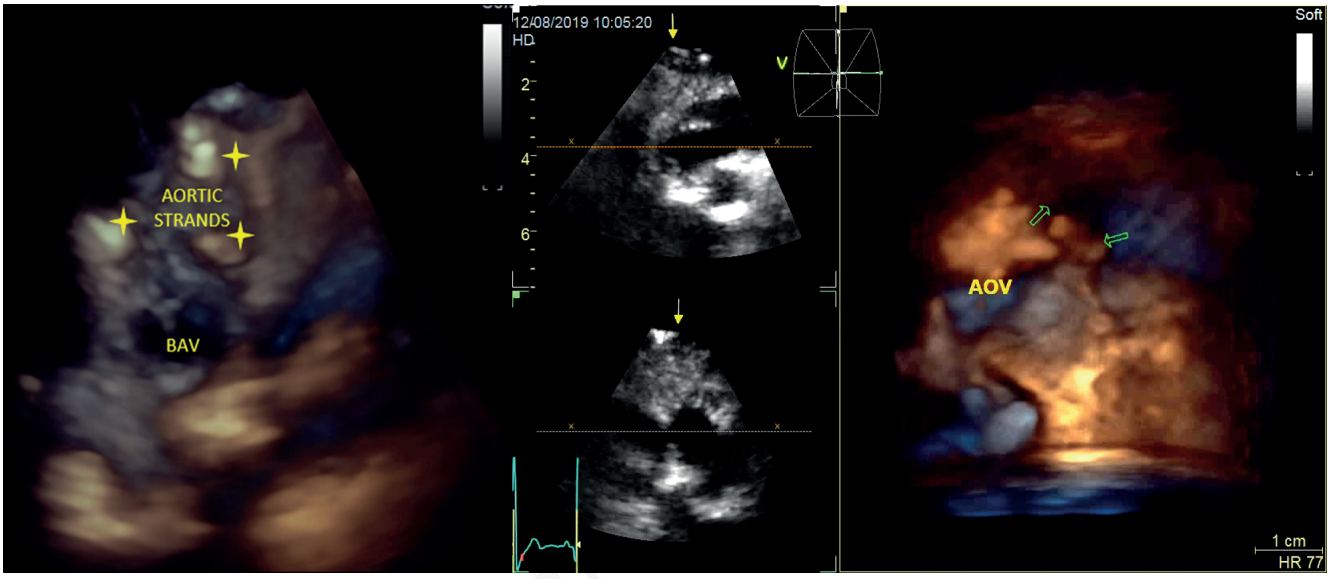

Figure 2. 3D echocardiography visualizing the ruptured aortic strands.

AR due to fibrous strands rupture is rare and 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 mostly described in the right and noncoronary cusps [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 with strands rupture in pediatric patients.

Conclusions

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

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