## FLOPPY MITRAL VALVE SYNDROME

## (MITRAL VALVE PROLAPSE)

### DEFINITION

- 1. **Floppy mitral valve syndrome** is the commonest cause of pure mitral regurgitation. The condition is probably responsible for a significant proportion of patients described as having **mitral valve prolapse** or the **mid-systolic click late systolic murmur syndrome**. The diagnosis can usually be confirmed by echocardiography.
- 2. The enlarged, lax leaflets of the mitral valve may billow into the left atrium during ventricular systole, not always with regurgitation. Prolapse implies systolic displacement of commissural surfaces of one or both leaflets towards the atrium, usually accompanied by regurgitation.
- 3. Prevalence varies between 1% and 6% in otherwise normal populations. The condition is more common in people above 50 years of age, with a female preponderance. Mitral valve prolapse is the most common form of congenital valvular heart disease.

#### **CLINICAL MANIFESTATIONS**

- 4. The clinical features are very variable depending on the degree of mitral regurgitation and the presence or absence of left ventricular disease. The pure lesion is well tolerated. Regurgitation is not always present and, even when it does occur, mild cases may still be asymptomatic for many years. In about 40% of cases of mitral valve prolapse, tricuspid valve prolapse is also present. The pulmonary and aortic valves are affected in about 10% and 2% of cases respectively.
- 5. The early symptoms are palpitations, fatigue and dyspnoea. Dizziness, orthostatic hypotension, non-anginal chest pain and latent tetany may also occur. If mitral regurgitation becomes severe, congestive heart failure develops.
- 6. Complications are less common than in mitral stenosis, but mitral prolapse is associated with an increased incidence of infective endocarditis. There is an increased incidence of cardiac arrhythmias which are usually relatively benign, but may be frequent and bizarre in a small minority of patients. Patients with mitral prolapse are also at a slightly increased risk of embolic strokes and transient ischaemic attacks.
- 7. Where atrial fibrillation occurs, anticoagulants are given to prevent emboli. Betablocking drugs may be used to raise the arrhythmia threshold in some cases. Prophylactic administration of antibiotics to guard against endocarditis, eg. prior to dental extraction, is recommended in all cases of mitral valve abnormality. In cases where heart failure ensues or is imminent, mitral valve replacement is the definitive treatment.

# AETIOLOGY

- 8. Mitral valve prolapse may be congenital, either familial, non-familial or acquired. Where inheritance is familial, there is autosomal dominance with variable penetrance and between 30% and 50% of first degree relatives are affected. In most cases, the basic aetiology is obscure with no identifiable underlying process other than ageing. In such cases, there is myxomatous degeneration in the valve leaflets, which become voluminous and redundant due to conversion of fibrous tissue into weaker, mucinoid material.
- 9. Deficiencies of magnesium and carnitine, a stimulant of fatty acid oxidation, have been postulated as aetiological factors, but these theories are not generally accepted.
- 10. Mitral prolapse may also be caused by inherited generalised connective tissue disorders and occurs with:
  - Marfan's syndrome, Ehlers-Danlos syndrome and osteogenesis imperfecta, in all of which the collagen is of abnormal structure.
  - Pseudoxanthoma elasticum, which affects the elastic fibres
  - Skeletal deformities including scoliosis, straight back and depressed sternum. About 25% of patients have a high-arched palate.
  - Hypomastia
- 11. Causal associations for mitral prolapse also exist with:
  - Rheumatic heart disease
  - Ischaemic heart disease, particularly acute inferior infarction with papillary muscle dysfunction
  - Congenital atrial septal defect
  - Hypertrophic obstructive cardiomyopathy (HOCM)
  - Papillary muscle damage during mitral valvotomy
  - Spontaneous rupture of the chordae tendineae of unknown aetiology.
- 12. Mitral prolapse is also more common in patients with thyrotoxicosis and sickle cell disease.
- 13. Sporadic cases of mitral prolapse occur in the absence of any of the above causative factors or associations.

## CONCLUSION

14. Floppy mitral valve syndrome, which is relatively common above the age of 50, is a non-inflammatory process, which affects either cusp, partially or completely. It is the commonest cause of mitral valve prolapse and pure mitral regurgitation. It is usually due to degenerative changes associated with ageing or a congenital abnormality of the heart. Mitral valve prolapse may occur in association with other syndromes, which are listed above or, in individual cases, may have no identifiable cause.

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