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CASE REPORT
Congenital heart disease

Unicuspid aortic valve replacement with development of complete heart block: a case report

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Background Aortic stenosis is a progressive disease that frequently remains undiagnosed until late in the disease course. In patients that present with symptoms of heart failure and a systolic murmur at a young age, a congenital valvular abnormality must be on the differential. With patients that have accelerated symptoms of aortic stenosis and valvular dysfunction, a unicuspid aortic valve (UAV) could be present. A UAV is often difficult to distinguish from a bicuspid aortic valve (BAV) on transthoracic echocardiography. In patients with congenital valvular abnormalities an ascending aortic aneurysm can also be present. Aortic stenosis changes the jet of fluid emerging from the aortic valve leading to an increased risk for aortic aneurysm dissection and rupture. The gold standard treatment for aortic stenosis secondary to a congenital valvular abnormality is valve replacement. A known risk of aortic valve replacement is conduction abnormalities. In this case, we present a patient with a unicuspid valve who postoperatively develops complete heart block leading to pacemaker implantation.

Case summary We present a case of a 46-year-old Caucasian male with no prior medical history who presented with progressively worsening exertional dyspnoea and palpitations for 7 months. Transthoracic echocardiogram showed a BAV, however, further work up confirmed a unicommissural aortic valve with severe aortic stenosis and moderate regurgitation along with an ascending aortic aneurysm. Aortic valve replacement and aortic aneurysm repair via the Bentall procedure was successfully completed with postoperative course being complicated by a complete heart block and subsequent permanent pacemaker placement.

Discussion When assessing patients with symptoms of heart failure with a systolic murmur that suggests aortic stenosis at a young age, a UAV must be kept on the differential. The symptoms of aortic stenosis and valvular dysfunction are accelerated in UAVs when compared with BAVs. Currently, the treatment for patients with congenital valvular abnormalities presenting with aortic stenosis is aortic valve replacement using traditional open surgery. A known sequelae of isolated aortic valve replacement is conduction abnormalities that can sometimes lead to permanent pacemaker placement. After the confirmation of unicuspid or bicuspid valve postoperatively, it is important to report any postoperative conduction abnormalities. This is because, currently, there is no literature that compares the incidence of conduction abnormalities after unicuspid replacement to that of other BAV syndromes.

Keywords Unicuspid aortic valve • Complete heart block • Ascending aortic aneurysm • Transthoracic echocardiography • Permanent pacemaker • Congenital valvular abnormality • Case report

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Learning points

- In patients that present with symptoms of heart failure and a systolic murmur at a young age, a congenital valvular abnormality such as a unicuspid or bicuspid aortic valve (BAV) must be on the differential.
- A unicuspid aortic valve is often difficult to distinguish from a BAV on transthoracic echocardiography and can be better diagnosed by transoesophageal echocardiogram.

Introduction

Aortic stenosis is a progressive disease that frequently remains undiagnosed until late in the disease course. In patients that present with symptoms of heart failure and a systolic murmur at a young age, a congenital valvular abnormality must be on the differential. With patients that have accelerated symptoms of aortic stenosis and valvular dysfunction, a unicuspid aortic valve (UAV) could be present. A UAV is an extremely rare valvular abnormality with it only being found in 0.02% of the adult population when compared with bicuspid aortic valves (BAVs) that are found in about 1–2% of the adult population.[1,2] Accompanying ascending aortic aneurysms can be a common non-valvular abnormality found in patients with aortic stenosis secondary to a congenital aortic valve abnormality.[3] Aortic stenosis changes the jet of fluid emerging from the aortic valve leading to an increased risk for aortic aneurysm, dissection, and rupture. A known sequelae of isolated aortic valve replacement is conduction abnormalities that can sometimes lead to permanent pacemaker placement. Currently, there is little literature about the complications of UAV replacement compared with that of bicuspid. In this case report, we will discuss a unique case of a UAV (*Figure 1*) with successful aortic valve replacement and an ascending aortic aneurysm repair via the Bentall procedure with the postoperative course being complicated by a complete heart block and subsequent permanent pacemaker placement.

Timeline

Initial presentation	Presented with chief complaints of 7 months of dyspnoea and fatigue.
1-month prior to operation	Transthoracic echocardiogram is ordered, and patient is preliminary diagnosed with aortic stenosis due to a bicuspid aortic valve with additional aortic root dilation.
	Computed tomography angiogram was completed showing a 5.5 cm ascending aortic aneurysm at the sinotubular junction.
Operation	Patient underwent successful Bentall procedure for replacement of aortic valve and aorta repair. Patient was confirmed to have a unicuspid aortic valve.
Postoperative Day 1	Patient was found to be in junctional rhythm.
	Temporary pacemaker was set to pace at a heart rate of 70 b.p.m.
Postoperative Day 5	Patient was diagnosed with complete heart block.
	Permanent pacemaker was placed.
Postoperative Day 9	Patient was discharged.

Case presentation

A 46-year-old Caucasian male presented with occasional palpitations and shortness of breath with exertion for 7 months. The patient denied any past medical history and had not seen a physician since age 16. The patient's vitals on presentation was a heart rate of 75 b.p.m., with a blood pressure of 135/95. Physical exam was positive for a Grade II/IV systolic ejection murmur and delayed upstroke of the peripheral pulses. The patients point of maximum apical impulse was laterally displaced and on chest auscultation there were faint bibasilar crackles. Initial electrocardiogram on presentation was sinus rhythm with a heart rate of 90 b.p.m., right bundle branch block present with a QRS duration of 126 ms, and no left ventricular (LV) hypertrophy by voltage criteria. The patient underwent a transthoracic echocardiogram (TTE) and during the original report was found to have a BAV with aortic stenosis. His echocardiogram revealed mild LV hypertrophy with mildly reduced LV end-diastolic dimension. The patient was found to have a preserved ejection fraction of 61% and a peak/mean gradient of 42.53 mmHg/24.14 mmHg across his aortic valve with an aortic valve area (AVA) of 1.1 cm^2 and AVA index to be 0.56 cm^2/m^2. LV inner diastolic diameter was measured to be 3.8 cm with the interventricular septal diastolic thickness of 1.4 cm and a LV posterior wall diastolic thickness of 1.3 cm. Additionally, his two-dimensional echo showed mild to moderate aortic regurgitation with a pressure half time of 547 ms as well as a >5 cm aortic aneurysm. *Figure 2* below shows a highly calcified aortic valve, which at the time was interpreted to be a BAV. *Figure 3* notes the aortic valve stenosis with a dilated aortic root. Additionally, the patient underwent dobutamine stress echo that showed no areas of stress induced ischaemia. These findings were unique in this patient

had a reverse area gradient mismatch for the diagnosis of aortic stenosis, which will be discussed later.

A computed tomography (CT) angiography was requested to assess the size and location of the patient's aortic aneurysm. He was found to have a 5.55 cm anterior to posterior diameter ascending thoracic aneurysm at the sinotubular junction with normal caliber aortic arch and patent branching vessels and no obvious coronary anomalies (*Figure 4*). Subsequent myocardial perfusion imaging revealed no definite focal areas of pharmacologically induced stress ischaemia with LV ejection fraction of 52%.

At this time, a decision was made to proceed with an aortic valve replacement and aortic aneurysm repair via the Bentall procedure. This recommendation was supported by both the 2017 ESC/EACTS and the 2014 AHA/ACC valvular heart guideline, as well as the 2016 update to the thoracic aortic disease guidelines both being Class IIa level C evidence recommendation for tube graft repair for ascending aortic aneurysm with congenital valvular abnormalities.[4] Of note the patient did not undergo three-dimensional transoesophageal echocardiogram (TOE) before surgery.

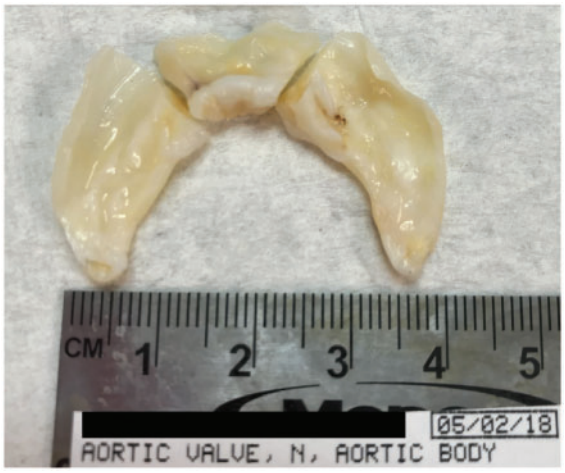

Figure 1 Segments of the unicuspid valve with a single commissure between the left and non-coronary cusps and two raphes between the region of the normal left and right commissures and between the region of the normal right-non-commissure.

The patient underwent surgery with the placement of a 25 mm On-X valve as well as a 32 mm Gelweave Dacron tube graft. Upon removal of patients native aortic valve, he was found to have a UAV. During the surgery, the patient's anatomical coronary placement was assessed and found to be normal. There were no surgical complications and the patient was transferred to the cardiovascular intensive care unit for recovery. During the surgery, the patient prophylactically had a temporary epicardial pacemaker placed and in recovery was found to be in an accelerated junctional escape rhythm with a high degree atrioventricular block (*Figure 5*) and occasional heart rates in the 40's. Temporary pacing was set to occur if the patient's heart rate dropped below 70. Over the next few days, the patient's junctional rhythm did not improve, and it was determined that the patient had postoperative complete heart block with need for a permanent pacemaker. Permanent pacemaker was placed on postoperative Day 5, and patient was discharged on postoperative Day 9.

Discussion

This case report is unique as it carries several important teaching points. (i) A UAV is a rare congenital abnormality that can be easily missed or confused with a BAV. (ii) The treatment for aortic stenosis due to congenital valvular abnormalities such as UAV's and BAV's can have post-procedural complications.

UAV's are a rare valvular abnormality found in around 0.02% of the general population.[1,2] Although this type of congenital malformation is rare in the general population, it is found in 4–6% of the population who undergoes isolated aortic valve replacement.[5] Patients with UAV's often present with dyspnoea due to aortic stenosis as

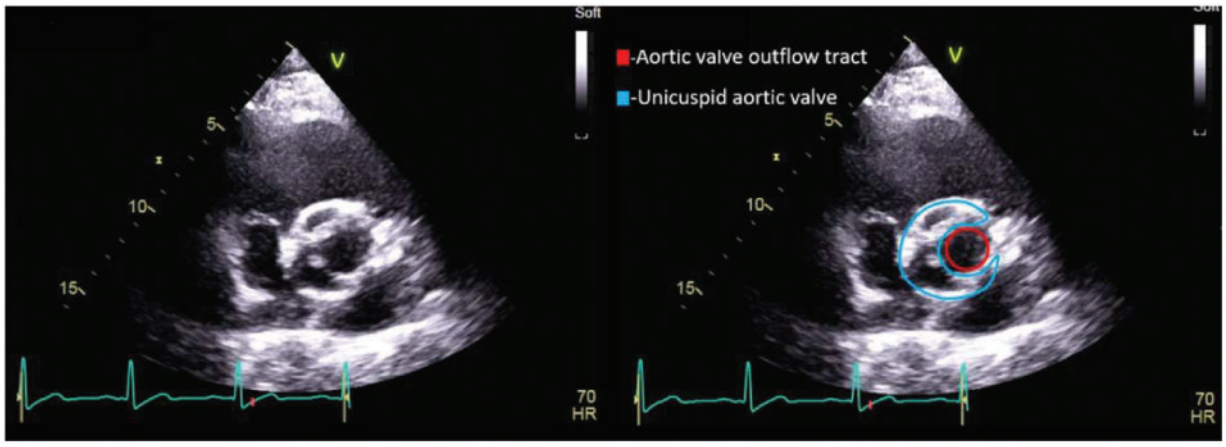

Figure 2 Transthoracic echocardiogram parasternal short-axis view demonstrating a unicuspid aortic valve.

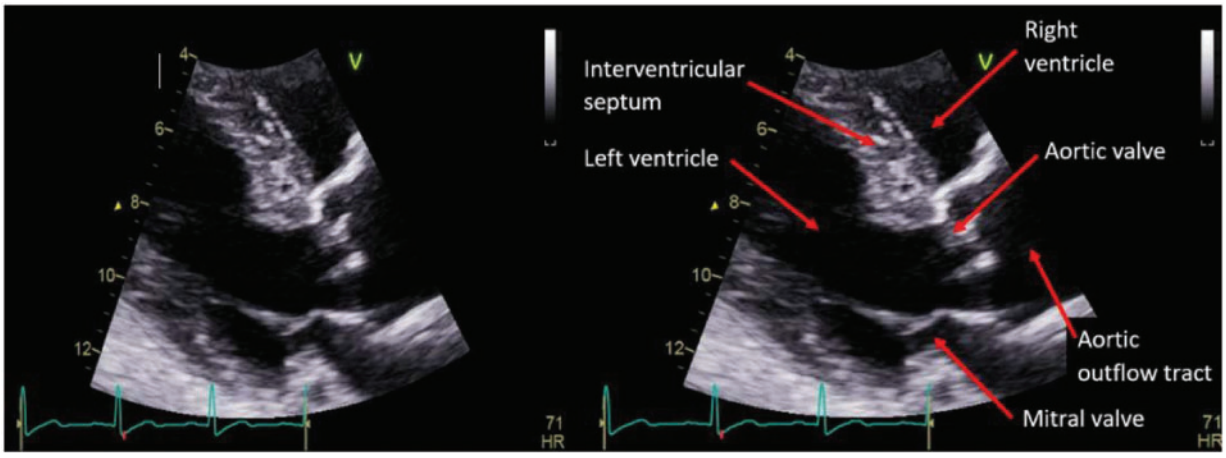

Figure 3 The anatomy of the sclerotic aortic valve as well as a dilated aortic outflow tract.

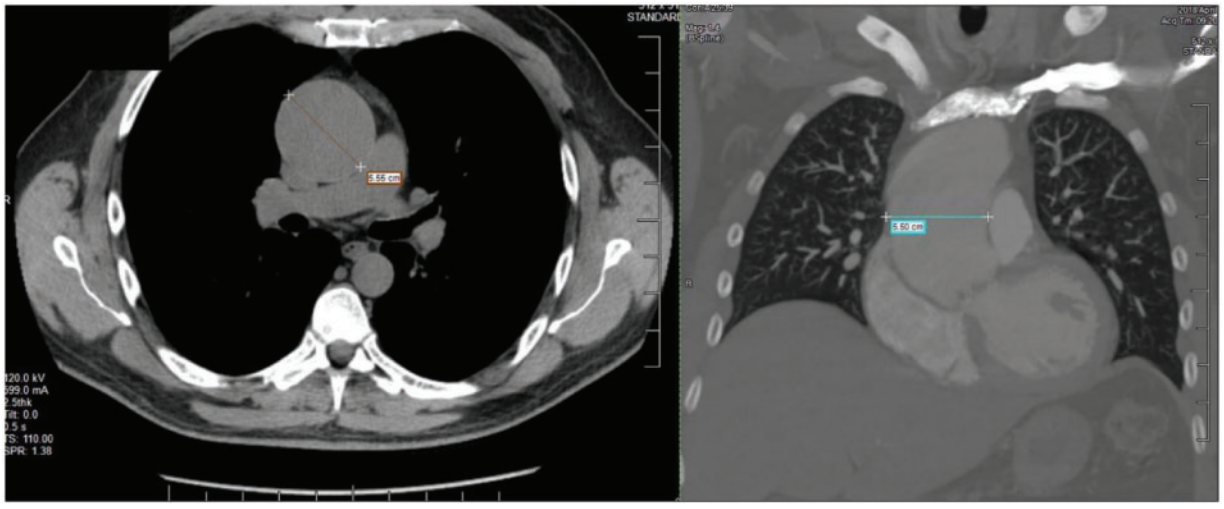

Figure 4 Computed tomography angiogram showing a 5.55 cm ascending aortic aneurysm at the sinotubular junction as well as a sagittal view of the ascending aortic aneurysm.

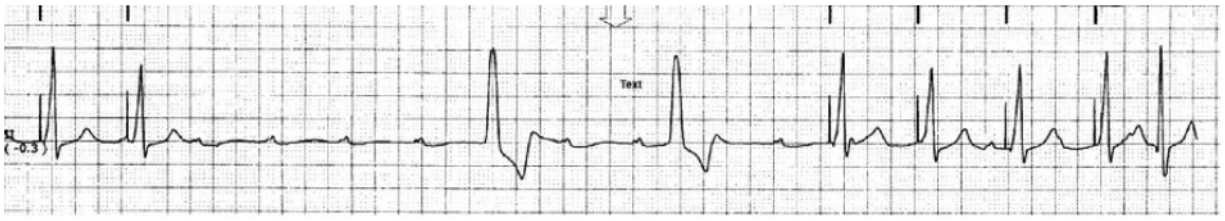

Figure 5 Postoperative rhythm strip showing complete heart block with a junctional rhythm with subsequent pacing.

Table 1 Summary of two-dimensional transthoracic echocardiographic findings of unicuspid vs. bicuspid aortic valve[8]

Type of valve	Unicuspid aortic valve	Bicuspid aortic valve
Echo findings	• One area of contact from valve commissure to the aortic root. • Heavily calcified valve in younger patients. • Low cusp height. • Eccentric valve during systolic opening 'systolic doming'. • Eccentric coaptation during valve closure in the parasternal long axis. • Two regurgitation jets. One eccentric jet through the middle of the orifice, while the other is at the level of both the non-coronary and left cusp.	• Two areas of contact from valve commissures to the aortic root. • Football shaped systolic opening seen in the parasternal short axis.

seen in our patient. When comparing patients with UAV's to those with BAV, patients with UAV present at a younger age with faster progressing symptoms. Other abnormalities associated with UAV's can be anomalous coronary artery anatomy, a single coronary artery, aberrant right subclavian artery, ventricular septal defects, aortic coarctation, and most commonly aortopathy.[6] In addition to UAV's, BAV's are associated with aortopathy, although they can have subtle differences. When comparing aortopathy in both UAV's and BAV's, they both often present with a dilated aortic annulus but more commonly patients with UAV will have less dilation of the ascending aorta compared with BAV's.[7]

Specifically, when looking at UAV's there are two forms, acommissural and unicommisural. They are differentiated by the presence or absence of lateral attachment of the valve commissure to the aorta. In acommisural, UAV's all three raphae are almost completely fused leading to a central orifice that can be seen as a pin hole on echocardiography and often presents at a younger age due to severe stenosis. In contrast, unicommissural UAV has a lateral commissural attachment to the aorta leading to a slit like opening seen on echocardiography. This and other echo findings summarized in *Table 1* are often difficult to find and differentiate from a BAV.

For this reason, modalities such as three-dimensional echocardiography (TOE) allow for better visualization than two-dimensional echocardiography (TTE) of the aortic valve to determine the type of congenital valvular malformation. In suspected cases of UAV's 14–25% of TTEs were able to diagnose the valvular malformation preoperatively, while for TOEs, 69–75% were able to correctly diagnosis a UAV.[7] *Table 1* highlights the differences between tricuspid and BAVs. In addition, patients with a unicommisural UAV's, the lateral commissural attachment leads to a larger aortic valve opening leading to development of symptomatic aortic stenosis later in life, often between the 3rd and 5th decade as seen in this patient.[1]

When looking specifically at our patients TTE, he had an AVA of 1.1 cm^2, which falls in the moderate stenosis range according to the 2017 ESC/EACTS guidelines. When looking at the peak/mean gradient of 42.53 mmHg/24.14 mmHg across his aortic valve according to the same guidelines, he falls in the severe aortic stenosis range. This is referred to a reverse area gradient mismatch or discordance. This phenomenon is known to occur in patients with alterations in pressure recovery which can be seen in congenital valvular disease, such as a UAV or BAV. Additionally, errors of measurements as well as other causes such as alternations in trans-aortic valve flow should be kept in mind.

In addition to echocardiograms such as the TTE and TOE to diagnose a UAV, other modalities do exist. For instance, cardiac CT and magnetic resonance imaging (MRI) can diagnose UAV's with great accuracy. In addition to seeing the valve morphology with CT and MRI other important information can be determined that can help management of valve replacement. For example, cardiac CT allows for better visualization and measurement of an ascending aortic aneurysm, if present, as well as the anatomy of the coronaries and great vessels. As mentioned before abnormalities of these vessels occur more commonly in patients with a UAV.

When looking at patients with a UAV's they often undergo valve replacement at a young age. Studies have shown that there is minimal difference in long-term outcomes of mechanical vs. bioprosthetic valves and that mechanical valves have a slightly longer replacement time.[9] There are two main components that play a role in patients with UAVs that can lead to valve replacement. One being aortic stenosis with regurgitation which is seen in our patient, and the rarer being an isolated aortic regurgitation. In both cases underlying conduction abnormalizes can be seen. It is well known the aortic valve has a close proximity to the atrioventricular conduction system. In patients with UAV's, there is often invading calcification that can disrupt this conduction system leading to bundle branch blocks and even complete heart block. In addition, replacement of the aortic valve holds inherent risks of causing conduction abnormalities. Dawkins *et al.*[10] demonstrated in a study of 342 patients who underwent an isolated aortic valve replacement that one in 12 patients would need a permanent pacemaker implantation (PPI) due to a conduction abnormality.

When looking at patients who underwent aortic valve replacement, the most common cause of PPI was a complete heart block.[9] In a study of 138 patients who underwent PPI secondary to complete heart block, the mean time to implantation was 6 days.[9] The assessment for PPI is done by turning down temporary pacemaker pacing rate to 40 b.p.m.[9] If there is no intrinsic pacing, a PPI is indicated. While conduction abnormalities after aortic valve repair that lead to haemodynamic instability are rare, it is recommended to have temporary placement of pacemakers to reduce chances of sudden death. With studies showing conduction abnormalities happening in both minimally invasive and traditional aortic valve replacement, it is

important for patients to know the risks and benefits of both procedures.

Conclusion

A UAV is a rare but severe valvular abnormality. Patients often undergo aortic valve replacement at a young age due to symptoms of heart failure secondary to aortic stenosis and regurgitation. Additionally, patients often have accompanying aortic root aneurysms that must be addressed at the time of surgery. With the known sequelae of conduction abnormalities after isolated aortic valve replacement, it is important for placement of a temporary pacemaker and preoperative education of the patient on possible need for PPI. Currently, there is no literature that compares conduction abnormalities in patients with UAVs to those of BAVs, but it has been shown that patients with congenital valvular abnormalities are at a higher risk for condition abnormalities than those without. Overall with there being little literature about the complications of UAVs, it is important for clinicians to have a high index of suspicion in patients who present with heart failure and a systolic murmur at a young age to have a UAV. In these patients, the use of three-dimensional transoesophageal echocardiography to assess the valvular structure is beneficial as two-dimensional transthoracic echocardiography often does not show enough detail to differentiate between a bicuspid and unicuspid valve like seen in our patient. Once diagnosed, it is important for physicians to look for other congenital abnormalities such as anomalous coronary artery anatomy and aortopathy as they are more common in patients with congenital valvular abnormalities and can have an impact on treatment. In addition, it is important to follow and report the presentation, if conduction abnormalities were present pre- or post-aortic valve replacement and their postoperative complications as there have been few studies on this rare valvular abnormality.

Supplementary material

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

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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