

- ECG: Myocardial ischemia; possible CHF if volume overload and LV dysfunction is present
- Temperature monitoring

**Induction**

- Consider rapid sequence induction.
- Swallowing secretions: Consider anticholinergic agents.
- Cave hypovolemia; CV dysfunction makes BP and HR fluctuate.

**Maintenance**

- Inhalational anesthetics (sevoflurane and nitrous oxide) and sedatives (midazolam and propofol) were used.

- >2.0 MAC sevoflurane and hyperventilation can be associated with epileptiform activity on EEG.

**Extubation**

- CV and pulm-drive insufficiencies (common with neuropathies)
- Aspiration risk

**Adjuvants**

- Regional/neuraxial anesthesia possible despite neurodegeneration

**Postoperative Period**

- Possible tracheal reintubation, hypothermia, and seizure
- Can keep pt in ICU/PACU overnight

**Anticipated Problems/Concerns**

- Increased intraabdominal pressure and decreased FRC
- Decreases in oxygen saturation
- Hypothermia
- Gastroparesis

## Noonan Syndrome

Jiri Horak | Alexander Fort | Lee A. Fleisher

**Risk**

- Incidence between 1:1000-2500 live births
- Incidence consistent worldwide
- Equal distribution between genders

**Perioperative Risks**

- Airway
- Cardiovascular
- Hematologic
- Infectious

**Worry About**

- Difficult airway
- Cardiovascular complications
- Bleeding
- Endocarditis

**Overview**

- Key features include facial anomalies, neck webbing, short stature, chest deformity, spinal deformity (e.g., scoliosis, atlanto-occipital fusion, cervical fusion), congenital heart disease (e.g., pulmonic stenosis, hypertrophic obstructive cardiomyopathy), bleeding diathesis, mental retardation.
- Congenital heart disease may include pulmonic stenosis (in 80% of cases); hypertrophic obstructive cardiomyopathy (20–30%). Less common lesions include ventricular septal defect, tetralogy of Fallot, aortic stenosis, coarctation of the aorta, Ebstein malformation, total anomalous pulm venous return, and patent ductus arteriosus.
- Increased incidence of cancers, especially hematologic, with roughly a 3.5-fold increased risk.

**Etiology**

- Primarily an autosomal dominant disorder; however, sporadic cases are reported.
- Mutation on the PTPN11 gene on chromosome 12 in roughly 50% of cases.
- Also associated with mutations in genes that are part of the RAS/RAF/MEK/ERK signal transduction pathway (regulators of cell growth).

**Usual Treatment**

- Repair congenital cardiac defects.
- Administer growth hormone.
- Treat hematologic disorders.

**Assessment Points**

System	Effect	Assessment by Hx	PE	Test
HEENT	Facial deformity Atlanto-occipital or cervical instability or fusion	Instability Pain	Limited neck range of motion	Cervical spine x-ray
RESP	Chest wall deformity Restrictive lung disease Pulm edema	Dyspnea	Tachypnea Crackles	CXR CT scan PFTs ABG
CV	Congenital heart disease (repaired or unrepaired, most commonly HOCM or PS)	Dyspnea Orthopnea Exercise intolerance Syncope Arrhythmias	Tachycardia Murmur S <sub>3</sub> /S <sub>4</sub> Displaced PMI JVD	ECG ECHO Cardiac cath
GI	Hepatic congestion Hepatosplenomegaly Decreased appetite Gastroparesis	Abd pain N/V Failure to thrive Weight loss	RUQ tenderness/fullness Jaundice	LFTs RUQ US Albumin/prealbumin
ENDO	Growth hormone supplementation	Glucose intolerance Hypertension Dyslipidemia		BMP Lipid profile
CNS	Mental retardation Seizures	Developmental delay		Mental status exam
HEME	Bleeding disorder Hematologic malignancy	Easing bruising Epistaxis Bleeding gingiva GI bleed Fatigue	Pallor Petechiae Hematochezia Melena	CBC with differential Coagulation profile Bleeding time Fibrinogen vWF Factor levels
MS	Scoliosis Joint laxity	Restrictive lung disease	Spinal curvature	Lumbar/thoracic spine x-ray PFTs
METAB	Lyte abnormalities from malnutrition	Fatigue Weight loss ECG changes		BMP/Mg/Ph ECG

**Key Reference:** Aggarwal V, Malik V, Kapoor PM, et al.: Noonan syndrome: an anesthesiologist's perspective, *Ann Card Anaesth* 14(3):214–217, 2011.

**Perioperative Implications****Preoperative Preparation**

- Assessment of cardiac function (ECG, ECHO).
- Pulm function testing if significant chest deformity.
- Thorough airway evaluation.
- Prophylaxis for potential subacute bacterial endocarditis.
- Appropriate blood products and adjuncts should be available (e.g., desmopressin).
- Prevent preop anxiety.

**Monitoring**

- Arterial line, central line, and PA cath if indicated
- TEE if there are significant cardiovascular abnormalities

**Airway**

- Advanced airway equipment must be available, including fiberoptic bronchoscope.

**Preinduction/Induction**

- In case of hypertrophic obstructive cardiomyopathy, maintain preload/afterload and avoid tachycardia.
- Developmental delay may make IV placement or awake intubation challenging or impossible; consider preop sedatives when appropriate.

**Maintenance**

- Inhalational agents or total IV anesthesia depending on the case

**Extubation**

- Assess adequacy of ventilation/oxygenation if significant cardiac/pulmonic abnormalities.

**Adjuvants**

- Regional anesthesia when appropriate; thorough hematologic assessment prior to any intervention

**Postoperative Period**

- Cardiovascular monitoring
- Close monitoring for bleeding
- Increased risk for subacute bacterial endocarditis

**Anticipated Problems/Concerns**

- In many pts, Noonan syndrome may go undiagnosed or misdiagnosed; therefore thorough preop assessment is vital.
- Thorough cardiovascular assessment is vital, as many pts undergo surgery for congenital cardiac defects.
- Difficult airway is common; advanced airway devices must be readily available.

## Obsessive-Compulsive Disorder

Andrew J.D. Cameron | Bradley K.W. Ng

**Risk**

- Lifetime prevalence is 1–3%, with bimodal onset in childhood and late adolescence/early adulthood
- Males and females affected equally

**Perioperative Risks**

- Drug interactions, notably risk of serotonin syndrome in pts on serotonergic agents such as SSRIs and clomipramine
- Anxiety/panic attacks
- OCD-related behavior interfering with preop and postop instructions

**Worry About**

- Serotonin syndrome
- Coexistent psychiatric illness
- Anxiety/panic attacks
- Behaviors triggered by OCD complicating recovery (e.g., noncompliance with bedrest, interference with dressings.)

- Nutritional status if coexistent with an eating disorder

**Overview**

- Characterized by obsessions and/or compulsions (both usually occur together).
- Obsessions: Repetitive and persistent behaviors and unwanted thoughts or urges that pts recognize as being a product of their own mind, which cause distress and/or anxiety.
- Compulsions: Ritualistic behaviors or mental acts, which aim to either abolish or decrease the obsessions or prevent a dreaded event; they are excessive and not realistically relevant to the adverse event.
- Often associated with comorbid psychiatric illness (approximately 50%), including anxiety disorders, mood disorders, body-dysmorphic disorder, eating disorders, and alcohol dependence.

- Low rates of remission if untreated; significant numbers of pts do not receive adequate and evidence-based pharmacotherapy and psychotherapy.

**Etiology**

- Twin/family studies show some genetic predisposition.
- Exact etiology remains unclear.

**Usual Treatment**

- Psychotherapy; The best evidence is for exposure and response-prevention therapy and cognitive therapy.
- SSRIs (usually at high doses): Fluoxetine, paroxetine, sertraline, and fluvoxamine.
- Clomipramine, a tricyclic antidepressant.
- For treatment-resistant cases: Other antidepressants, antipsychotics (e.g., risperidone, aripiprazole) and anticonvulsants (e.g., lamotrigine, topiramate), which are used off license.

**Assessment Points**

System	Effect	Assessment by Hx	PE	Test
NEURO	Anxiety	Subjective anxiety	Vigilance, tachycardia	Not required

**Key References:** Grant JE: Obsessive-compulsive disorder, *N Engl J Med* 371(7):646–653, 2014; Buckley NA, Dawson AH, Isbister GK: Serotonin syndrome, *Br Med J* 348:g1626, 2014.

**Perioperative Implications****Preoperative Preparation**

- Review mental status, including any concurrent psychiatric illness.
- Review nutritional status if pt has a concurrent eating disorder.
- Review pt's psychiatric medications, including high-dose antidepressants.
- Review pt's ability to follow and comply with any preop instructions and preparations; a family member or support person may be helpful in some circumstances.
- Ask about stressors and discuss how to minimize them (e.g., pt obsessed with cleanliness could be shown equipment in sterile packs).
- Ask about compulsive behaviors and plan accordingly (e.g., pts with a background of excoriation [skin picking] may interfere with their wounds; wounds should be dressed in a way that makes them inaccessible to pt).

- Consider liaison with a psychiatrist or a psychiatric review in pts with severe OCD or if a number of challenges are identified preop.

**Intraoperative Period**

- Pts on medication for OCD are at risk of serotonin syndrome if given a second serotonergic medication. These include opioids (fentanyl, hydrocodone, meperidine, morphine, oxycodone, and tramadol), cocaine, methylene blue, and ondansetron.
- Consider strategies to avoid opioids (regional anesthesia and nonopioid analgesics).

**Postoperative Period**

- If pt is on serotonergic medication, monitor for serotonin syndrome; watch for altered mental state (e.g., agitation, confusion), neuromuscular excitation (e.g., clonus, hyperreflexia, myoclonus, rigidity), and autonomic excitation (e.g., hyperthermia, tachycardia).

- In addition to the above-listed drugs, avoid dextromethorphan and ergotamine.
- Consider closer nursing observation and/or a private room.
- Consider short-term anxiolytics or sedation if the pt's rituals or anxiety pose a major barrier to recovery.
- Consider early involvement of a psychiatrist or psychiatric team if available in the hospital.

**Anticipated Problems/Concerns**

- Preop anxiety: Consider premedication with midazolam 7.5 mg PO 1 h before the procedure.
- Postop anxiety/panic attacks or problematic rituals: Administer prn benzodiazepines and arrange a psychiatric consultation.
- Serotonin syndrome if the pt is on medication; the risk is generally higher than for pts being treated for anxiety/depression due to the high doses required to treat OCD.