Rupture of Aneurysm of Valsalva Sinus and Aortopathy in Bicuspid Aortic Valve: A Case Report and Review of Literature

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Introduction
The bicuspid aortic valve (BAV) is one of the most common forms of congenital heart defects. It is a complex entity, in which involvement of the thoracic aorta forms part of the disease spectrum. The ascending aorta is the most common site involved in this process. We report the extremely rare case of a patient with true BAV associated with a diffuse aortopathy, involving the aneurysm of the non coronary sinus, that suddenly rupted into the right atrium and required urgent cardiac surgery, and the aneurismal dilation of a (non aberrant) left subclavian artery, resembling a Kommerell’s diverticulum.

Case Report
We present the case of 58 years-old patient who came to our attention for acute onset of chest pain, dyspnea and...
The bicuspid aortic valve (BAV) is one of the most common form of congenital heart defects. The clinical manifestations usually relate to function of the aortic valve and acquired complications such as endocarditis. Although much of the original interest has focused on abnormal bileaflet valve, BAV is a more complex entity, whose involvement of the thoracic aorta forms part of the disease spectrum. The most common comorbidity is dilation of aorta. BAV is also associated with other congenital vascular (in particular coarctation of aorta) and cardiac defects (i.e. hypoplastic left ventricle, reversal of dominance of coronary artery, atrial septal defect, ventricular septal defect). This suggests a more global development disorder. A genetic predisposition has been reported, and in familial cases, an autosomal dominant inheritance with variable penetrance appears to be advocated. We report the extremely rare case of a patient with true BAV associated with a diffuse aortopathy, involving the aneurysm of the non coronary sinus, that suddenly rupted into the right atrium and required urgent cardiac surgery, and the aneurysmal dilation of a (non aberrant) left subclavian artery, resembling a Kommerell’s diverticulum. In the emergency care unit, TEE easily identified the aortic-to-right atrium fistulous tract in the life-threatening setting of an acute right failure and allowed the prompt choose of the appropriate repair strategy. CT (although carrying disadvantages of irradiation and use of iodine contrast media) due to its availability, short scanning time and high spatial resolution, is of great value in evaluation and follow-up of patients with suspected extensive aortic pathology due to the diagnosis of bicuspid aortic valve.

Declarations of Interest
The authors declare no conflicts of interest.

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The authors state that they abide by the “Requirements for Ethical Publishing in Biomedical Journals” [10].
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