ESC European Society of Cardiology

European Heart Journal - Case Reports (2024) **8**, 1–5
https://doi.org/10.1093/ehjcr/ytae097

CASE REPORT
Cardiovascular imaging

Unicuspid aortic valve by cardiac computed tomography: the best view is from the mountaintop—a case report

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Received 2 September 2023; revised 12 February 2024; accepted 15 February 2024; online publish-ahead-of-print 19 February 2024

Background Unicuspid aortic valve (UAV) is a rare valvular heart disease and a challenging diagnosis. Advanced imaging techniques, particularly cardiac computed tomography (CT), appear to be invaluable tools to correctly identify this disease pre-operatively, as this may have an impact on the optimal surgical treatment.

Case summary We describe the case of a young patient admitted with heart failure, due to a severely stenotic UAV. Cardiac CT allowed adjusting the imaging plane to the best view in two orthogonal planes to identify the top of the 'dome' and to accurately measure the smallest valve opening by planimetry. Surgical inspection confirmed a rare case of acommissural UAV.

Discussion Cardiac CT angiography is crucial to understand the complexity of UAV disease and to differentiate the acommissural from the unicommissural type. Accurate positioning of the imaging plane through the smallest valve opening in systole reduces the risk of missing the diagnosis of this rare disease.

Keywords Case report • Unicuspid aortic valve • Cardiac CT angiography • Valvular heart disease

ESC curriculum 2.4 Cardiac computed tomography • 2.1 Imaging modalities • 2.2 Echocardiography • 4.2 Aortic stenosis • 9.7 Adult congenital heart disease

Learning points

A young patient presented with severe aortic stenosis secondary to a unicuspid aortic valve (UAV).

- To understand the morphology of UAV.
- To be able to differentiate the acommissural type from the unicommissural type of UAV.
- To emphasize the importance of cardiac computed tomography angiography to fully explore the UAV.

Introduction

Bicuspid aortic valve (BAV) is the most common congenital heart defect, with a prevalence rate estimated between 0.5 and 2%. In contrast, the prevalence rate of unicuspid aortic valve (UAV) is far lower (around 0.02%) with a male-to-female ratio of 4/1.[1–3] As it is a rare condition, the diagnosis can be challenging. Herein, we describe the case of a young patient admitted with heart failure, due to a severely stenotic UAV. This case highlights the importance of cardiac computed tomography (CT) to differentiate the acommissural type from the unicommissural type of UAV.

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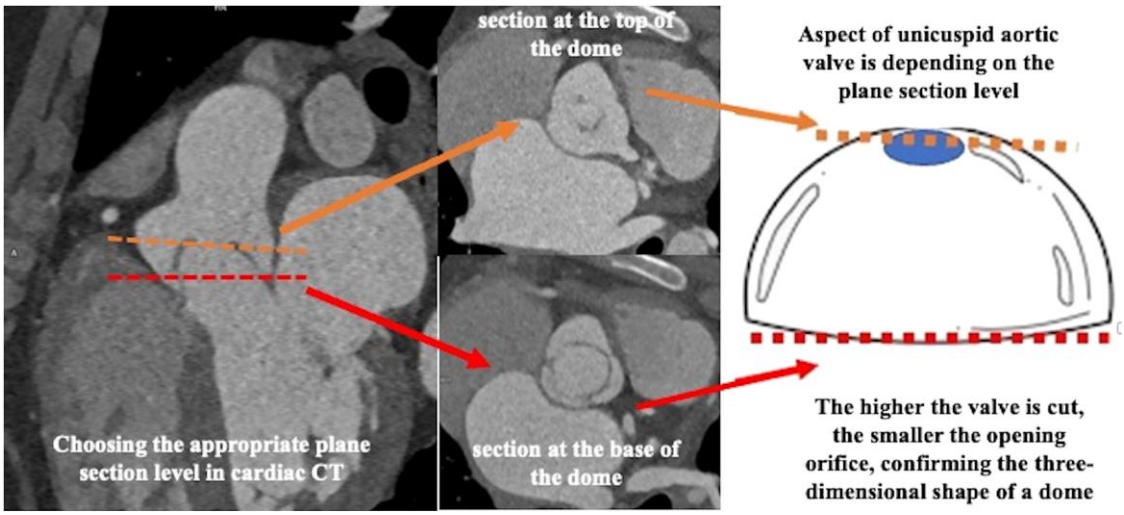
Handling Editor: Matthew Williams
Peer-reviewers: Sarv Priya; Enrique Garcia-Sayan
Compliance Editor: Nikolaos Spinthakis

Summary figure



Case presentation

A 23-year-old man was referred to our hospital with progressive dyspnoea on exertion over a course of several months. In his past medical history, we found that he was operated upon in childhood for an aortic coarctation with concomitant closure of a ventricular septal defect. A mildly stenotic aortic valve (AV, initially labelled as BAV) and a parachute mitral valve were also diagnosed in childhood. He was lost to cardiologic follow-up since the age of 12.

On admission, his blood pressure was 100/65 mmHg, heart rate 105 b.p.m., and oxygen saturation 96% on room air. A physical examination revealed a Grade 4/6 systolic ejection murmur at the right upper sternal border and a diminished S_2. The electrocardiogram was remarkable for sinus tachycardia with repolarization abnormalities. Transthoracic echocardiography (TTE) showed a dilated left ventricle (LV, end-diastolic volume index: 110 mL/m^2) with moderate LV systolic dysfunction: an ejection fraction of 35%. The AV was not calcified, but its morphology was not clearly seen by TTE. A low-flow/low-gradient severe aortic stenosis was suspected with a cardiac index of 2.3 L/min/m^2, a peak aortic jet velocity (V_{max}) of 3.2 m/s, a mean gradient of 28 mmHg, and a calculated AV area of 0.7 cm^2. At low-dose dobutamine stress echocardiography, the V_{max} increased to 4.1 m/s, the mean gradient rose to 49 mmHg, and the valve area remained at 0.7 cm^2, demonstrating the presence of true severe aortic stenosis.

Diagnostic assessment

Trans-oesophageal echocardiography (TEE) suggested the diagnosis of UAV with an eccentric opening, severe valve stenosis, and mild regurgitation (see Supplementary material online, *Videos S1–S3*).

To better understand the functional anatomy of this non-calcified AV stenosis and to assess the thoracic aorta, a gated cardiac CT was performed. In the diastolic phase, the AV appeared like a BAV, with a fusion of the left coronary and non-coronary cusps, without visible raphe (*Figure 1*). The ascending aorta was not dilated (39 mm), and there was no residual stenosis at the coarctation site. No aspect of hypertrophic cardiomyopathy, sub-aortic membrane, or supravalvular stenosis was found.

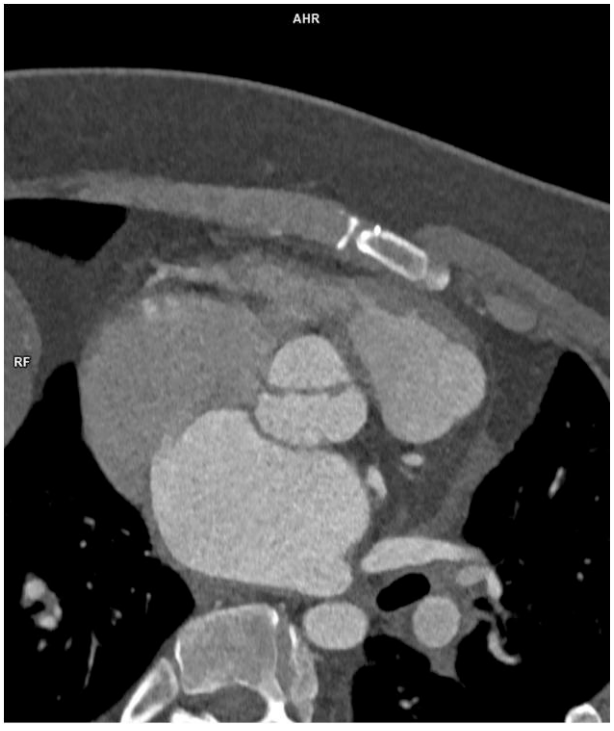


Figure 1 Cardiac computed tomography angiography: a diastolic transverse view of the valve. In diastole, the valve looks like a bicuspid aortic valve with left coronary and non-coronary cusps fusion.

The coronary arteries were normal. Other associated congenital anomalies were ruled out.

In the systolic phase, AV opening was typically like a dome in the sagittal plane (*Figure 2*; Supplementary material online, *Video S4*). In the transverse

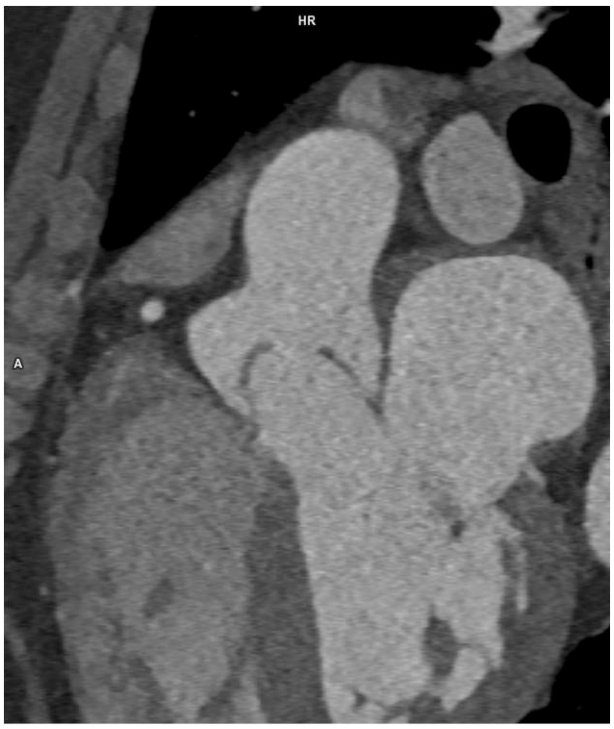


Figure 2 Cardiac computed tomography angiography: a systolic sagittal view of the valve showing the dome shape of the valve.

plane, the valve opening was central and rather circular, without any functional commissure (*Figure 3A*; Supplementary material online, *Video S5*). These findings were consistent with the diagnosis of acommissural UAV. Of note in this case, the CT imaging plane was critical: when the imaging plane passed through the base of the 'dome', the valve opening looked almost normal, with the exception of three tiny raphes (*Figure 3B*). As the section plane is moved towards the top of the dome, the valve opening becomes central, severely reduced, and without any visible raphe. Thus, cardiac CT makes it possible to move the imaging plane to the best view in two orthogonal planes to accurately measure the smallest valve opening by planimetry.

Interventions

The patient underwent surgical AV replacement with a mechanical (bi-leaflet) AV, based on his informed treatment choice. Surgical inspection confirmed a UAV without any functional commissure but with three thin raphes (*Figure 4*). The post-operative course was uneventful.

Follow-up

Surgery led to the resolution of all symptoms. More than 2 years after surgery, the patient is still asymptomatic from the cardiovascular point of view. His bi-leaflet aortic prosthesis is perfectly functioning, without significant regurgitation or stenosis. The left ventricular ejection fraction has improved to 50%.

Discussion

The diagnosis of UAV is challenging. In our case, the use of multi-planar reconstruction by cardiac CT established this diagnosis. Unicuspid aortic

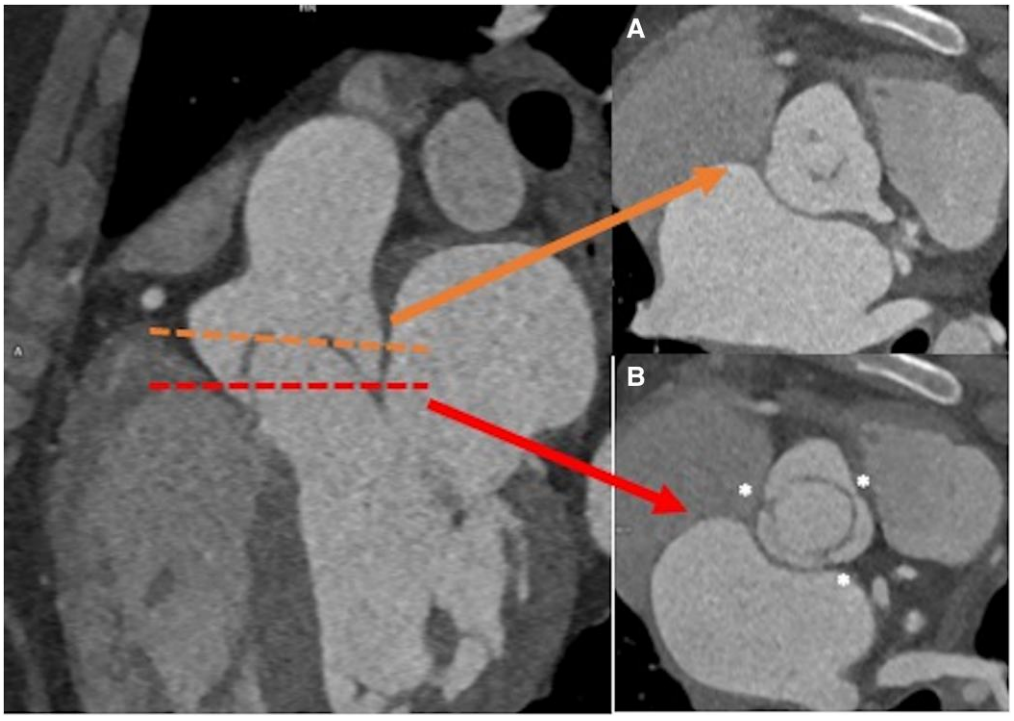


Figure 3 The aspect of valve opening depends on the section plane. The higher the valve is cut, the smaller the opening orifice, confirming the three-dimensional shape of a dome. (*A*) Section at the top of the dome. (*B*) Section at the base of the dome, where three tiny raphes can be seen (*).

valve may be associated with other congenital defects such as an anomalous connection of the coronary arteries, aortic aneurysm, or coarctation. Two types of UAV are commonly described: the unicommissural type and the acommissural type (*Figure 5*).[1] By TEE or cardiac CT, a unicommissural AV has two raphes with an eccentric valve opening limited to the functional commissure, which is 'slit-shaped'. Given the presence of two raphes, the unicommissural AV was labelled as 'Type 2-BAV' in the Sievers and Schmidtke classification, which is controversial.[4] Of note, the unicommissural AV is not so rare: in adult patients with surgically excised stenotic AVs, the estimated prevalence rate of the unicommissural AV is up to 5%.[2] When compared with the acommissural type, the clinical progression of the unicommisural AV is usually slower, and patients may remain asymptomatic until the 3rd to 4th decade of life. An acommissural AV is exceptionally seen in adulthood because it leads to severe aortic stenosis at an early age with the need for surgical correction in childhood. However, 4 cases of acommissural UAV were previously reported in a series of 932 adult patients operated upon for aortic stenosis.[2] It is commonly assumed that the acommissural type has no visible raphe or any functional commissure: valve opening is central and 'pinhole shaped'. Our case supports the fact that three raphes might be present in the acommissural type. The AV, excised in one piece by the surgeon, nicely demonstrates the central 'pinhole' orifice, the lack of any true commissure, and the three raphes in our patient (*Figure 4*). Moreover, our patient's surgical piece is very similar to that of the four latter cases of acommissural AV described by Roberts and Ko.[2] By TEE or cardiac CT, the 'central dome' opening is characteristic of an acommissural AV. It can be imagined like the Dome of the Pantheon in Rome with a very restricted opening at its top (see Supplementary material online, *Figure S1*). Guidelines make no specific mention of the management of UAV patients. However, it is important to consider the therapeutic challenge of this pathology, given the young age of the patients. In the presence of UAV, the most frequently discussed therapeutic options are mechanical valve replacement, surgical valve repair by bicuspidization (a few cases of tricuspidization have also been described), or the Ross procedure.[5] Identifying unicuspid valves prior to surgery is therefore essential, since in some cases, a thorough analysis of the valve anatomy leads to specific repair techniques, with potential outcome improvement.

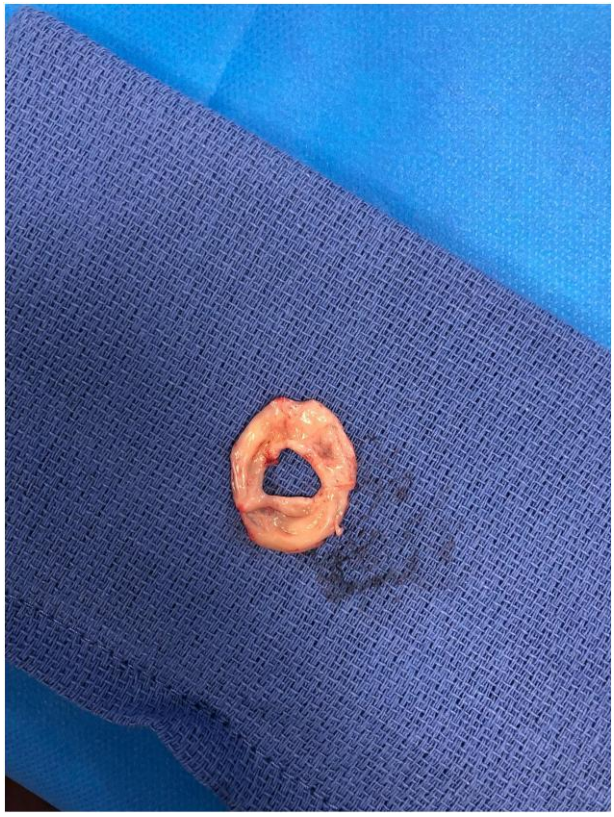


Figure 4 Surgical piece (ventricular aspect). The aortic valve was excised in one piece, confirming the acommissural unicuspid aortic valve where three tiny raphes can be seen.

Conclusion

We present the case of aortic stenosis in a young adult, secondary to an acommissural UAV. Our case highlights the crucial role of advanced

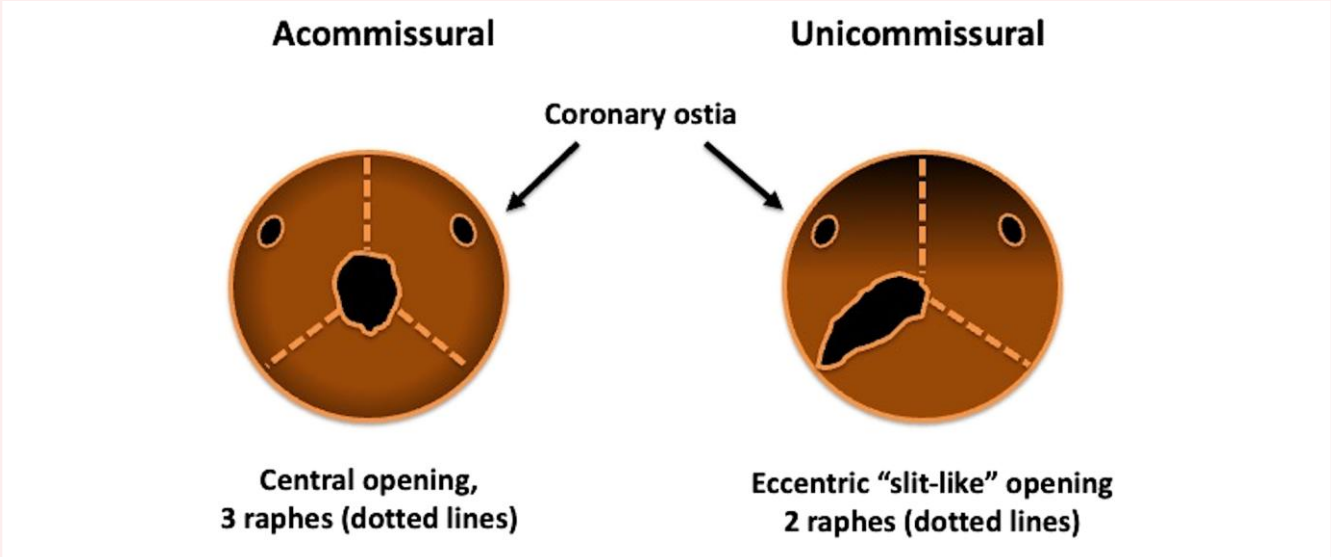


Figure 5 A diagram illustrating the two types of unicuspid aortic valve: acommissural vs. unicommissural.

imaging techniques, particularly cardiac CT: accurate positioning of the imaging plane through the smallest valve opening in systole might reduce the risk of missing the diagnosis of UAV. The importance of correctly identifying this valve disease pre-operatively is emphasized, as this may have an impact on the optimal surgical procedure. Cardiac CT is key to overcome the complexity of UAV and to differentiate the acommissural type from the unicommissural type.

Lead author biography

Julien Rosencher is working in the Catheterization laboratories and in the Cardiovascular Imaging laboratories at the Groupe Hospitalier Privé Ambroise Paré—Hartmann, France. Dr Rosencher has a strong interest in clinical education with mentorship in both laboratories. He is also involved in the French liberal cardiologist association (CNCF).

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Consent: Informed consent for patient information and images to be published was provided by the patients in accordance with COPE guidelines.

Conflict of interest: None declared.

Funding: None declared.

Ethics approval: This material is the authors' own original work, which has not been previously published elsewhere.

Data availability

The numerical data and all images used to support the findings of this case series report are included within this article.

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