

VALVULAR HEART DISEASE - MITRAL INCOMPETENCE



“The Favourites of the Emperor Honorius”, oil on canvas, 1883, John William Waterhouse. Art Gallery of South Australia.

Honorius was only in the fourteenth year of his age; Serna, the mother of his bride, deferred, by art or persuasion, the consummation of the royal nuptials; Maria died a virgin, after she had been ten years a wife; and the chastity of the Emperor was secured by the coldness, or perhaps the debility, of his constitution. His subjects, who attentively studied the character of their young sovereign, discovered that Honorius was without passions, and consequently without talents; and that his feeble and languid disposition was alike incapable of discharging the duties of his rank. In his early youth he made some progress in the exercises of riding and drawing the bow; but he soon relinquished these fatiguing occupations, and the amusement of feeding poultry became the serious and daily care of the monarch of the West, who resigned the reigns of empire to the firm and skilful hand of his guarding Stilicho. The experience of history will countenance the suspicion that a prince who was born in the purple received a worse education than the meanest peasant of his dominions, and that the ambitious minister suffered him to attain the age of manhood without attempting to excite his courage or to enlighten his understanding. The

predecessors of Honorius were accustomed to animate by their example, or at least by their presence, the valour of the legions; and the dates of their laws attest the perpetual activity of their motions through the provinces of the Roman world. But the son of Theodosius passed the slumber of his life a captive in his palace, a stranger in his country, and the patient, almost the indifferent, spectator of the ruin of the Western empire, which was repeatedly attacked, and finally subverted, by the arms of the barbarians. In the eventful history of a reign of twenty-eight years, it will seldom be necessary to mention the name of the emperor Honorius.

Edward Gibbon, "The History of the Decline and Fall of the Roman Empire", Volume 3, 1781.

When the valve is permanently open, admitting of regurgitation, the first sound is intended with a murmur. It may be rough (rasping) or smooth, (bellows -murmur) according to the nature of the contraction (the force of the circulation and the character of the blood), & c (p.107). Its key is low - more or less like a whispering "who" (.110); yet it sounds louder and near if explored about the apex of the heart, and a little to the sternal side of the nipple. It may thus be easily distinguished from a direct semilunar murmur, which, in this low situation, always sounds feeble and distant. The murmur in some cases completely drowns the natural first sound on the left side; in others, the sound can be distinguished at the commencement of the murmur.

I have found perceptible purring murmur to be produced more frequently by regurgitation through the mitral valve than by any other valvular lesion - especially when the ventricle was hypertrophous and dilated, by which the refluent current was rendered stronger.

If the regurgitation be considerable, but not otherwise, the pulse is more or less small, weak, intermittent, irregular and unequal (p.359); and this, even though the impulse of the heart be violent.

James Hope, "Diseases of the Mitral Valve", in "Treatise on the Diseases of the Heart", Philadelphia 1842.

The last great Western Emperor of Imperial Rome was Theodosius I. He was the last emperor to rule over both the West and the East, and when he died in 395 A.D, he left behind him two sons, Arcadius, who was eighteen years of age and Honorius aged just twelve years. To Arcadius, Theodosius betrothed the East, and to Honorius, the West. Although minors, it was an astonishing tribute to the respect and the esteem to which Theodosius was held, that his two sons were allowed to remain on their respective thrones. Their power however was a complete illusion, both being weak willed and ill educated and both being dominated by a series of powerful generals of their respective armies, initially Rufinus in the East and the brilliant Stilicho in the West. The empire had been divided in the past, either by design or by usurpers. When by design, there was always an understanding that one of the two Emperors held the senior position, but when Arcadius and Honorius took their thrones, there was no such understanding. With the sons of Theodosius came the final and definitive split between east and west and it would eventually prove a fatal one for the west. From this moment, the priority of the two empires, lay not in the common defense of the Roman world from internecine external

barbarian invasion, but rather the struggle for supremacy of the one over the other. Stilicho and Rufinus, not only intent on invading the lands of their rival, even resorted to the mercenary use of barbarian tribes, the Goths in particular, who by this time had thoroughly infiltrated the lands of the empire, to attack one another - the Goths happily changing sides according to the highest bidder or the most advantageous of terms.

The division of the empire however was not the only factor in the demise of the west. A second crucial factor was by this time the complete infiltration of the army by barbarians, - German and Goth in the main. Stilicho who held supreme power in the west, was in fact half Roman and half Vandal - his sympathies often confused and ambiguous. As the generals of the west fought those of the east, external barbarian hordes took full advantage of the chaos that ensued. Vast "Volkswanderungs" of northern tribes would cross the frozen Rhine and Danube in the winter months to maraud virtually unmolested through the empire in the first half of the Fifth century. The outlying regions of the empire were simply left to their fate, often defended by usurpers who took local defense into their own hands, after having been proclaimed emperor by their own desperate troops. In all of these momentous events the two boy emperors appeared to have played no part - being puppets to their generals and in the case of Arcadius, even to his formidable wife Aelia Eudoxia, who had herself proclaimed "Augusta" of the East. Gibbon gives a dismal final assessment of the career of Honorius - "But the son of Theodosius passed the slumber of his life a captive in his palace, a stranger in his country, and the patient, almost the indifferent, spectator of the ruin of the Western empire, which was repeatedly attacked, and finally subverted, by the arms of the barbarians. In the eventful history of a reign of twenty-eight years, it will seldom be necessary to mention the name of the emperor Honorius".

In 410 A.D the Visigothic general, Alaric, exasperated with Honorius, who had fled to Ravenna, from where he "ruled" the empire, entered Rome and put it to the sack. It was the first time a foreign army had desecrated the eternal city in almost 800 years. The shock of this event finally brought to the attention of all, the realization that Rome could no longer defend itself. Procopius relates the story, that Honorius was so dim-witted that on being given the news that Rome had fallen, he thought that he was being told that his favourite pet cock (whose name was "Roma") had succumbed. He reportedly exclaimed in astonishment, ".....but I was only just feeding him!" The east would survive, but in the west, Honorius would be succeeded by an increasingly miserable line of feeble emperors playing puppet to barbarian generals. The charade abruptly came to an end in 476 A.D when the Germanic general, Odoacer, deposed the last emperor of the west - the poignantly named Romulus Augustulus.

When we read the 1842 description of mitral regurgitation by the eminent Dr James Hope of Philadelphia we are reminded of the sad demise of the Western Empire. Ominous murmurings are heard, indicating internal disease lying at the very heart. Violent internal agitations cross the fractured border of the mitral valve - in both directions - and even though its central impulses be violent - the peripheral pulse is weak. And as with the shocking sack of Rome, when overt symptoms begin, the first inkling of the full extent of inner decay finally becomes clear for all to see.

VALVULAR HEART DISEASE - MITRAL INCOMPETENCE

Introduction

Mitral incompetence (MI or mitral regurgitation - MR) is now the second most frequent valve disease after AS.

The treatment has been re-orientated as a result of the good results of valve repair.

Causes can be grouped into **organic, ischemic and functional** and within these groups acute or chronic disease may occur.

Organic MI covers all aetiologies in which *leaflet abnormality* is the primary cause of the disease, in opposition to ischaemic and functional MR, in which MR is the secondary consequence of *LV disease*.

Natural History

Primary MI: Intrinsic Valve Disease

Acute MI is poorly tolerated and carries a poor prognosis in the absence of intervention.

In asymptomatic MI, the estimated 5 year rates of death from any cause, death from cardiac causes, and cardiac events (death from cardiac causes, heart failure, or new AF) with medical management are approximately 22%, 14%, and 33%, respectively.

Secondary MI:

Ischaemic:

Acute MI, secondary to papillary muscle rupture, has a dismal short-term prognosis and requires urgent treatment.

Patients with chronic ischaemic MI have a poor prognosis.

Although coronary artery disease and LV dysfunction have prognostic importance, the presence and severity of MI are independently associated with increased mortality.

Functional:

The data on the natural history and results of surgery are limited.

A precise analysis is difficult because of the limited number of series including small numbers of patients and mixing patients with or without revascularization.

Several observational studies have shown the high prevalence of significant MI in chronic heart failure, as well as its independent association with a poor prognosis. However, its true prevalence and its pathogenic contribution to prognosis remain uncertain.

Pathophysiology

Causes

1. Primary Intrinsic Valve Disease:

Intrinsic Valve Disease MI covers all aetiologies in which *leaflet abnormality* is the primary cause of the disease.

These include:

- Degenerative, (most common).
- Rheumatic fever.
- Infective endocarditis.
- Progression of mitral valve prolapse.
- Connective tissue diseases, (eg Marfans & SLE - Libman-Sacks lesion)

2. Secondary MI:

This can be:

- **Ischaemic:**

MI due to ischaemic papillary muscle rupture or dysfunction is a relatively frequent entity.

It can occur in an ACS or in chronic ischemia It is frequently overlooked in the setting of both acute or chronic coronary disease.

Chronic ischaemic MI is the consequence of a restriction in leaflet motion, which is due to tethering by the subvalvular apparatus in patients who have LV enlargement and/or dysfunction, in particular of the posterolateral wall.

- **Functional:**

In this group, mitral valves are also structurally normal and MI is secondary to the changes in the overall LV geometry resulting from impaired LV function.

It includes MI seen in:

- ♥ Dilated cardiomyopathies in general

- ♥ Ischaemic heart disease with severe LV dysfunction.

Acute MI can be due to:

1. Bacterial endocarditis
2. Acute ischemic papillary muscle rupture or dysfunction/ chordae tendinae rupture.
3. Occasionally trauma.

Complications

1. Infective endocarditis.
 - Note that mitral incompetence may also be **caused by** endocarditis, as well as predisposing to it.
2. Left ventricular failure.
3. AF can develop in the longer term due to chronic atrial dilatation
4. Longer term, pulmonary hypertension with right ventricular failure.

Clinical Features

In acute lesions there will be rapid onset of dyspnoea/ pulmonary edema.

In **chronic cases** the following is seen:

Symptoms

In the **chronic setting** there will be progressive dyspnea on exertion and features of LVF in general.

Signs

1. Pulse:
 - May be reduced in severe cases.
2. Blood pressure:
 - May be reduced in severe cases
3. Apex beat:
 - Displaced

- Diffuse
 - Hyperdynamic.
4. Thrill:
- May be detected in severe cases.
- There may be a left parasternal heave if RVH/ pulmonary hypertension is present.
5. Heart sounds:
- Soft S1 if severe.
6. Murmur:
- Harsh pan-systolic murmur best heard at the apex with radiation to the axilla. Intensity does *not* correlate with severity of the lesion.

Clinical Indicators of Severity

SIGN	SEVERITY INDICATOR
Pulse	<i>Small pulse volume.</i>
Heart sounds	<i>S3 and early diastolic flow rumble.</i>
Murmur	<i>Enlarged LV</i>
Heart failure	<i>Signs of pulmonary hypertension.</i>

Investigations

CXR

Check for:

- Cardiomegaly
- Signs of pulmonary congestion.

ECG

- Check for signs of LHV and strain.

Echocardiography

Indications of severe MI on echocardiography include: ¹

- Patients with severe LV dysfunction:
 - ♥ LVEF < 30%
 - ♥ End systolic dimension > 55 mm
- Size of regurgitant jet on colour flow doppler.
- Quantification of regurgitant volume as a proportion of the stroke volume:
 - ♥ Regurgitant volume > 60 mls per beat.
 - ♥ Regurgitant fraction > 50%
- Effective regurgitant orifice area $\geq 0.4 \text{ cm}^2$
- Systolic flow reversal in pulmonary veins
- Pulmonary hypertension

Coronary angiography:

- Coronary angiography is indicated in selected cases to detect associated coronary artery disease when surgery is planned.

Knowledge of coronary anatomy improves risk-stratification and determines whether coronary revascularization is indicated in association with valvular surgery.

Management

1. Medical therapy:

Acute MI:

- In acute MR, reduction of filling pressures can be obtained with nitrates and diuretics.
- Sodium nitroprusside reduces afterload and regurgitant fraction, as does an intra-aortic balloon pump.

- Inotropic agents and intra-aortic balloon pump should be added in cases of hypotension.

Chronic MI:

- There is no evidence to support the use of vasodilators, including ACE inhibitors, in chronic MR without HF and they are therefore not recommended in this group of patients.
- However, when HF has developed, **ACE inhibitors** are beneficial and should be considered in patients with advanced MR and severe symptoms, who are not suitable for surgery or when there are still residual symptoms following surgery.
- Beta-blockers (carvedilol) may also be considered as appropriate.

2. Surgical therapy:

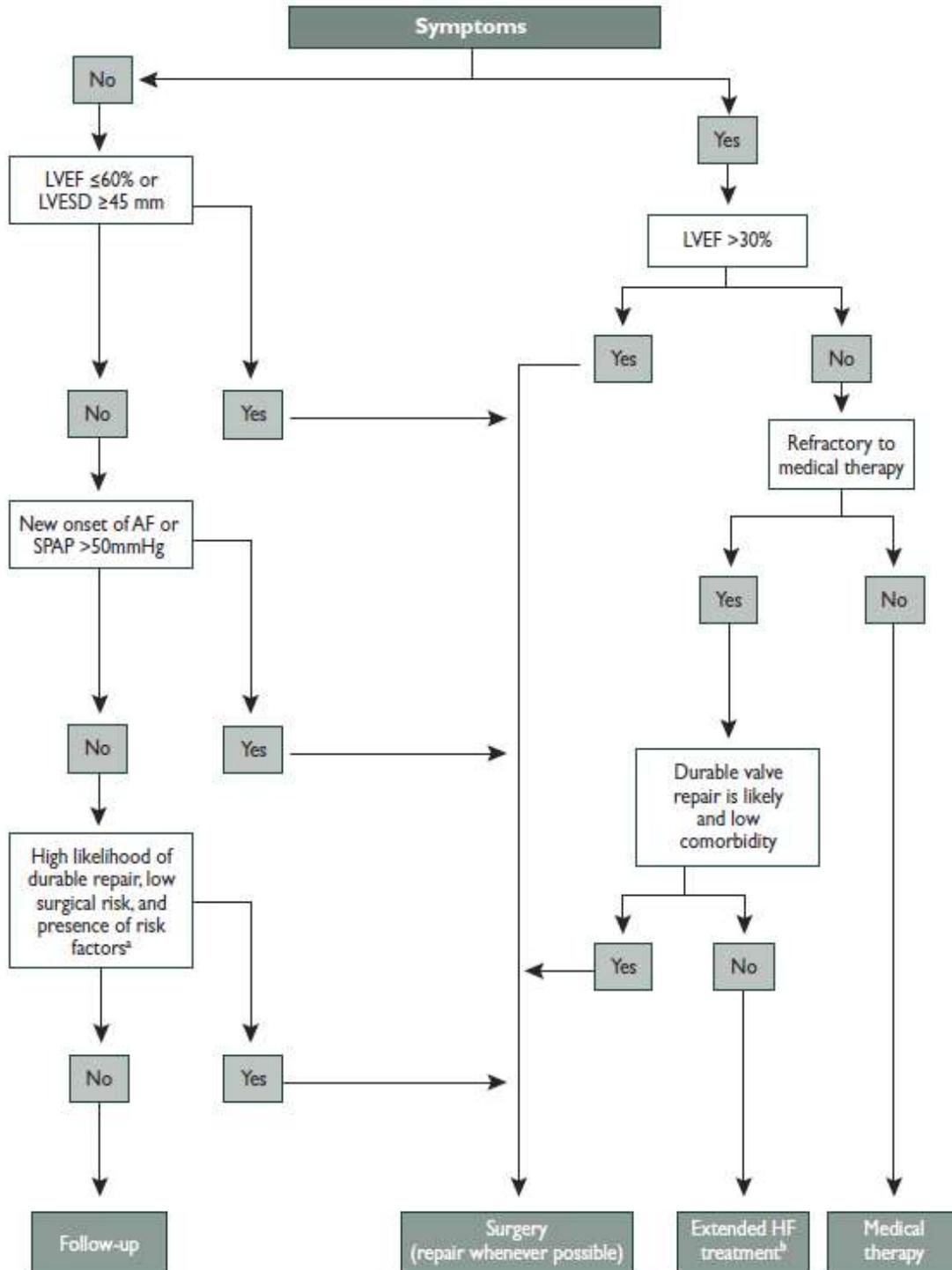
Mitral valve repair/ replacement will be indicated in:

- Acute MI
- Chronic MI:
 - ♥ **The general approach to the need for surgery in chronic cases can be summarized by the flow chart in Appendix 1 below.**

The general approach to surgery in patients with severe **chronic organic MI** is summarized in the flow diagram below:¹

3. Antibiotic prophylaxis for surgical procedures, (see latest edition of Antibiotic guidelines)

Appendix 1



AF = atrial fibrillation; BSA = body surface area; HF = heart failure; FU = follow-up; LA = left atrium; LV = left ventricle; LVEF = left ventricular ejection fraction; LVESD = left ventricular end-systolic diameter; SPAP = systolic pulmonary arterial pressure.

*When there is a high likelihood of durable valve repair at a low risk, valve repair should be considered (IIaC) in patients with fail leaflet and LVESD ≥ 40 mm; valve repair may be considered (IIbC) if one of the following is present: LA volume ≥ 60 mL/m², BSA and sinus rhythm or pulmonary hypertension on exercise (SPAP ≥ 60 mmHg).

†Extended HF management includes the following: cardiac resynchronization therapy; ventricular assist devices; cardiac restraint devices; heart transplantation.



“The Emperor Honorius”, oil on canvas, Jean-Paul Laurens 1880. Chrysler Museum of Art, Norfolk, USA.

Honorius became Emperor of the West in 395 A.D, at the age of 12 years. His reign marked the beginning of the end of the Western Roman Empire.

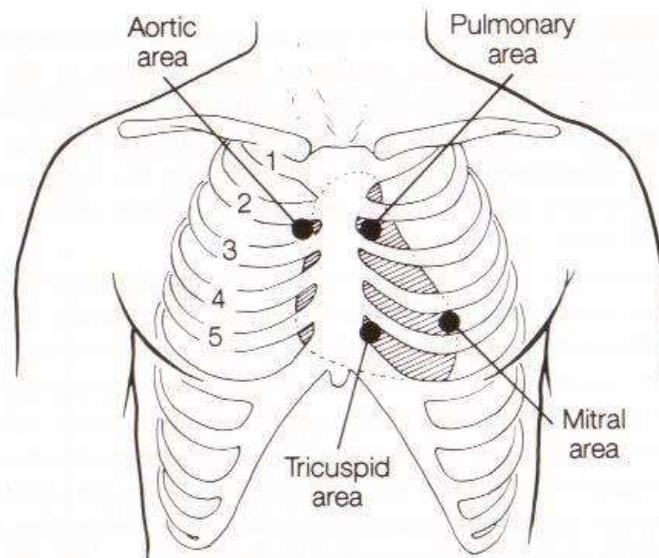
Appendix 2

Grading of the loudness of murmurs:

- Grade 1 *Very soft, requires an experienced listener.*
- Grade 2 *Soft.*
- Grade 3 *Moderate and without a thrill*
- Grade 4 *Loud with thrill just palpable.*
- Grade 5 *Very loud and thrill easily palpable.*
- Grade 6 *Very loud, may be heard without the aid of a stethoscope.*

Appendix 3

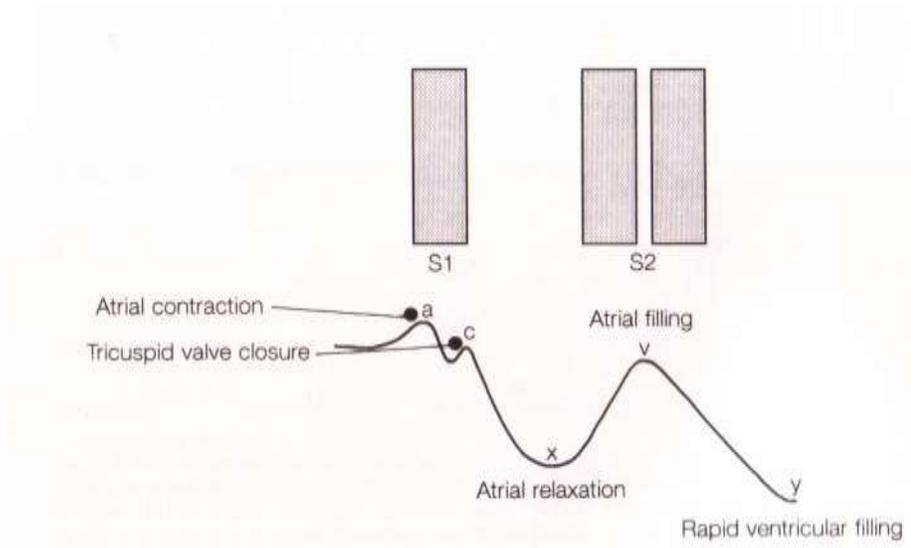
Auscultatory regions of the heart:



Note, these regions show the optimal areas for listening to the heart valve indicated, they do not exactly correlate with surface anatomy of the anatomical location of the valve.

Appendix 4

The JVP wave form:



Components of the jugular venous pressure wave with relationships to the first and second heart sounds.

References

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