38 - 276 Mitral Valve Prolapse

276 Mitral Valve Prolapse

edge-to-edge repair (TEER) versus surgical repair are ongo ing to determine if the extension of clip technology to low- and intermediate-risk surgical patients with significant degenerative MR is appropriate. The TEER system is also approved for the treatment of heart failure patients with secondary MR. The results of transthoracic and transesophageal echocardiographic imaging are critical to patient selection, along with an assessment of the adequacy of GDMT for heart failure. The use of TEER with a clip device in addition to medical therapy was shown to be superior to medical therapy alone in a trial involving symptomatic heart failure patients with reduced EF and at least moderately severe secondary MR followed through 5 years. Patients treated with the clip device had significantly lower rates of heart failure hos pitalizations and all-cause mortality than those treated medically. This was the first randomized trial to show a survival benefit in patients with heart failure and secondary MR. There has been a rapid expansion over the past 5 years in both the number of sites offering mitral valve TEER and the number of cases reported to the Society of Thoracic Surgeons/American College of Cardiology Transcatheter Valve Therapy Registry. Other transcatheter approaches to mitral valve repair have included the deployment of a device within the coronary sinus that can be adjusted to reduce mitral annular circumference and the effective orifice area of the valve much like a surgically implanted ring. Variations in the anatomic relationship of the coronary sinus to the mitral annulus and circumflex coronary artery have limited the applicability of this technique. Attempts to reduce the septal-lateral dimension of a dilated annulus using adjustable cords placed across the LV in a subvalvular location have been investigated. Construction of neochords to the mitral leaflets under TEE guidance using a system delivered via the cardiac apex has also been studied. Investigational experience with transcatheter mitral valve replacement systems remains in early clinical stages, although the field is evolving rapidly. Many high surgical risk patients are not candidates for transcatheter mitral valve repair, and thus, there is keen interest in refining this technology. Challenges with transseptal delivery and LV outflow tract obstruction from the devices used have prompted iterative changes in the systems utilized in early feasibility studies. ■ ■FURTHER READING Badhwar V et al: Risk of mitral valve repair for primary mitral regur gitation. J Am Coll Cardiol 81:636, 2023. Bonow RO et al: 2020 focused update of the 2017 expert consensus decision pathway on the management of mitral regurgitation. J Am Coll Cardiol 75:2236, 2020. Lim DS et al: Randomized comparison of transcatheter edge-to-edge repair for degenerative mitral regurgitation in prohibitive surgical risk patients. J Am Coll Cardiol Interv 15:2523, 2022. Otto CM et al: 2020 ACC/AHA guideline for the management of patients with valvular heart disease: A report of the American Col lege of

Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. Circulation 143:e72, 2021. Stone GW et al: Five-year follow-up after transcatheter repair of sec ondary mitral regurgitation. N Engl J Med 388:2037, 2023. von Bardeleben RS et al: 1-year outcomes with fourth generation mitral valve transcatheter edge-to-edge repair from the EXPAND G4 study. J Am Coll Cardiol Interv 16:2600, 2023. Zahr F et al: 1-year outcomes following transfemoral transseptal transcatheter mitral valve replacement: Intrepid TMVR early feasibil ity study results. J Am Coll Cardiol Interv 16:2868, 2023.

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Mitral Valve Prolapse Mitral Valve Prolapse CHAPTER 276 The role of the physical examination in the evaluation of patients with valvular heart disease is also considered in Chaps. 44 and 246; of elec trocardiography (ECG) in Chap. 247; of echocardiography and other noninvasive imaging techniques in Chap. 241; and of cardiac catheter ization and angiography in Chap. 249. MITRAL VALVE PROLAPSE Mitral valve prolapse (MVP), also variously termed the systolic clickmurmur syndrome, Barlow's syndrome (Fig. 276-1), floppy-valve syndrome, and billowing mitral leaflet syndrome, is a relatively common but highly variable clinical syndrome resulting from diverse pathologic mecha nisms affecting the mitral valve apparatus. Among these are excessive or redundant mitral leaflet tissue, which is commonly associated with myxomatous degeneration and greatly increased concentrations of certain glycosaminoglycans. MVP is the most common abnormality leading to primary mitral regurgitation (MR) and mitral valve repair surgery (see Chap. 275). In most patients with MVP, the cause is unknown, but in some, it appears to be genetically determined. A reduction in the production of type III collagen has been implicated, and electron microscopy has revealed fragmentation of collagen fibrils. A meta-analysis of six B A C FIGURE 276-1 Mitral valve prolapse. Myxomatous thickening and prolapse of the mitral valve can occur in isolation in 2–3% of the general population or may be associated with heritable connective tissue disorders, such as Marfan syndrome. Myxomatous degeneration of the valve predisposes to severe regurgitation and chordal rupture and is a frequent indication for mitral valve repair or replacement. Prolapse can affect one or both leaflets, to varying degrees. A. Three-dimensional transesophageal echocardiogram showing a myxomatous mitral valve from the left atrial en face aspect. There is billowing and prolapse of the entire middle scallop of the posterior leaflet (asterisk). (Figure courtesy of Douglas C. Shook, MD, Department of Anesthesiology, Perioperative and Pain Medicine, Brigham and Women's Hospital.) B. The posterior leaflet of the mitral valve demonstrates marked prolapse and hooding in all segments and severe redundancy in this postmortem photograph taken from the vantage point of the left atrium. C. Opening the left heart reveals prominent mitral leaflet hooding (arrows). The chordae are focally thickened but are not fused as would be the case in rheumatic valve disease. (Used with permission from JC Wu, RF Padera: Clinicopathologic correlates, in Atlas of Echocardiography, 2nd ed, SD Solomon [ed], E Braunwald [series ed]. Philadelphia, Current Medicine Group LLC, 2008. p 363.)

PART 6 Disorders of the Cardiovascular System genome-wide association studies and 4884 MVP cases identified 14 genetic loci associated with MVP. Candidate genes included LMCD1, SPTBN1, LTBP2, TGFB2, NMB, and ALPK3. A polygenic risk score improved the performance of a clinical risk prediction model for the development of MVP, although overall performance was modest. Other work has identified genetic associations with LMNA, FLNC, and FLNA, which are often expressed as cardiomyopathies. MVP is a frequent finding in patients with heritable disorders of connective tissue, including Marfan syndrome (Chap. 425), osteogen esis imperfecta, and Ehlers-Danlos

syndrome. MVP may be associated with thoracic skeletal deformities similar to but not as severe as those in Marfan syndrome, such as a high-arched palate and alterations of the chest and thoracic spine, including kyphosis and the so-called straight back syndrome. Other associated features can include a his tory of inguinal hernias, joint dislocations, meniscal tears, and easy bruisability. In most patients with MVP, myxomatous degeneration is confined to the mitral valve, although the tricuspid and aortic valves may also be affected. Prolapse can affect one or both leaflets. The posterior mitral leaflet is usually more affected than the anterior, and the mitral valve annulus is often dilated. In many patients, elongated, redundant, or ruptured chordae tendineae cause or contribute to the regurgitation. MVP also may occur rarely as a sequel to acute rheumatic fever, in ischemic heart disease, and in various cardiomyopathies (see above), as well as in 20% of patients with ostium secundum atrial septal defect. MVP may lead to excessive stress on the papillary muscles, leading to localized ischemia, infarction, and replacement fibrosis. The latter, which may be a nidus for ventricular arrhythmias, may be visible on cardiac magnetic resonance imaging as late gadolinium enhancement and occurs in the absence of coronary artery disease. Rupture of chordae tendineae and progressive annular dilation and calcification contribute to valvular regurgitation, which then places more stress on the diseased mitral valve apparatus, thereby creating a vicious cycle. ■ ■CLINICAL FEATURES MVP is more common in women than men and occurs most fre quently between the ages of 15 and 30 years; the clinical course is most often benign. MVP may also be observed in older (>50 years) patients, often men, in whom MR is often more severe because of chordal rupture and requires surgical treatment. There is an increased familial incidence for some patients, suggesting an autosomal dominant form of inheritance with incomplete penetrance. MVP varies in its clinical expression, ranging from only a systolic click and murmur with mild prolapse of the posterior leaflet to severe MR due to chordal rupture and leaflet flail. The degree of myxomatous change of the leaflets can also vary widely. In many patients, the condition progresses over years or decades; in others, it worsens rapidly as a result of chordal rupture or endocarditis. Most patients are asymptomatic and remain so for their entire lives. However, in North America, MVP is now the most com mon cause of isolated severe MR requiring surgical treatment. Arrhythmias, most commonly ventricular premature contractions and paroxysmal supraventricular and ventricular tachycardia, as well as atrial fibrillation (AF), have been reported and may cause palpitations, light-headedness, and syncope. Sudden death is a very rare complication and occurs most often in patients with severe MR and depressed left ventricle (LV) systolic function, although it can occur in individuals with normal LV size and function. A small subset of MVP patients with high-grade ventricular ectopy has been identified with phenotypic features including electrocardiographic inferior-apical T-wave abnormalities, high-density premature ven tricular complexes at rest, mitral annular disjunction (defined as abnormal atrial displacement of the mitral valve leaflet hinge point), and papillary muscle fibrosis on cardiac magnetic resonance imaging (see above). In addition, there may be an excess risk of sudden death among patients with a flail leaflet. Most of these patients have severe MR. Many patients with MVP have chest pain that can be difficult to evaluate; it is often substernal, prolonged, and not related to exertion, FIGURE 276-2 Barlow’s valve with classic mitral valve prolapse, as seen on transthoracic echocardiogram in parasternal long-axis windows. Left: parasternal long-axis window, showing both myxomatous leaflets (arrows) billowing into the left atrium in late systole. Right: same window with color Doppler showing significant mitral regurgitation (arrow) in systole. (Courtesy of Justina Wu, MD, PhD.) but may rarely resemble angina pectoris. Transient cerebral ischemic attacks secondary to emboli from the mitral valve due to endothelial disruption have been reported. Infective endocarditis may occur in patients with MR and/or leaflet thickening. Auscultation A frequent finding is the mid- or late (nonejection) systolic click, which occurs 0.14 s

or more after S1 and is thought to be generated by the sudden tensing of slack, elongated chordae ten dineae or by the prolapsing mitral leaflet when it reaches its maximal excursion. Systolic clicks may be multiple and may be followed by a high-pitched, mid-late systolic crescendo–decrescendo murmur, which occasionally is “whooping” or “honking” and is heard best at the apex. Radiation of the murmur will depend on the involved leaf let. With posterior leaflet prolapse, the jet of MR is directed anteriorly and the murmur will radiate to the base of the heart. With anterior leaflet involvement, the jet of MR is directed posteriorly and the mur mur will radiate to the axilla and back. The click and murmur occur earlier with standing, during the strain phase of the Valsalva maneu ver and with any intervention that decreases LV volume (preload), exaggerating the propensity of the leaflet to prolapse. Conversely, squatting and isometric exercises, which increase LV volume, dimin ish MVP; the click-murmur complex is delayed, moves away from S1, and may even disappear. Some patients have a mid-systolic click without a murmur; others have a murmur without a click. Still others have both sounds at different times. LABORATORY EXAMINATION The ECG most commonly is normal but may show biphasic or inverted T waves in leads II, III, and aVF and, occasionally, supra ventricular or ventricular premature beats. Transthoracic echocar diography (TTE) is particularly effective in identifying the abnormal position and prolapse of the mitral valve leaflets. A useful echocar diographic definition of MVP is systolic displacement (in the para sternal long axis view) of the belly of the mitral valve leaflets by at least 2 mm into the left atrium (LA) superior to the plane of the mitral annulus. There can be prolapse of one or both leaflets (Fig. 276-2). Color flow and continuous wave Doppler imaging is helpful to evaluate the associated MR and provide estimates of severity. The jet lesion of MR due to MVP is most often eccentric, and assessment of the effective regurgitant orifice area and regurgitant volume can be difficult with standard techniques. Both three-dimensional echocardiography and cardiac magnetic resonance imaging can provide more precise deter minations of LV volumes. Transesophageal echocardiography (TEE) is indicated when more accurate anatomic information is required and is performed routinely for intraoperative guidance during surgi cal or transcatheter valve repair. Exercise testing can be performed when there is uncertainty regarding functional capacity. It is often combined with rest and immediate poststress TTE to assess LV and right ventricular (RV) function and the dynamic nature of MR and pulmonary artery pressures. Left ventriculography done at the time

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