Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	First-degree heart block, pericarditis, myocarditis and valvulitis. Criteria: Major: Migratory polyarthritis, carditis, Sydenham chorea, erythema marginatum, and subcutaneous nodules Minor: Arthralgias, fever, elevated serum acute-phase reactants, and first degree heart block	Dyspnea Palpitations Chest pain	Cardiac murmur Chest pain CHF, edema, orthopnea, DOE	ECHO Cardiac cath ECG (first-degree block)
RESP	Pulm edema	DOE Orthopnea Paroxysmal nocturnal dyspnea Hemoptysis	Tachypnea Rales Wheezing	CXR
GI	Hepatomegaly from right heart failure		Enlarged liver	LFTs
RENAL	Fluid retention		Edema	Serum lytes
CNS	Embolic stroke	Sudden unilateral neurologic deficits TIA	Focal, unilateral neurologic deficits	CT or MRI of the head TEE Carotid US

Key References: Seckeler MD, Hoke TR: The worldwide epidemiology of acute rheumatic fever and rheumatic heart disease, *Clin Epidemiol* 3:67–84, 2011; Moore RA, Martin DE: Anesthetic management for the treatment of valvular heart disease. In Hensley FA, Martin DE, Gravlee GP, editors: *A practical approach to cardiac anesthesia,* ed 3, Philadelphia, 2003, Lippincott Williams & Wilkins.

Perioperative Implications

Preoperative Preparation

- A determination of the specific cardiac lesions need to be made so that the ideal hemodynamic profile can be decided upon. Choice of anesthetic approaches and drugs will be largely determined by the desired hemodynamic profile.
- Note that with aortic stenosis, there is a need to keep a lower heart rate and a higher SVR. These are critical hemodynamic considerations and require special attention.
- Assess fluid status. If pt is dehydrated, liberal IV fluids should be provided preop, since with all lesions preload should be maintained. If fluid overload exists, concern for the development of CHF may direct treatment by fluid restriction and diuresis.
- If a decrease in PVR is desirable hemodynamically, avoid premedication causing hypoventilation.
- It is rare that a single valvular heart defect exists in RHD. The most common combined valvular defect is mitral stenosis with mitral regurgitation.

Monitoring

- Depending on the severity of the cardiac disease and the extent of surgery, have a low threshold for invasive monitoring. Consider arterial cath and CVP cath.
- Use caution when placing a pulm artery cath. Excessive force when placing the cath may rupture the pulm artery due to the combination of long-standing pulm Htn and thin pulm arterial walls.
- TEE is helpful for assessment of worsening valvular regurgitation and left ventricular dysfunction.

Airway

- In deciding between deep sedation and general anesthesia with a controlled airway, worsening pulm Htn with hypercapnia should be considered.
- If a difficult airway is anticipated, the initial intubation attempt should be aided by video or fiberoptic laryngoscopy.

Induction

- With mitral and aortic stenosis, tachycardia must be controlled upon induction and emergence from anesthesia.
- Induction agents should be chosen to minimize cardiovascular changes that would adversely affect the optimal hemodynamic profile.
- A potent alpha-adrenergic agonist such as phenylephrine should be on hand.

Maintenance

- Adequate fluids should be given to maintain adequate preload, but care should be taken to avoid fluid overload and the resultant CHF.
- Positive-pressure hyperventilation is an adjunct for decreasing PVR.
- Avoid long-acting narcotics that might depress ventilation postop.
- Use high FIO_2.
- Avoid hypothermia.

Extubation

- Do not attempt deep or early extubation.
- Prior to extubation, it is important to assess the adequacy of ventilation by measuring inspiratory pressure and ensuring adequate tidal volumes.

Postoperative Period

- Close monitoring of ventilation and pulse oximetry.
- Active warming.
- Be prepared for immediate reintubation.

Adjuvants

- Pulm vasodilators may be indicated for pts with severe pulm Htn. Nitric oxide, prostacyclin, and milrinone are all possible adjuvants, but use of these medications should be balanced with their effects on the hemodynamic profile, such as falls in SVR, before instituting treatment.

Rheumatoid Arthritis

Nathan Kudrick | Pedro Orozco | Lee A. Fleisher

Risk

- Internationally the prevalence of RA is believed to range from 0.4% to 1.3%.
- In 2005, an estimated 1.5 million (0.6%) of USA adults >18 y had RA.
- Male-female ratio: 1:2.

Perioperative Risks

- Risk of neurologic injury is increased due to possible occult damage to the cervical spine.
- Associated cardiac disease may be present but not clinically apparent.
- Pulm complications arise secondary to possible pulm fibrosis and restrictive lung disease.

Worry About

- Visualization of glottis and tracheal intubation may be difficult due to rheumatoid-associated damage to the cervical spine.
- Former successful ET intubation does not reliably eliminate existing airway abnormalities.
- Occult coronary vascular disease, pericardial effusion, pericardial thickening, rheumatoid nodules in the cardiac conduction pathway, valvular fibrosis.
- Iatrogenic injury to the cervical spinal cord during laryngoscopy and tracheal intubation.
- Chronic corticosteroid use may necessitate intraop steroid administration.
- Mental health conditions.

Overview

- Chronic systemic inflammatory disease involving diffuse joints and organ systems.
- The natural history of RA varies considerably with at least three possible disease courses:
  - Monocyclic: Have only one episode that ends within 2–5 y of initial diagnosis. This may result from early diagnosis or aggressive treatment.
  - Polycyclic: The level of disease activity fluctuates over the course of the condition.
  - Progressive: RA continues to increase in severity and does not go away.
- Systemic effects include pericardial effusion, tamponade, pleural effusion, pulm fibrosis, anemia, keratoconjunctivitis, and renal failure.

Etiology

- Autoimmune disorder triggered by an antigen in genetically predisposed persons.
- Clinical variability may stem from differences in triggering antigens and immune response.
- Pathology: Synovial cellular hyperplasia, synovial infiltration by lymphocytes, plasma cells, and fibroblasts leading to degeneration of cartilage and articular surfaces.

Treatment

- Aspirin and NSAIDs: Ibuprofen, indomethacin, naproxen, piroxicam, sulindac, and tolmetin
- Nonbiologic DMARDs: Methotrexate, hydroxychloroquine, sulfasalazine, leflunomide, azathioprine, cyclosporine, penicillamine, and gold
- Biologic agents, sometimes called biologic DMARDs: Etanercept, adalimumab, infliximab, certolizumab pegol, and golimumab

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