Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Edematous mucosa Arthritis of larynx	Epistaxis Hx of voice change	Friable mucosa Voice, airway exam	Direct laryngoscopy
CV	LV dysfunction Aortitis Pericarditis	Dyspnea Orthopnea Reduced exercise Reduced exercise Dyspnea	S_3 Rales Diastolic murmur (A1) Distant heart sounds Friction rub	ECG Stress ECG ECHO ECHO ECHO
RESP	Fibrosis	Dyspnea	Dry rales	CXR, PFTs
GI	Peptic ulcer	Epigastric pain, N/V		
RENAL	Renal dysfunction	Drug induced		Cr
CNS	Spinal cord compression Neurologic dysfunction	Neck pain Numbness	Sensory deficits Motor deficits ROM of neck	Radiography
MS	Arthritis	Joint pain	Swelling Pain with motion Restricted motion	Radiography

Key References: Lisowska B, Rutkowska-Sak L, Maldyk P, et al.: Anaesthesiological problems in patients with rheumatoid arthritis undergoing orthopaedic surgeries, *Clin Rheumatol* 27(5):553–556, 2008; Samanta R, Shoukrey K, Griffiths R: Rheumatoid arthritis and anaesthesia, *Anaesthesia* 66(12):1146–1159, 2011.

Perioperative Implications

Preoperative Evaluation

- Thorough airway evaluation is a priority. If atlantoaxial instability exists, flexion of the neck can compress the spinal cord. Radiating pain to the occiput may be an indication of cervical cord involvement. Imaging—such as x-ray, CT, or MRI—may be indicated if the amount of cervical involvement is not known.
- Cardiopulmonary status needs to be evaluated. If severe restrictive lung disease is suspected, preop pulm function tests may be indicated. Anticipation of postop ventilatory support should be considered.
- Must have adequate knowledge of the pt's current medications. Stress-dose corticosteroid supplementation may be indicated for pts being treated chronically with these drugs. Anti-inflammatory medications, aspirin, and other rheumatoid drugs can interfere with platelet function, clotting, and formation of RBCs.
- Joint mobility and restriction should be assessed to determine appropriate intraop positioning.

Monitoring

- Standard monitors
- Further invasive monitoring depending on pt's disease state and the anticipated procedure

Airway

- Presence of atlantoaxial instability involvement assessed. Cervical collar placement to minimize movement during direct laryngoscopy considered. Awake fiberoptic laryngoscopy may be best method.
- TMJ disease can limit mouth opening and ability to perform direct laryngoscopy adequately.
- Cricoarytenoid involvement can decrease size of the glottic inlet and necessitate use of a smaller ETT.

Preinduction/Induction

- Preinduction and induction agents/techniques dependent on pt's specific associated comorbidities.

Maintenance

- CV effects from induction agents and volatile anesthetics potentially more pronounced; risks of hemodynamic instability, cardiac conduction abnormalities, and myocardial ischemia increased.
- Pulm disease may be associated with restrictive changes leading to decreased lung volumes and vital capacity, V/Q mismatch, and poor arterial saturation.
- Hematologic abn such as anemia can be evident intraop.
- Appropriate extremity positioning; padding and manipulation assessed throughout procedure

Extubation

- Post-extubation laryngeal obstruction secondary to edema and erythema possible from cricoarytenoid involvement.
- With severe restrictive lung disease, postop ventilatory support is anticipated.

Adjuvants

- Regional and neuraxial techniques can be utilized assuming no significant thoracic, lumbar, and sacral spine involvement as well as normal coagulation studies

Anticipated Problems/Concerns

- Tracheal intubation difficulties secondary to cervical spine and TMJ involvement
- Intraop CV instability and restrictive pulm disease issues
- Associated side effects of current drug therapy (e.g., anticoagulation, anemia, poor wound healing).
- Multiorgan system involvement
- Intraop positioning concerns secondary to advanced joint involvement and decreased ROM
- Potential need for postop ventilatory support

Riley-Day Syndrome (Familial Dysautonomia, Hereditary and Sensory Autonomic Neuropathy Type III)

Elvedin Luković | H. Thomas Lee

Risk

- Incidence: 1:3700 live births among American and Israeli Jews.
- Since original description in 1949, more than 600 pts have been identified and registered with the Dysautonomia Center at New York University.

Perioperative Risks

- Prior to 1960 there was a 50% probability of death before age 5 y; currently a newborn with FD has 50% probability of reaching age 40 y, although many require multiple surgical interventions.
- Mortality is primarily due to pulmonary complications (26%, decreasing with aggressive treatment of aspirations). Some deaths are unexplained (38%, possibly due to unopposed vagal stimulation or sleep abnormality). Others are due to sepsis (11%), bradycardia/CHB, hyponatremia, or renal failure.

Worry About

- Labile blood pressures exacerbated by physical and/or emotional stress
- Dysrhythmias, especially bradycardia, which can lead to asystole/CHB
- Compromised respiratory function at baseline due to chronic aspirations and severe thoracic kyphosis/scoliosis
- Hyponatremic seizures secondary to hypertensive vomiting, which is associated with excessive secretion of vasopressin and water retention
- Advancing renal failure due to progressive denervation of renal arteries, leading to poor regulation of RBF during paroxysmal hypertensive and hypotensive episodes

Overview

- FD is characterized by poor development and poor survival of autonomic and sensory neurons; motor neurons are typically spared; intelligence is usually normal.
- Signs and symptoms of FD are usually apparent at birth and tend to progress with age.
- Diagnosis is based on documentation of mutation(s) in the *IKBKAP* gene. There is high suspicion for disease if five cardinal criteria are present: Absence of overflow emotional tears (after age 7 mo), absent

lingual fungiform papillae, depressed patellar reflexes, lack of an axon flare following intradermal histamine, documentation of Ashkenazi Jewish extraction.

- Affected individuals are hypersensitive to sympathomimetic and parasympathomimetic drugs (due to upregulation of adrenergic and cholinergic receptors) but have decreased endogenous catecholamine levels at baseline.
- During physical and emotional stress, plasma epinephrine, NE, and DA are relatively elevated and precipitate dysautonomic crises: intractable emesis, diaphoresis, tachycardia, Htn, and personality changes.
- Additionally there is a decreased response to temperature and somatic pain (palms, soles of feet, neck, and genital areas are spared). Visceral pain perception is normal or heightened.
- Pts also experience orthostatic and paroxysmal Htn (due to failure of the afferent baroreflex), arrhythmias/CHB, prolonged QT interval, erythematous skin rash, central sleep apnea/disordered sleeping, altered response to hypoxia and hypercarbia (due to malfunctioning chemoreceptors), dry-eye optic neuropathy with retinal injury, decreased blink rate, ocular anesthesia and corneal damage, oropharyngeal incoordination, GI dysfunction and bleeding, progressive glomerulosclerosis, hypotonia (in infants) and gait ataxia (in adults), and spinal deformities.
- Fixed Htn can develop chronically, secondary to advancing renal failure.
- Hyponatremic seizures may occur with hypertensive vomiting, excessive sweating, and poor fluid/salt intake. 10% of pts have a seizure disorder.

Etiology

- Autosomal recessive disease with complete penetrance, but variable expression.
- Relatively low carrier frequency (1:3000) in non-Jewish individuals; significantly higher carrier frequency (~1:30) among people of Ashkenazi/Eastern European Jewish ancestry.
- Single noncoding mutation (base pair 6 change from T to C on chromosome 9q31) in the *IKBKAP* gene leads to expression of truncated IKAP (IκB kinase–associated protein), responsible for >99% of all cases of FD.
- IKAP is believed to regulate gene transcription and expression during neuronal development and myelination in embryogenesis.

Usual Treatment

- Treatment is supportive and preventative.
- Careful hydration (pts tend to become dehydrated easily because of excessive sweating and drooling, fever associated with aspiration pneumonia, and vomiting).
- Gastrostomy and fundoplication allow improved nutrition and reduction of pneumonia.
- Pulmonary hygiene by bronchodilation, postural drainage, suction of tracheal secretions.
- Noninvasive positive-pressure ventilation during sleep because of tendency to hypoventilate. Make sure that mask fits properly to avoid corneal damage.
- Dysautonomic crisis is treated with centrally acting agents such as BZDs and clonidine; these must be used cautiously as they can precipitate hypotension and respiratory depression.
- Hypertensive vomiting can also be treated with carbidopa.
- Hyponatremic seizures are treated with slow correction of serum sodium.
- Blood pressure: Htn is treated with fluids, fludrocortisone (may exacerbate Htn and renal disease) and midodrine; Htn is treated with BZDs, α_2 agonists, CCBs, and positional changes, such as sleeping with head of the bed raised 20–40 degrees.
- Asymptomatic Htn is not treated because it is usually transient.
- Treatment with artificial tears, tarsorrhaphy, corneal surgery.
- Anemia of chronic disease is treated with erythropoietin.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CNS	Decreased somatic pain and temperature perception	Hx of injuries or self-injurious behavior	Assess extremities/back for injury/skin breakdown Assess for self-injurious behavior	
	Hypotonia (infants) Broad-based, ataxic gait (adults)	Delayed motor developmental milestones	Positive Romberg test, decreased vibrational sense and joint position	
	Catecholamine surge	Personality changes during dysautonomic crises, psychiatric disorders may also be present		
	Seizure disorder (10% of pts)			EEG
	Central sleep apnea	NIPPV	Baseline oxygen saturation	Polysomnography
HEENT	Altered sweet sensation		Absence of fungiform papillae on tongue	
	Reduced tear volume, corneal anesthesia, reduced blink rate, incomplete lid closure during sleep	Eye dryness, use of artificial tears, absence of overflow tears with emotional crying	Ocular injury/ulcers, infection, optic neuropathy, retinal detachment	
CV	BP lability (supine Htn, orthostatic hypotension, dysautonomic crises) due to dysfunction of baroreceptors and hypersensitivity to catecholamines	BP variation throughout the day, frequency of dysautonomic crises Nocturia may indicate Htn episodes during sleep	BP supine/standing	Ambulatory BP monitoring
	Dysrhythmias (bradycardia/CHB) Prolonged QT Conduction abnormalities	Syncope, DOE, poor exercise tolerance		ECG Holter monitoring, PPM placement/interrogation, BMP
	LVH/CHF	DOE, poor exercise tolerance	JVD, edema, rales, cardiomegaly, murmurs	CXR, TTE/TEE
GI	Incoordinated swallowing Excessive salivation GI dysmotility GI bleeding Emesis	Feeding and drinking difficulty, daily vomiting, recurrent aspiration, severe dysphagia and GERD	FTT, drooling Assess intravascular fluid status (decreased skin turgor/dry mucous membranes)	BMP Proper hydration
RESP	Chronic aspiration leading to recurrent infections, atelectasis, bronchiectasis Restrictive lung disease due to kyphosis/scoliosis	Difficulty breathing, DOE Last pneumonia episode SOB, increased work of breathing	Auscultation Assess for signs and symptoms of current infection (febrile response may be severely altered)	CXR PFTs
	Dysfunction of chemoreceptors	Hypoxia can induce hypotension and bradycardia leading to syncope	Baseline SpO_2/PaO_2/$PaCO_2$	ABG
RENAL	Glomerulosclerosis Anemia of chronic disease	UO, AKI on CKD, ESRD		BMP CBC
ENDO	Hyponatremia	History of emesis, poor fluid intake, excessive diaphoresis in hot weather, fevers		BMP
MS	Decreased DTRs			
	Spinal deformities (kyphosis/scoliosis)			

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 PaO_2/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 ± 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

Sinisa Markovic | Paul R. Knight III

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.