

Perioperative Implications**Preoperative Preparation**

- Stabilize vascular bed by hydration (NS/LR) prior to induction of anesthesia.
- Obtain ABG to correct, if able, pH and electrolyte (Na^+ , K^+) abnormalities.
- Anxiolysis (pharmacologic or presence of parent in OR for children) to prevent dysautonomic crisis.
- Vent gastrostomy.

Monitoring

- Consider arterial BP for intraop and postop management. FD pts have lower resting $\text{PaO}_2/\text{SpO}_2$ and higher PaCO_2 . Most pts have compromised pulm function and Hx of sleep apnea. Their BP is variable and extreme hypotension tends to occur with induction of anesthesia, especially if pt was not prehydrated.
- Consider central access with CVP monitoring in surgical procedures with significant hemodynamic shifts, as FD pts are more susceptible to extreme BP swings associated with intravascular volume.
- Consider TEE/CO monitoring for assessment of cardiac function and as guidance for rational fluid management.
- Consider placing defibrillation/external pacing pads.
- Maintain normothermia as temperature regulation is affected at baseline.

Airway

- Sialorrhea: consider glycopyrrolate/atropine prior to induction (carefully titrate, as pts are sensitive to cholinergic and adrenergic agents).

Induction

- Ensure eye lubrication and protection, as corneal epithelium is extremely prone to injury.
- Propofol has been used with preop hydration. Consider ketamine \pm propofol when unable to prehydrate. Ketamine may exacerbate oral secretions.
- Both depolarizing and nondepolarizing neuromuscular blockers have been used successfully.
- Consider rapid sequence induction with cricoid pressure even in pts with Nissen fundoplication (studies

have shown that 15% of pts had malfunctioning fundoplication after 5 y).

Maintenance

- Maintain normocapnia to decrease BP lability.
- Inhalational agents can be used, but consider adding opioid/dexmedetomidine infusion or TIVA to smooth out BP variation and minimize PONV.
- Consider EEG-based depth-of-anesthesia monitoring to help minimize exposure to anesthetics.

Extubation

- Titrate analgesics carefully. Consider alternatives to opioids (NSAIDs, IV acetaminophen, dexmedetomidine infusion).
- If able, use nonanticholinergic reversal of NMB, as pt are hypersensitive to cholinergic/adrenergic agents.
- Return of spontaneous breathing may be delayed due to chemoreceptor dysfunction (PaCO_2 is not a trigger for the brain stem to initiate spontaneous breathing).
- Due to a blunted ventilatory response to hypoxia, pts may experience hypercapnic-induced Htn. Apnea is associated with severe desaturation and hypotension.
- Gentle suction (as to not precipitate dysautonomic crisis) is important as pts are at increased risk for aspiration pneumonia.
- Prior to extubation, it is important to ensure that airway reflexes have returned, but conservative extubation criteria could create anxiety and precipitate dysautonomic crisis. Consider coating ETT with lidocaine jelly prior to intubation.

Adjuvants

- Consider regional anesthesia either alone or with GA for intraop and postop pain control.
- Successful use of epidural anesthesia has been reported (and is the preferred analgesic for labor and cesarean delivery). Spinal anesthesia should probably be avoided as it would likely precipitate severe and refractory hypotension due to sympathectomy.
- MAC sedation has also been safely administered in ambulatory settings with midazolam and propofol.

Postoperative Period

- Care must be taken in providing supplemental oxygen, as it may accentuate eye dryness and increase the risk of epithelial breakdown.
- Although somatic pain sensation is diminished, visceral pain sensation is intact. Pain must be well controlled so as to avoid precipitating dysautonomic crisis.
- Respiratory status should be carefully monitored in PACU/ICU, especially if respiratory depressants are administered (e.g., opioids for pain control, BZDs for dysautonomic crises).
- NIPPV may ameliorate respiratory depression and prevent respiratory failure and reintubation. However, properly fitted masks must be used to avoid eye injury.
- Pulm hygiene should be instituted, as most pt have chronic lung disease secondary to repeated aspirations.
- Manage dysautonomic crises with diazepam (first-line drug) or clonidine (second-line drug); may also use hydralazine and labetalol if Htn is refractory to initial treatments.
- Elevate head of bed to 30 degrees and encourage early sitting to avoid supine Htn.
- Treat hypotension with fluids/blood products and fludrocortisone.

Anticipated Problems/Concerns

- Dysfunction of chemoreceptors leads to altered response to hypoxia and hypercapnia. Low PaO_2 does not stimulate tachypnea and can cause syncope, as hypoxia induces both hypotension and bradycardia.
- Presence of spinal deformities may make neuraxial anesthesia more technically challenging.
- PONV precautions should be employed.
- Increased sensitivity to exogenous adrenergic and cholinergic agents; minimal doses may produce exaggerated responses.
- Avoid drugs that prolong QT interval.
- Pts may be on chronic BZDs and may have developed tolerance and dependence.

Rocky Mountain Spotted Fever

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Risk

- Incidence in USA: In most states, most commonly in the southeastern and south central states, there are ~250-2200 cases per y.
- Exposure to tick-infested terrain or dogs.
- Severe infection; very young (<4 y), males and those with G6PD deficiency are at risk for death.
- Mortality is 23% when untreated, 0.3-4.0% even with early treatment (within first 5 d).
- Mortality increases with delay in Dx, older age (>60 y), male sex, very young age (<4 y), in blacks, chronic alcohol abuse, and those with G6PD deficiency.

Perioperative Risks

- Increased mortality secondary to CV instability and noncardiogenic pulm edema
- Increased risk of organ injury due to compounded insults
- Increased bleeding tendency

Worry About

- Severe intravascular volume depletion leading to shock
- Lyte disturbances

- Cardiac arrhythmias
- Microvascular hemorrhage
- Consumptive coagulopathy
- Intraop respiratory and renal failure

Overview

- Uncommon but severe; pathophysiology primarily due to endothelial cell prostaglandins, resulting in increased vascular permeability, edema, hypovolemia, and ischemia.
- Initial symptoms appear in 1-3 d: Nonspecific, mimicking a viral syndrome with fever, headache, malaise, myalgias, arthralgias, and nausea; specific symptoms appear in 2-14 d, most in 5-7 d, mostly in the spring and summer months; pts generally have a known or possible tick bite.
- Rash appears in most pts in 3-5 d, after onset of fever, initially maculopapular and progressing to petechiae; usually starts on the ankles and wrists, then palms and soles; finally spreads to the body and face; rash absent in 10-12%
- Disease progression (more likely with delay in treatment) results in multiorgan involvement: Noncardiac pulm edema, encephalitis, myocarditis, hepatitis, bleeding (secondary to

thrombocytopenia and direct vessel damage), and acute renal failure.

Etiology

- *Rickettsia rickettsii* is transmitted via the saliva of ticks after 6-10 h of attachment and feeding or by exposure to infected tick hemolymph during the removal of ticks.
- Incubation period ~7 d (2-14 d).
- Obligatory intracellular bacterium that replicates in vascular endothelial cells, causing direct cell injury with loss of vascular integrity.

Usual Treatment

- Dx is difficult, made primarily from clinical and epidemiologic (potential tick exposure) evidence and serologic testing or biopsy of skin lesion to confirm.
- Doxycycline, chloramphenicol (for pregnant women in the first two trimesters of pregnancy or severe adverse reaction to doxycycline); therapy within first 5 d is important (mortality 6.5 vs. 22.9).
- Correct hypovolemia, coagulation defects, thrombocytopenia; provide intensive, supportive care for various organ system failures.

Assessment Points				
System	Effect	Assessment by Hx	PE	Test
CV	Extensive microvascular leak, interstitial myocarditis	Rash, swelling	Rash, edema, arrhythmias	ECG, CXR, lytes, BP
RESP	Noncardiac pulm edema, interstitial pneumonitis	Reduced exercise tolerance, dyspnea, cough	Rales by auscultation	CXR, spirometry
GI	Gastroenteritis; liver, spleen, and pancreatic microvascular hemorrhage and edema	N/V, abdominal pain, diarrhea	Abdominal tenderness Hepatosplenomegaly	SGOT, bilirubin
HEME	Thrombocytopenia, anemia	Easy bleeding, malaise	Rash	Hct/Hgb, plts/PT, PTT, BUN
RENAL	Microvascular hemorrhage and edema, interstitial nephritis, prerenal azotemia	Lumbar pain		Cr, lytes
CNS	Meningoencephalitis	Focal defects, deafness, confusion, meningismus, photophobia, seizures		CSF: Checking WBCs and protein
MS	Microvascular hemorrhage, edema	Myalgia, arthralgia	Reduced ROM	

Key References: Walker DH, Blanton LS: *Rickettsia rickettsia* and other spotted fever group rickettsiae (Rocky Mountain spotted fever and other spotted fevers). In Bennett JE, Dolin R, Blaser MJ, editors: *Mandell, Douglas, and Bennett's principles and practice of infectious diseases*, ed 8, Philadelphia, PA, 2014, Elsevier, pp 188, 2198–2205.e4; Walker DH: Rocky Mountain spotted fever: a disease in need of microbiological concern, *Clin Microbiol Rev* 2(3):227–240, 1989.

Perioperative Implications

Preoperative Preparation

- Antibiotic therapy and correction of underlying organ system dysfunction
- Surgery only for emergency
- Assess volume, respiratory, and renal status

Monitoring

- Consider PA cath, arterial line, UO.
- Intraop ABGs and lytes.
- Platelets and other coagulation variables.

Airway

- Severe edema of oropharynx and increasing bleeding tendency can lead to difficult intubation.

Induction

- Hypovolemia can cause hypotension.

- Microvascular leak in lung can cause rapid desaturation.
- Increased cardiac arrhythmias.

Maintenance

- Owing to CV instability, volume status is key.
- Possibility of respiratory failure and constant volume resuscitation should be anticipated in selecting an anesthetic technique.

Extubation

- Oropharyngeal edema and increased bleeding tendency may make reintubation very difficult.

Adjuvants

- Vasoactive drugs used in acute resuscitation should be readily available.
- Lidocaine for treatment of cardiac arrhythmias.

Postoperative Period

- Intravascular volume shifts; coagulation defects, respiratory failure, CV instability, renal failure

Anticipated Problems/Concerns

- Owing to the possibility of multisystem failure, prolonged postop management in the ICU may be required.
- Because early treatment with antibiotics is curative and highly successful in preventing complications, a high index of suspicion (e.g., after tick exposure in endemic areas) is needed.

Rubella and Congenital Rubella Syndrome

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Risk

- A rubella epidemic in USA in 1964–1965 resulted in 12.5 million cases of rubella infection and about 20,000 newborns with CRS.
- The number of reported cases of rubella in USA remains low, with a median of 11 cases annually in 2005–2011 because of vaccination.
- The overall burden of CRS is still high in developing countries. There were 66 cases reported in Bangladesh, 26 in Romania, 16 in Nepal, 10 in Zambia, 9 in Japan, and 4 in Sri Lanka in 2014.
- Incidence of cardiac defects in CRS with eye involvement could be as high as 95%. Most common cardiac anomaly in CRS is PDA.

Perioperative Risks

- CRS is a constellation of multisystem abnormalities
- Such pts may require cardiac surgery like congenital cardiac septal defect correction and/or other non-cardiac surgery like cleft lip/cleft palate repair and a variety of eye procedures under anesthesia. Cataract extraction is an urgent vision saving procedure, so complete optimization (correction of cardiac defects, adequate weight gain) of neonate may not be possible.

Worry About

- Unexpected difficult intubation in various upper airway anomalies like subglottic stenosis, shortened trachea, and short glottis carinal length associated with many congenital syndromes.

- Hypothermia.
- Hypoglycemia: Exogenous sodium, water and glucose should be provided periop, as they have low GFR and are more prone for hypoglycemia.
- Balance of PVR and SVR.
- Drug metabolism may be deranged because of associated liver abnormalities and hypothyroidism.
- IE prophylaxis is essential, as the turbulent flow produced by the high velocity systolic jet in pulm artery stenosis increases the potential for development of endocarditis.

Overview

- Rubella is a viral illness characterized by a mild, maculopapular rash. The rubella rash occurs in 50–80% of rubella-infected persons and is sometimes misdiagnosed as measles or scarlet fever.
- Rubella is contagious disease which spreads in droplets. The respiratory secretion, cataractous lens is one of the most infectious materials hence warrants universal precaution.
- CRS is an illness resulting from rubella virus infection during pregnancy. When rubella infection occurs during early pregnancy, serious consequences—such as miscarriages, stillbirths, and a constellation of severe birth defects in infants—can result. The risk of congenital infection and defects is highest during the first 12 wk of gestation and decreases after the 12th week of gestation, with defects rare after the 20th wk of gestation.

- Common congenital defects of CRS include cataracts; congenital heart disease, including PDA, coarctation of aorta, VSD, ASD, and pulm artery stenosis; hearing impairment and developmental delay; brain damage (microcephaly, mental retardation, meningoencephalitis); hepatosplenomegaly; thrombocytopenia; and neonatal jaundice. Other manifestations are type I diabetes mellitus, growth retardation, transient hemolytic anemia, metaphyseal “celery stalk” changes in long bones, transient pneumonitis, transient generalized lymphadenopathy, cryptorchidism, inguinal hernia, and dermal erythropoiesis (“blueberry muffin syndrome”). Infants with CRS usually present with more than one sign or symptom consistent with congenital rubella infection. However, infants may present with a single defect. Hearing impairment is the most common single defect.

Etiology

- Rubella is a viral illness caused by a togavirus of the genus *Rubivirus*, which is most closely related to group A arboviruses, such as eastern and western equine encephalitis viruses.
- It is an enveloped RNA virus with a single antigenic type that does not cross-react with other members of the togavirus group.
- Rubella virus is relatively unstable and is inactivated by lipid solvents, trypsin, formalin, ultraviolet light, low pH, heat, and amantadine.