

Assessment Points				
System	Effect	Assessment by Hx	PE	Test
CV	Atrial myxoma (ball-valve effect)	Sx of CHF, pulm edema	Rales S ₃	CXR ECHO CT angio
	Vena cava obstruction	RV failure, CV collapse	Possible caput medusae, venous engorgement, edema	
	SVC syndrome	Head swelling, airway edema, increased ICP	Venous congestion of head and neck	CT angio
RESP	Pulm embolus	Dyspnea		PE protocol CT
GI	Gastroparesis Bowel obstruction Hepatic metastases Sarcoma of ampulla of Vater	Early satiety Vomiting Obstructive jaundice Hepatic dysfunction	Abdominal distention Jaundice	X-ray of abdomen EGD, bilirubin levels
HEME	Hypercoagulable Pancytopenia due to chemotherapy Anemia due to GI hemorrhage	Opportunistic infections Bruising, bleeding	Gross rectal bleeding	PT/PTT Leukocyte and plt counts CBC Stool guaiac
RENAL	Compression of ureters by retroperitoneal tumor	Sx renal failure		BUN/Cr Renal US or CT
CNS	CN compression	Various symptoms Dysphagia Loss of sensation, motor function	Neurologic exam	EMG Head CT
MS	Bone sarcomas Limb loss	Hypercalcemia	Chvostek's sign	Blood Ca ²⁺

Key References: Burningham Z, Hashibe M, Spector L, et al.: The epidemiology of sarcoma, *Clin Sarcoma Res* 2(1):14, 2012; Makela J, Kiviniemi H, Laitinen S: Prognostic factors predicting survival in the treatment of retroperitoneal sarcoma, *Eur J Surg Oncol* 26(6):552–555, 2000.

Perioperative Implications

Preoperative Preparation

- Use aspiration precautions for pts with gastroparesis, including prokinetics and NG decompression.
- Assess end-organ impairment (especially heart and lungs) secondary to chemotherapeutic agents.

Monitoring

- Use arterial line and CVP or PA cath for resection of large tumors.

Airway

- Risk of aspiration with large abdominal mass or brainstem compression.
- Possibility of difficult airway with SVC syndrome.

Induction

- Hemodynamic instability with cardiac involvement or caval compression.

Maintenance

- Potential blood loss

Extubation

- Awake if at risk for aspiration due to GI or CNS involvement.

Adjuvants

- Altered pharmacokinetics with hepatic or renal involvement.

Postoperative Period

- Pulm embolism, coagulopathy

Anticipated Problems/Concerns

- Adverse effects of chemotherapeutic agents
- Respiratory compromise due to pulm metastases
- Mass effect and/or organ compression and functional impairment
- Effects of prolonged anesthesia
- In prolonged abdominal cases, hypothermia, complications of massive transfusion

Scheie Syndrome (Mucopolysaccharidosis Type IS)

Arun K. Gupta

Risk

- Incidence: 1:500,000
- Inherited as autosomal recessive

Perioperative Risks

- Difficult intubation
- Positioning difficulty
- Cardiac problems

Worry About

- Macroglossia leading to difficult intubation
- Valvular heart disease, cardiomyopathy
- Stiff joints

- Spastic paresis
- Poor IV access

Overview

- Described by American ophthalmologist Harold Glendon Scheie in 1962.
- Mildest form of mucopolysaccharidosis type (1) and a rare lysosomal storage disease.
- Pts are of normal height and do not show intellectual deficiency.
- Corneal clouding and glaucoma.
- Coarse facial features.
- Normal life expectancy, stiff joints, and aortic regurgitation.

Etiology

- Caused by mutations in genes that control production of enzyme alpha-L-iduronidase (IDUA).
- The gene known as IDUA causes deficiency of enzymes and results in buildup of undigested mucopolysaccharides in the cells.
- Diagnosis occurs by elevated GAG in urine and demonstration of IDUA deficiency in leukocytes.

Usual Treatment

- Enzyme replacement
- Hematopoietic stem cell transplant
- Bone marrow transplant and cord blood transplant

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Difficult airway anticipation Coarse facial features Small chin, corneal clouding	Pain	Neck ROM	Neck x-ray Neck US
CV	Aortic regurgitation Cardiomyopathy Difficult IV access	Poor exercise tolerance Angina Hx		ECG, CXR, ECHO
RESP	Restrictive lung disease Obstructive sleep apnea			PFT, sleep studies
GI	Hepatosplenomegaly Umbilical hernia, inguinal hernia			
CNS	Many have normal intelligence Cervical cord compression Hydrocephalus leads to increased ICP, deafness			CT/MRI
MS	Stiff joints Carpal tunnel syndrome Spondylolisthesis	Joint mobility	Decreased ROM of joints	

Key References: Clarke LA, Heppner J: Mucopolysaccharidosis Type I. October 31, 2002 [Updated July 21, 2011]. In Pagon RA, Adam MP, Ardinger HH, et al. editors: *GeneReviews®*, Seattle, WA, 1993–2015, University of Washington. <<http://www.ncbi.nlm.nih.gov/books/NBK1162/>> (Accessed 06.06.16); Nakayama H, Arita H, Hanaoka K: Anesthesia in a patient with Scheie syndrome, *Masui* 43(9):1385–1388, 1994.

Perioperative Implications

Preoperative Preparation

- Anticipate upper airway obstruction.
- Excessive secretions (need for antisialagogue).
- Antibiotic prophylaxis for valvular heart disease.

Monitoring

- Routine

Airway

- Abnormal airway and short chin predispose to difficult airway.
- Large tongue and secretions leads to airway problems.

- Small size of ETT needed.
- Fiberoptic bronchoscopy.
- Use LMA and other supraglottic devices.

Induction

- IV access before induction
- Proper positioning with padding
- Spontaneous breathing

Maintenance

- Avoid myocardial ischemia.

Extubation

- Pt should be fully conscious, with intact airway reflexes.

Adjuvants

- Local anesthetics infiltration and regional anesthesia where needed

Postoperative Period

- Delayed emergence
- Pneumonia, bronchospasm, and laryngospasm apnea

Anticipated Problems/Concerns

- Management of difficult airway
- Cardiopulmonary problems common

Schizophrenia

Alan David Kaye | Christopher J. Cullom

Risk

- Most common psychotic disorder with a lifetime worldwide prevalence of 1%
- Increased risk of suicide (5–10%)

Perioperative Risks

- Marked by deterioration of function and self-care
- Exacerbation of psychosis with abrupt discontinuation of medications

Worry About

- Pt being uncooperative, combative, or catatonic.
- Increased morbidity and mortality due to poorly controlled coexisting systemic disease and increased incidence of alcohol and substance abuse.
- Drug interactions and side effects:
 - Cardiogenic side effects include hypotension, tachycardia, prolonged QT interval, VFIB, and torsades-de-pointes.
 - EPS include muscle rigidity and laryngospasm.
 - Use of metoclopramide may worsen schizophrenic symptoms.

Overview

- Schizophrenia is a psychiatric disorder that may be characterized by thought disorders, hallucinations, and fixed false beliefs.
- Antipsychotic medications are the mainstay treatment for schizophrenia.
- Antipsychotics have anticholinergic effects (dry mouth, blurry vision, urinary retention, constipation, tachycardia), histamine antagonism (sedation), and $\alpha 1$ antagonism (orthostatic hypotension).
- First-generation antipsychotics have strong dopamine antagonism leading to EPS, such as tardive dyskinesia.
- Second-generation or atypical antipsychotics have serotonin antagonism and less dopamine antagonism leading to less EPS.
- EPS can be treated with anticholinergics such as benztropine 2 mg or diphenhydramine 50–100 mg.
- NMS is a rare but potentially fatal syndrome occurring after an increase in dosage of antipsychotic medications or abrupt D/C of dopamine agonist. The syndrome is marked by muscle rigidity, hyperthermia, altered consciousness, and autonomic instability. It is clinically similar to malignant hyperthermia and may be related to dopamine blockade.

- Autonomic instability presents as labile blood pressure, tachycardia, diaphoresis, incontinence, and flushing.
- Treatment of NMS includes hydration and cooling measures, IV dantrolene, and dopamine agonists such as bromocriptine.
 - Bromocriptine reduces mortality by 50% and is only available orally; thus NGT may be required.
 - Dantrolene is a skeletal muscle relaxant and will reduce heat production.
 - Benzodiazepines may also be used to alleviate catatonic symptoms.
- Avoid dopamine antagonists, such as metoclopramide, if NMS is suspected.

Etiology

- Functional hyperactivity of dopamine transmission may play a role.
- Genetic and environmental factors are unclear and controversial.

Usual Treatment

- Antipsychotic medications
- Psychotherapy
- ECT