Accessory mitral valve tissue (AMVT) is a rare congenital cardiac anomaly sometimes responsible for left ventricular outflow tract (LVOT) obstruction. It is diagnosed during both neonate–childhood and adult periods in patients usually symptomatic for dyspnoea, chest pain, palpitations, fatigue, or syncope. Nevertheless, AMVT is often an incidental finding. AMVT is most often associated with other cardiac and vascular congenital malformations, such as septal defects and transposition of the great arteries. Surgery is indicated only in cases of significant LVOT obstruction and in patients undergoing correction of other cardiac malformations or exploration of an intracardiac mass. Two-dimensional echocardiography, both transthoracic and transoesophageal, is considered the main imaging modality for AMVT diagnosis and patient follow-up. The recent introduction of three-dimensional echocardiography allows a more realistic characterization of this entity. We present three clinical cases in which AMVT was incidentally diagnosed during standard echocardiography and an updated review of the literature highlighting the usefulness of echocardiography for AMVT morphological and functional characterization as well as the most relevant clinical implications due to its discovery.

Keywords  Accessory mitral valve tissue  •  Left ventricular outflow tract obstruction  •  Echocardiography

Introduction

Accessory mitral valve tissue (AMVT) is a rare congenital cardiac malformation that sometimes causes left ventricular outflow tract (LVOT) obstruction and is commonly associated with other congenital cardiac anomalies.1,2 The first case of this peculiar lesion and first surgical treatment were described in the literature in 1842 and 1963, respectively.3,4 However, the prevalence of AMVT and the age in which it is most frequently diagnosed are still debated.1,2,5 Two-dimensional (2D) echocardiography, both transthoracic and transoesophageal, and the recent introduction of three-dimensional (3D) echocardiography play an important role in the diagnosis, management, and follow-up of patients with this abnormality.6–11 We describe three cases of AMVT, incidentally diagnosed during 2D TTE. We performed a review of the literature and identified 104 cases dating back to 1963 (Supplementary data online, Table S1) to clarify some debated epidemiological concerns and to underline the usefulness of echocardiography for the characterization of AMVT.

Case reports

Case 1

A 30-year-old woman presented with syncope. She had a history of frequent dizziness. Cardiac examination and rest electrocardiogram (ECG) were unremarkable. Two-dimensional TTE performed with a Vivid 7 ultrasound scanner (GE Vingmed Ultrasound AS, Horten, Norway) in the emergency department revealed a mobile spherical cyst-like mass attached to the tip of the anterior MV leaflet (Figure 1 and Supplementary data online, Video S1). A non-echogenic central area delimited by hyperechogenic edges moved into the LVOT during systole, but did not obstruct flow. No impairment in MV leaflet mobility or other associated abnormalities were observed. Three-dimensional echocardiography was used to study mass morphology, size, and motion (Figure 2) and to confirm the absence of LVOT obstruction. Morphological characteristics and motion were suggestive of AMVT. Furthermore, the gradient across the LVOT did not change with exercise, thereby excluding
AVMT as the cause of the patient’s symptoms. Instead, some episodes of non-sustained ventricular tachycardia (Figure 3) were recorded during Holter monitoring. Anti-arrhythmic drugs and annual follow-up of the AMVT with serial echocardiography were recommended for the young patient. To date, she remains asymptomatic without echocardiographic evidence of LVOT obstruction.

Figure 1 Case 1. (A) AMVT with spherical cyst-like shape delimited by hyperechogenic edges attached to the tip of the anterior mitral valve leaflet. The same image depicted in the (B) early diastolic and (C) end-diastolic frames. (D) Colour Doppler echocardiography detects normal laminar diastolic flow across the valve.

Figure 2 Case 1. Real-time 3D echocardiography (A) demonstrates the relationship between the AMVT and subvalvular apparatus and (B) is useful for studying mass morphology and size.
Case 2
A 22-year-old man with a bicuspid aortic valve and history of aortic coarctation underwent echocardiographic examination. Clinical history revealed surgical repair of the aortic coarctation at Age 3 and successful transcatheter stent implantation in the aorta at Age 16. The aortic valve was preserved given its normal function. Since the last operation, he had been asymptomatic. Two-dimensional TTE performed with a Vivid 7 ultrasound scanner (GE Vingmed AS) showed spherical remodelling of the left ventricle but normal systolic function (ejection fraction: 60%) with the exception of a septal dyskinesia. An echogenic serpiginous structure that changed in shape during the cardiac cycle, thus appearing in some frames as a circular ring, was attached to the anterior MV leaflet. The structure poorly projected into the LVOT during systole and prolapsed into the LV cavity during diastole (Figure 4 and Supplementary data online, Video S2). Based on its echocardiographic appearance, AMVT was diagnosed even though it was not recognized in previous echocardiographic examinations, and considered an incidental finding without indication to surgery. At 1-year follow-up, the patient remained stable without new cardiac symptoms or major events.

Case 3
A 52-year-old man with angina and dyspnoea for the past 3 months underwent coronary angiography after stress scintigraphy suggested myocardial ischaemia. Prior to angiography, 2D echocardiography performed with a Vivid 7 ultrasound scanner (GE Vingmed AS) revealed asymmetrical LV hypertrophy of the interventricular septum with preserved regional wall motion and ejection fraction. Systolic anterior motion of the anterior MV was associated with significant LVOT obstruction at rest (60 mmHg) and severe mitral regurgitation. Due to poor image resolution, the patient underwent TEE, which provided a more accurate anatomical depiction of the mechanism underlying LVOT obstruction and mitral regurgitation (Figure 5 and Supplementary data online, Videos S3 and S4). Attached to the ventricular aspect of the posterior MV leaflet, AMVT, which most likely contributed to LVOT obstruction, was clearly identified. Surgery was scheduled due to the presence of symptoms.

Discussion
In our case series, AMVT was an incidental finding in patients undergoing TTE for different indications: syncope (1 patient), post-cardiac surgery follow-up (1 patient), and ECG alterations (1 patient). In only one patient, a significant LVOT obstruction owing to AMVT was discovered. The most significant finding of this anomaly in our case series was the frequent association with other cardiac malformations. Surgery was recommended only for the symptomatic patient (Case 3), whereas asymptomatic subjects remained stable with no clinical deterioration or LVOT obstruction during follow-up (Table 1). Also, we recently demonstrated a case of AMVT associated with an anomalous origin of the left coronary artery in an elderly patient with ischaemic ECG alterations.12 Our experience particularly highlights the key role of echocardiography in the diagnosis, characterization, and functional evaluation of this structure.

Epidemiological and demographic concerns
More frequently identified in men, AMVT has a male-to-female ratio of 1.75 : 1.2 It was first described in 1842 with the first surgical management introduced by MacLean in 1963.3,4 The exact prevalence of this entity is not well known because several cases of AMVT were incidentally diagnosed intraoperatively.5 The largest review, based on 63 reports of 90 patients published from 1963 to 2002, noted that the age of AMVT diagnosis ranged from newborns to 77 years.
(mean 8.6 years), but did not provide information regarding the incidence in childhood and adults, respectively. With fewer than 100 cases of AMVT reported in the literature and <30% of which described the adult population, the estimated incidence of AMVT in adults was 1/26,000 echocardiograms. Therefore, AMVT was no longer considered a clinical entity typical of the first two decades of life. An increasing number of diagnoses in older patients was recently demonstrated.

Our experience with AMVT involves four adults, including a previously published case, with a higher prevalence in males (3 of 4 patients, 75%). Patient age at the time of diagnosis ranged from 22 to 73 years (Table 1), suggesting a benign course of AMVT, which frequently is an incidental diagnosis in asymptomatic subjects. These results reflect literature data, showing a larger prevalence in young men, with a male-to-female ratio of 1.5 : 1 and a prevalence of 54.6% in patients <18 years vs. 45.4% in older persons (Figure 6). This may be due both to the high frequency of symptomatic AMVT types with other complex congenital cardiac anomalies in the young, and the use of echocardiography to detect a heart murmur in asymptomatic children, resulting in earlier diagnosis.

Figure 4 Case 2. (A–D) Echogenic serpiginous AMVT that becomes circular in shape during the cardiac cycle. (E) Bicuspid aortic valve in the same patient.

Aetiology, pathophysiology, and associated co-morbidity
Accessory MV tissue may affect one or both of the atrioventricular valves simultaneously; however, the MV is most often involved. Although the exact embryologic mechanism of AMVT formation is not clear, it may stem from the abnormal or incomplete separation of the MV from the endocardial cushions. In favour of this theory, AMVT is commonly associated with other congenital intracardiac and vascular malformations, with a higher incidence of ventricular septal defects, subaortic membrane, and transposition of great arteries (Figure 7 and Supplementary data online, Table S1). The function of the MV is almost always preserved; however, two cases of AMVT producing congenital mitral stenosis were recently described.

From a pathophysiological point of view, AMVT is recognized as a cause of LVOT obstruction. Two mechanisms underlying obstruction in patients with AMVT were recognized: (i) mass effect of the accessory tissue and (ii) progressive deposition of fibrous tissue due to the turbulent flow created by AMVT. In the first case, obstruction can be immediately significant while, in the other case, outflow gradient increases progressively, probably due to...
AMVT enlargement and LVOT narrowing. However, cases with no obstruction also were reported (Supplementary data online, Table S1), as was the case in the patients of our series (Table 1). Based on the literature data, we estimate that the obstructive form of AMVT has a prevalence of 86.5% (Figure 6); nevertheless, clear information regarding this complication was not available in 13% of the reviewed cases.

With regard to associated lesions, three patients in our case series (Cases 2–4) had other structural cardiac abnormalities as previously described and one patient (Case 1) showed alterations of electrical cardiac activity (Table 1). In the reviewed cases, AMVT was associated with other cardiac abnormalities in the majority of patients (65.4%), whereas it was isolated in 14.4%. However, no information was provided in this context in 20.2% of cases (Figure 7). The most frequently anomalies encountered in the literature are summarized in Figure 7. Sajja and Mannam described the only case of AMVT associated with congenital LV apical aneurysm. The frequent coexistence of AMVT and other cardiac lesions suggests that AMVT detection during standard TTE should lead to careful cardiac exploration to rule out any congenitally associated anomalies potentially susceptible for surgical treatment.

Clinical presentation
Patients with AMVT may be asymptomatic with the presence of a murmur, but, more frequently, they experience symptoms (Figure 8 and Supplementary data online, Table S1). Usually patients become symptomatic when the mean gradient across the LVOT reaches 50 mmHg. Signs of LVOT obstruction generally develop during the first decade of life in one-third of the patients and include typical symptoms. In this review, we found a prevalence of 86.6% for obstructive forms (71/82 out of 104 patients in whom detailed information about LVOT obstruction was available). Low cardiac output due to subaortic obstruction and congestive heart failure also were reported in the literature. Because of the frequent association with other cardiovascular malformations, it is possible that symptoms of

Table 1

<table>
<thead>
<tr>
<th>Age/sex</th>
<th>Clinical presentation</th>
<th>Echocardiographic findings</th>
<th>LVOT obstruction</th>
<th>Associated lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>30 y/F Syncope, non-sustained ventricular tachycardia</td>
<td>Mobile spherical cyst-like mass attached to the tip of the anterior mitral valve leaflet</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Case 2</td>
<td>22 y/M Asymptomatic</td>
<td>Echogenic serpiginous structure attached to the anterior mitral valve leaflet</td>
<td>No</td>
<td>Bicuspid aortic valve, aortic coarctation</td>
</tr>
<tr>
<td>Case 3</td>
<td>52 y/M Dyspnoea</td>
<td>Thin echogenic string-like structure attached to the posterior mitral valve leaflet</td>
<td>Yes</td>
<td>Hypertrophic cardiomyopathy</td>
</tr>
<tr>
<td>Case 4a</td>
<td>73 y/M Asymptomatic, systolic murmur</td>
<td>String-like structure attached to the anterior mitral valve leaflet</td>
<td>No</td>
<td>Anomalous origin of the left coronary artery</td>
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LVOT, left ventricular outflow tract.

*Ref. [12].
AMVT are due to the concomitant abnormalities. Furthermore, AMVT is a potential source of cardioembolism, probably because of the highly mobile and often fragile tissue or owing to accumulated platelets with intermittent embolization. Four cases in the literature described the probable association between AMVT and cardioembolic transient ischaemic attacks.

In our cases series, one patient had syncope. However, LVOT obstruction was excluded at rest and during exercise, but a diagnosis of ventricular tachycardia was made. Whether syncope was definitely related to the arrhythmia or AMVT is not demonstrable. In the other three patients, clinical presentation was unremarkable, confirming the high probability of incidental discovery of AMVT in adults with no signs or symptoms of LVOT obstruction. According to the literature review, 37.5% of cases were asymptomatic. The prevalences of the symptoms described in the other published cases are detailed in Figure 8.

**Imaging modalities for the diagnosis of accessory mitral valve tissue**

Before the widespread use of 2D echocardiography, the diagnosis of AMVT was difficult to determine, typically by angiography or,
sometimes, as an unsuspected finding at surgery. Angiography can visualize a mass in the subaortic area, but allows the diagnosis of AMVT only if the mass is bulky. Cardiac catheterization may provide the measurement of LVOT gradient and may be required to investigate other associated malformations. For its own characteristics, echocardiography is currently the favoured imaging modality for both the diagnosis and management of AMVT. However, there is recent growing interest in other non-invasive imaging modalities, i.e. computed tomography (CT) and magnetic resonance imaging (MRI), particularly for differential diagnosis of cardiac masses.

**Echocardiography**

Since the first report of AMVT diagnosed by echocardiography dating back to 1985, 2D echocardiography can be considered the gold standard modality for the evaluation of AMVT. Two-dimensional echocardiography plays a fundamental role as it enables anatomical characterization of AMVT and, also, shows possible associated lesions and complications such as LV hypertrophy, dilatation, and systolic dysfunction. Furthermore, Doppler evaluation of the LVOT allows a non-invasive assessment of the degree of obstruction.

With TTE, AMVT may look like a mobile parachute-like leaflet floating in the LVOT and usually undergoes a progressive thickening or a fixed structure attached to the interventricular septum by a short chordal apparatus. Large redundant AMVT may appear as a globular or even cystic mass, as in Case 1. The AMVT typically prolapses into the LVOT or aortic valve during systole and retracts during diastole owing to its attachment to various MV and subvalvular apparatus.

Our experience confirms the literature data, which shows a high prevalence of the mobile type of AMVT, in most cases originating from the anterior MV leaflet, with a higher incidence of parachute-like or balloon-like structures. In addition, the literature review showed an 18.2% prevalence of parachute-like AMVT (Figure 6). To our knowledge, there is only one report describing AMVT not connected to the MV, which makes it ectopic AMVT. A variable degree of mitral regurgitation may be often present.

With regard to echocardiographic tissue characterization, the AMVT has usually the same echogenicity of normal MV tissue, but echocardiography cannot provide an accurate tissue characterization as other imaging modalities, i.e. cardiac MRI, may be able to do. As for differential diagnosis, echocardiography may differentiate AMVT from redundant MV chordae, which are attached to the MV leaflets and do not prolapse into the LVOT unless there is chordal rupture. Since these redundant chordal structures may be involved in chordal systolic anterior motion with dynamic LVOT obstruction, it may be possible to screen out an erroneous diagnosis of obstructive AMVT through TEE. Furthermore, during echocardiographic examination, a careful differential diagnosis should be made between AMVT and other causes of LVOT obstruction (hypertrophic cardiomyopathy, subaortic stenosis, etc.). Sometimes dynamic and fixed LVOT obstruction co-exist, as in Case 3 from our cohort, but in most of the cases, AMVT is the unique cause of LVOT obstruction.

Echocardiography is important for differential diagnosis of ventricular masses, such as tumours and vegetations. However, these lesions more commonly originate from cardiac muscle or build up directly upon the low-pressure side of a heart valve. Although TTE is usually sufficient for diagnosis and evaluation of AMVT, TEE can define details, such as morphology and attachment points of the accessory tissue, particularly in patients scheduled for surgery. Intraoperative TEE is also necessary to follow MV function. With the recent introduction of 3D echocardiography, it is possible to obtain additional information for the evaluation of mass shape, dimension, and complications (Figure 2). Furthermore, real-time 3D TEE is effective in defining difficult spatial relationships and anatomical anomalies, allowing successful treatment of obstructive types of AMVT with alcohol septal ablation.

In summary, echocardiography is the preferred tool for the morphological and functional study of AMVT, with no need for other imaging modalities in the majority of cases.
Other imaging modalities

Although echocardiography provides a “gold standard” for the evaluation of AMVT, the high temporal and spatial resolutions of CT and MRI makes these two modalities useful for the assessment of accessory tissue structure and function. Cardiac MRI may overcome echocardiographic limitations in both differential diagnosis of cardiac masses (i.e. doubtful cases by echocardiography) and tissue characterization. Moreover, MRI provides an important input for volume quantification and treatment of obstructive types of AMVT due to the 3D reconstruction of anatomy and connections. However, there is little evidence for using cardiac MRI and CT in routine clinical practice. To date, one case of AMVT was diagnosed by cardiac CT and, in another case, a tricuspid accessory valve was identified by cardiac MRI. It is likely that these limited data are owing to the rarity of the disease.

Surgical insights

Cardiac surgery is not indicated in every case of AMVT. The overall mortality after surgery is 8.9% of all reported cases. The main factors affecting mortality and the presence of a residual LVOT gradient appear to be incomplete removal of AMVT and the presence of other congenital cardiac defects. The current approach is to submit for intervention only patients with a significant LVOT gradient (mean gradient of ≥ 25 mmHg) and those undergoing correction of other congenital malformations or exploration of an intracardiac mass. For patients without significant LVOT obstruction, a follow-up with serial echocardiography to assess progression of the gradient is indicated.

It should be noted that identification of AMVT during surgery might not always be possible. The thin structure is commonly collapsed in the empty and arrested LV on bypass. The location of AMVT attachment is usually on the ventricular side. In addition, AMVT may lie below any discrete fibrous obstruction, as when accompanied by subaortic stenosis. Thus, aortotomy may not be sufficient, and left/right atriotomy may be employed as an additional and helpful approach to localize and fully examine AMVT. Systemic ventriculotomy provides good access, but it compromises cardiac function and causes scar-induced arrhythmias. Accessory MV tissue may be successfully removed through a ventricular septal defect. Usually excision of AMVT and division of the accessory chordae is the preferred treatment. Accessory chordae are left intact if there is a concern that disruption of the native subvalvular mitral apparatus may cause mitral regurgitation. If AMVT is present as an isolated finding, surgical repair is fairly successful with good long-term surgical results.

Prifti et al. using a surgically excised specimen of AMVT, provided an anatomical classification of this anomaly: fixed and mobile. Fixed AMVT include the most primitive form, a nodular mass (Type IA), which is normally attached to the mitroaortic continuity or ventricular side of the anterior MV leaflet, and membranous (Type IB), which is normally attached to the mitroaortic continuity. The location of the membrane attachment differs from case to case. Mobile AMVT includes a pedunculated form (Type IIA), which is normally attached through a fibrous “neck” to the mitroaortic continuity or ventricular side of the anterior MV leaflet, and a leaflet-like form (Type IIB), which is the most frequent presentation. This anatomical classification is further divided into those with rudimentary chordae (free-edge chordae) (Type IIB1) or well-developed chordae (Type IIB2). In Type IIB2, it is important to identify the chordae attachment. Preoperative findings showed six different locations of chordae insertion: LV wall, interventricular septum, accessory papillary muscle, anterolateral papillary muscle, anterior MV leaflet, and the anterior MV chordae. Sometimes the shape of AMVT is identical to that of the anterior MV leaflet, which seems to be duplicated.

Histological analysis, performed in patients undergoing surgery, showed both fibrous and myxoid dysplasia rather than normal, mature valve tissue (Figure 6 and Supplementary data online, Table S1). In patients not surgically treated because of the risk of a neurological event, anti-coagulant treatment should be considered.

Conclusion

AMVT is a congenital cardiac lesion with uncertain prognosis depending on LVOT obstruction and/or concomitant anomalies. We presented a case series of three adult patients with different clinical presentations in which AMVT was diagnosed using echocardiography. The literature review highlights a high prevalence in young patients with symptoms of LVOT obstruction and as an incidental finding in adults. Other cardiac congenital abnormalities are consistently associated with AMVT. Echocardiography plays a pivotal role in the diagnosis and follow-up of patients with this rare congenital lesion.

Supplementary data

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.

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