Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Dysphagia	Coughing, choking, aspiration with feeding	Sialorrhea	Swallow study
CV	Cardiomyopathy Conduction defects (KSS)	Symptoms of CHF	Murmur, gallop, crackles	CXR, ECHO ECG, exercise testing (VO_2 max)
RESP	Disorganized respiratory muscle effort	Hypoventilation, hypoxia following sedative use	Rhonchi	CXR
GI	Chronic diarrhea Exocrine pancreatic failure (PS)	Dehydration Steatorrhea		Serum lytes
ENDO/METAB	Lactic acidosis Hepatic insufficiency	N/V Prolonged Rx effects	Hyperventilation	Serum lactate, CSF pyruvate/lactate ratio
GU	Renal tubular defects (PS), nephropathy	Urinary changes		Urinalysis Serum BUN, Cr, lytes
CNS	Encephalopathy (MILS) Ophthalmoplegia (CPEO, KSS) Stroke (MELAS) Seizure (MELAS, MERRF) Retinopathy, ataxia (NARP), blindness (LHON), deafness	Developmental delay Poor visual tracking Poor coordination Vision loss	Decreased ROM of extraocular mm Decreased visual acuity, ptosis Focal neurologic deficits Signs of seizures Pigmented retinas	Head CT or MRI Ophtho exam
PNS	Peripheral neuropathy	Weakness, clumsiness	Decreased strength	
MS	Hypotonia, weakness Myoclonus (MERRF)		Decreased strength	Muscle biopsy (ragged red fibers)

Clinical findings listed above may be characteristic of one or more mitochondrial myopathies. A specific disorder may follow in parentheses if the finding is a primary feature.
Key Reference: Niezgoda J, Morgan PG: Anesthetic considerations in patients with mitochondrial defects, *Paediatr Anaesth* 23(9):785–793, 2013.

Perioperative Implications

Preoperative Preparation
- Assess for cardiomyopathy and conduction defect.
- Preop anticholinergic for excessive oral secretions.
- Avoid prolonged fasting and dehydration, which can worsen acidosis.
- When possible, start IVF at NPO time, allow for late (2 h prior) clear fluid intake, and book as first case.

Airway
- Possible aspiration risk

Monitoring
- Routine, assuming no severe cardiomyopathy or CHF
- Consider BIS monitor prior to induction for possible increased anesthetic sensitivity

Induction
- Avoid lactate-containing IVF (e.g., lactated Ringer).
- Consider dextrose-containing IVF (e.g., 2–5% dextrose in normal saline).
- Avoid succinylcholine for uncharacterized myopathy or in face of neuropathy.

Maintenance
- Many techniques have been used safely.
- Avoid prolonged infusion of IV anesthetics, especially propofol, which is a known electron transport chain decoupler, due to worsened acidosis and reduced ATP production.
- Hepatic and renal insufficiency may increase IV anesthetic half-life and prolong elimination.
- If NMB agent is required, consider careful titration with shorter-acting agents.
- Implement aggressive temp control; recommend active warming techniques.
- Avoid tourniquets.

Extubation
- Muscle weakness and anesthetic sensitivity may delay extubation.

Regional Anesthesia
- Used successfully, but caution in those with underlying cardiac conduction block.
- Local anesthetics have potential to decouple electron transport chain.

Postoperative Period
- Close monitoring of respiratory function.
- For cases of longer duration, consider serum electrolytes or blood gas to assess acidosis.
- Some have reported increased incidence of PONV.

Anticipated Problems/Concerns
- Generally not associated with MH; however, scenario of critical ATP depletion may lead to muscular contraction mimicking MH.
- Although succinylcholine is not contraindicated as in Duchenne or Becker MD, acidosis and neuropathy may predispose to accentuation of hyperkalemia.

Mitral Regurgitation

Luiz Maracaja | Raj K. Modak

Risk
- Mitral regurgitation affects more than 2 million people in USA.
- Incidence of moderate/severe mitral regurgitation: Nearly 20% for age >55 y.
- Mitral valve prolapse is the primary form of myxomatous degeneration of the valve.
- Mitral valve prolapse is the most frequent diagnosed valve abnormality.
- Incidence in females is slightly higher than in males.

Perioperative Risks
- Acute mitral regurgitation
- Atrial arrhythmias (tachycardia, atrial fibrillation, atrial flutter)
- LV dysfunction yielding reduced cardiac output, acute CHF, pulm edema, and acute RV failure
- Bacterial endocarditis

Worry About
- Worsening symptoms of fatigue, orthopnea, dyspnea on exertion
- Acute or chronic mitral regurgitation
- New-onset atrial fibrillation
- Hemodynamic instability in setting of poor LV function and acute MI

Overview
- The mitral valve allows one-way blood flow through the left heart.
- During diastole, it acts as an open conduit for blood flow from the LA to the LV During systole, it closes preventing backflow while the heart contracts.
- With mitral regurgitation, retrograde flow occurs from the LV to the LA during systole. This can occur as an acute or chronic process.
- The acute form results in sudden elevations in LA pressure. Elevated pressures in the pulm vasculature resulting in pulm edema and RV strain and possible failure.
- Chronic mitral regurgitation is tolerated well. LV hypertrophy is followed by dilation and failure. Similar changes in the RV and pulm circulation occur, as in the acute form, but are better tolerated over the longer time period.
- As a general rule, the more precipitous the onset, the more significant the sequelae.

Etiology
- Acute: Myocardial ischemia or MI causing papillary muscle dysfunction, ruptured chordae causing a flail mitral valve from infarction or endocarditis, trauma, prosthetic valve dysfunction.

- Chronic: Include acute processes over longer time, myxomatous degeneration, ischemic heart disease, dilated cardiomyopathy, rheumatic disease, lupus, congenital valvular disease, LA myxoma. All forms can be accelerated by systemic Htn.

Usual Treatment

- Medical therapy: Afterload reduction, CHF regimens, arrhythmia control, endocarditis prophylaxis
- Pharmacology: Includes Angiotensin inhibitors, hydralazine, cardiac glycosides, diuretics, nitrates, antibiotics
- Surgical therapy: Mitral valvuloplasty (repair), annuloplasty, mitral valve replacement (mechanical and tissue)
- Transcatheter mitral valve interventions: Edge-to-edge repair/enhanced coaptation, chordal repair, annuloplasty, mitral valve implantation

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Mitral regurgitation LA enlargement	Fatigue, exertional or nocturnal dyspnea	Pansystolic and late systolic murmur, rales	Doppler ECHO, 2D/3D ECHO 2D-ECHO ECG
	AFIB	Palpitations, defibrillation, anticoagulation	Irregular rhythm, bruises	ECG, PT/INR
	RV failure	Peripheral swelling, RUQ pain, tenderness	Ankle edema, hepatomegaly, hepatojugular reflux	Cardiac cath 2D and Doppler ECHO
	Cardiomegaly		Displaced posterior MI	CXR, 2D ECHO
RESP	CHF, pulm edema	Dyspnea, orthopnea	Gallop, rales	CXR
GI	Congestive hepatopathy	Bleeding with minor trauma	Bruises	PT, PTT, LFTs
RENAL	Decreased perfusion Diuretic-induced Decreased K^+, Mg^{2+}	Oliguria Palpitations	 Muscle weakness Decreased reflexes	Decreased BUN, Cr Serum K^+, Mg ECG
MS	Cachexia	Weight loss	Muscle wasting	Decrease weight

Key References: Bhattacharyya S, Khattar R, Chahal N, et al.: Dynamic mitral regurgitation: review of evidence base, assessment and implications for clinical management, *Cardiol Rev* 23(3):142–147, 2015; Al-Atassi T, Malas T, Mesana T, et al.: Mitral valve interventions in heart failure, *Curr Opin Cardiol* 29(2):192–197, 2014.

Perioperative Implications

Preoperative Preparation

- Antibiotic prophylaxis.
- Manage anticoagulation issues related to atrial fibrillation and the possible use of regional techniques.
- Optimize HR issues related to AFIB.
- Optimize symptoms related to CHF.
- TEE with 3D imaging is essential for disease classification, surgical planning, and post-intervention valve assessment.

Monitoring

- In procedures with expected wide variations in BP, direct arterial BP monitoring should be considered, especially with moderate or severe mitral regurgitation.
- In settings of LV failure, a pulm artery cath or TEE may be useful in assessing changes and guiding pharmacologic therapy.

Airway

- Avoid hypoxemia and hypercarbia, which maintains the lowest pulm vascular resistance and reduces risk of RV failure.

Preinduction/Induction

- "Faster, Fuller, Forward."
- Avoid bradycardia, maintain high-normal preload, reduce afterload.
- Maintain stoke volume by avoiding myocardial depression and AFIB.

Maintenance

- Cardiac and pulm goals, same as induction.
- Avoid excessive PEEP, which reduces preload.
- If possible, follow cardiac output, using pharmacology as needed.
- Regional anesthetic techniques may be considered because they help to reduce afterload; however, caution is recommended in the setting of impaired LV function.

Extubation

- Airway management to avoid hypoxia and hypercarbia inducing RV strain and failure
- Requires vigilance on BP management to avoid Htn
- Transcatheter MV procedures slowly evolving to monitored anesthetic delivery without need for general endotracheal anesthesia

Adjuvants

- No known drug interaction problems

Postoperative Period

- Pain management critical to avoid hypertensive episodes.
- Both scheduled and pt-controlled analgesia useful for pain control.
- Fluid shifts and intraop volume management may alter LV function and antiarrhythmic blood concentrations.
- New onset AFIB.
- Consideration for pacemaker use: new-onset bradycardia and heart block related to manipulation and device insertion via direct pacing wires or transvenous pacing device.
- Consideration for restarting anticoagulation for chronic AFIB.

Anticipated Problems/Concerns

- High periop risk is best predicted by impaired LV function, symptoms of both LV and RV dysfunction.
- Htn can acutely worsen mitral regurgitation, causing CHF and pulm edema.

Mitral Stenosis

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Risk

- Bimodal age distribution: 20–39 y and 50–60 y.
- Mitral stenosis is 2–3 times more common in women and is the most common valve disease affecting pregnant women.
- Most common among immigrants to USA from regions where rheumatic fever is prevalent (e.g., Middle East, Asia, Latin America).

Perioperative Risks

- Increased risk of periop cardiac complications, including infectious endocarditis, pulm edema, resp failure, HF, tachyarrhythmias, new-onset AFIB or atrial flutter, embolic stroke of cardiac origin (0.7–0.9% risk of stroke after cardioversion)

Worry About

- Fluid status
- Paroxysmal AFIB or flutter
- Pregnancy
- Limited ability to increase cardiac output in response to increased metabolic demands or intravascular volume expansion
- Tachycardia, AFIB, or atrial flutter decreases atrial emptying by decreasing the duration of diastole
- Cardiomyopathy, pulm Htn, RV failure, hepatic dysfunction, tricuspid regurgitation, and associated aortic valve disease
- Pulm edema

Overview

- The normal mitral valve has an area of 4–6 cm^2. Symptoms start when the mitral valve area is reduced to 1.5 cm^2. Diastolic emptying of blood from the LA into the LV is impaired critically when the mitral valve area is <1 cm^2.
- MS can be classified as at risk for MS (Stage A), progressive MS (Stage B), asymptomatic severe MS (Stage C), or symptomatic severe MS (Stage D), based on the presence of dyspnea on exertion, exercise intolerance, diastolic doming and commissural fusion of the mitral valve leaflets, left atrial enlargement, and pulm Htn.
- Transmitral pressure gradient varies directly with blood flow across the valve; acute increases