

Perioperative Implications**Preoperative Preparation**

- Oxygen therapy if decreased arterial oxygen saturation present
- Antibiotics for bronchopneumonia

Monitoring

- Bilateral upper extremity SpO₂ and Doppler probes are useful for assessing subclavian, carotid, and temporal pulses during temporary occlusion of the arch that is to be resected. Regional tissue oxygenation of the brain can be monitored via bilateral cerebral NIRS probes, which can reveal unilateral desaturation if carotid flow is compromised during arch manipulation and the patient's circle of Willis anatomy is not adequately providing collateral circulation.
- Potential for hemodynamic and respiratory instability warrant placement of arterial cath; presence of

an aberrant subclavian artery may affect appropriate cath site.

- Large-bore IV access is essential; central venous line should be considered for pts with poor vascular access and those who require extensive repair on CPB.

Airway

- Dynamic and static airway obstruction likely; significant tracheal compression may require smaller ETT size than predicted.

Induction

- Inhalation induction without neuromuscular blockade until airway maintenance is documented by mask and/or ETT is placed distal to area of obstruction.
- Bronchoscopy during spontaneous ventilation allows for direct assessment of tracheal pathology and degree of dynamic airway collapse, thus identifying pts at risk for postop respiratory compromise.

Maintenance

- Balanced technique of narcotics and volatile agent is usually well tolerated.

Extubation

- Extubation at end of case if tracheomalacia and stenosis absent

Postoperative Period

- Good pain control essential for stable hemodynamics and avoidance of respiratory complications; IV opioids, rectal acetaminophen, intercostal nerve blocks, one-shot caudal, and caudal epidural cath have all been used with success.

Anticipated Problems/Concerns

- Despite surgical correction, persistent postop airway obstruction requiring prolonged mechanical ventilation and CPAP can occur secondary to edema, mucosal friability/reactivity, and long-segment tracheomalacia.

Stephanie Black

Down Syndrome**Risk**

- Trisomy 21 is the most common autosomal aneuploidy; approximately 1:1000 live births.
- 80% of children with this condition survive beyond 1 y; average life expectancy 60 y.
- Increased incidence in mothers >35 y, but most are born to younger mothers, owing to higher fertility rates.
- Incidence decreased by elective termination of pregnancy from prenatal screening: high beta-hCG, low AFP, cell-free DNA, thickened nuchal fold, abnormal ductus venosus waveforms, absent nasal bone—cell-free DNA can now increase the sensitivity and specificity of these tests.

Perioperative Risks

- Airway obstruction
- Cardiac dysfunction due to CHD
- Cervical spine instability
- Immune and endocrinologic dysfunction

Worry About

- Airway obstruction:
 - Upper airway obstruction common immediately on induction of GA due to macroglossia, midface crowding, small mandible, short neck.
 - Subglottic stenosis in 20–25%; may cause postop stridor in children.

- Obstructive sleep apnea in 30–50%. Central apnea also common.
 - Chronic hypoxemia may contribute to pulm Htn risk and increased opioid sensitivity.
- Congenital cardiac dysfunction:
 - 40% are born with CHD.
 - Most common: Complete atrioventricular canal defect (40%), VSD (25%).
 - Cyanotic CHD in 4% (usually tetralogy of Fallot).
 - Risk for pulm Htn because of pulm overcirculation.
 - May develop R-to-L shunting with profound hypoxemia.
 - Risk for paradoxical/systemic air emboli (coronary or cerebral vessels).
 - Bradycardia with inhalational induction with sevoflurane.
- Cervical spine instability:
 - Extension during intubation can cause neurologic symptoms (neck pain, arm pain, upper extremity weakness, torticollis) from atlanto-occipital instability.
- Generalized joint laxity; TMJ may sublux with jaw thrust.
- Endocrine: hypothyroidism (4–6% in children; 15–20% in adults), hypothermia.
- Immune dysregulation causes higher rates of certain cancers (ALL and AML) and respiratory infections.
- GI: Duodenal atresia in 4%; recurrent aspiration may cause pneumonia.

- Developmental delay:
 - May have fears of the unknown; can become physically resistant to entering OR.
 - Alzheimer disease and other mental illnesses (depression, psychosis) may coexist.

Overview

- Most common autosomal aneuploidy with an increasing life expectancy because of early interventions for multiple comorbidities
- Concerns for congenital cardiac disease, hypotonia, immune dysregulation, airway obstruction, recurrent pneumonia, oncologic predisposition, and GI disorders
- Physical exam findings: Midface hypoplasia, brachycephaly, epicanthal folds, simian crease, downward medial slant of eyes, high-arched palate, glossoproposis, and murmur
- May require surgery for tympanostomy, strabismus, CHD repair, duodenal/esophageal atresia, marrow aspiration/biopsy, cervical spine fusion

Etiology

- Genetic: Trisomy 21

Usual Treatment

- Depends on penetrance and pathophysiology; may include the use of CPAP, thyroid hormone replacement, OT/PT

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Redundant tissue Midface hypoplasia Subglottic stenosis	Sleep apnea Intubation Hx Hearing deficit	Macroglossia Glossoproposis "Down's facies"	Audiology Polysomnography
CV	CHD in 40% CAVC most common Tetralogy of Fallot in 4% Bradycardia on induction Risk for pulm Htn	Symptoms of CHF "Tet spells" Hx of CHD repair	Cyanosis Murmur Clubbing	ECHO ECG
ENDO	Hypothyroidism Metabolic syndrome	Thyroid hormone replacement Hypothermia	Obesity	Thyroid hormone levels
IMMUNE/ONC	Oncologic predisposition Immune dysregulation	Respiratory infections AML and ALL	Cough Lymphadenopathy	Auscultation Bone marrow biopsy/aspiration
MS	Hypotonia Subluxation of C1/C2	Upper motor neuron symptoms	Joint laxity	Cervical spine radiographs (controversy over whether these should be routine)

Key References: Arumugam A, Raja K, Venugopalan M, et al.: Down syndrome—a narrative review, *Clin Anat* 29(5):568–577, 2016; Maxwell LG, Goodwin SR, Mancuso TJ, et al.: Systemic disorders: down syndrome. In Davis PJ, Cladis FP, Motoyama EK, editors: *Smith's anesthesia for infants and children*, ed 8, Philadelphia, PA, 2011, Elsevier, pp 1172–1174.

Perioperative Implications

Monitoring

- Temperature (hypothermia).
- ECG (arrhythmias, ischemia); consider IM antimuscarinic drug to treat bradycardia from inhalational induction with sevoflurane, avoid hypercarbia and hypoxia to prevent PHTN.

Airway

- Have variety of devices available (e.g., oral and nasal airways, laryngeal mask, glidescope, fiberoptic) to manage airway obstruction.

- Avoid neck extension during laryngoscopy if possible.
- Smaller endotracheal tube may be necessary for narrowed subglottic space.

Vascular Access

- Allow more time for IV placement.
- Meticulously avoid injected air.

Patient Management

- Soft, warm, kind pt approach along with caregiver known to pt to help with initial management; warm, quiet OR

Anticipated Problems/Concerns

- Refractory hypoxia if R-to-L shunting develops
- Bradycardia with inhalational induction
- Resistance to separation from caregiver
- Life-threatening upper airway obstruction with difficult vascular access
- Spinal cord ischemia with neurologic damage

Drug Abuse, Lysergic Acid Diethylamide

Alan David Kaye | Burton D. Beakley

Risk

- “Hallucinogen” with primary effects of heightened or distorted mood, thought, and sensory perception. The hallucinogen class includes LSD, mescaline, phencyclidine, and psilocybin. These drugs cause tolerance and psychological drug dependence but not physical drug dependence or withdrawal.
- Initially marketed as an anesthetic agent; people began using it for recreational and spiritual purpose in the 1960s. LSD is still illegally used as a major hallucinogen worldwide.
- LSD use peaked in the late 1960s, and use has been declining since. The National Survey on Drug Use and Health reports more than 200,000 people using LSD for the first time yearly.
- LSD-related hospital visits remain low compared with those related to other major illicit drugs. In 2011, Drug Abuse Warning Network reported more than 1 million emergency department visits for nonalcohol illicit drug use; of these only 4819 were related to LSD.
- LSD is semisynthetic and produces psychedelic effects, including distortion of time and perceptions of colored visual patterns and abnormal movements. Psychological effects include dysphoria, euphoria, and changes in emotion and moods. LSD also causes multiple physical effects, including dilation of the pupils, salivation, dry mouth, loss of appetite, nausea, blurred vision, perspiration, hyperglycemia, Htn, tachycardia, and hyperthermia. The mechanism of action of LSD is thought to be predominantly by serotonin neurotransmitter interactions. Hallucinogen persisting perception disorder, also known as flashbacks, and psychosis are two long-term effects

that can be exacerbated by other drugs, such as sertraline, fluoxetine, and marijuana.

Perioperative Risks

- Acute intoxication produces a sympathomimetic effect, including mydriasis, increased body temperature, systemic Htn, tachycardia, anxiety, agitation, vomiting, aspiration, apnea, and unrecognized injuries.
- May prolong succinylcholine neuromuscular blockade and delay metabolism of ester local anesthetics (speculated inhibition of plasma cholinesterase).
- May potentiate analgesics.

Worry About

- Systemic: Htn, tachycardia, hyperthermia, hyperglycemia, salivation, nausea, vomiting, seizures, and apnea
- Serotonin syndrome: Triad of altered mental status, neuromuscular abnormalities, and autonomic hyperactivity
- Psychiatric: Hallucinations (visual, auditory, and tactile), labile mood, acute panic attacks, agitation, and hypertension

Overview

- LSD is a semisynthetic odorless and colorless product of lysergic acid, a natural substance from the parasitic rye fungus *Claviceps purpurea*. It is also found naturally in several species of morning glory and Hawaiian baby woodrose plants.
- LSD is physiologically well tolerated; severe symptoms from recreational use are uncommon. Only in the setting of large ingestion (>400 mcg) has

life-threatening toxicity occurred due to cardiovascular collapse and hyperthermia.

- There is high degree of psychological dependence but no evidence of physical dependence or withdrawal symptoms when acutely discontinued.
- Classified under Schedule I of the Controlled Substance Act.
- Psychological effects begin in 30–60 min and may last 8–12 h.

Etiology

- LSD displays both agonist and antagonist properties at the serotonin (5-HT) receptors, which are similar structurally with dopamine D2 receptors and have clinically related overlap.
- The most common route of exposure is via oral with rapid GI absorption.
- LSD is not associated with a physical or psychological addiction. Long-term use can result in persistent psychosis and hallucinogen persisting perception disorder (“flashbacks”).

Usual Treatment

- Supportive reassurance; transfer pt to calm, quiet area with minimum external stimuli.
- Benzodiazepines seem to be the most effective agents for treating LSD psychosis and visual disturbances. If psychotic features persist after appropriate benzodiazepine treatment, then neuroleptics can be used as adjunct treatment.
- Rare cases require hemodynamic control, intubation, and ventilatory and supportive care.

Assessment Points

System	Effect	Assessment by Hx	PE
HEENT			Dilated, reactive pupils
CV	Sympathetic nervous system stimulation	Palpitations Sweating	Htn Tachycardia
RESP	No consistent changes	Diaphoresis	Tachypnea, apnea
ENDO	Hyperglycemia Mild hyperthermia		Elevated body temperature
CNS	Euphoria Anxiety, labile mood Tremors Visual hallucinations and illusions Synesthesia Distorted sense of time	Hx of drug ingestion	Altered mental status Hypertonia

Key References: Abraham HD, Aldridge AM, Gogia P: The psychopharmacology of hallucinogens, *Neuropsychopharmacology* 14(4):285–298, 1996; Passie T, Halpern JH, Stichtenoth DO, et al.: The pharmacology of lysergic acid diethylamide: a review, *CNS Neurosci Ther* 14(4):295–314, 2008.