Worry About

- Unrecognized inborn errors of metabolism that produce Reye-like syndromes, such as fatty-acid oxidation defects, carnitine deficiency, and amino and organic acidopathies
- Recurrent liver dysfunction
- Permanent neurologic sequelae

Overview

- An acute, noninflammatory encephalopathy with hepatic dysfunction predominantly in children; typically starts several days after a viral illness, usually influenza or varicella.
- Encephalopathy heralded by protracted severe vomiting, with abnormal behavior and combativeness that may progress to coma.
- Dx is made by unexplained encephalopathy with one or more of following: Serum transaminases elevated to at least 3 times normal, blood ammonia levels at least 3 times normal, or hepatic microvesicular fatty infiltration on liver biopsy. There should be no other reasonable explanation for the cerebral or hepatic abnormalities.
- The CDC use a classification for progressive disease severity of stages 1–6. Mortality has decreased from 50% to 20% as a result of recognition of early phases and aggressive treatment.
- Prognosis depends on severity and duration of cerebral dysfunction. Severe disease can lead to subtle neuropsychological defects.

Etiology

- Abnormal reaction to viral illness modified by exogenous toxin in a susceptible host.
- Most frequently linked to influenza A and B and varicella. The exogenous toxin is aspirin in most cases. Salicylates were detectable in >80% of cases.

Usual Treatment

- Early recognition of mild cases and maintenance of fluid, lyte, acid-base, urine output and glucose balance.
- ET intubation may be required to ensure airway protection and control of ventilation to reduce ICP.
- Fluids restricted in pts with cerebral edema; ICP monitoring helps to improve cerebral perfusion pressure and manage ICP.
- Mannitol to induce cerebral dehydration and barbiturates to decrease cerebral metabolic demand.
- Coagulopathies treated with vitamin K and/or FFP.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
GI	Hepatic dysfunction	Severe vomiting	Hepatomegaly	Hepatic transaminases, ammonia levels Liver biopsy PT, PTT
CNS	Delirium Combative behavior Seizures Lethargy Coma	Alteration in mental status	No focal signs	CT scan ICP monitor

Key References: Goetz CG: Aminoacidopathies and organic acidopathies, mitochondrial enzyme defects, and other metabolic errors. In Goetz C, editor: *Textbook of clinical neurology*, ed 3, Philadelphia, PA, 2007, Saunders; Tasker RC: Update on pediatric neurocritical care, *Paediatr Anaesth* 24(7):717–723, 2014.

Perioperative Implications

- Surgery should not be undertaken except in life-or-death emergencies.

Adjuvants

- Ondansetron to decrease vomiting.
- Treat seizures with phenytoin.
- Correct hyperammonemia with sodium phenyl-acetate/sodium benzoate.

Anticipated Problems/Concerns

- Hepatic dysfunction
- Neurologic sequelae
- Underlying inborn errors of metabolism, particularly in children under 5 y

Rheumatic Fever (Acute) and Rheumatic Heart Disease

Mark S. Weiss | Victoria M. Bedell | Roger A. Moore

Risk

- Common illness among children and young adults.
- Primary chronic sequelae is RHD.
- Worldwide estimate is over 15 million cases of RHD, with 282,000 new cases and 233,000 deaths annually.
- Up to 1% of all school-age children in Africa, Asia, Latin America, and the eastern Mediterranean show signs of the disease.

Perioperative Risks

- In ARF with acute cardiac manifestations (including first-degree heart block and pericarditis), medications and equipment for maintaining heart rhythm and function during anesthesia should be available.
- An actively febrile pt's surgery should be delayed unless it is urgent or emergent.
- The approach to anesthesia in RHD must be tailored to the pt's specific physiologic parameters. Control of the pt's hemodynamic profile to optimize cardiovascular stability will depend on which valves are damaged and the extent of the myocardial compromise, either from RHD or from secondary cardiovascular effects due to chronic valvular disease.
- In the presence of valvular disease, prophylactic antibiotics should be given to prevent bacterial endocarditis.
- Periop assessment of cardiac status, including direct and indirect effects of chronic valvular disease on cardiac function, must be performed prior to surgery. Pts may be clinically asymptomatic for 20 y after developing RHD owing to compensatory alterations in cardiac structure and function. Knowledge of the cardiac compensatory changes in heart function is essential.

Worry About

- If valvular damage is present, maintain tight control of cardiac rate and rhythm, pulmonary and systemic vascular resistance, and intravascular fluid volume.
- If pulm hypertensive crisis occurs, hyperventilate and increase inspired O_2 to 100%.

Overview

- ARF is primarily due to a pharyngeal infection with *Streptococcus pyogenes* or group A beta hemolytic streptococcus, which is a common cause of throat infections in children. If left untreated, the child can develop ARF, which is an inflammatory response occurring 2–3 wk after the initial infection.
- RHD is an autoimmune reaction with cardiac tissue, resulting in permanent deformities of heart valves or chordae tendineae.
- Scarring leads to valvular stenosis, classically in the mitral valve followed by the aortic valve. However, all cardiac valvular defects can occur.
- Joint pain and carditis with valve damage are the major clinical manifestations of ARF. Carditis can occur in up to 80% of people with ARF, leading to mitral or aortic valvular disease. About half of those affected will develop chronic RHD. Death from ARF is not common, but chronic rheumatic heart disease can lead to morbidity from arrhythmias, endocarditis, and stroke.

Usual Treatment

- ARF is easily treated with penicillin if initiated within 9 d of onset of pharyngitis; this will prevent the development of chronic manifestations, such as carditis. Corticosteroids can be used in the acute inflammatory stage. Bed rest for several wk is required, followed by gentle ambulation. Diuretics and vasodilators are given for severe carditis as well as furosemide for mild to moderate CHF. ACE inhibitors are given for severe aortic regurgitation. Serial ECHO will determine cardiac dimensions and function. Valve repair surgery should be delayed until after the acute inflammatory stage.
- Chorea: Treatment is supportive, including psychosocial support.
- Arthritis and arthralgias: NSAIDs may mask signs of acute rheumatic fever. By clinical consensus, paracetamol is preferred. After Dx has been made, naproxen twice daily is the drug of choice.

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