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PRACA POGLĄDOWA
REVIEW

Mitral annular disjunction and ventricular arrhythmia: A rare arrhythmic syndrome

Separacja pierścienia mitralnego i komorowe zaburzenia rytmu serca – rzadki zespół arytmiczny

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ABSTRACT

Mitral annular disjunction (MAD) is a structural anomaly of the heart characterized by the displacement of the posterior mitral leaflet's attachment toward the left atrial wall. MAD can lead to mitral valve prolapse, ventricular arrhythmia, and sudden cardiac death. In the general population, MAD can be found in 7%–9% of individuals. Transthoracic echocardiography is the primary imaging assay for detecting MAD, while cardiac magnetic resonance imaging remains the gold standard. The treatment strategy depends on the severity of mitral regurgitation, the presence of symptoms, and the risk of arrhythmia. Further research is necessary to better characterize this condition and to define optimal diagnostic and therapeutic strategies.

KEYWORDS

mitral annular disjunction, ventricular arrhythmia, sudden cardiac death

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STRESZCZENIE

Separacja pierścienia mitralnego (*mitral annular disjunction* – MAD) to strukturalna anomalia w obrębie serca, polegająca na przemieszczeniu przyczepu tylnego płatka zastawki mitralnej w kierunku ściany lewego przedsionka. MAD może powodować wypadanie płatka zastawki mitralnej, zaburzenia komorowe rytmu oraz nagłą śmierć sercową. W populacji ogólnej MAD występuje u 7–9% osób. Podstawowym badaniem obrazowym w wykrywaniu MAD jest echokardiografia przezklatkowa, natomiast złotym standardem pozostaje rezonans magnetyczny. Strategia leczenia zależy od stopnia nasilenia niedomykalności mitralnej, obecności objawów i ryzyka wystąpienia arytmii. Konieczne są dalsze badania, które dokładniej scharakteryzują to schorzenie i określą optymalne strategie diagnostyczne i terapeutyczne.

SŁOWA KLUCZOWE

separacja pierścienia mitralnego, komorowe zaburzenia rytmu, nagły zgon sercowy

Introduction

Mitral annular disjunction (MAD) is a structural developmental anomaly of the mitral valve annulus. It involves the attachment point of the posterior mitral valve leaflet (PML) being displaced toward the left atrial wall. This results in a gap between the mitral annulus and the left ventricular myocardium. MAD is often associated with abnormal movement, excessive mobility, or reduced stability of the mitral annulus during cardiac systole.

Historically, MAD was often ignored, but in recent years interest in this morphological feature has increased due to its association with dangerous ventricular arrhythmias and sudden cardiac death (SCD) [1,2,3]. Currently, MAD is increasingly recognized as a clinically significant problem that can lead to dangerous ventricular arrhythmia.

Although it is an anatomical condition, its association with ventricular arrhythmia and SCD makes its proper diagnosis crucial. The use of multimodal imaging, with a particular focus on cardiac magnetic resonance (CMR), is essential for precise diagnosis and risk assessment. Krawczyk-Ożóg et al. [4] and Faletra et al. [5] represent two extremes, questioning the unequivocally pathological nature of MAD. These studies, along with analyses of CMR imaging and three-dimensional (3D) echocardiography, focus on establishing a precise definition, the true prevalence, and the prognostic significance of MAD. A distinction is made between "true MAD" (superior displacement of the posterior leaflet attachment) and "pseudo-MAD" (associated with mitral valve prolapse [MVP], where the leaflet rests against the atrial wall during systole). MAD often occurs in isolation, and in healthy individuals it is most leftcommonly located non--posteriorly (the so-called "evolutionary MAD"). "Pathogenic MAD" – associated with arrhythmia – is most frequently located posteriorly [5].

Epidemiology

The exact prevalence of MAD is difficult to determine due to a lack of a unified definition and differences in imaging methods. In the general population, MAD may occur in 7%–9% of individuals [1], although autopsy studies have indicated significantly higher values, even above 90% [6].

Studies based on advanced imaging (computed tomography [CT] or CMR) suggest that MAD is a more common phenomenon than previously thought, which undermines its automatic recognition as a purely pathological feature. Using cardiac CT, Krawczyk--Ożóg et al. [4] identified the presence of atrial MAD (a-MAD) in as many as 20% of healthy hearts and noted that it occurs more frequently in the leaflets than in the commissures. In a study utilizing CMR, it was found that MAD of ≥1 mm affected nearly half (49%) of the consecutive patients referred for examination, which means that the mild form of MAD is very common and has a benign medium-term prognosis [4]. MAD is common in MVP patients, having been found in 20%–58% of this group [2,7], which suggests that the two conditions are related [8]. Sonaglioni et al. [9] showed a prevalence of about 40% in MVP patients. It has been found that up to 30% of them also have MAD, with the prevalence being significantly higher in some subgroups (e.g., patients with Barlow's disease) [1].

Research consolidates the view that although MAD occurs in healthy hearts, its greater extent and association with are crucial for the malignant phenotype. Patients with MVP have a significantly higher prevalence and greater dimension of MAD (MAD ≥1 mm in 90% of patients with MVP vs. 46% without MVP) [8].

MAD is more frequently observed in women and younger individuals [1]. This phenomenon is also sometimes associated with connective tissue diseases, such as Marfan syndrome or Ehlers–Danlos syndrome [10].

Pathophysiology

The mitral annulus has a dynamic, three-dimensional structure. Under normal conditions, during systole, the curvature of the annulus deepens and its circumference is reduced, which stabilizes the leaflets and ensures valve competence. In the case of MAD, the posterior part of the annulus undergoes paradoxical

widening and displacement toward the atrium, leading to the mitral valve leaflets and chordae tendineae stretching [1,2].
MAD can cause the progression of mitral regurgitation, left ventricular volume overload, and repetitive micro--trauma to the papillary muscle and left ventricular myocardium. Repeated micro-trauma can lead to the development of fibrosis, which is detected on CMR as "late gadolinium enhancement" (LGE) [11]. Fibrosis provides a substrate for ventricular arrhythmia – from single premature ventricular contractions to malignant tachyarrhythmias [2,12].

Diagnostics

Echocardiography

Transthoracic echocardiography (TTE) is the primary tool for detecting MAD. During TTE, the systolic separation of the annulus from the left ventricular myocardium can be observed in the parasternal long--axis view. However, because the sensitivity of TTE is limited, MAD is often overlooked [2,8]. In two--dimensional imaging, MAD is often difficult to visualize because it is mainly visible during the systolic phase of the heart. Therefore, a full assessment requires the use of 3D echocardiography, which allows for more accurate visualization and measurement of the degree of disjunction. Additionally, transesophageal echocardiography – especially in 3D – offers a more precise anatomical assessment.

Computed tomography

With CT, the anatomical structure can be assessed, but due to the lack of dynamic systole-diastole characteristics, its diagnostic role is limited [1].

Cardiac magnetic resonance

CMR is considered the gold standard. This examination allows for not only the identification of MAD, but also an assessment of the presence and extent of fibrosis [10]. This data has crucial prognostic significance in arrhythmia risk stratification. CMR is ideally suited for ventricular arrhythmia risk stratification in MAD patients due to its unique ability to non-invasively identify focal myocardial fibrosis using LGE. A link is postulated between complex ventricular arrhythmias and LGE in the mid-portion of the papillary muscles or focal myocardial fibrosis in the inferobasal region.
Any standard CMR protocol can identify MAD. A comprehensive MAD assessment should include a detailed description of the mitral valve, the degree of MAD, left ventricle remodeling, and fibrosis. The degree of MAD includes its extent around the mitral annulus and its width. The relationship between the extent of MAD and ventricular arrhythmia is still debated [13,14].

Clinical significance

Although most patients with MVP have a good prognosis, a small subgroup – especially young women – is burdened with a higher risk of ventricular arrhythmia and SCD [12,15].
High-risk features include:
– the presence of MAD >8–10 mm
– papillary muscle and/or inferolateral wall fibrosis on CMR
– frequent ventricular premature beats and episodes of non-sustained ventricular tachycardia (VT)
– history of syncope
– family history of SCD [2,7,9,12].
The co-occurrence of MAD and MVP increases the probability of arrhythmia compared to isolated MVP [1,2,14].
Below is an EKG showing a complex ventricular arrhythmia from a 12-lead Holter recording (Figure 1) and a TTE examination (Figure 2) of a 24-year-old female patient with MAD.

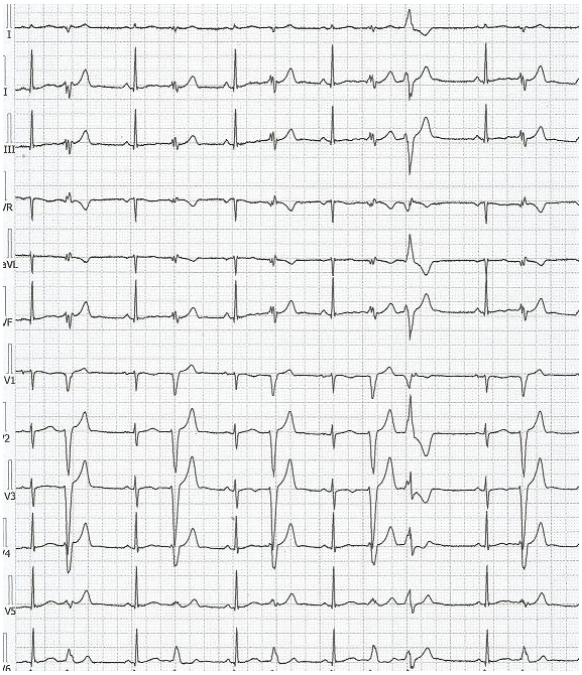

Fig. 1. Multiform ventricular premature beats (ventricular bigeminy and couplet) recorded during 24-hour 12-lead Holter monitoring in a 24-year-old female patient with mitral annular disjunction and numerous ventricular premature beats (over 7,000/day; paper speed: 25 mm/s)

CMR studies suggest that larger dimension (MAD ≥ 4−6 mm) correlates with a greater burden of ventricular ectopic beats on Holter monitoring and a tendency for a worse prognosis (SCD/VT). It has been concluded that defining a "malignant MAD dimension threshold" is necessary to improve risk stratification [14].

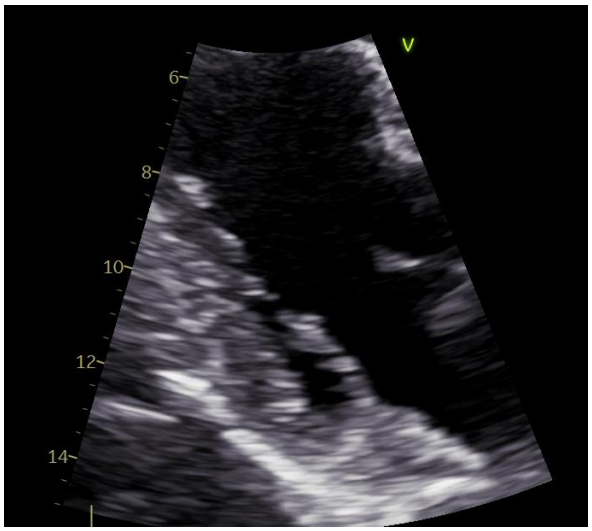

Fig. 2. Echocardiography examination, parasternal long-axis view, showing mitral annular disjunction

MAD is postulated as a contributing factor to electrical instability through:

– altered mechanics – MAD causes dynamic annular enlargement during systole and "annular slippage" [15], which may generate excessive mechanical stress at the atrioventricular junction
– myocardial fibrosis – a hypothesis proposed by Faletra et al. [5], among others, suggesting that continuous stress can lead to fibrosis in the adjacent myocardium, which becomes a substrate for arrhythmias.

Furthermore, MAD is strongly associated with primary atriopathy (left atrial dysfunction), especially in the case of bileaflet MVP, irrespective of the severity of mitral regurgitation [14], which further increases the risk of cardiovascular events.

Therapeutic management

The treatment strategy depends on the severity of mitral regurgitation, the presence of symptoms, and the risk of arrhythmia.

– Observation – for patients without symptoms and high-risk arrhythmias
– Pharmacotherapy – beta blockers and/or other antiarrhythmic drugs
– Percutaneous procedures – e.g., transcatheter edge-to-edge repair (TEER) or a MitraClip procedure (transcatheter mitral valve repair using a MitraClip device) improve regurgitation but do not correct MAD itself; their effect on arrhythmia risk requires further research [2,16]
– Ablation – used for symptomatic arrhythmias resistant to pharmacotherapy [17]
– Cardioverter-defibrillator implantation – for secondary prevention after cardiac arrest or VT with hemodynamic instability; for primary prevention, decisions about cardioverter-defibrillator implantation should be made individually
– Surgery – classic valve repair with ring annuloplasty may eliminate the MAD phenomenon and reduce arrhythmia risk [18]

Conclusions

Although MAD was first described over 50 years ago by Bharati et al. [19] as an abnormal connection of the PML to the left atrial wall, it remains a significant and insufficiently understood diagnostic and therapeutic problem. Despite its most frequent co-occurrence with MVP, a growing body of evidence indicates its independent arrhythmogenic effect. The diagnosis of MAD requires multimodal imaging, with CMR being the gold standard.

Jaworski et al. [3] propose a multiparametric approach to the assessment of arrhythmogenic MVP, suggesting that MAD is only one – albeit a key – component in a broader set of SCD risk markers, which also includes echocardiographic features, clinical symptoms, and myocardial fibrosis.

MAD is increasingly recognized not only as a pathology that contributes to mechanical dysfunction, but also as a factor that can cause electrical instability, raising concerns about the development of dangerous ventricular arrhythmias [20].

The presence of MAD should prompt a thorough risk stratification for arrhythmias and consideration of more intensive monitoring.

There is still a lack of clear guidelines for management; further prospective studies and a unified definition of MAD are needed. Data on the progression of MAD and associated ventricular arrhythmia is lacking.

The recent papers (2022–2025) are characterized by intensive efforts aimed at understanding MAD and separating its benign, anatomical variants from the malignant arrhythmic phenotype. MAD is a common phenomenon, with low-grade variants (e.g., ≥1 mm) – that are frequent in patients without MVP and often carry a benign prognosis – and advanced MAD. The latter is a risk marker because there is a strong correlation between larger MAD dimension (especially ≥4–6 mm) and the presence of MVP, severe ventricular arrhythmias, and dynamic changes in annular and left atrial geometry [14].

The debate entitled "MAD or MADness?" [5] suggests that the clinical evaluation of patients with MAD must move away from a binary classification (present/absent) and focus on quantitatively assessing the dimension and associated pathological features (MVP, fibrosis, or atriopathy) for optimal stratification of the risk of SCD. Future research should aim to establish reliable measurement thresholds that unequivocally define a malignant MAD dimension [3].

MAD is a phenomenon that is gaining increasing importance in cardiology. The future of treating patients with MAD will depend on the results of ongoing studies, which aim to better characterize this condition and define optimal diagnostic and therapeutic strategies [13,21].

Authors' contribution
Study design – A. Pawlus, K. Kolebacz, B. Średniawa
Data collection – K. Kolebacz, A. Pawlus, B. Średniawa
Manuscript preparation – A. Pawlus, K. Kolebacz, B. Średniawa
Literature research – B. Średniawa, K. Kolebacz, A. Pawlus
Final approval of the version to be published – B. Średniawa, A. Pawlus, K. Kolebacz

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