

VALVULAR HEART DISEASE

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These notes discuss mitral stenosis (MS), chronic and acute mitral regurgitation (MR), aortic stenosis (AS), chronic and acute aortic regurgitation (AI) and idiopathic hypertrophic subaortic stenosis (IHSS). Endocarditis and antibiotic prophylaxis will not be discussed, but should be kept in mind whenever a murmur is heard. Even if a lesion is not hemodynamically significant, the abnormal valve is more susceptible to infection. The emphasis of the discussion is on pathophysiology and bedside clinical assessment in order to gauge severity and to aid in decisions about monitoring, anesthesia and consults.

Most of the important variables subject to manipulation in the management of valvular heart disease can be organized into four categories:

1. Rate and rhythm
2. Preload
3. Afterload
4. Contractility

If you can remember these four items and the pathophysiology of the lesion, the management of each item should become apparent.

Abbreviations and Definitions:

RA - Right Atrium

RV - Right Ventricle

LA - Left Atrium

LV - Left Ventricle

MAP - Mean Arterial Pressure

CVP - Central Venous Pressure, a measure of RV filling pressure

HR - Heart Rate

SV - Stroke Volume

CO - Cardiac Output (= HR x SV)

PAW - Pulmonary Artery Wedge Pressure

PA - Mean PA pressure

SVR - Systemic Vascular Resistance = $80 \times (\text{MAP} - \text{CVP})/\text{CO}$

PVR - Pulmonary Vascular Resistance = $80 \times (\text{PAW} - \text{CVP})/\text{CO}$

JVD - Jugular Venous Distention

PMI - Point of Maximum Impulse - where the left ventricle pushes against the chest, usually felt in the left 5th intercostal space at the mid-clavicular line

Heave - When the movement of the heart against the chest is so diffuse, prolonged and pronounced as to push a substantial portion of the chest outwards

S1 - First heart sound, has two components - mitral and tricuspid valve closure

S2 - Second heart sound, has two components - aortic (A2) and pulmonic (P2) valve closure

Physiologic S2 Splitting - A2 followed by P2 - Aortic valve closes first, split widens with inspiration. With inspiration, blood return to the right heart is enhanced, so RV ejection is prolonged and pulmonic closure delayed. The delay increases the time separation between the two valve closures.

Paradoxical S2 Splitting - P2 followed by A2 - Split narrows with inspiration. Whenever A2 occurs after P2 (as in left bundle branch block), the delay in P2 with inspiration brings A2 and P2 closer together.

S3 (Gallop) - Occurs with very rapid flow into a ventricle, or normal flow into a dilated, stiff ventricle (i.e., failure)

S4 (Gallop) - When the atrial kick ejects a lot of blood into the LV (Even more likely to be heard if the LV is not compliant)

Murmurs - Caused by turbulent blood flow. MS, MR, AS, AI, IHSS cause turbulent blood flow by a narrowed orifice. Functional murmurs occur with relatively rapid blood flow through normal valves, typically the soft (I-II/VI) systolic ejection murmur of young people.

Dyspnea - Difficulty breathing, usually with shortness of breath.

Orthopnea - Difficulty breathing on assuming a level position.

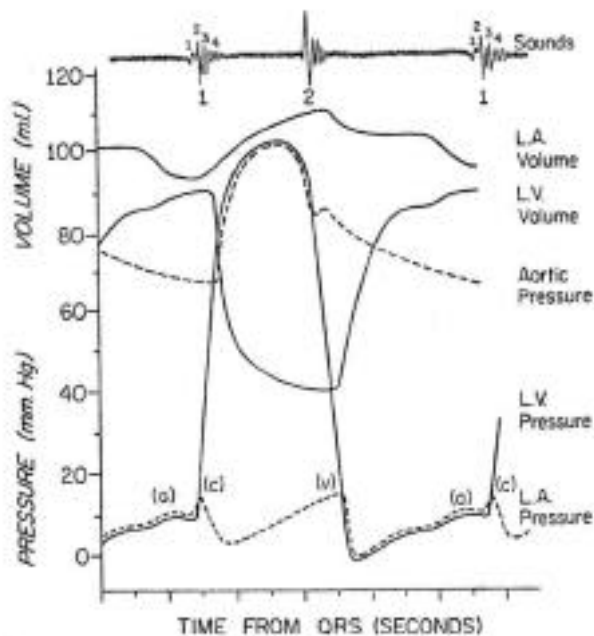
Paroxysmal Nocturnal Dyspnea - Waking up short of breath.

Compliance - The relationship between ventricular end-diastolic pressure and ventricular end-diastolic volume. Compliance is not constant, it is altered by acute and chronic processes such as changes in heart rate or contractility, edema from acute myocardial infarction, hypertrophy, fibrosis or infiltrates.

Preload - Refers to the degree of sarcomere stretch at the beginning of contraction and that, in turn, designates the position on the Frank-Starling relationship between the degree of stretch versus the strength of contraction. Preload is indirectly assessed by PAW or CVP but such measurements ignore pressure gradients across the atrioventricular valve and ignore changes in ventricular compliance. In the management of valvular heart disease, control of preload not only refers to control of end-diastolic volume, but also refers to the effects of left atrial pressure on the lungs and the effects of right atrial pressure on the body.

Afterload – Technically, afterload refers to the pressure that the ventricle must overcome in order to eject blood. Clinically this translates into aortic pressure unless a pressure gradient exists (e.g. aortic stenosis). In common parlance, afterload is also used in reference to SVR. In the management of valvular heart disease, control of afterload entails not only the control of MAP but also control of SVR. SVR not only directly affects MAP (by Ohm's law, $MAP = CO \times SVR$), but SVR affects CO as well via the principle of afterload reduction.

Contractility - Refers to the intrinsic squeeze of the heart. The total effort of a ventricle (peak pressure and stroke volume) is a function of both contractility and preload.



Normal Heart
(figure courtesy Dr. E. O. Feigl)

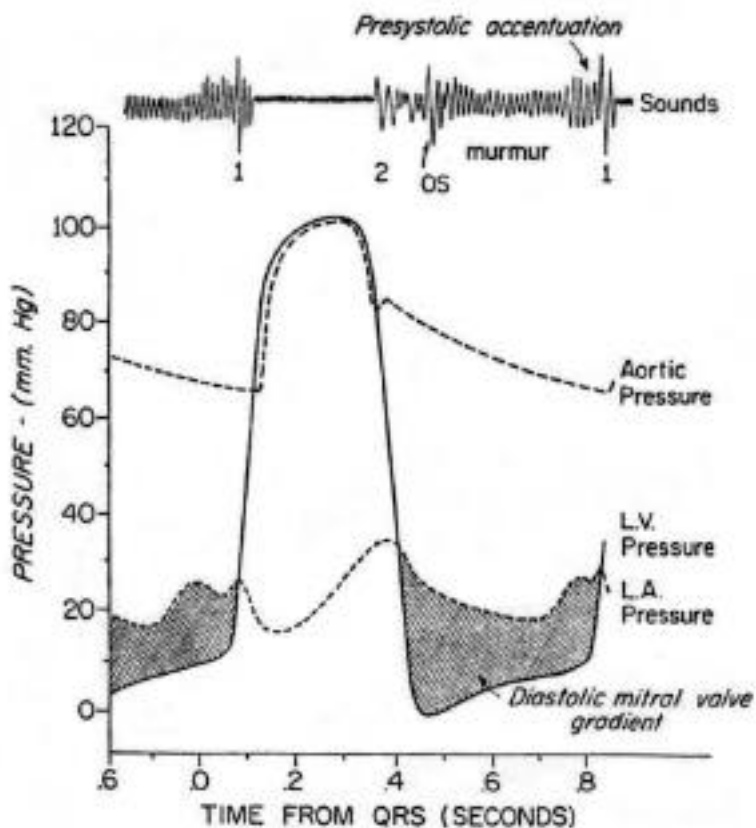


Location of Heart Sounds
(figure courtesy Dr. E. O. Feigl)

MITRAL STENOSIS

Etiology:

Usually rheumatic heart disease. Four times more common in females than males. After rheumatic carditis, the valve thickens and fibroses over decades. Commissures fuse, and the chordae tendinae shorten.



(figure courtesy Dr. E. O. Feigl)

Gorlin formula

$$\text{valve area in cm}^2 = \frac{\text{CO}}{38 \cdot \text{DFT} \cdot \text{HR} \cdot \text{PG}}$$

where

- CO = total cardiac output (forward and regurgitant)
- DFT = diastolic filling time
- PG = average pressure gradient across the mitral valve during diastole

Pathophysiology:

As valve area lessens, a pressure drop develops across the valve during diastole. Severe stenosis results in a 25 mm Hg gradient or higher. In consequence, the high left atrial pressures back up throughout the pulmonary circulation. The left atrium responds to the increased pressure by gradual dilation which predisposes to atrial fibrillation. In the lungs, the high pressure encourages capillary fluid leak causing the lungs to stiffen, increasing the work of breathing and leading to dyspnea.

PAW will reflect LA pressures but not LV end-diastolic pressures because of the pressure drop across the mitral valve. The relationship between LAP and LVEDP will depend on the pressure gradient across the valve and the presence or absence of the atrial kick. The atrial kick helps to hold LAP down by pushing blood into the LV, yet the duration of the kick is so short that mean LA pressure is barely increased. With the onset of atrial fibrillation, mean LAP must rise in order to maintain blood flow across the mitral valve and therefore the symptoms of MS dramatically and acutely worsen with the loss of the atrial kick.

Exercise normally leads to an increase in heart rate and cardiac output. The rate of blood flow across the mitral valve thereby increases (I CO and L diastolic filling time) and leads to a rise in atrial pressure and dyspnea worsens. Functional tricuspid regurgitation is common in severe mitral stenosis. As shown above, at any given mitral orifice size, the transvalvular gradient is directly proportional to the square of transvalvular blood velocity. Therefore, small increases in cardiac output (i.e., flow) cause increases in the valve gradient and therefore increase symptoms. In summary, left atrial and pulmonary pressures (and therefore capillary fluid leak) increase with:

1. Increased cardiac output - higher flow across the valve requires higher driving pressure.
 - a. Exertion - increased CO causes increased LA pressure, patient has dyspnea on exertion.
 - b. Chronic high cardiac output states - i.e., pregnancy, fever, postop major surgery - all can lead to dyspnea at rest or pulmonary edema in a patient previously okay at rest despite severe stenosis.

NOTE: In severe stenosis cardiac output cannot increase very much because the high LA pressures that would result would not be tolerated. Cardiac output is therefore relatively fixed at a constant low value in severe MS.

2. Tachycardia or atrial fibrillation - with tachycardia cardiac output may not fall, but overall diastolic filling time is reduced so diastolic flow increases. With loss of the atrial kick, LA pressure must rise to push the blood into the LV that had been previously pushed in by the atrial kick.
3. Supine posture - increases pulmonary blood volume and that increases static pulmonary pressures. Patient notes orthopnea and paroxysmal nocturnal dyspnea (PND).
4. Increasingly narrowed valve - eventually dyspnea at rest despite chronic low cardiac output.

The elevated pulmonary pressure is a passive phenomenon in most patients, that is, pulmonary vascular resistance is normal. However, a subset (15%) of patients with MS also develop increased PVR so their PA pressure becomes further elevated and frequently causes RV failure because the geometry of the RV is not designed to handle high pressure. RV failure and LV failure have the common characteristics of a low resting cardiac output and high sympathetic tone. In RV failure, CVP rises to utilize the Frank-Starling mechanism but the elevated CVP also leads to jugular vein distension, hepatomegaly, peripheral edema, ascites and possibly defects in coagulation secondary to liver dysfunction.

Symptoms of Mitral Stenosis:

1. Dyspnea on exertion or even at rest, PND, orthopnea, pulmonary edema - all due to the high pulmonary capillary pressures.
2. Hemoptysis, frequent "bronchitis" can also occur at least in part from high pulmonary pressures. Bronchitis is due to a bronchial hyperemia that leads to increased mucous secretion. (Passive congestion leads to hyperemia.)
3. Fatigue - occurs in severe MS and suggests a chronically low cardiac output, often accompanied by RV failure.
4. Stroke or other embolic events - due to clots originating from the large LA or vegetations in endocarditis breaking loose.
5. Initial symptom may be CHF from new onset of atrial fibrillation.

Physical Findings in Mitral Stenosis:

1. Pulse - may be weak due to low SV - atrial fibrillation in 40% of patients.
2. Jugular veins - normal unless RV failure.
3. PMI - normal position, normal or decreased intensity. Apical diastolic thrill with severe disease.

KEY POINT: In mitral stenosis the LV is spared, it is not subjected to high filling pressures. Lateral displacement of the PMI indicates that some additional lesion (e.g. ischemic heart disease, other valvular disease) must also exist. Realize too, that MS protects the LV from the pathophysiology of AS should AS also be present.

4. Auscultation:
 - a. Loud S1 (high LA pressures and a stiff valve lead to an abrupt valve closure). The combination of atrial fibrillation plus a loud S1 is MS until proven otherwise.
 - b. S2 normal, S3 and S4 not present unless other disease exists.
 - c. Diastole - Opening snap followed by a low pitched rumble that is mid-diastolic in mild disease and lasts to S1 in severe disease. If the murmur disappears before S1, it suggests that LV filling has had time to be complete (i.e. LAP and EDP have equalized). The murmur is heard best at the apex with the stethoscope bell with the patient in the left lateral decubitus position, and it radiates to the axilla. The opening snap is the abrupt opening of the stiff mitral valve that is pushed open rapidly by the high left atrial pressure once the ventricle relaxes. The S2-OS interval decreases as MS worsens. Mitral valve calcification may lead to an absent opening snap. The murmur, if pansystolic, will get louder just before S1. Such presystolic accentuation rules out significant mitral regurgitation.

5. RV Failure - accompanied by peripheral edema, JVD, hepatomegaly, possible RV heave (felt over sternum). Parasternal heave suggests pulmonary hypertension with chronic right ventricular failure which may lead to cardiac cirrhosis and ascites. Hoarseness can occur from entrapment of the left recurrent laryngeal nerve between a tense, large pulmonary artery and the aorta at the ligamentum arteriosum. Right sided S3 may appear.

EKG in Mitral Stenosis:

1. Atrial fibrillation in nearly half of the patients with mitral stenosis.
2. If in NSR, you will see a widened P-wave in lead II (P-mitrale).
3. Possible R axis deviation or R ventricular hypertrophy.

X-Ray:

1. Left atrial enlargement. Early on you will see an increased left atrial appendage with straightening of the left heart border. Extreme left atrial enlargement causes double density on the pulmonary artery with posterior displacement of the esophagus laterally and left mainstem bronchus elevation.
2. Increased cephalad pulmonary blood flow.
3. Calcified mitral valve. (Note: Calcification of the annulus but not the leaflets may lead to regurgitation but not stenosis.)
4. Kerley B lines.
5. PA and RV enlargement.

Catheterization Results:

Will provide values for cardiac output, the valve gradient at that output and the estimated valve area. Also included should be CVP and PA pressures.

Severity of Mitral Stenosis:

Can be largely assessed by history and physical exam. Asymptomatic to a little dyspnea on exertion suggests adequate cardiac reserve. Symptoms of dyspnea at rest, PND, orthopnea or pulmonary edema suggest no reserve and probably chronic low cardiac output with high resting sympathetic tone. A short time interval between S2 and the opening snap, rales in the lungs, and signs of RV failure also suggest severe disease. Even more worrisome is the presence of pulmonary hypertension; patients so affected may experience low cardiac output and/or RV failure when they experience surgical and anesthetic stress. A cardiac consult before elective surgery is needed to assess severity and decide if valve replacement may be more important. Most patients with severe mitral regurgitation have a III/VI or louder murmur.

The normal valve has an area of about 4.0 cm². The severity of disease correlates closely with the reduction in valve area.

- STAGE I. Minimal: > 2.5 cm² - no symptoms.
- STAGE II. Mild: 1.4-2.5 cm² - dyspnea with severe exertion.
- STAGE III. Moderate: 1.0-1.4 cm² - dyspnea, PND, Orthopnea, > pulmonary edema.
- STAGE IV. Severe: < 1.0 cm² - resting dyspnea, disabled - Class IV.
- STAGE V. Reactive Pulmonary Hypertension: Class IV, plus fatigue and ventricular failure.

Indications for Open Mitral Commissurotomy (OMC):

1. Pure MS
2. Minimal calcification
3. Non-rigid leaflets
4. Mobile, normal chordae
5. Non-infected valve.

Turbulence and fibrosis of the valve surface following OMC leads to restenosis. 5-10% will have restenosis within 5 years. 10% of patients with commissurotomies will need an operation in 5 years, and 60% will need operations by 10 years.

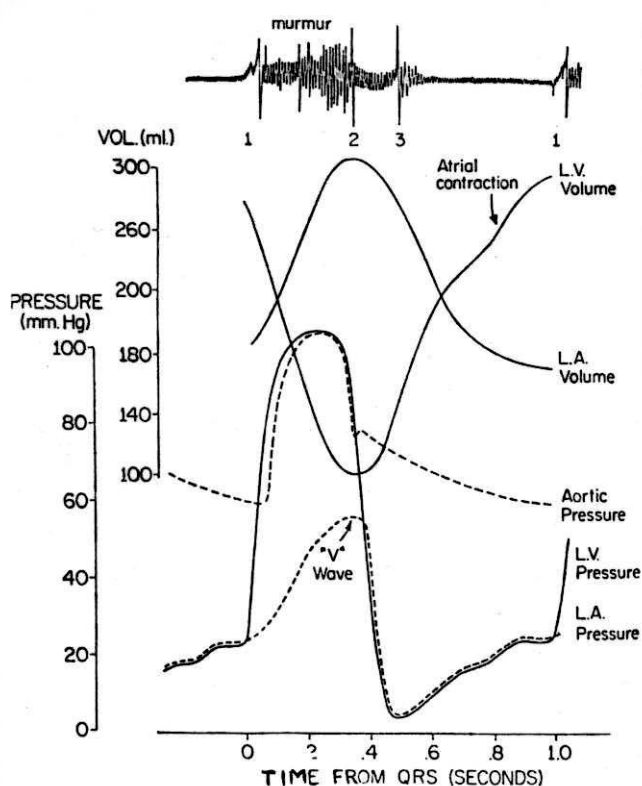
Anesthetic Management of Mitral Stenosis:

1. Heart rate should be in the low normal to normal range (70-90). High heart rates are deleterious because diastolic filling time falls as HR increases. This leads to a fall in CO if LV filling diminishes and/or an increase in pulmonary congestion if LA pressure rises. Low heart rates may provide adequate LV filling, but CO may be limited by the low HR since in all of us SV cannot increase sufficiently to maintain CO at very low heart rates.
2. Maintain sinus rhythm. The atrial kick is important because it supports LV filling more than it raises LA pressure. Atrial fibrillation not only eliminates the atrial kick but may also cause tachycardia.
3. Preload must be kept high to support LV filling, but pulmonary edema must be avoided, too. Trendelenburg position may support preload in an emergency, but it may also promote pulmonary edema.
4. SVR must be kept at the patients usual level, if not higher. A fall in SVR may not be compensated by a proportional rise in CO because CO is limited by the mitral stenosis. In MS a fall in SVR usually causes a fall in BP. Alpha agonists will support SVR adequately in most cases, but beware the patient with pulmonary hypertension who also respond with increased PVR; RV failure may result. For such patients inotropic support (or mixed beta and alpha agonism) may be best.
5. Contractility - Should not be depressed. If the LV is forced to maintain CO via the Frank-Starling mechanism, LA and pulmonary capillary pressure will also have to rise.
6. Patients with reactive pulmonary vasculature (generally those with chronically elevated PVR) can be very difficult to manage. Avoid anything that raises sympathetic tone above the patients usual level (e.g. light anesthesia, N2O). The use of pure alpha agonists is relatively contraindicated in patients with elevated PVR. Sudden increases in PVR can lead to higher PA pressures and RV failure. PVR may be decreased by NTG, isoproterenol or phentolamine via the RA, while systemic BP can be supported via epinephrine or norepinephrine via the left atrium. In general, narcotic anesthesia is effective for these patients. Hypocapnia lowers PVR, whereas hypercapnia raises PVR.
7. Give drugs with CNS effects carefully because in low output states flow is preferentially distributed to the brain. Premeds should be small, if any, if the patient appears at all ill.

CHRONIC MITRAL REGURGITATION

Etiology:

Most often rheumatic heart disease, but also isolated chordae tendinae rupture, mitral valve prolapse, ischemic papillary muscle dysfunction. Usually the posterior papillary muscle is at risk because it usually gets blood from the PDA, whereas the anterior papillary muscle will receive blood from the LAD and circumflex coronary arteries. LV dilatation can cause mitral insufficiency via stretching of the mitral annulus such that the leaflets are unable to appose properly. Mitral valve prolapse is seen in 5-8% of the population and of these perhaps 10-15% have some MR.



(figure courtesy Dr. E. O. Feigl)

Pathophysiology and Symptoms:

Regurgitation into the LA forces both the LA and LV to enlarge in order to accommodate not only normal forward cardiac output, but also the regurgitant volume. The dilated LA absorbs the regurgitant volume and in so doing minimizes the rise in LA pressures. Consequently the pulmonary vasculature is not subjected to high pressures. LV dilation, plus modest muscle hypertrophy, maintains a normal forward output but the LV cannot enlarge indefinitely. Eventually forward output suffers and the patient notes fatigue or easy fatigability as opposed to SOB. Dyspnea on exertion is common however. As the LV myocardium deteriorates LVEDP

rises and congestive symptoms (PND, orthopnea, dyspnea) appear as the pulmonary capillary pressures rise with LVEDP. Such symptoms suggest at least some, if not severe, LV failure (impaired contractility). Angina is not common unless there is associated CAD.

Mitral valve incompetency makes it easy for the LV to empty, and therefore the ejection fraction is normally very high at first, then falls with diminished contractility. An ejection fraction of 50% represents severe myocardial depression in MR.

Chronic MR leads to an enlarged annular circumference plus enlargement of the mural leaflet and both commissures. The anterior leaflet is unchanged in size. As MR worsens, the chordae elongate and finally rupture leading to increased MR and a prolapsed leaflet.

Secondary MR (e.g. from acute LV distension) is usually not severe (40% regurgitant fraction or less).

Physical Findings in Chronic Mitral Regurgitation:

1. Pulse - Irregularly irregular if in atrial fibrillation (due to large LA).
2. Jugular veins - Normal, unless RV failure occurs (rare in chronic MR).
3. PMI - In moderate MR, the PMI is hyperdynamic and displaced to the left and downwards. With severe MR, the PMI becomes prolonged and is more of a heave, because of the long time needed to eject the high stroke volume.
4. Auscultation:
 - a. S1 - normal or soft.
 - b. S2 - widely split (rapid LV unloading leads to an early A2). May not be heard at all if drowned out by the murmur.
 - c. S3 - frequently present, does not necessarily imply LV failure.
 - d. S4 - rare (not at all in atrial fibrillation).
 - e. Murmur - high frequency murmur lasting throughout systole, heard best at apex, radiates to axilla and back, intensity of murmur crudely correlates with severity of regurgitation. In mitral valve prolapse and papillary muscle dysfunction, the murmur doesn't begin until well after S1. A click precedes the murmur in mitral valve prolapse.

EKG:

Atrial fibrillation, left atrial enlargement, left ventricular hypertrophy.

CXR:

LVE, LAE, pulmonary venous redistribution, calcified mitral valve in rheumatic heart disease.

Severity of Chronic Mitral Regurgitation:

Mild - regurgitant fraction of 30% or less. Mild MR is usually asymptomatic and poses only a minimal increased risk for anesthesia.

Moderate - regurgitant fraction of 30% to 60%. With moderate MR the patient notes fatigue and dyspnea on exertion. The patient has myocardial dysfunction and is at modestly increased risk but remember that, in general, mitral regurgitation is not as dangerous as mitral stenosis.

Severe - regurgitant fraction greater than 60%. With severe MR the symptoms of LV failure are present. By now contractility is quite impaired; the patient is at high anesthetic risk.

Anesthetic Management of Chronic Mitral Regurgitation:

The main concern is promoting forward cardiac output and minimizing regurgitant flow. The division of flow depends on the sizes of the regurgitant orifice and the aortic valve, and the pressures in the left atrium and the aorta. Aortic pressure is the product of SVR and CO, so if SVR is lowered then CO can rise to maintain a similar pressure.

1. Normal to high heart rates are preferable to low heart rates. High heart rates will promote cardiac emptying and tend to lower LVEDV and so reduce the valve area during systole. The atrial kick is not important in MR because flow through the valve is not limited.
2. Preload will probably already be high because the body retains fluid in response to impaired forward output. May have to lower preload if pulmonary congestion is severe.
3. Avoid high SVR. Afterload reduction (e.g. nifedipine) will promote forward flow. It is especially important to beware events that cause vasoconstriction (e.g. high sympathetic outflow with surgical stimulation or intubation under light anesthesia) because a rise in BP may be at the cost of a marked fall in CO.
4. Avoid decreases in contractility. High levels of contractility promote cardiac emptying (lessen valve area) and improve total cardiac output.
5. A PA catheter is useful to follow height of V wave (rough gauge of severity of regurgitation), CO and SVR. The V wave may be absent if LA very compliant and dilated.
6. Patients with mitral valve prolapse are prone to atrial and ventricular arrhythmias. In prolapse the heart rate should be kept low because large ventricular volumes pull the papillary muscles away from the valve and decrease the degree of prolapse.)

ACUTE MITRAL REGURGITATION

Etiology:

1. Papillary muscle dysfunction due to ischemia/infarction (usually posterior papillary because of a single vessel supply - RCA or circumflex) or due to marked LV dilation. Up to 30% of patients with coronary artery disease have MR from either LV dilation OR ischemia of a papillary muscle.
2. Chordae tendinae rupture.
3. Leaflet destruction (e.g. endocarditis).
4. Leaky prosthetic valve (usually paravalvular).

Pathophysiology:

Acute regurgitation doesn't give time for the LA to dilate so the LA is small and thick walled and not compliant; in consequence the V wave is large and LA pressures become more elevated than in chronic MR. RV failure is common in acute MR because of the high LAPs. The degree of regurgitation worsens with LV dilation so systemic hypertension may lead to RV failure via increased MR causing increased LAP causing increased PA pressure causing RV decompensation. Unloading (systemic vasodilation) therefore leads to a decreased regurgitant fraction by shrinking the LV and allowing closer apposition of the mitral valve apparatus. 50% regurgitation in acute MR is much more serious than 50% regurgitation on a chronic basis because the LV is smaller in acute MR. The non-enlarged LV has a limited total stroke volume so forward stroke volume is reduced and CO is impaired much in acute MR more than in chronic MR.

Symptoms:

Easy fatigability (low CO), dyspnea (pulmonary congestion), pedal edema (RV decompensation) are common. Frank pulmonary edema occurs with severe dysfunction (e.g., papillary muscle rupture) due to the very high LA pressures that accompany severe acute MR. Cardiogenic shock may be the consequence of an impaired forward cardiac output.

Physical Findings:

1. Pulse shows sinus tachycardia with a rapid upstroke and falloff (MR unloads the LV quickly). The tachycardia results from sympathetic nervous system activation that, if severe, leads to a shock-like appearance of cool, ashen, clammy skin.
2. Jugular veins may be distended, especially if RV failure develops.

3. PMI may be prominent but not displaced or sustained (ejection is rapid, LV not dilated in acute MR).
4. Auscultation.
 - a. S1-normal or increased.
 - b. S2-widely split (A2 early due rapid LV unloading). P2 may be loud (if increased PA pressure).
 - c. S3 and S4 often present.
 - d. Holosystolic murmur, loudest at the apex and may radiate widely.

The murmur may decrease in intensity during late systole (decrecendo) because the LAP may approach LV pressure, or the murmur may mimic an ejection type murmur. In papillary muscle rupture LV function may be so poor that flow is too slow to produce a murmur. An S4 suggests acute MR because a chronically dilated LA has too weak an atrial kick to yield an S4. The severity of the regurgitation can be estimated by the duration of the murmur (the shorter the more severe) rather than by the intensity of the murmur (as in chronic MR).

EKG:

No consistent findings.

CXR:

Pulmonary vascular engorgement/pulmonary edema.

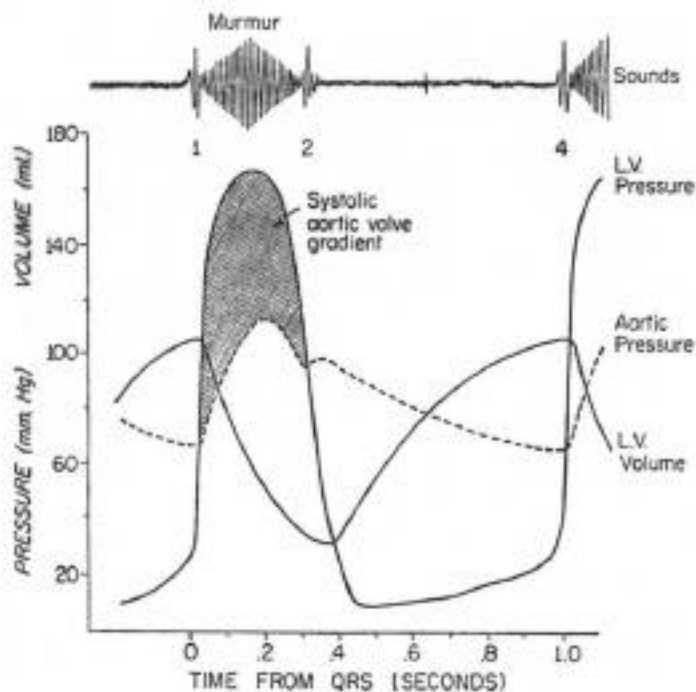
Management:

Even if patient needs surgery, medical stabilization with vasodilator therapy will help. Some patients will also need inotropic support and/or IABP. Treatment for acute MR is similar to chronic MR - except that contractility may need more active support. -Vasodilator therapy may have limited effectiveness because the total cardiac output of a normal sized ventricle is smaller than that of a chronically enlarged ventricle. Vasodilation may lead to hypotension if the rise in CO fails to match the drop in SVR. Induction of anesthesia must proceed slowly and drugs must be given in small increments. Loss of sympathetic tone may rapidly lead to a fall in SVR and BP. Operative mortality for emergent valve replacement correlates with the end-systolic volume (ESV). If the ESV is normal (30-90 cc/M²), MVR is well tolerated. If ESV is more than 90 cc/M², then high operative mortality is seen. Patients with acute MR secondary to infarction are usually in a low output state and require inotropic support, IABP and emergent CABG plus MVR.

AORTIC STENOSIS

Etiology:

The most common form of valvular aortic stenosis is calcification and/or fibrosis of a congenital bicuspid valve (rarely get regurgitation). Rheumatic fever may account for 1/3 of all cases of AS, but the mitral valve will also be diseased. Senile calcific stenosis of the aortic valve in older patients is a common form of valvular calcification in the United States. Subvalvular and supralvular narrowing can also occur.



(figure courtesy Dr. E. O. Feigl)

Pathophysiology:

The narrowed aortic valve creates a resistance to flow and causes a drop in pressure from the left ventricle to the aorta. In response, the left ventricle hypertrophies as all muscle does in response to increased tension. The wall thickens to keep wall tension constant, [Law of Laplace: $Tension = (Pressure \times Radius) / (2 \times Wall \text{ Thickness})$]. The thick muscle leads to a fall in diastolic ventricular compliance and therefore end-diastolic pressure must rise in order to maintain the same end-diastolic volume. In AS the atrial kick is responsible for up to 40% (normal 15-20%) of filling but the kick is too brief to increase mean left atrial pressure significantly. Loss of the atrial kick must be met by a drop in ventricular filling (causing decreased cardiac output) or a rise in mean left atrial pressures (causing pulmonary congestion/edema). When the atrial kick is present, an S4 is heard because of the relatively large amount of blood pushed into a stiff LV. Although the wall thickens, end-diastolic volume is largely unchanged (concentric hypertrophy). Because of the narrowed opening and the limitations on maximum chamber pressure (260 to 300 mm Hg), it takes longer to eject the blood and systole represents a greater percentage of total cardiac cycle time. A massive muscle that

contracts with great strength over a long period of time causes a pronounced and prolonged movement of the lower precordium, i.e., a heave.

As in mitral stenosis, it is difficult to increase cardiac output. Increased output requires the valve gradient to rise by roughly the square of the output. The ventricle may not be able to generate such pressures so the increase in cardiac output may be limited.

As compensatory hypertrophy reaches its limits, muscle degeneration occurs, contractility falls and the left ventricle dilate (enlarges without an appropriate increase in muscle mass) to maintain chamber pressure via the Frank-Starling mechanism. High PA pressures, a low ejection fraction and a below normal cardiac output complete this picture of LV failure.

Ischemia and angina are common in AS, even in the absence of coronary artery disease. Myocardial oxygen demand is high because of the high intraventricular pressure, but oxygen delivery is impaired by the low aortic pressure, the high end-diastolic pressure, the thick myocardial wall (increased intercapillary diffusion distance) and the shortened diastolic time relative to the time of systole. Chronic ischemia may lead to patchy necrosis and speed the decline in contractility. Ischemia/necrosis of the conduction system may lead to A-V block or bundle branch block. Episodic ischemia may cause ventricular arrhythmias or impaired contractility, either of which can lead to light-headedness, syncope or even sudden death.

Symptoms of Aortic Stenosis:

The first symptoms to appear often occur with exertion. An increase in cardiac output worsens the pressure drop across the obstruction and as described above, may lead to angina, palpitations, dizziness/syncope or dyspnea on exertion. Once failure develops, fatigue, cough, PND, orthopnea become evident.

Sudden Death - 0.4% per year in asymptomatic patient.

Physical Findings in Aortic Stenosis:

1. Pulse - of prolonged duration with a gradual upstroke.
2. Jugular veins - normal unless failure present.
3. Palpation - Unless failure present, PMI will be minimally displaced. The PMI will be diffuse, prolonged and prominent (LV heave).
4. Auscultation:
 - a. S1 - normal.
 - b. S2 - usually only single sound (P2) because A2 is lost as the excursion of the aortic valve diminishes. If A2 present, prolonged LV systole makes A2 late, even to the point of occurring after P2 (so get paradoxical S2 splitting).
 - c. S3 - with LV failure or decompensation.
 - d. S4 - expect to hear it if in sinus rhythm.

e. Systolic ejection murmur - Begins after S1, crescendo - decrescendo (starts soft, builds to a peak, then trails off), harsh or raspy, usually loudest at the second right sternal edge but often heard across the chest and in the carotids. May hear a click between S1 and the murmur onset. As AS gets more severe, the murmur tends to peak later and get louder, but in failure the murmur may soften due to low SV.

EKG:

Left ventricular hypertrophy is commonly observed. Left atrial enlargement and/or conduction disturbances may be present. The rhythm is usually sinus.

CXR:

Rounding of the lower left border of the heart shadow, poststenotic aortic dilatation, valve cusp calcification - or CXR may look entirely normal.

Associated Features:

1. More than 50% of adults with AS have CAD.
2. Cerebral emboli from vegetations off of a bicuspid valve.
3. GI bleeding secondary to angiodysplasia in the right colon (rare).

Severity of Aortic Stenosis:

Anyone with symptoms should be considered to be at high risk. This is reflected in the mortality figures. Mean life expectancy after onset of angina is 5 years, with syncope it is 3 years and once failure develops, only a 2 year 50% survival. Aortic stenosis is probably the most dangerous valvular lesion, in large part because once hypotension develops, it quickly leads to a vicious circle of ischemia, to more hypotension, to more ischemia. When these patients arrest, chest compressions are largely ineffective because the stenosis limits flow.

A pressure gradient of 50 mm Hg suggests a severe stenosis but valve area provides a more accurate assessment of severity. Less than 1.5 cm^2 is severe and below $.75 \text{ cm}^2$ is defined as critical.

Indications for Surgery in A.S.:

1. All children with noncalcified bicuspid valves and CHF require urgent commissurotomy.
2. Valve area $< 0.75 \text{ cm}^2$ (or $< 0.4 \text{ cm}^2/\text{M}^2$). [Normal area of AV is $2.5\text{-}3.5 \text{ cm}^2$].
3. Severe associated AI with an aortic diastolic pressure $< 50 \text{ mm Hg}$.

4. Any symptoms of CHF, angina, syncope.
5. Patients with noncritical AS and no symptoms and a normal heart size but in whom any physically demanding work cannot be performed.

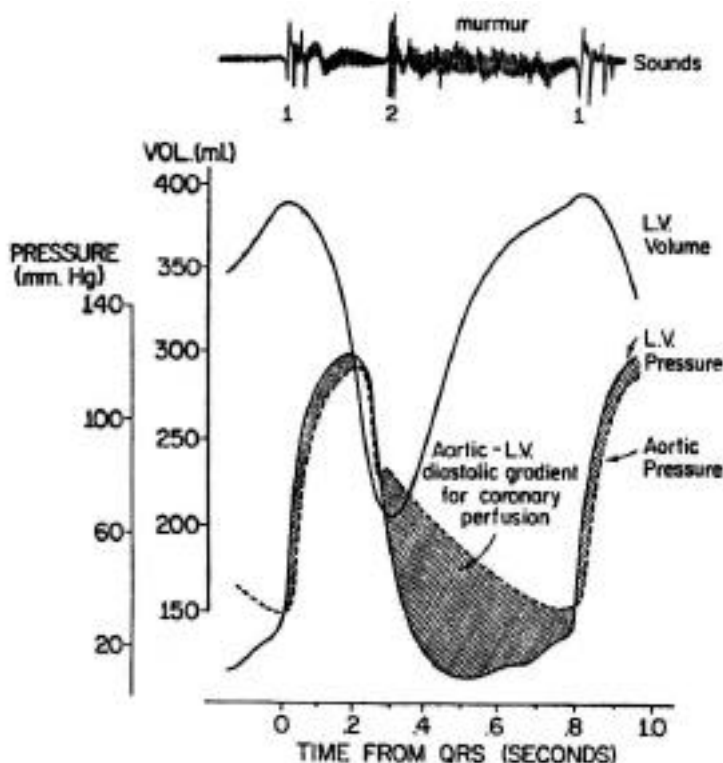
Management of Aortic Stenosis:

1. Maintain sinus rhythm because of the important atrial kick. Halothane and enflurane are relatively contraindicated because they tend to cause nodal rhythms.
2. Avoid tachycardia - A normal heart ejects blood faster as HR rises. In AS, ejection rate is limited so high heart rates may not increase CO. High heart rates may aggravate ischemia. Very high heart rates promote failure by inadequate diastolic filling times and inadequate LV emptying.
3. Maintain a high normal preload because a non-compliant LV requires a high filling pressure in order to maintain a normal end-diastolic volume. Even higher LVEDPs are needed if the atrial kick is lost.
4. Avoid decreases in SVR because cardiac output won't rise much in compensation so BP will fall. In a normal heart CO will rise when SVR falls but in AS, the stenosis limits the rise in CO. Conversely, systemic hypertension may lead to failure.
5. Do not depress contractility.
6. Concomitant ischemic heart disease - Worsens an already impaired oxygen delivery system. Use nitroglycerin with caution because veno- and vasodilation can lead to worse hypotension. The first priority should be an adequate aortic diastolic pressure and this is achieved primarily by supporting blood pressure with inotropes, vasoconstrictors and fluid, as needed.
7. PA catheter can help sort out hypotension due to low SVR versus low CO due to failure versus low CO due to inadequate preload. Balance this benefit against the increased risk of arrhythmias during catheter placement. CVP is an inadequate measure of filling of a stiff LV. The A-wave may be prominent in PAW tracing - if so, the peak of the A-wave is a better representation of LVEDP than PAW.
8. Generous premed to prevent anxiety from causing tachycardia.
9. If BP falls, treat first then figure out why (must avoid vicious downhill spiral).

CHRONIC AORTIC REGURGITATION

Etiology:

Rheumatic fever, congenital defects (e.g., unicuspid valve), endocarditis, aortic dissection or aortic lesions such as cystic medial necrosis or Takuyasu's aortitis, connective tissue disorders, hypertension, syphilis, just to name a few. Acquired AI much more common than AI on a purely congenital basis.



(figure courtesy Dr. E. O. Feigl)

Pathophysiology:

Inadequate valve closure allows blood to enter the LV from the aorta during diastole. With regurgitation, aortic pressure falls rapidly and aortic diastolic pressure is often very low. LVEDP is elevated because of the regurgitant flow that tries to equilibrate LVEDP with diastolic aortic pressure, but a short diastolic time period and a large, compliant LV may prevent the equalization of pressures from occurring. LA pressures (and PAW) may not be very elevated. Forward cardiac output is preserved by increases in stroke volume via LV dilation. LV wall thickness is maintained at normal levels (eccentric hypertrophy). BP, CO and SVR are well preserved until compensation can go no further and contractility declines. As in MR, an ejection fraction of 50% or less suggests decompensation with diminished contractility. Once the limits of compensation are achieved and contractility begins to decline, forward (systemic) cardiac output declines.

Symptoms of Chronic Aortic Insufficiency:

With elevated pulmonary pressures, fatigue, PND and orthopnea appear. Angina may be present and exacerbated by the low diastolic pressure. Dyspnea on exertion is not prominent, because the tachycardia that accompanies exertion causes the diastolic time to fall and this limits regurgitant flow and tends to raise diastolic pressure.

Physical Findings in Chronic Aortic Regurgitation:

1. Pulse - low diastolic pressure. Very brisk upstroke followed by a rapid trailing off is a key finding (Corrigan's pulse). The pulse may be so dynamic that with each pulse, the head may bob (de Musset's sign), and the liver pulsate (Rosenbach's sign) (spleen pulsation is Gerhardt's sign). Other findings include systolic and diastolic femoral bruits with pressure placed (Durozier's sign) on the artery (to and fro blood flow) and nailbed pulsations with pressure on the nail (Quincke's sign). Landolfe's sign: pupil size change with each heart beat. Hill's sign: popliteal systolic BP 60 mm Hg higher than brachial systolic BP; is supposed to pathognomonic of severe AI.

2. Jugular veins - normal.

3. Palpation - marked lateral and inferior displacement of the PMI. PMI forceful and brisk.

4. Auscultation:

a. S1 - soft or normal.

b. S2 - usually normal.

c. S3 - with LV failure.

d. S4 - with prominent LV hypertrophy.

e. Diastolic murmur - soft, high-pitched, blowing in character, most prominent in early diastole and then trails off (decrescendo murmur) or may last throughout diastole, heard best along the left sternal border or 2nd right sternal edge. May also hear an Austin-Flint murmur (soft, low pitched, mid-diastolic to late diastolic, sounds like MS, due to the aortic regurgitation pushing the anterior mitral leaflet upward and/or noise of the aortic regurgitation jet itself).

f. Systolic murmur - the increased aortic valve flow may create an audible ejection murmur. If murmurs suggesting AS and AI are present, must turn to peripheral exam to decide which lesion is the more serious. Severe AI is associated with a large pulse pressure, Hill's sign, a large, active and displaced PMI, an Austin-Flint murmur, and a diastolic murmur lasting $> 2/3$ of diastole.

Management of Chronic Aortic Insufficiency:

Management centers around improving forward output. The degree of regurgitation depends on (1) the size of the incompetent aorta, (2) the pressure difference between the aorta and the ventricle, and (3) the duration of systole. Regurgitation increases with an increase in the area of valvular regurgitation, with diastolic hypertension, and with bradycardia.

1. Avoid bradycardia (low HR means long diastoles, more regurgitation and lower diastolic pressure). The atrial kick is relatively unimportant. Increased HR may decrease regurgitant flow by decreasing diastolic time. Even if increased HR fails to lower the regurgitant fraction, total cardiac output and therefore forward output may be improved. Tachycardia may also tend to reduce LVEDV.

2. Preload should be high but without causing pulmonary edema.

3. Keep SVR low to enhance forward output. A given aortic pressure can be achieved with a low or a high CO depending on SVR. A low SVR allows CO to rise without increasing aortic pressure. Decreased SVR tends to limit regurgitation by lowering diastolic pressure but the degree of afterload reduction may be limited by the decline in diastolic pressure.

4. Do not depress contractility. Isoproterenol may improve contractility and cause vasodilation and increased HR as well.

5. In the absence of failure, CVP is likely to be an adequate measure of LV filling pressures. However, close control of SVR can only be achieved with a PA catheter and thermodilution CO.

6. A rise in PAW pressure suggests LV dysfunction because one expects good LV compliance and therefore minimally elevated PAW. Be especially watchful for a rise in PAW in patients ill enough for AVR because their contractility is probably depressed (see pre-op EF).

ACUTE AORTIC REGURGITATION

Etiology:

Most often endocarditis. Extension of the infection may lead to 1, 2, or 3 heart block, PVCs, pericarditis, ventricular rupture. Embolization of infected vegetation may occur.

Other Causes:

1. Aortic dissection or aortic aneurysm.
2. Closed chest trauma.
3. Spontaneous rupture of bicuspid cusp or myxomatous valve.

Pathophysiology:

Acute aortic regurgitation does not permit time for the LV to turn into a high-volume, high-compliance pump. Acute regurgitation therefore results in a marked increase in LVEDP as LV filling is increased by the regurgitation. The large rise in LVEDP can only increase end-diastolic volume a little, so stroke volume is minimally raised. (The LV is normally close to being maximally full, hence little reserve.) Since total output is only modestly raised, the regurgitant flow leads to a decrease in forward output. Low CO causes high sympathetic tone. The pulse pressure is not widened because stroke volume is not large. The high LVEDP leads to elevated LA pressures, although the regurgitant flow may raise LV diastolic pressure so quickly as to cause premature closure of the mitral valve. Premature closure prevents the LAP from equalling the high LVEDP, but it also prematurely stops filling of the LV from the LA. Note that this means that PAW will be less than LVEDP. (If MR is present, this protection is lost.) Some acute compensation is achieved by high sympathetic tone and tachycardia, the former preserves BP while the latter improves total CO and limits diastolic pressure falloff.

Symptoms:

Mainly those of CHF: dyspnea at rest and/or exertion, PND, orthopnea, fatigue. If asymptomatic at rest, will often show severe congestion with exertion. In general, organ perfusion is poor.

Physical Findings:

1. General - low forward output plus high sympathetic tone may cause cool, pale extremities and tachycardia.
2. Pulse - the signs of large pulse pressures (as in chronic AI) are not present.

3. PMI - is essentially normal.
4. S1 - may be absent if the mitral valve prematurely closes.
5. S3 present, S4 absent (premature mitral closure).
6. Murmur - begins at S2, is mid-pitched, soft, and short in duration. An Austin-Flint murmur is prominent in acute AI.

ECG:

May show heart block in endocarditis. Some repolarization abnormalities may be present.

CXR:

Pulmonary congestion, possible dissection.

Natural History:

Significant, acute AI rapidly leads to cardiac (and systemic) deterioration and death. This is especially true in patients with prior LVH (e.g., HTN, AS) as the non-compliant LV tolerates a volume overload less well than a normal heart does.

Therapy and Management:

Surgery is vital but medical stabilization may help. Diuretics for pulmonary congestion, inotropes for improved cardiac output, and vasodilation to reduce the regurgitant fraction may all help (but beware a fall in BP with vasodilators as there are limits to the reduction of regurgitant flow). IABP is not helpful as it enhances regurgitant flow.

Anesthetic management is similar to chronic AI, except that more active support of contractility may be necessary (beware in dissection, though) and some preload reduction may be necessary to treat pulmonary edema.

IDIOPATHIC HYPERTROPHIC SUBAORTIC STENOSIS

Etiology:

Muscular hypertrophy primarily of the ventricular septum (asymmetric septal hypertrophy - ASH). Histological exam reveals bizarre, disorganized muscle fibers in the affected region. The disorder is genetic (autosomal dominant) and can affect the entire myocardium, but LV outflow obstruction most often occurs in patients with disease localized to the septum.

Pathophysiology:

LV outflow obstruction occurs in some but not all patients with ASH. The LV outflow tract is bordered by the anterior mitral leaflet and the ventricular septum. When obstruction occurs, it is the result of the abnormally hypertrophied septum pushing against the anterior mitral leaflet. The anterior mitral leaflet may move anteriorly during systole (SAM: systolic anterior motion) and worsen the obstruction. The narrowing of the outflow tract creates a pressure gradient across the tract when ejection occurs. The aortic valve, however, is normal and has no pressure gradient.

The thickened ventricle is also stiffer than normal so compliance is reduced and end-diastolic pressure must increase to maintain normal end-diastolic volume. This feature is present even in the absence of significant outflow obstruction. Poor ventricular diastolic compliance can lead to pulmonary congestion even though CO is normal and the ejection fraction is high (i.e., no loss of contractility).

The key feature to IHSS is the fact that the stenosis is dynamic; the degree of obstruction varies with the size of the left ventricle. Maneuvers that dilate the heart (e.g. increased aortic pressure, volume expansion, decreased contractility) also distend the outflow tract and relieve the obstruction. Maneuvers that empty the heart (decreased afterload, decreased preload, increased contractility) allow the septum to close off the outflow tract even more. The obstruction and resulting pressure gradient develops as ejection proceeds. If the heart starts a contraction relatively empty, outflow obstruction starts early in systole and rapidly worsens. If contraction occurs with a full heart, more ejection can occur before the ventricle shrinks enough to create a significant obstruction. Hypovolemia during surgery should be avoided.

Although the hypertrophy progresses slowly (years to decades), symptoms may appear in young patients but usually well before myocardial damage has occurred. Indeed, the LV is hypercontractile with ejection fractions over 80%. Eventually muscle degeneration develops via mechanisms similar to those that operate in AS.

Symptoms of IHSS:

As in AS, pre-syncope, syncope (not as ominous as in AS), dyspnea, sudden death, and less frequently, angina. Palpitations are common.

Physical Findings in IHSS:

1. Pulse - brisk upstroke, often with a double peak (remember that in valvular AS the upstroke is slow). The second peak occurs as the ventricle begins to relax which lessens the obstruction and permits more rapid ejection. Post-extrasystolic beats do not have a stronger pulse (the high inotropy clamps down on the outflow tract and limits the ejection).
2. Palpation - vigorous, brief PMI
3. Jugular veins - normal unless elevated RV pressures
4. Auscultation
 - a. S1 - normal
 - b. S2 - usually single sound (P2) or paradoxically split
 - c. S3 - no
 - d. S4 - likely present (atrial kick important to LV filling)
 - e. Systolic ejection murmur - similar to character to AS, but loudest at the lower left sternal border or the apex, not at the base. Murmur gets louder with amyl nitrite, sudden standing, Valsalva, beta agonism (all decrease ventricular size). Murmur gets softer with vasopressors, handgrip, sudden squatting, beta blockade (all increase ventricular size).

Key point - All systolic ejection murmurs should be examined a) with the patient supine and b) immediately on standing. Lack of an increase in murmur intensity should rule out IHSS. Note: The murmur of a ventricular septal defect is also heard during systole and heard best at the left sternal border. Unlike IHSS, the VSD murmur will be pansystolic. In addition, in VSD, S2 will be normal, a heave will not be present nor will S4, and the murmur gets louder with ventricular dilation. A VSD murmur is similar to that of mitral regurgitation except that in MR the murmur is most prominent at the apex and radiates to the axilla. In severe VSD, pulmonary congestion is prominent.

f. A murmur of MR may accompany IHSS because the septum may push against and distort the anterior mitral leaflet.

Severity of IHSS:

The severity of the dynamic obstruction can be lessened by medical therapy, namely, beta blockade. Prior to elective surgery, such therapy should be optimized. In the absence of failure, adequate beta blockade likely means complete beta blockade achieved by at least 320 mg Inderal daily. Beta blockade not only dilates the heart but blunts the rise in contractility associated with events that raise sympathetic tone (e.g. intubation).

Management of IHSS:

1. Maintain sinus rhythm. As in AS and MS the atrial kick contributes a lot to LV filling.
2. Avoid tachycardia. High heart rates result in diminished EDV.
3. Maintain normal to high levels of end-diastolic pressure (PAW). Beware drops in preload that may occur with increased intrathoracic pressure (as with positive pressure ventilation, PEEP or air trapping in bronchoconstriction) or body positional changes (e.g. reverse Trendelenburg).
4. Do not let SVR fall - unloading the aorta promotes more complete ventricular emptying which worsens the obstruction.
5. Maintain low contractility - Halothane, enflurane are good anesthetic choices (unlike in AS) and have Inderal handy.
6. Worsened obstruction will manifest itself as hypotension. It should be treated by fluids, Trendelenburg position, phenylephrine and halothane/propranolol. Note that the depression of contractility is not the usual treatment of hypotension.

Some Final Key Points:

1. Stenotic lesions are more dangerous than regurgitant lesions. Most dangerous is AS - if acute LV failure occurs it is difficult to treat and nearly impossible to perform adequate chest compressions.
2. Any lesion that has led to a decompensated LV (as MR, AS, AI can do) deserves invasive monitoring (A-line, Swan) and vasoactive drips (neo, nipride, NTG, dopamine) should be available if not already mixed.
3. Concomitant coronary artery disease complicates management, especially in AS. Maneuvers used to prevent ischemia (low HR, adequate diastolic pressure, decreased preload) may not be desirable for certain valvular lesions and vice versa. Therapy is usually based on which lesion is more pressing at the time.
4. Stenotic lesions depend on the atrial kick for adequate LV filling. Regurgitant lesions do better with high heart rates, stenotic lesions best at normal heart rates.
5. The character of the carotid pulses and the PMI often provide the best clues to the severity of MR, AS and AI. For MS, the history is best. Murmurs provide little more than identification of the lesion.
6. Hemodynamic stability is imperative because hypertension, hypotension, tachycardia and bradycardia are more likely to lead to a vicious circle and rapid deterioration in valvular disease (esp. AS) than in normal patients.
7. For any surgery requiring a CVP line in a normal patient, place a PA catheter instead. Measurements of CO and SVR (especially in AI) are useful in sorting out untoward events and gauging therapy. In addition, CVP is probably a poor correlate of PAW in the presence of significant mitral valve disease (MR or MS), a non-compliant LV (e.g. AS), RV decompensation, LV decompensation or pulmonary hypertension.
8. In anyone with chronic atrial fibrillation, do not try to cardiovert. Conversion to NSR may cause atrial mural thrombi to break free. Besides, NSR will likely only be transient.

Suggested General References:

Manual of Cardiac Anesthesia, Stephen J. Thomas, Ed., Churchill-Livingstone, New York, 1984, Chapter 9, pp. 231-258.

Valvular Heart Disease, James E. Dalen and Joseph S. Alpert, Little-Brown and Co., Boston, 1981.

Cardiac Anesthesia, Joel A. Kaplan, ed., Grune and Stratton, New York, 1979, Chapter 6.