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RESEARCH ARTICLE

MITRAL VALVE ANNULAR DISJUNCTION – A COMPILATION OF INFORMATION IN A MINI REVIEW

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ABSTRACT

Background: Mitral valve annulus disjunction is a structural anomaly characterized by excessive displacement of the mitral annulus in relation to the left ventricular myocardium. This condition can lead to mitral valve dysfunction, predisposing to mitral regurgitation and increasing the risk of ventricular arrhythmias. Clinically, patients may be asymptomatic or present symptoms such as palpitations, dyspnea, and fatigue. In more severe cases, there may be heart failure and an increased risk of sudden death. Diagnosis is made by echocardiography, especially with Doppler and advanced techniques such as three-dimensional echocardiography and cardiac magnetic resonance imaging. Treatment varies according to the severity of the disjunction and the presence of significant mitral regurgitation. Mild cases can be followed clinically, while severe cases may require surgical mitral valve repair or replacement. **Objective:** The objective of this mini narrative review is to highlight that early recognition of mitral valve annulus disjunction is essential to avoid complications and improve patient prognosis. **Conclusion:** Mitral valve annular disjunction, in combination with mitral regurgitation, is a condition that can lead to significant morbidity and mortality, particularly if not diagnosed early or treated appropriately. Appropriate management, which may involve medication, regular monitoring and, in severe cases, surgery to repair or replace the mitral valve, is essential to improve the prognosis and reduce the risks associated with the condition. Early detection and effective treatment can improve the patient's quality of life and reduce mortality related to mitral regurgitation.

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INTRODUCTION

Mitral valve annulus disjunction is a term used to describe a condition in which there is an abnormal increase in the distance between the mitral valve annulus and the walls of the heart, particularly in the area of the left ventricle, which can result in disturbances in the function of the mitral valve, which may predispose to mitral regurgitation. When the disjunction leads to mitral regurgitation, the symptoms may vary depending on the severity and progression of the condition[1]. It was described in the 1980s by Hutchins et al[2], and was identified in approximately 92% of cases of mitral valve prolapse. However, little attention was given to mitral valve annulus disjunction, as it was considered to have no significant clinical consequences[3]. In this mini narrative review, the clinical and complementary diagnostic aspects of mitral valve annulus disjunction were emphasized. Scientific databases, including PubMed, Scopus, Web of Science, Google Scholar, and Medline Plus using the terms "mitral annular disjunction; mitral valve prolapse; mitral valve; mitral valve insufficiency; arrhythmia" were searched for this review. Within these search terms, every subtitle in the text was searched individually. Full-text papers were reviewed after removing duplicates and screening titles and abstracts and the most relevant information found is presented in this mini narrative review.

DIAGNOSTIC CRITERIA FOR MITRAL VALVE ANNULAR DISJUNCTION

Clinical Diagnosis: Mitral valve annular disjunction is frequently asymptomatic and is often an incidental finding on cardiac imaging. However, some patients may present

cardiovascular symptoms and clinical signs suggestive of the condition[4-9].

Clinical History

Symptoms may range from nonspecific to more severe manifestations[4-9]:

- Asymptomatic: Many patients do not present symptoms, especially in the early stages.
- Palpitations and Arrhythmias: Ventricular extrasystoles and episodes of ventricular tachycardia may occur, being more common in patients with mitral valve annulus disjunction associated with mitral valve prolapse.
- Dyspnea and Fatigue: Related to progressive mitral insufficiency.
- Atypical Chest Pain: Common in patients with mitral valve annulus disjunction, usually unrelated to effort.
- Syncope or Pre-Syncope: May be associated with malignant ventricular arrhythmias.
- History of Sudden Death in the family: Important risk factor, especially in young people.

Physical Examination

Clinical findings may vary and depend on the presence of mitral valve prolapse and mitral insufficiency[4-9]:

- Midsystolic click: Present when there is mitral valve prolapse.
- Systolic murmur in the mitral area: If there is associated mitral insufficiency.
- Irregular heartbeats: Indicating ventricular arrhythmias.
- If the mitral valve annulus disjunction is isolated (without mitral valve prolapse), the findings on physical examination may be normal[4-9].

Differential Diagnosis

Mitral valve annulus disjunction can be confused with other cardiovascular conditions, such as[5,6,9,10]:

- Mitral valve prolapse without mitral valve annulus disjunction.
- Arrhythmogenic cardiomyopathy.
- Myocarditis.
- Long QT syndrome.
- Coronary artery disease (if chest pain is present)[5,6,9,10].

Diagnosis by Complementary Exams: Mitral valve annulus disjunction is diagnosed when there is a displacement ≥ 2 mm of the mitral annulus in relation to the left ventricular myocardium, usually in the posterolateral wall, observed on echocardiography or magnetic resonance imaging[9,11-15]:

- Electrocardiogram: To assess early repolarization, ventricular extrasystoles or polymorphic ventricular tachycardia.
- 24-hour Holter: To detect ventricular arrhythmias.
- Transthoracic Echocardiography: Initial exam to assess mitral valve anatomy. It can identify mitral annulus displacement and assess the presence of valve prolapse.
- Transesophageal Echocardiography: Considered the gold standard for detailed evaluation of the mitral annulus. It allows better visualization of the disjunction and its extent.
- Echocardiography with Strain (Speckle Tracking): Assesses the mechanics of the mitral annulus and the deformation of the adjacent myocardium. It can help in the early identification of disjunction of the mitral valve annulus before the onset of symptoms.
- Cardiac Magnetic Resonance Imaging: Highly accurate examination to evaluate the anatomy of the mitral annulus and the presence of myocardial fibrosis. Important for patients with suspected disjunction of the mitral valve annulus associated with ventricular arrhythmias.
- Cardiac Computed Tomography: Less commonly used for this diagnosis but can assist in the structural evaluation of the mitral annulus[9,11-15].

THERAPEUTIC APPROACH

Treatment of mitral valve annular disjunction depends primarily on the severity of the condition and the symptoms it is causing, as well as associated comorbidities, such as mitral regurgitation. Since mitral valve annular disjunction is often a risk marker for mitral regurgitation, management primarily involves controlling mitral regurgitation and preventing more serious complications[7,9,16-21]:

Clinical Treatment (Medications): If mitral valve annular disjunction is causing mitral regurgitation, the main goal of medical treatment is to relieve symptoms and improve heart function. This is done with medications that help control the workload on the heart and minimize progression of the condition. Some of the main medications include[7,9,16-21]:

- **Angiotensin-converting enzyme inhibitors (angiotensin receptor blockers):** These medications help reduce the workload on the heart by promoting vasodilation and lowering blood pressure, which can relieve the strain on the left ventricle.
- **Beta blockers:** These can be used to control heart rate and reduce the heart's demand for oxygen, especially if arrhythmias are present.
- **Diuretics:** If there is fluid retention, as in the case of congestive heart failure, diuretics can be administered to reduce fluid accumulation in the body, improving symptoms such as edema and difficulty breathing.
- **Anticoagulants:** If there is an increased risk of blood clots, especially if the patient has atrial fibrillation or other risk conditions, anticoagulant medications may be indicated to prevent thromboembolic complications.

Monitoring and Regular Follow-up: In mild cases of mitral valve annulus disjunction or when there are no significant symptoms, treatment may be more conservative, with regular monitoring. Periodic examinations, such as echocardiograms, are necessary to monitor mitral valve function and the progression of mitral regurgitation, if present. Monitoring aims to identify any changes in cardiac function early and adjust treatment as necessary[7,9,16-21].

Surgical Treatment: In more severe cases, especially when mitral regurgitation is significant and symptoms do not respond to medical treatment, surgery may be necessary. Options include[7,9,16-21]:

- **Mitral valve repair:** Mitral valve repair is preferred whenever possible because it preserves the original valve and its function. During the procedure, the surgeon may

correct the valve annulus disjunction, reconstruct the mitral valve, or adjust the shape of the valve to improve its closure. The goal is to restore normal valve function and reduce regurgitation.

- **Mitral valve replacement:** When valve repair is not feasible or when there is irreversible damage, it may be necessary to replace the mitral valve with a mechanical or biological valve. This is usually indicated in cases of severe mitral regurgitation with a high risk of complications, such as heart failure or persistent atrial fibrillation.

Treatment of Arrhythmias: If mitral valve annulus disjunction is associated with arrhythmias, such as atrial fibrillation, treatment may involve controlling the heart rate with medication, as well as anticoagulation to prevent blood clots and embolisms[7,9,16-21].

Approaches to Underlying Causes

Mitral valve annular disjunction can occur in conditions such as Marfan syndrome, dilated cardiomyopathy, or other connective tissue diseases. In these situations, in addition to treatment directed at the mitral valve, it is important to treat the underlying condition that may be contributing to the disjunction[7,9,16-21]:

- Marfan syndrome: Treatment may involve the use of beta-blockers or other medications to control the risk of complications associated with connective tissue, such as aortic dilation.
- Dilated cardiomyopathy: Management of cardiomyopathy, often with the use of heart failure medications, is also crucial.

Treatment of mitral valve annulus disjunction is highly individualized and depends on the severity of symptoms and mitral valve function. In many cases, regular monitoring and medication control are sufficient, but when mitral regurgitation becomes severe or symptoms worsen, surgical intervention may be necessary to restore heart function and improve the patient's quality of life. It is essential that patients diagnosed with mitral valve annulus disjunction are closely monitored by cardiologists to ensure adequate management and avoid or minimize complications and long-term morbidity, with the consequent reduction in mortality[7,9,16-21].

MORBIDITY

The morbidity associated with mitral valve annulus disjunction is mainly related to mitral regurgitation, which is the most common clinical consequence of this condition. The most common complications that impact morbidity include[7,16,19,22]:
- Congestive Heart Failure: Severe mitral regurgitation can lead to blood pooling in the lungs (pulmonary edema), which impairs gas exchange and results in shortness of breath, extreme fatigue, and decreased functional capacity. This can lead to congestive heart failure, a chronic condition that affects the patient's quality of life.

- **Cardiac Arrhythmias:** Patients with mitral valve annulus disjunction, especially those with severe mitral regurgitation, have an increased risk of developing arrhythmias such as atrial fibrillation. Atrial fibrillation can further worsen heart function and increase the risk of blood clots, which can result in stroke or embolisms.
- **Peripheral Edema:** The accumulation of fluid in the lower limbs, due to the heart's failure to pump blood adequately, is a common manifestation in patients with severe mitral regurgitation. Edema can affect mobility and quality of life.
- **Reduced Functional Capacity:** Increased workload of the heart due to mitral valve annulus disjunction and mitral regurgitation may limit the patient's ability to perform physical activities. Shortness of breath, fatigue, and related symptoms hinder the ability to perform daily tasks.

MORTALITY

Mortality associated with mitral valve annular disjunction depends on several factors, including the severity of mitral regurgitation, the presence of comorbidities, and the effectiveness of treatment. Although the condition may be asymptomatic in the early stages, as mitral regurgitation worsens, mortality can increase significantly if not treated appropriately[7,8,16,22,23]:

- Heart Failure and Mortality: Heart failure is one of the major causes of mortality in patients with severe mitral regurgitation and mitral valve annulus disjunction. Progression of heart failure can lead to collapse of the cardiovascular system, with heart failure, which is a potentially fatal condition. In addition, severe mitral regurgitation is associated with an increased risk of sudden cardiac death, especially in patients with arrhythmias such as ventricular fibrillation or ventricular tachycardia.
- Thromboembolic complications: Patients with severe mitral regurgitation and atrial fibrillation have a high risk of developing blood clots, which can travel and cause thromboembolism in vital organs, such as the brain (stroke) or lungs (pulmonary embolism). These thromboembolic events increase mortality in patients with this condition.
- Surgical Interventions and Mortality: Mortality associated with surgical interventions to correct the mitral valve (such as valve repair or replacement) has been decreasing over the years due to advances in surgical techniques and medicine. However, there is still a risk associated with any invasive cardiac procedure, especially in older patients or those with multiple comorbidities. Postoperative mortality may be higher in cases where heart function is already severely compromised or in patients with other health conditions, such as hypertension or diabetes.

PROGNOSTIC FACTORS

The prognosis for patients with mitral valve annular disjunction is closely related to the presence and severity of mitral regurgitation. Some factors that may influence prognosis and mortality include[8,13,23]:

- Severity of Mitral Regurgitation: Patients with severe or advanced mitral regurgitation have a much higher risk of serious complications, such as congestive heart failure, arrhythmias, and sudden death.
- Presence of Underlying Diseases: Patients with pre-existing conditions, such as dilated cardiomyopathy, Marfan syndrome, or other connective tissue diseases, have a higher risk of complications and mortality due to mitral valve annulus disjunction.
- Surgical Intervention: Patients who undergo mitral valve repair or replacement surgery generally have a more

favorable prognosis, provided the intervention occurs in a timely manner and cardiac function has not been irreversibly impaired. However, delayed intervention may be associated with an increased risk of complications.

- Age and Comorbid Conditions: Patients of older age or with other comorbidities (such as hypertension, diabetes, or lung disease) tend to have a higher risk of serious complications and early death.

CONCLUSION

Mitral valve annular disjunction, in combination with mitral regurgitation, is a condition that can lead to significant morbidity and mortality, particularly if not diagnosed early or treated appropriately. Appropriate management, which may involve medication, regular monitoring and, in severe cases, surgery to repair or replace the mitral valve, is essential to improve the prognosis and reduce the risks associated with the condition. Early detection and effective treatment can improve the patient's quality of life and reduce mortality related to mitral regurgitation.

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