



Challenges in Cardiology: Mixed Valvular Heart Disease

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Introduction

Valvular heart disease is either in form of stenosis or regurgitation that result from acquired pathology or related to congenital cardiac lesions. Mixed valvular disease is referred to having more than one valvular lesion together that may challenge the proper estimation of each valve severity. Previously, multivalvular disease with secondary effects on cardiac chambers, used to be attributed to rheumatic heart disease, which is the commonest cause of mitral stenosis. Degenerative changes related to aging are another important cause of significant calcification & valvular dysfunction. Severe mitral annular calcification can result in mitral regurge due to decreased annular shortening during systole or mitral stenosis caused by extension of the calcification onto the leaflets causing restricted valve opening. Other less common causes of multivalvular disease are valvular endocarditis, myxomatous degeneration, mediastinal radiation, carcinoid heart disease & marfan's syndrome [1,2].

Pathophysiology

The pathophysiologic derangements associated with the more proximal valve disease can mask the full expression of the more distal valve lesion. For example, in patients with rheumatic mitral & aortic valves disease, the reduction in cardiac output imposed by the mitral valve disease will decrease the magnitude of the hemodynamic derangements related to the severity of the aortic valve lesion. Alternatively, the development of atrial fibrillation during the course of mitral stenosis can lead to sudden worsening in a patient whose aortic valve disease was not previously felt to be significant [3].

The hemodynamically more severe lesion causes the predominant clinical features; however, assuming similar degrees of valvular severity, symptoms are related to the disease of the more proximal (upstream) valvular lesion. The primary valve lesion can create secondary

(“back pressure”) effects on the secondary valve lesion, for example chronic severe mitral regurge is leading to right ventricular dilation, secondary functional tricuspid regurge, and pulmonary hypertension. In advanced multivalvular disease, symptoms of heart failure are often present, and are accompanied by secondary hemodynamic effects on the cardiac chambers [1-3].

Clinical Features

Symptoms may develop at a relatively earlier stage in the natural history of their disease. Symptoms such as exertional dyspnea and fatigue are usually related to elevated filling pressures, reduced cardiac output, or their combinations. Palpitations may signify atrial fibrillation and identify mixed valvular disease as an important component of the clinical presentation, even when not previously suspected.

Chest pain compatible with angina could reflect: left or right ventricular oxygen supply & demand mismatch on a substrate of hypertrophy and pressure/volume overload, with or without superimposed coronary artery disease. Symptoms related to right heart failure abdominal fullness, bloating and edema are late manifestations of advanced disease [1].

Diagnosis

The diagnostic workup of multivalvular heart disease

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is not significantly different from that of individual valvular lesions.

Electrocardiogram signs indicative of right-sided cardiac abnormalities in patients with left-sided valve lesions should prompt additional assessment for pulmonary hypertension and/or right-sided valve disease. The presence of atrial fibrillation in patients with aortic valve disease may be a clue to the presence of previously unsuspected mitral valve disease in the appropriate context [1].

Echocardiography is the most commonly used imaging modality for the diagnosis and characterization of mixed valvular disease & may often demonstrate findings not clinically suspected. An integrated assessment of the clinical and transthoracic echocardiography findings is needed to help determine the dominant valve lesion(s) and establish an appropriate plan for treatment and follow-up.

Multivalvular Disease has a significant impact on other echocardiographic parameters. In any lesion with higher-pressure gradient will affect the color jet area for the other lesion: e.g., aortic stenosis will increase the mitral regurgite regurgitant volume. Increases pulmonary arterial pressure (from any lesion), will worsen tricuspid regurgite jet area. Severe aortic regurgite will further dilates left ventricle, increase functional mitral regurgite by increasing the size of the regurgitant orifice area. In contrast, severe aortic regurgite will further dilate left ventricle, making the mitral valve prolapse less pronounced. Methods of assessment as flow convergence and vena contracta should generally not be influenced by presence another regurgitant jet [4].

All the volumetric methods for quantifying valvular regurgitation require a reference stroke volume, in mitral & aortic regurgite the left ventricle stroke volume includes both the valves regurgitant volumes (RVols), subsequently reference stroke volume is from the right heart. If all four valves have important regurgitation, or a shunt is present from an atrial or ventricular septal defects, then there may simply be no reference systemic stroke volume to use. In such cases, direct measurement of stroke volume is most reliably performed with Cardiac magnetic resonance [4,5].

When severe aortic regurgite accompanies aortic stenosis, as the high trans-aortic volume flow rate, maximum velocity, and mean gradient will be higher than expected for a given valve area. In this situation, reporting accurate quantitative data for the severity of both stenosis and regurgitation is helpful for clinical decision-making. Patients with significant aortic stenosis, a non-dilated left ventricle chamber, and concentric hypertrophy will poorly tolerate the abrupt development of aortic regurgite, as may occur, for example, with infective endocarditis

or after aortic valve replacement (AVR) complicated by Para-valvular leakage. The noncompliant left ventricle is not prepared to accommodate the sudden volume load, and as a result, left ventricle diastolic pressure rises rapidly and severe heart failure develops [4].

In mixed aortic or mitral valve disease when aortic or mitral regurgite is the dominant lesion the left ventricle is dilated. While when aortic or mitral stenosis predominates, left ventricle size will be normal or small. Despite that it can sometimes be difficult to ascertain whether stenosis or regurgitation is the dominant lesion in patients with mixed valve disease, yet an integrated clinical and noninvasive assessment can usually help.

In severe mitral stenosis accompanies aortic valve disease, low cardiac output will result and, therefore, low flow low gradient (LFLG) aortic stenosis status is expected [4]. When severe mitral regurgite accompanies aortic stenosis, first, with severe mitral regurgite, trans-aortic flow rate may be low resulting in a low gradient even when severe aortic stenosis is present; however valve area calculations remain accurate. Second, CW Doppler may mistake a high-velocity mitral regurgite jet for the aortic stenosis jet, as both are systolic signals directed away from the apex. In contrast, Mitral regurgite signal is longer in duration, starting with mitral valve closure and continuing until mitral valve opening. Surgical AVR or trans catheter AVR often, but not always, results in reduction or elimination of the functional mitral regurgite. Persistence of significant mitral regurgite following AVR is associated with reduced survival. Most surgeons' advocate for repair of moderate-to-severe or severe functional mitral regurgite at time of surgical AVR [4].

Functional tricuspid regurgite in the setting of significant mitral valve disease is one of the most common examples of mixed valvular disease. Functional tricuspid regurgite occurs as a consequence of right ventricle & annular dilation as pulmonary hypertension is often present. The tricuspid leaflets are morphologically normal [2,4].

Exercise testing with or without imaging can be useful when the degree of functional limitation reported by the patient is not adequately explained by the findings on transthoracic echocardiography performed at rest. For more accurate assessment particularly in mitral valve disease, Trans-esophageal echocardiography may be required. Cardiac magnetic resonance provides additional anatomic and physiologic information when echocardiography proves suboptimal, but less well suited to the evaluation of valve morphology. Cardiac computed tomography is less used in valvular heart disease however it is useful in assessing intra-cardiac structures in patients with complicated endocarditis [4-6].

Right and left heart catheterization may be required to characterize more completely the individual contributions of each lesion. Measurement of pulmonary artery pressures and calculation of pulmonary vascular resistance can help inform clinical decision-making in certain patient subsets, such as those with advanced mitral and tricuspid valves disease. Attention to the accurate assessment of cardiac output is essential [4].

Management

Management can be challenging. Initial steps include determine the dominant valve lesion to proceed accordingly. Then To establish indications for surgical treatment, the clinical and hemodynamic significance of the multi-valvular heart disease components must be assessed. Any defect assessed separately may not be severe enough to warrant surgical treatment; however, when combined, these may have significant hemodynamic consequences that require surgery.

Double-valve replacement is generally associated with higher perioperative and long-term risks, owing to a more complex surgical technique and longer cardiopulmonary bypass times, compared to single valve repair or replacement with or without coronary artery bypass graft (CABG) surgery; thus, whenever feasible, repair is often preferred. Risk factors that reduce long-term survival after double-valve replacement include advanced age, less favorable functional status, decreased left ventricle ejection fraction, greater left ventricle enlargement, and accompanying ischemic heart disease requiring CABG. In view of the higher risks, a higher threshold is required for multivalvular versus single-valve surgery [4-6].

Trans-catheter treatments for double mixed valve disease can be considered provide no contraindication. Percutaneous mitral balloon valvuloplasty (PMBV) for mitral stenosis is helpful however it is contraindicated in presence of moderate or severe mitral regurgite. Trans-catheter AVR for aortic stenosis was not done in the initial PARTNER trials in the presence of severe, coexistent aortic regurgite. *Though, combining both Trans-catheter AVR for aortic stenosis and Mitra-Clip for functional mitral regurgite has been reported.* Further advances in trans catheter treatments for multiple and mixed valve disease are anticipated [5,6].

Triple-Valve Disease is Uncommon & typically caused by rheumatic heart disease. Patients with trivalvular disease may present in a picture of advanced heart failure with marked cardiomegaly and, in these cases, surgical correction of all three valvular lesions is impera-

tive. When multiple prosthetic valves must be inserted, it is logical to select two bio-prostheses or two mechanical prostheses for the left side of the heart [5].

It is important to recognize that management of multivalve disease is essentially an “evidence-free zone,” and thus recommendations in the most current 2017 American College of Cardiology and American Heart Association guidelines are brief and consensus-based.

Conclusion

The patho-physiologic derangements associated with the more proximal valve disease can mask the full expression of the attributes of the more distal valve lesion. Trans-esophageal echocardiography may be required for, more accurate assessment especially when endocarditis is considered. Despite a detailed noninvasive and invasive workup, the decision to treat more than one valve is challenging and often is made on the basis of findings on palpation or direct inspection at the operating table. We think that a higher threshold is required for multivalvular versus single-valve surgery. Given the lack of high-quality data on mixed and multiple valve diseases and the virtual absence of randomized clinical trials on this topic, evidence-based recommendations for double valve intervention cannot be made.

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