Mitral Stenosis

Shah SN, Sharma S.

Continuing Education Activity

Mitral stenosis (MS) is a form of valvular heart disease characterized by the narrowing of the mitral valve orifice. The most common cause of mitral stenosis is rheumatic fever, though the stenosis typically does not become clinically relevant until several decades later. Uncommon causes of mitral stenosis are calcification of the mitral valve leaflets and congenital heart disease. Other causes of mitral stenosis include infective endocarditis, mitral annular calcification, endomyocardial fibroelastosis, malignant carcinoid syndrome, systemic lupus erythematosus, Whipple disease, Fabry disease, and rheumatoid arthritis. This activity reviews mitral stenosis and the role of the interprofessional team in its management.

Objectives:

- Review the causes of mitral stenosis.
- Describe the characteristic exam of a patient with mitral stenosis.
- Outline the treatment for mitral stenosis.
- Explain modalities to optimize care coordination among interprofessional team members to improve outcomes for patients affected by mitral stenosis.

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Introduction

Mitral stenosis (MS) is a form of valvular heart disease. Mitral stenosis is characterized by narrowing of the mitral valve orifice. Today, the most common cause of mitral stenosis is rheumatic fever, but the stenosis usually appears clinically relevant only after several decades.

Etiology

The most common cause of mitral stenosis is rheumatic fever. Uncommon causes of mitral stenosis are calcification of the mitral valve leaflets and congenital heart disease. Other causes of mitral stenosis include infective endocarditis, mitral annular calcification, endomyocardial fibroelastosis, malignant carcinoid syndrome, systemic lupus erythematosus, Whipple disease, Fabry disease, and rheumatoid arthritis.

Epidemiology

The prevalence of rheumatic disease in developed countries is declining, with an estimated incidence of 1 in 100,000. The prevalence is higher in developing nations than in the United States. In Africa, for example, the prevalence is 35 cases per 100,000.

Rheumatic mitral stenosis is more common in females. The onset is usually between the third and fourth decades of life.[1][2]

Pathophysiology

The mitral valve is a bileaflet valve positioned between the left atrium and left ventricle. The normal mitral orifice area is 4 to 6 square centimeters. Under normal physiologic conditions, the mitral valve opens during the left ventricular diastole to allow blood to flow from the left atrium to the left ventricle. The pressure in the left atrium and the left ventricle during diastole are equal. The left ventricle gets filled with blood during early ventricular diastole. Only a small amount of blood remains in the left atrium. This small amount of blood fills the left ventricle with the contraction of the left atrium (the "atrial kick") during late ventricular diastole.[3][4][5]

Mitral valve areas less than 2 square centimeters cause an impediment to the blood flow from the left atrium into the left ventricle. This creates a pressure gradient across the mitral valve. As the gradient across the mitral valve increases, the left ventricle requires the atrial kick to fill with blood.

Mitral stenosis causes an increase in left atrial pressure. The normal left ventricular diastolic pressure is 5 mmHg. A pressure gradient across the mitral valve of 20 mmHg due to severe mitral stenosis will cause a left atrial pressure of about 25 mmHg. This left atrial pressure is transmitted to the pulmonary vasculature resulting in pulmonary hypertension.

As left atrial pressure remains elevated, the left atrium will increase in size. As the left atrium increases in size, there is a greater chance of developing atrial fibrillation. If atrial fibrillation develops, the atrial kick is lost.

Thus, the left ventricular filling depends on the atrial kick in severe mitral stenosis. With the loss of the atrial kick, there is a decrease in cardiac output and sudden development of congestive heart failure.

Mitral stenosis progresses slowly from initial signs of mitral stenosis to NYHA functional class II symptoms to atrial fibrillation to NYHA functional class III or IV symptoms.

History and Physical

Mitral stenosis presents 20 to 40 years after an episode of rheumatic fever. The most common symptoms are orthopnea and paroxysmal nocturnal dyspnea. Patients may have symptoms of palpitations, chest pain, hemoptysis, thromboembolism when the left atrial volume is increased, ascites, edema, and hepatomegaly (if right-side heart failure develops).

There is also an increase in symptoms of fatigue and weakness with exercise and pregnancy.

On auscultation, the first heart sound is usually loud and maybe palpable due to increased force in closing the mitral valve.

The P2 (pulmonic) component of the second heart sound (S2) will be loud if severe pulmonary hypertension is due to mitral stenosis.

An opening snap (OS) is an additional sound that may be heard after the A2 component of the second heart sound (S2). This is the forceful opening of the mitral valve when the pressure in the left atrium is greater than the pressure in the left ventricle.

A mid-diastolic rumbling murmur with presystolic accentuation is heard after the opening snap. This murmur is a low-pitched sound. It is best heard with the bell of the stethoscope at the apex. The murmur accentuates in the left lateral decubitus position and with isometric exercise. (Listen to audio)

Advanced mitral stenosis presents with signs of right-sided heart failure (jugular venous distension, parasternal heave, hepatomegaly, ascites) and/or pulmonary hypertension.

Other signs include atrial fibrillation, left parasternal heave (right ventricular hypertrophy due to pulmonary hypertension), and tapping the apical beat.

Evaluation

Mitral stenosis is evaluated using noninvasive and invasive measures. Noninvasive tests are electrocardiograms (ECG), chest X-rays, echocardiograms, and exercise echocardiograms. An invasive test for mitral stenosis would include a cardiac catheterization.[6][7][8]

On the ECG, the P wave changes suggest left atrial enlargement. The presence of right axis deviation and right ventricular hypertrophy suggests severe pulmonary hypertension. ECG frequently detects atrial arrhythmias such as atrial fibrillation.

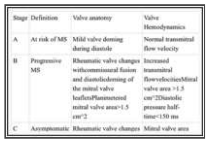
On the chest X-ray, the early stages of mitral stenosis findings are normal heart size, straightening of the left border of the cardiac silhouette, prominent main pulmonary arteries, dilatation of the upper pulmonary veins, and displacement of the esophagus by an enlarged left atrium. During the severe chronic stage of mitral stenosis, the chest X-ray will have enlargement of all the chambers, pulmonary arteries, and pulmonary veins.

An echocardiogram is useful for assessing mitral stenosis etiology, morphology, severity, and treatment intervention. The analysis of the morphology of the mitral valve apparatus includes leaflet mobility and flexibility, leaflet thickness, leaflet calcification, subvalvular fusion, and the appearance of commissures. The Wilkins score grades each of the components of the mitral apparatus from 1 to 4: leaflet mobility, thickness, calcification, and impairment of the subvalvular apparatus. The Padial score grades the leaflet thickening (separately), the commissural calcification, and the subvalvular disease from 1 to 4. The Wilkins score of less than 8, Padial scores of less than 10, and less than moderate regurgitation have better outcomes after a catheter-based balloon mitral valvotomy procedure.

An exercise echocardiogram is performed using an upright treadmill or supine bicycle with Doppler recording of transmitral and tricuspid valve velocities. This measures the transmitral gradient and pulmonary artery systolic pressure at rest and with exercise.

Cardiac catheterization is an invasive procedure. Cardiac catheterization should be performed to assess the severity of mitral stenosis when noninvasive tests are inconclusive or when there is a discrepancy between noninvasive tests and clinical findings regarding the severity of mitral stenosis (Class I, Level of Evidence C).

Classification of Severity of Mitral Valve Stenosis (Adapted from the ACC/AHA 2020 Valve Guidelines)



Stage	Definition	Valve anatomy	Valve Hemodynamics
A	At risk of MS	Mild valve doming during diastole	Normal transmitral flow velocity
B	Progressive MS	Rheumatic valve changes with commissural fusion and diastolic doming of the mitral valve leaflets Planimetered mitral valve area >1.5 cm^2	Increased transmitral flow velocities Mitral valve area >1.5 cm^2 Diastolic pressure half-time <150 ms
C	Asymptomatic	Rheumatic valve changes	Mitral valve area

Table

Mitral Valve Anatomy According to the Wilkins Score

Grade 1

- Mobility: Highly mobile valve with only leaflet tips restricted
- Thickening: Leaflet near normal in thickness (4 mm to 5 mm)
- Calcification: A single area of increased echo brightness
- Subvalvular Thickening: Minimal thickening just below the mitral leaflets

Grade 2

- Mobility: Leaflet mid to base portions have normal mobility
- Thickening: Mid leaflets normal, considerable thickening of margins (5 to 8 mm)
- Calcification: Scattered areas of brightness confirmed to leaflet margins
- Subvalvular Thickening: Thickening of chordal structures extending to one of the chordal length

Grade 3

- Mobility: Valve continues to move forward in diastole, mainly from the base
- Thickening: Thickening extending through the entire leaflet (5 mm to 8 mm)
- Calcification: Brightness extending into the mid portions of the leaflets
- Subvalvular Thickening: Thickening extended to the distal third of the chords

Grade 4:

- Mobility: No or minimal forward movement of the leaflets in diastole
- Thickening: Considerable thickening of all leaflet tissue (more than 8 mm to 10 mm)
- Calcification: Extensive brightness throughout much of the leaflet tissue
- Subvalvular Thickening: Extensive thickening and shortening of all chordal structures extending down to the papillary muscles.

Treatment / Management

Treatment for mitral stenosis involves medical therapy, percutaneous mitral valvuloplasty, and surgical therapy. Currently, no medical therapy can relieve a fixed obstruction of the mitral valve. Medical therapy is focused on preventing endocarditis, decreasing new cases of rheumatic fever, improving symptoms, and decreasing the thromboembolic risk.[9][10][11]

Endocarditis prophylaxis should only be given to high-risk patients before dental procedures involving gingival tissue manipulation or perforation of the oral mucosa. High-risk patients are those patients with a prosthetic heart valve or prosthetic material used for valve repair, previous history of infective endocarditis, and cardiac valvuloplasty.

Rheumatic fever prophylaxis with Benzathine penicillin is the primary prevention treatment in patients with streptococcal pharyngitis.

If the rhythm is normal sinus, medical therapy is used to improve symptoms. Diuretics are utilized to help relieve congestion. Beta-blockers and/or calcium channel blockers help with exertional symptoms associated with elevated heart rate.

If the rhythm is atrial fibrillation, the first step is to control the rate using AV node blocking agents such as beta-blockers, calcium channel blockers, and/or digitalis. In an unstable patient, perform direct current cardioversion. If one cannot convert atrial fibrillation to normal sinus rhythm, then the primary goal is rate control. In a stable patient, restoring normal sinus rhythm is preferred over rate control to improve functional capacity and quality of life.

Anticoagulation prevents thromboembolic events. Anticoagulation is indicated in patients with mitral stenosis and atrial fibrillation (paroxysmal, persistent, or permanent), previous embolic events, and the presence of left atrial thrombus. At present, Warfarin is the anticoagulation of choice.

Aspirin or other antiplatelet drugs are not approved to decrease thromboembolic risk in mitral stenosis. Warfarin should be monitored using the international normalized ratio (INR) to target 2.5.

Percutaneous mitral balloon valvuloplasty (PMBV) is an invasive procedure to manage mitral stenosis. PMBV improves symptoms by increasing the mitral valve area and reducing the mitral valve gradient. PMBV is indicated in symptomatic patients (New York Heart Association functional class greater than II) or asymptomatic patients with pulmonary hypertension with moderate or severe stenosis and favorable valve morphology in the absence of left atrial thrombus or moderate to severe mitral regurgitation.

Mitral valve replacement surgery is indicated in patients with symptomatic moderate or severe mitral stenosis when percutaneous mitral balloon valvuloplasty is contraindicated or unfavorable valve morphology (Class I, Level of Evidence B).

Differential Diagnosis

- Left atrial myxoma
- Endocarditis

Prognosis

Mitral valve stenosis can remain asymptomatic for years, especially when resulting from rheumatic fever. It can take decades following rheumatic fever before mitral valve stenosis develops.[12]

Once symptoms become apparent, the progression of the disease generally accelerates, particularly when it is secondary to rheumatic fever. Approximately 80% of patients will not survive ten years from symptomatic onset. In patients with pulmonary hypertension secondary to mitral valve stenosis, survival is around three years. Heart failure often accompanies advanced cases.

The outlook for children born with mitral valve stenosis depends large on the severity of their condition, often requiring screening for related heart problems throughout their lives.[13]

Complications

- Heart failure
- Stroke
- Failure to thrive
- Pulmonary hypertension
- Endocarditis

Consultations

Cardiac surgery consultations should be obtained in cases of severe mitral stenosis to evaluate the patient for mitral valve replacement if the patient is ineligible for the PMBV procedure.

Deterrence and Patient Education

Patients with advanced mitral valve stenosis may need to limit their activity to prevent overtaxing their hearts. Patients may also be given certain dietary restrictions, such as a low-sodium diet.

All patients with mitral valve stenosis, regardless of severity, may require medications to prevent complications like strokes, hypertension, and heart failure; these are usually necessary for the individual's lifespan, and the importance of medication adherence must be stressed to these patients.[14]

Pearls and Other Issues

Pregnancy during mitral stenosis will increase the patient’s symptoms by one New York Heart Association class. Medical therapy is attempted first to improve symptoms. If symptoms do not improve with medical treatment, refer the patient for percutaneous mitral balloon valvuloplasty.

Enhancing Healthcare Team Outcomes

Mitral stenosis is a relatively common disorder, which, if left untreated, is associated with high morbidity and mortality. Mitral stenosis rates had decreased in the US four decades ago, but with the mass migration of individuals from other countries, a resurgence of cases has been observed. The

number one cause for most cases of mitral stenosis is rheumatic fever. The key is to prevent the valvular disorder by ensuring that patients with strep throat are adequately treated. Because of the high morbidity of the disorder, the condition is best managed by an interprofessional team.

Once the diagnosis of mitral stenosis is made, the patient should be educated about the need for surgery. Those who remain asymptomatic will need annual exams, including echocardiograms. Patients with palpitations may require a Holter monitor to confirm the presence of atrial arrhythmias. Many of these patients will need anticoagulation with warfarin. Hence, the pharmacist should ensure that monthly blood work is done to ensure therapeutic anticoagulation. A dietary consult is often important as failure to thrive is common. A cardiology nurse should monitor the patient for symptoms as they will require surgery. The type of prosthetic valve used depends on patient factors.

Cardiac surgery nurses should educate the patient on the type of prosthetic valves available and which may be best suited for them. Finally, the cardiologist, primary care provider, pharmacist, and nurse should discuss with the patient the possibility of developing an infection of the valve and the need for prophylaxis when undergoing any invasive procedure. Because atrial fibrillation is a persistent problem, anticoagulation will be required in most patients. Hence, close monitoring of the INR by a dedicated nurse is necessary.[15][16] All interprofessional team embers must monitor the patient's progress and status and record any changes in the patient's condition in their permanent medical record. Further, it is incumbent on them to reach out to the appropriate team embers in case additional interventions are necessary. This interprofessional paradigm will drive improved patient outcomes. [Level 5]

Outcomes

Prior to the era of open-heart surgery, the prognosis for most patients with mitral stenosis was poor. In the era of mitral valve replacement, the prognosis is excellent. Survival is significantly better for patients undergoing an open mitral valve replacement compared to a commissurotomy. Today, there is an 80% survival at ten years, but in patients who have developed pulmonary hypertension, the survival is less than three years. Other complications that may result in high morbidity include stroke and persistent atrial fibrillation.[17][18] [Level 5]

Review Questions

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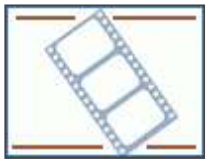

Figure
Mitral Stenosis. Contributed by Katherine Humphreys

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Author Information

Authors

Sandy N. Shah; Saurabh Sharma[1].

Affiliations

[1] Guthrie Robert Packer Hospital

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